UNIVERSIDADE FEDERAL DE UBERLÂNDIA
FACULDADE DE MEDICINA

MEMORIAL DESCRITIVO

LOURDES DE FÁTIMA GONÇALVES GOMES

UBERLÂNDIA
2023

LOURDES DE FÁTIMA GONÇALVES GOMES

MEMORIAL DESCRITIVO

Memorial descritivo apresentado à Faculdade de Medicina da Universidade Federal de Uberlândia, como requisito para a Promoção do Professor Integrante da Carreira do Ensino Básico, Técnico e Tecnológico do nível IV da Classe Associado IV para Classe Titular nos termos da Resolução Nº 03/2017, do Conselho Diretor (CONDIR – UFU).

UBERLÂNDIA

2023

Dados Internacionais de Catalogação na Publicação (CIP)
Sistema de Bibliotecas da UFU, MG, Brasil.

G633m 2023

Gomes, Lourdes de Fátima Gonçalves.
Memorial descritivo [recurso eletrônico] / Lourdes de Fátima Gonçalves Gomes. - 2023.

Memorial Descritivo (Promoção para classe E - Professor Titular) - Universidade Federal de Uberlândia, Faculdade de Medicina.
Modo de acesso: Internet.
Disponível em: http://doi.org/10.14393/ufu.di.2023.7052
Inclui bibliografia.
Inclui ilustrações.

1. Medicina - Estudo e ensino (Superior). 2. Professores universitários. I. Universidade Federal de Uberlândia. Faculdade de Medicina. II. Título.

CDU: 61:378

Glória Aparecida
Bibliotecária Documentalista - CRB-6/2047

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FACULDADE DE MEDICINA

MEMORIAL

LOURDES DE FÁTIMA GONÇALVES GOMES
UBERLÂNDIA, 2023

BANCA EXAMINADORA

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Representante Interno Titular

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Representante Externo Titular

Profa. Dra. Virgínia Paes Leme Ferriani
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Prof. Dr. Luiz Carlos Marques de Oliveira
Representante Interno Suplente

“O Senhor é meu pastor,
nada me faltará
Deitar-me faz em verdes pastos e
guia-me mansamente em águas tranquilas,
refrigera minha alma
e guia-me pelas veredas da justiça
Por amor de seu nome,
ainda que eu andasse pelo vale da sombra da morte,
não temerei mal algum,
Porque tu estás comigo.”
Salmo de Davi 23 (1-4)

“Ensinar não é transferir conhecimento, mas criar as possibilidades para a sua própria produção ou a sua construção.”

(Paulo Freire)

AGRADECIMENTOS

Obrigada,
meu Senhor e meu Tudo! Obrigada pela generosidade de me permitir a vida.

A Elvis,
obrigada pelo amor intenso e incondicional o tempo todo!

A Guilherme,
obrigada pelo seu amor e alegria, juventude, companheirismo e gosto pela vida!

Aos meus pais, Alcindo e Josina,
pelo amor que me deram na vida e por me guiarem. Obrigada por me ensinarem a essência da existência, do conhecimento, da realidade, foco e a ver a simplicidade no amor a Deus! . A Sra Albertina Gomes pelo apoio, carinho e sabedoria

Aos meus irmãos e irmãs e a todos os meus familiares
(Inez e Reginaldo; Lucia Aparecida, Alcindo e Márcia; José Roberto e Geane; João e Aurélia e seus filhos, Hélia Gomes, Maria Gomes , Elvécio Silva e Eliane Silva),
cujo amor, amizade e harmonia fortalecem cada vez mais nossos laços de vida. Obrigada pelo entendimento pela minha ausência de suas vidas durante toda esta trajetória de vida. A Sra Sirleide Simões pelo cuidado, disponibilidade e afeto.!

A Guilherme Silva Mendonça,
pela amizade e por ter me conduzido na organização da trajetória que me levou à realização deste memorial. Um Professor nato na construção acadêmica. Deus abençoe você e sua família sempre!
Muito Obrigada.

Ao Professor Dr. Carlos Henrique Martins Silva,
pela sua amizade e por ser meu inspirador em todas as etapas de minha vida profissional. Deus ilumine e guarde-o sempre!

Ao Professor Dr. Werther Brunow de Carvalho e a todos os colegas da UTI Pediátrica do HSP-EPM-UNIFESP, obrigada por todos os ensinamentos, especialmente no incentivo inicial à produção científica.

Ao Professor Dr. Antônio Carlos de Camargo Carvalho,
serei eternamente grata por toda a confiança, seu apoio incondicional e por me guiar nesta vida de aluna, médica, pesquisadora e professora. O senhor foi o meu tutor e guia acadêmico. Que Deus o tenha em um bom lugar!

À professora Dra. Célia Maria Camelo da Silva,
obrigada por todo ensinamento e tutoria em minha vida profissional, pela sua amizade, carinho, e apoios preciosos e constantes. !

Aos professores , doutores, e colegas da UNIFESP
(Dr Valdir Ambrósio Moisés, Dr. Victor Manoel Oporto, Dr. Ranulfo Pinheiro Matos Neto, Dra. Fernanda Maria Rebouças, Dra. Maria de Fátima Freire, Dra. Maria Suely B. Dyógenes, Dra Patricia Belo e todas Secretarias – Edileuza , Roseli e Solange), obrigada pela amizade demonstrada e por todos os ensinamentos compartilhados!

A todos os médicos da UTI Pediátrica e Neonatologia do Hospital São Paulo – Escola Paulista de Medicina – UNIFESP, representados na pessoa do professor e Dr. Benjamin Israel Kopelman, Dra Maria Fernanda Branco e Dra Ruth Gunsburgh, obrigada pelo acolhimento em todo este período e por todoapoio à minha trajetória!

A todos os professores médicos e funcionários do Departamento de Pediatria, representados na pessoa do Dr Hélio Lopes e Chefia atual Dra Lídia Mayrink, obrigada por toda amizade, carinho, apoio e ajuda em meu transitar pela vida acadêmica e profissional!

A todos os médicos, equipes de enfermagem, manutenção, integrantes da pediatria do Hospital de Clínicas da Universidade Federal de Uberlândia, obrigada pelo companheirismo e pelo entendimento por todos esses anos, especialmente, no acolhimento da criança cardiopata!

A todas as equipes que compõem a UTI Pediátrica e UTI Neonatal do Hospital de Clínicas da Universidade Federal de Uberlândia, obrigada pela acolhida e por propiciarem tantos momentos maravilhosos de trabalho intensivo!

Ao Professor Dr. Vilmar José Pereira, a todas as equipes envolvidas na abordagem hemodinâmica e intervencionista da criança cardiopata, obrigada pela amizade e pelos anos de trabalho construído em todos os sentidos, incluindo a formação dos novos cardiologistas!

Ao professor Dr. Claudio Ribeiro, Dr. Daniel Oliveira De Conti, Dr. Paulo César Santos e toda a equipe de Cirurgia Cardiovascular, obrigada pelo apoio e por realizarem tão bem o tratamento cirúrgico da criança cardiopata! . Pelo milênio de crianças operadas

À Dra. Neide Aparecida Faria e à Dra. Viviane Athadeu Gontijo,
obrigada pela amizade e pelo respeito conquistados em todos esses anos, representando todos os Cardiologistas Pediátricos envolvidos na abordagem global da criança cardiopata! Ambas “Prata da Casa” – Agora o cardiopata está caminhando mais em direção a vocês – acolham-no como ele deve ser acolhido – global e integralmente!

Em especial a Nilma Aparecida de Assunção,
Ana Aparecida Domingues Melo
Elena Yara de Oliveira do Nascimento
Vera Lúcia Vieira Mendes,
representantes de toda a parte executiva operacional da Cardiologia Pediátrica, Congênita e Fetal Cardiológica, obrigada pela competência , carinho apoio e constância diários.

À Leda Maria Viana Borges e à Juliene Cristine de Oliveira
(toda a equipe do Projeto Amigos do Coração, Posso Ajudar e Instituto Mãos Dadas),
obrigada por todo apoio recebido em todos esses anos de trabalho realizado na humanização da criança e dos familiares de portadores de Cardiopatia Congênita!

À Ana Flávia Sena Barbosa e à Silvia Regina de Lima,
que me impulsionaram e por todo auxílio e entendimento necessários, para que terminasse este trabalho começado! Pelo seu carinho, leveza e acolhimento com doçura e precisão. Deus as abençoe!

A Fernanda dos Santos Abreu, Roberta Cristina Silva e Moreira, que sempre me acolhem na FAMED, pela educação e pela fineza no trato, pela atenção e pelo carinho que recebo vindos de vocês. Deus esteja sempre com vocês!

A todos os alunos que passaram pela Universidade Federal e Hospital de Clínicas durante a minha trajetória, muito obrigada! Vocês foram importantes na construção da minha carreira profissional!

A todas as pessoas que me auxiliaram em minha trajetória e estão no anonimato, na construção deste memorial,
muito obrigada! Vocês foram essenciais na construção da minha carreira profissional!

Ao paciente que foi, é e será a causa de todo o progresso e o aprendizado.
Muito obrigada!

Aoa Acadêmicoss, Internos e Residentes e em memória a os nossos antepassados que trabalharam e conseguiram melhorar o mundo para podermos viver melhor e com mais conhecimento, Muito obrigada!

À Exma. Diretora Professora Dra. Catarina Machado Azeredo – Diretora da Faculdade de Medicina da Universidade Federal e Uberlândia –
por sua sutileza, educação, meiguice e ternura diante de tantos desafios! Deus a abençoe em seu cargo e a proteja! Muito obrigada!

Ao professor reitor Dr. Valder Steffen Júnior e Vice-Reitor professor Dr. Carlos Henrique Martins Silva e toda a reitoria,
obrigada por disponibilizarem suas vidas para dirigirem esta universidade.
E a todos os dirigentes pregressos e nossos antepassados.

A todos os Professores, Médicos e Titulares da FAMED – UFU,
representados pela professora Dra. Valéria Bonetti e professor Dr. Orlando Cesar Mantese e Professor Dr. Carlos Henrique Martins Silva,
obrigada por me inspirarem a evoluir em minha carreira profissional.

A todas as equipes médicas e não médicas,
UTI Pediátrica, Neonatologia, Enfermaria de Pediatria, Pronto Socorro Infantil, Ambulatório de Pediatria Geral e Especialidades, Cardiologia, Ecocardiografia, Hemodinâmica e Intervenção, Cirurgia Cardiovascular, Centro cirúrgico, Cardiologia, Pronto Socorro Adulto, Clínica Médica, Portaria Hospitalar, Manutenção, Radiologia, Tomografia e Ressonância, Ginecologia e Obstetrícia, Ultrassonografia,
sem a integração e a força dessas equipes, nada existiria, nem o paciente.

À Banca Examinadora,
todos aqueles que compõem as estruturas das universidades UFU e UNIFESP, pela presença, pelo carinho e pela disponibilidade por terem se voltado para este exame.
Sem vocês, nada seria possível.

A Deus,
por ter sido tão generoso comigo, por ter me brindado com todas essas pessoas que com seu amor e dedicação me iluminam em todo o caminho percorrido!

“A felicidade é feita de coisas pequenas e simples e quase sempre estão perto de nós”
(Anderson Cavalcante)

RESUMO

Neste relato, descrevo os principais caminhos percorridos na vida de professora e meu desenvolvimento profissional acadêmico, como integrante do Departamento de Pediatria da Universidade Federal de Uberlândia. Este memorial é requisito para progressão no Magistério de Professora Associada IV para Professora Titular da Carreira de Magistério Superior, de acordo com a Resolução Nº 03/2017, do Conselho Diretor da Universidade Federal de Uberlândia. Minha vida escolar pré-universitária, graduação em Medicina e residência em pediatria geral foi desenvolvida em Uberlândia/MG. Decidi fazer Medicina e ser professora antes mesmo de entrar na escola, pois venho de família de professores dedicados ao ensino. Em 1979, mediante o concurso vestibular, fiz graduação no curso de Medicina da Faculdade de Medicina na Universidade Federal de Uberlândia e me formei em 1984. Em 1985 e 1987, cursei a Residência Médica em Pediatria Geral, por concurso público, no Hospital de Clínicas da Faculdade de Medicina na Universidade Federal de Uberlândia. Após o período de Residência, fui contratada via Departamento de Pediatria da Universidade Federal de Uberlândia com funções de ensinos teóricos e práticos aos novos acadêmicos e residentes recém-admitidos no Pronto Socorro Infantil, na Enfermaria de Pediatria, na Unidade Neonatal e nos plantões de Pediatria, de janeiro a julho de 1987. Após concurso como docente substituta, de agosto de 1987 a dezembro de 1988, mantive-me nas funções de ensino na graduação e na residência médica em pediatria, na coordenação do ambulatório de pediatria geral para internos e residentes e nas atividades assistenciais na enfermaria de pediatria, no pronto socorro e no atendimento de pacientes graves. Em 1998, após prova, obtive o título de Especialista em Pediatria Geral, pela Sociedade Brasileira de Pediatria. Nesse período, via concurso pela Prefeitura Municipal, trabalhei na Unidade Básica de Saúde do Município de Uberlândia, onde aprendi muito sobre saúde pública. Eu coordenava as reuniões de pré-natal com grupos de gestantes (saúde fetal e pré-natal, puericultura com as mães e pais e atendimento ambulatorial de pediatria geral). Graças à minha necessidade de aprender mais, ao interesse pelo paciente grave e ao início da Unidade de Terapia Intensiva Pediátrica, pelo professor Dr. Orlando Cesar Mantese, em 1989, via concurso, iniciei Residência em Terapia Intensiva Pediátrica no Hospital São Paulo, na Escola Paulista de Medicina da Universidade Federal de São Paulo, com o professor Dr. Werther Brunow de Carvalho. Concluí a Residência de Terapia Intensiva Pediátrica em 1990 e obtive o título de Especialista em Terapia Intensiva Pediátrica pela Associação de Medicina Intensiva Brasileira. Nos dois anos seguintes, especializei-me em Neonatologia e Terapia Intensiva Neonatal na Unidade de Neonatologia do Hospital São Paulo, sob a coordenação do professor Dr. Benjamin Israel Kopelman, e obtive o título de Especialista em Neonatologia, em 1993. Em fevereiro de 1992, um sonho realizado, como contrato emergencial (retomado após concurso em 1995), assumi como intensivista e preceptora com funções assistenciais, didáticas e pesquisas, cumprindo carga horária, nos fins de semana, na UTI Pediátrica de Uberlândia, no Hospital de Clínicas de Uberlândia, sob a chefia do professor Dr. Orlando César Mantese. Em novembro de 1995, na defesa do mestrado na Universidade Federal de São Paulo, orientada pelo professor Dr. Antônio Carlos Camargo Carvalho, obtive o título de Mestre em Pediatria. Em 1996, iniciei a especialização em Cardiologia Pediátrica e Congênita no Hospital de São Paulo, assim obtive o título de Especialista em Cardiologia Pediátrica e

Congênita na prova de títulos, em 2004, pela Sociedade Brasileira de Cardiologia e Sociedade Brasileira de Pediatria. Após aprovação no concurso público, iniciei o cargo de docente na Universidade Federal de Uberlândia, em agosto de 1998. Como itinerante, concluí a especialização em Ecocardiografia Pediátrica, Congênita e Fetal. Avançamos na assistência, no ensino e na pesquisa (primário ao terciário) em pediatria e UTI Pediátrica e em cardiologia pediátrica e ecocardiografia na graduação e residência médica. Assim, com a UTI pediátrica e neonatal já bem estabelecidas, iniciamos com o programa de cirurgias cardiovasculares no Hospital de Clínicas de Uberlândia em 2004. Em 2005, tornamo-nos Centro de Referência credenciado pelo Ministério da Saúde, Ministério da Educação e Cultura e Sociedade Brasileira de Hemodinâmica, SBC e SBP, em Assistência e Tratamento em Cardiologia Pediátrica (clínico, cirurgia cardiovascular, intervenções hemodinâmicas). Assim, iniciamos a especialização (2006) e Residência Médica em Cardiologia Pediátrica e Congênita (2007) e a pós em ECO, em 2008. Em 2012, obtive o título de Doutora em Cardiologia Pediátrica e Congênita no HSP-EPM-UNIFESP, com a defesa de tese sob orientação do professor Dr. Antônio Carlos de Camargo Carvalho, Titular de Cardiologia. Assim, a formação da especialidade em cardiologia pediátrica, congênita, fetal, cirurgia cardiovascular e intervenções foi muito positiva e válida por resultar em trabalhos científicos, artigos para a pediatria e em outras áreas do Hospital de Clínicas de Uberlândia. Foram realizadas dissertações de mestrado e doutorado (Cirurgião Cardiovascular Dr. Cláudio Ribeiro da Cunha, responsável pelas cirurgias cardiovasculares e congênitas; e professora Dra. Alessandra Carla Ribeiro, informações científicas de qualidade com apresentações, participações em congressos de UTI Pediátrica, Neonatologia, Pediatria e Cardiologia Pediátrica e Congênita, ECO, Cirurgia Cardiovascular). Na minha vida acadêmica, participei, ainda, como autora e coautora de capítulos de livros, pesquisas científicas publicadas em periódicos nacionais, internacionais, apresentação de trabalhos em eventos científicos, conferências, aulas, palestras em reuniões, congressos regionais, nacionais e internacionais. Participei de bancas de mestrado, doutorado e coordenação/organização de eventos científicos, desde a pós-graduação até o mestrado e doutorado, além de pequenos seminários, reuniões científicas e projetos sociais. Incentivamos acadêmicos e alunos em pesquisas, apresentações em congressos, seminários. Construímos linha de pesquisa em Doença de Kawasaki, cardiopatia congênita cianótica e acianótica, insuficiência cardíaca ECO fetal e neonatal, miocardiopatia dilatada, cirurgia cardiovascular, cardiopatias neonatais, hipertensão pulmonar, atenção às famílias atendidas cardiopatas ou não e intervenções hemodinâmicas cardiovasculares. Tornamo-nos Centro de Referência nos protocolos estaduais de Palivizumabe, Sildenafil e Bosentana no tratamento de hipertensão pulmonar pré e pós-operatório de cardiopatia congênita. Coorientei 5 alunos em projetos de pesquisa, 10 alunos em Residência de Cardiologia Pediátrica e Congênita, 8 alunos em ECO. Atualmente, eles são profissionais e exercem a especialidade em diferentes localidades. Assim, na realização deste memorial, o desafio esteve em como construí-lo. Foi necessário um exercício longo, que resultou neste trabalho cuidadoso. Ele reflete a disposição em resgatar fatos vividos, interpretá-los e entender que as derrotas aqui não relatadas são muito mais numerosas e constituem um fundamento para as possíveis vitórias. Esta versão final do Memorial passou por revisão técnica do texto no sentido de adequação às normas vigentes da língua portuguesa, sendo realizado por três revisores de gramática e ortografia, conforme declaração em anexo. O capítulo final desta jornada ainda está por ser escrito.

“Não há saber mais ou saber menos, há saberes diferentes”

(Paulo Freire)

SUMÁRIO

1 INTRODUÇÃO

A Resolução Nº 03/2017, do Conselho Diretor (CONDIR – UFU) veio contemplar o anseio de vários docentes desta instituição na promoção da carreira de associado IV para a classe titular. A construção de um memorial com esta finalidade não é uma exigência apenas da Universidade Federal de Uberlândia (UFU), pois outras Instituições de Ensino Superior (IES) a utilizam para este fim.

O memorial (do latim *memoriale*) é uma produção escrita das memórias de fatos marcantes de seu autor. Uma particularidade do memorial consiste na permissão da construção de uma autoavaliação, na qual são incluídos comentários pessoais, atribuindo-lhes julgamento de valor, o que o difere basicamente do *curriculum vitae*.

Segundo França e Vasconcellos (2007), memorial é o relatório exigido em universidades para obtenção de progressão vertical na carreira dos docentes, cuja estrutura básica consiste no relato resumido dos feitos marcantes da vida acadêmica, científica e profissional do autor. Essa estrutura é semelhante à das dissertações e teses. A decisão da melhor estratégia narrativa (ordem cronológica ou outra) fica a critério do escritor.

Com base nesses conceitos, este memorial descritivo tem o objetivo de apresentar os principais caminhos da minha trajetória acadêmica e profissional até a presente data. Neste trabalho, pretendo fundamentar a expectativa do cumprimento de mais esta etapa de minha carreira como docente.

Este memorial relata o desenvolvimento profissional de Lourdes de Fátima Gonçalves Gomes no ensino, na pesquisa, na extensão e na gestão de forma integrada, como membro integrante do Departamento de Pediatria da Faculdade de Medicina da Universidade Federal de Uberlândia. Ele requisita a progressão no Magistério de Professora Associada IV para Professora Titular da Carreira de Magistério Superior, de acordo com a Resolução Nº 03/2017, do Conselho Diretor (CONDIR – UFU).

Acredito que ocupar o cargo de Professora Titular do Departamento de Pediatria da Faculdade de Medicina da Universidade Federal de Uberlândia não é apenas um prêmio, uma função privativa, mas principalmente é receber um dever. Este fato indica que sou uma pessoa capaz de pensar na Universidade com objetivo

de buscar a excelência na construção do ensino, da pesquisa, da gestão e da estratégia de orientação da gestão como futuro com formação capaz de influenciar a tomada de decisões de políticas acadêmicas e a instituição. Esse dever é mais do que uma posição pessoal, pois é fundamentalmente uma posição institucional.

A aptidão em agrupar indivíduos em torno de um objetivo pode ser um modelo viável de ser seguido. Uma liderança política na universidade acadêmica não pode ser isolada e, para chegar à excelência, não deve esquecer as pessoas que trabalham para tal intento. Por isso, aqui agradeço a todas as pessoas que de qualquer forma me auxiliaram a trilhar este caminho, especialmente às que ficaram no anonimato, o meu reconhecimento, o meu muito obrigada. E a excelência em tratar o paciente e o nosso aluno é a meta que devemos perseguir e alcançar independentemente de onde nos encontramos. Assim, o capítulo final desta jornada está ainda por ser escrito.

Grata,

Lourdes de Fátima Gonçalves Gomes

"Feliz aquele que transfere o que sabe e aprende com o que ensina"

(Cora Coralina)

2 IDENTIFICAÇÃO

Eu vivi grande parte da minha vida em Uberlândia, onde meus pais residem. Fui criada em uma família de professoras e, em todas as brincadeiras de infância, a escolinha fazia parte, bem como o cuidar recíproco dos meus irmãos, em que um cuidava do outro e todos andávamos juntos. Sou muito grata aos meus pais e aos familiares, que me acompanharam sempre.

Meu nome é Lourdes de Fátima Gonçalves Gomes, nasci na cidade de Uberlândia, em Minas Gerais, em 11 de outubro de 1959. Filha de Alcindo Gonçalves Cunha e Josina Aparecida Naves Gonçalves. Sou casada com Elvis Gomes, desde 1986.

Meus dados documentais são:

- Registro Nascimento: Cartório Antonino Martins da Silva Nº 46950. Talão: 238. Folha: 195º Livro Nº A94. Data Emissão: 13 de outubro, 1959;
- RG: 1415884 Órgão Emissor: SSP / MG Data de emissão: 26/08/2014;
- Certidão de casamento: Cartório Antonino Martins da Silva Nº 14680.0 Folha: 154º Livro Nº 37-B Data Emissão: 17 de julho, 1986;
- Passaporte de Imunização: COVID 19 Imunização atualizada – 2020;
- Título de Eleitor: 2202912700167 – Sessão 374 Seção: 0239 Município/UF: São Paulo/SP Data de emissão: 23/09/2016;
- E-mail: lourdes.gomes@ufu.br e lourdes.gomes@uol.com.br.

3 OBJETIVOS

3.1 Objetivo geral

Alcançar a promoção da classe de Professora Associada IV para a classe de Professora Titular da Carreira de Magistério Superior, por meio de avaliação do desempenho acadêmico, acompanhado do relatório individual de atividades.

3.2 Objetivos específicos

- Apresentar um relato resumido de vida;
- Descrever a trajetória acadêmica e profissional;
- Contribuir como documento para alcançar a promoção da Classe de Professora Associada IV para a Classe de Professora Titular da Carreira de Magistério Superior, nos termos da Resolução Nº 03/2017, do Conselho Diretor (CONDIR – UFU).

4 FORMAÇÃO

Minha vida escolar foi desenvolvida na cidade de Uberlândia/MG. Inicialmente do primeiro ao terceiro ano na Escola Estadual Honório Guimarães, e a minha primeira professora foi a Senhora Maria Tereza. Marcou-me nela a letra bonita no quadro e sua postura elegante. Daí por diante, foi com muito entusiasmo que frequentei todas as escolas. Uma das minhas lembranças mais marcantes é a da minha mãe sempre lendo livros e jornais, também era uma verdadeira artista dos tecidos. Meu pai lia todas as noites por pelo menos duas horas e sempre se interessou muito pelo nosso estudo, além de ser um artista e arquiteto da madeira. Na minha casa, a ordem era harmonia, amizade, cuidado, zelo mútuo entre os irmãos, companheirismo, felicidade e amor. Foi nesse cenário de vida que cresci e desenvolvi minha vida escolar até o vestibular. Desde então, já havia me decidido por fazer o curso de Medicina, ser professora e seguir a carreira do magistério, como tradição em minha família.

4.1 Formação Escolar Pré-universitária

O Ensino Fundamental e Médio foi cursado em instituições diferentes, conforme apresentado abaixo:

- 1ª a 3ª séries: Escola Estadual Honório Guimarães;
- 4ª série: Escola Estadual Coronel José Teófilo Carneiro;
- 5ª série ao 3º colegial: Instituto Teresa Valsé;

No ano de 1978, concluí o colegial – hoje Ensino Médio – e, no ano de 1979, fui aprovada no Concurso Vestibular da UFU e iniciei a graduação no curso de Medicina.

4.2 Formação universitária em nível de Graduação

Enquanto eu progredia em meus estudos, paralelamente, nascia a Universidade Federal de Uberlândia, que foi criada pelo decreto Lei nº 762, de 14 de agosto de 1969, e alterada para Fundação de Ensino Superior (Lei nº 6532, de 24 de maio de 1978), com sede em Uberlândia, estado de Minas Gerais.

Avançando nessa meta, fui aprovada no Concurso Vestibular 1979/1º Semestre Unificado, na Universidade Federal de Uberlândia (UFU), obtendo 68.467

pontos e, no mesmo ano, iniciei o Curso de Graduação em Medicina, com o número de matrícula 2791020. Concluí o curso em 1984, na mesma universidade.

Mas foi muito marcante especialmente para a minha turma (13ª) a Federalização da Universidade Federal de Medicina, elevada à categoria Federal pelo Exmo. Sr. Presidente João Baptista de Oliveira Figueiredo, durante sua visita ao Hospital de Clínicas da Faculdade de Medicina e Universidade Federal de Uberlândia em maio de 1979. Assim, foi possível continuar e me formar médica por essa faculdade. Fiz o curso médico com muito entusiasmo, fascínio e empolgação e fui transitando pelas cadeiras básicas, hospitalares e me identifiquei e gostei de todas as áreas e dos estágios desenvolvidos de 1979 a 1984. Envolvi-me completamente na Medicina e admirei e admiro muito, respeito e agradeço todos os meus professores pela disponibilidade, pelo carinho, pela atenção, pelo compromisso e pela dedicação à minha formação. No período de férias, me dedicava a fazer cursos extracurriculares e acompanhava os professores nas visitas clínicas e cirúrgicas aos pacientes. O lugar que mais frequentava durante o curso básico era a biblioteca e estudava rotineiramente até o fechamento dela (às 22 horas). Segui na plena certeza de que Medicina é o que me fascina. Fui monitora nas disciplinas de Histologia, Cirurgia geral, Pediatria, Clínica Médica, Imunologia, Semiologia Pediátrica e Puericultura.

No internato médico, realizei muitos estágios e cada um mais fascinante que o outro, incluindo Ginecologia e Obstetrícia, Ortopedia e Traumatologia, Cirurgia Geral, Clínica Médica, Cardiologia, Gastroenterologia, Radiologia, Nefrologia, Grande Queimado, Otorrinolaringologia, Saúde Pública, Medicina Preventiva, Dermatologia, Hematologia, Oftalmologia, Neonatologia, Pediatria, Proctologia, Oncologia, Emergência e Cirurgia Pediátrica com o professor Dr. Nilson de Abreu. Durante o curso, todos os professores foram marcantes e aqui os represento por Dra. Claudia Lúcia de Matos, Dr. Delcides Faleiros, Dr. Elisio de Castro, Dr. Enio Avelar Naves, Dr. Evando Guimarães Filho, Dr. Hélio Teixeira, Dr. Gladstone Rodrigues da Cunha, Dra. Maria José Junho Sologuren, Dr. Melicégenes R. Ambrósio, Dr. Renato E. Sologuren Acha, Dr. Rimmel A. Guzmana Heredia, Dr. Silésio do Prado, Dr. Toshirico Hashimoto, Dr. Walter Manhães, Dr. Aguinaldo Coelho da Silva, Dr. Elmiro Resende Santos, Dr. Takeo Iwace, Dr. Nilson de Abreu, Dr. Antonio Geraldo D. Roquete, Dr. Ben Hur Braga Taliberti, Dra. Valéria Bonetti, Dra. Vania Olivetti Steffen Abdallah, Dr. Marcelo Simão Ferreira, Dr. Nestor Barbosa

de Andrade, Dr. Eduardo de Andrade, Dra. Leda Maria F. da Silva Lima, Dr. Paulo Tannus Jorge, Dr. Divino Prudente, Dr. Hélio Lopes da Silveira. Foi nesse cenário que concluí o curso de Medicina em 1984. Para mim, foram professores excelentes no ensino da Medicina.

Sei que fiz a escolha certa em estudar Medicina e na Faculdade de Medicina da Universidade Federal de Uberlândia. Eu tenho muito respeito pelos professores em sua totalidade e reconhecimento de que a Universidade Federal de Uberlândia ocupa destaque na formação dos seus alunos. O curso de Medicina é excelente e contribuiu muito para minha realização pessoal e profissional e norteou minha vida. Muito obrigada a todos os seus professores, passados e atuais.

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4.3 Formação universitária em nível de Pós-graduação

4.3.1 Residência Médica em Pediatria Geral

Como eu gostei de todas as especialidades vivenciadas na graduação, tive muita dificuldade na escolha da que iria fazer. Inclinava-me para área cirúrgica, mas não suportava ficar muito tempo paramentada e de pé. Decidi-me por fazer Pediatria, por ser uma especialidade ampla, que comtempla uma clínica forte abrangente e não segmentava o paciente. Realizei o concurso para Residência Médica em Pediatria Geral, em Brasília, Goiânia, Ribeirão Preto e Uberlândia, que eram, naquela época, os lugares considerados de excelência no ensino e tinham mais disponibilidade e rapidez de locomoção para viagens. Foi muito importante, pois vivi outras realidades de avaliação do conhecimento em Medicina.

Após conclusão da Graduação, também fiz o concurso de Residência Médica na Faculdade de Medicina da Universidade Federal de Uberlândia (FAMED-UFU) e, quando aprovada, desisti de fazer residência em outras localidades. Iniciei o Programa de Residência Médica (R1 e R2) na área de pediatria, sendo cursada no período de 3 de janeiro de 1985 a 3 de janeiro de 1987.

Escolhi Uberlândia para não me afastar de minha família, especialmente de meus pais. Com muita felicidade e vínculo ao Hospital de Clínicas da Faculdade de Medicina, fiz Residência Médica em Pediatria na UFU, no período de 1985 a 1987. Meu envolvimento foi muito grande e minha felicidade era tal que eu estudava os casos e, além de discutir, participava com a enfermagem e ficava junto ao paciente até que tudo fosse realizado.

O que mais me marcou na Pediatria foi a proximidade que tínhamos com os professores e a disponibilidade deles em ensinar e nos acompanhar. O programa teórico era executado por eles com muito rigor sob a supervisão do professor Dr. Elísio de Castro. Dediquei-me de corpo e alma à residência e discutia todos os casos com todos os especialistas. Envolvia-me com tudo e com todos relacionados aos pacientes. Como eu gostava de cirurgia, acompanhava os procedimentos cirúrgicos realizados nos meus pacientes e foi assim que aprendi e realizei muitos procedimentos, como intubação, flebotomia, drenagem de abscessos, drenagem de tórax, punção lombar, acessos vasculares, pericardiocentese, outras paracenteses, traqueostomia, exsanguineotransfusão. Era uma seguidora do cirurgião e professor Dr. Nilson de Abreu e acompanhava todos os procedimentos após o término de minhas atividades diárias de residência médica, usualmente à noite e fins de semana. Terminava o ambulatório e me dirigia ao Pronto Socorro Pediátrico. Aprendi muito com todos os professores e colegas médicos administrativos: Dr. Marco Antônio, Dra. Maria Inez, Dra. Kátia, Dr. Oliveira e neurologia pediátrica com o professor de Neurologia Pediátrica e pós-graduação na França, Dr. José Martins Borges – com ele, todos os dias tínhamos visitas e discussões clínicas no Pronto Socorro Infantil. Assim, além de ver os pacientes, discutíamos artigos e livros, entre eles, o *Neurologia Infantil* Lefréve, de Aron Diament & Saul Cypel, por seminários diários. Havia muitos casos de neuropatias, especialmente meningites, convulsões, malformações cerebrais, trauma, síndromes neurológicas. Sentimos muito sua falta e sua morte foi uma perda irreparável para todos nós, alunos, e para o DEPED-UFU. Tudo era resolvido no Pronto Socorro Infantil. Como não tínhamos terapia Intensiva, discutíamos todos os casos entre os professores e nós residentes (residentes do 2º ano: Dra. Aglai Arantes, Dr. Abelardo José Carvalho Campos, Dra. Luzinete da Silva Santiago, Dra. Márcia Berbet Ferreira, Dra. Márcia Aparecida Mendes, Dra. Maria Neuza Gomes da Silva Lobato, Dra. Neuza Helena M. de Melo Fernandes) e os residentes do primeiro ano (Dr. Fernando Jorge, Dra. Gilca Ribeiro Starling Diniz Marra, Dra. Lilian Sanchez Lacerda, Dra. Maly de Albuquerque, Dra. Rossane Cristina Dalia de Melo, Dra. Vera Lucia Barra Bisnoto); e os plantonistas do Pronto Socorro, que eram os próprios professores. Com a chegada do professor Dr. Orlando César Mantese (UTI Pediátrica), da professora Dra. Vania Olivetti Steffen Abdallah (Neonatologia), da professora Dra. Valéria Bonetti (Nefrologia) e da professora Dra. Virgínia Paes Lemes Ferriani (Pediatria Geral), professor Dr. Hélio

Lopes da Silveira (Pneumologia) e, posteriormente, professor Dr. Carlos Henrique Martins, novos horizontes foram se abrindo na Pediatria. Assim, as especialidades foram se construindo pelo paciente que precisava de tratamento, aliado aos professores capacitados e, em consequência, capacitando os residentes até alcançarem o formato estruturado e reconhecido pelas respectivas Sociedade Brasileira de Pediatria, Ministério da Saúde e Ministério da Educação e Ensino.

Paralelamente, foram sendo ocupados espaços nas atividades de gestão e de pesquisa na universidade, como a semana científica: aulas, palestras, conferências com professores de outras universidades, jornadas, encontros, seminários e congressos e pequenas publicações. Desse modo, o foco tornou-se o progresso com a melhora do ensino, também a formação de outras Especialidades, Gestão e Pesquisa Científica. Nesse contexto, terminei a minha Residência Médica em Pediatria. No segundo ano de Residência em Pediatria na Unidade Neonatal da UFU, fui indicada pela professora Dra. Claudia Matos e pela professora Dra. Vania Stefen, para conduzir um neonato portador de cardiopatia congênita canal dependente para tratamento cirúrgico cardíaco para o Hospital Beneficência Portuguesa de São Paulo, Equipe do Cirurgião Cardiovascular, professor Dr. Miguel Lorenzo Barbeiro Marcial. Fiquei maravilhada ao entrar no Hospital Beneficência Portuguesa. Eram 23 horas, mas pareciam 7 horas da manhã, tal era o movimento de todos profissionais. Aí sim, naquele momento, tive rapidamente uma ideia do que é uma estrutura potente e rica em tudo. Foi nesse contexto que eu persisti em seguir a carreira acadêmica e acendeu mais forte a chama que me chamava para Cardiologia Pediátrica e Congênita. Como residente, foi uma das melhores e mais ricas experiências vividas e norteou, de certa forma, minha vida futura. Era preciso aprender muito até chegar à criança com cardiopatia congênita e adquirida.

Esses dois anos de residência médica foram bem vividos enquanto executava as atividades teóricas e práticas determinadas pelo programa do DEPED-UFU nas salas de aulas e nas atividades práticas nos ambulatórios e unidades hospitalares da pediatria e emergência com o paciente grave do HC-FAMED-UFU.

Após a Residência, fui contratada via Departamento de Pediatria/Universidade Federal de Uberlândia (DEPED/UFU), com funções de ensinos teóricos e práticos aos novos acadêmicos e residentes recém-admitidos no Pronto Socorro Infantil, Enfermaria de Pediatria e Unidade Neonatal e plantões de Pediatria de janeiro de 1987 a julho de 1987. Após concurso para docente substituta,

de agosto de 1987 a dezembro de 1988, eu mantive-me nas funções de ensino na graduação e na residência médica em pediatria, na coordenação do ambulatório de pediatria geral para os internos e residentes e nas atividades assistenciais na enfermaria de pediatria, pronto socorro e pacientes graves. Nesse período, via concurso pela Prefeitura Municipal, trabalhei na Unidade Básica de Saúde do Município de Uberlândia, onde aprendi muito sobre saúde pública. Eu coordenava as reuniões de pré-natal com grupos de gestantes (saúde fetal e pré-natal) e puericultura com as mães e os pais e fazia atendimento ambulatorial de pediatria geral.

Na data de 23 de março de 1988, fui aprovada na prova de Títulos de Especialista em Pediatria – TEP, em conformidade com o convênio entre a Associação Médica Brasileira (AMB), a Sociedade Brasileira de Pediatria (SBP) e o Conselho Federal de Medicina (CFM). Em 31 de maio de 1988, foi-me conferido o título de Especialista em Pediatria.

A minha crescente necessidade de aprender mais, o interesse pelo paciente grave e o fato de que, em outubro de 1988, ter sido iniciada a construção da Unidade de Terapia Intensiva Pediátrica (UTI Pediátrica) pelo professor Dr. Orlando César Mantese no HC-FAMED-UFU, incentivaram-me ainda mais a me especializar em UTI Pediátrica.

4.3.2 Especialização em Terapia Intensiva Pediátrica

No ano de 1989, iniciei a Residência Médica em Terapia Intensiva Pediátrica na Universidade Federal de São Paulo (UNIFESP), sob a orientação do Professor Dr. Werther Brunow de Carvalho. O período de fevereiro de 1989 a fevereiro de 1990, em regime integral, foi intenso e extraordinariamente rico de novos aprendizados em todos os aspectos. O diploma foi emitido pela Escola Paulista de Medicina em 6 de fevereiro de 1990.

Para minha felicidade, era quase inacreditável que eu estava na Escola Paulista de Medicina especializando-me em Terapia Intensiva Pediátrica no Hospital São Paulo, cercada de pessoas capacitadas e brilhantes. Foi nesse período que, além dos objetivos de aprendizado, me dediquei à pesquisa mais intensamente. A busca por aprender sempre mais me levou a participar da apresentação de aulas, encontros, jornadas, congressos, tanto nacionais quanto internacionais, que foram a base para caminhar nas especializações. Percebi o quanto é importante continuar a

me especializar: por mais que eu estudasse, havia muita coisa a aprender para favorecer o paciente, a fim de evitar que sofresse ainda mais pela minha ignorância e pelo meu desconhecimento científico. Foi o atendimento ao paciente crítico, incluindo neonato e criança crítica no pré e pós-operatório de cirurgia cardíaca, que me conduziu às trajetórias seguintes.

Em 22 de abril de 1990, fui aprovada na prova de título como primeira nota (82) pela Associação de Medicina Intensiva Brasileira (AMIB) para obtenção do título de Especialista em Terapia Intensiva Pediátrica. Recebi o referido Título de Especialista e Certificado de Atuação na Área de Medicina Intensiva Pediátrica, emitido pela Associação Médica Brasileira/Associação de Medicina Intensiva Brasileira (AMB/AMIB) e Sociedade Paulista de Terapia Intensiva (SOPATI), em 25 de abril de 1990.

4.3.3 Especialização em Neonatologia e Terapia Intensiva Neonatal

No final da especialização em UTI Pediátrica me direcionei para a capacitação em Neonatologia sob a coordenação do Professor Dr. Benjamin Israel Kolpeman e toda a Equipe Neonatal. Mediante seleção interna para Neonatologia, pude me dedicar à especialização em neonatologia por mais dois anos e meio de muito estudo e encantamento neonatal em todos os segmentos desde a concepção. Nesse período, a Professora Dra. Ruth Guinsburg regressou dos Estados Unidos, então realizei e participei do primeiro curso de reanimação neonatal.

Assim, de 20 de janeiro de 1990 a 31 de março de 1992, realizei estágio com Especialização em Neonatologia e Terapia Intensiva Neonatal, cumpri o programa de especialização desenvolvido na Neonatologia com atividades teóricas e práticas, sob a supervisão da Dra. Maria Fernanda Branco, Dr. Milton Harumi Miyoshi e Dr. Bejamin Israel Kopelmman. As atividades de especialização foram realizadas no Berçário e Terapia Intensiva Neonatal (UTI Neonatal) na Unidade de Neonatologia no Hospital São Paulo na Escola Paulista de Medicina, da Universidade Federal de São Paulo (HSP-EPM- UNIFESP), finalizando esse estágio no ano de 1992.

Esse período foi muito profícuo pelo aprendizado e pela experiência médica em Neonatologia, em que eu pude viver também as atividades exercidas como pediatra neonatologista e plantonista do Amparo Maternal. Inicialmente como bolsista pela Sociedade Paulista para o Desenvolvimento da Medicina, de 1º a 30 de setembro de 1990, posteriormente, no HSP-EPM-UNIFESP, como neonatologista

especializada em exercício de 1º de outubro de 1990 a 31 de março de 1992. Contei, ainda, com atividades de plantonista em sala de parto desde 15 de maio de 1989 até 11 de janeiro de 1990, com registro de 714 horas de sala de parto e, entre setembro de 1990 a 31 de janeiro de 1991, mais 840 horas realizadas na sala de parto pelo Amparo Maternal. Também, atuei como Pediatra Chefe de Plantão da Sala de Parto da Unidade Neonatal e Terapia Intensiva Neonatal desde 1º de fevereiro de 1990 até 31 de março de 1992, contando com 1.104 horas de atividades vivenciadas.

Em abril de 1993, fui aprovada pela Sociedade Brasileira de Neonatologia, SBP e obtive o Título de Especialista em Neonatologia.

4.3.4 Especialização em Cardiologia Pediátrica e Congênita

Em fevereiro de 1996, iniciei a especialização em Cardiologia Pediátrica e Congênita pela Cardiologia no HSP-EPM-UNIFESP, sob a coordenação do professor Dr. Antônio Carlos de Camargo Carvalho e supervisão da professora Dra. Célia Maria Camelo da Silva, até 1998.

Quando comecei as atividades em cardiologia pediátrica, eu percebi que uma nova medicina se apresentava a mim e que não sabia nada sobre o paciente portador de cardiopatia congênita e de cardiopatias. Constatei que a criança cardiopata descompensada é um dos pacientes mais graves e críticos e que morre tão rápido como um piscar de olhos. Assim, meu compromisso com a minha formação foi muito maior. A residência em cardiologia pediátrica foi a que mais exigiu de mim. Por isso, não é infrequente muitos alunos desistirem da residência antes da metade do seu curso. Descobri, também, que a ignorância em cardiologia pediátrica é muito grande e que o cardiologista pediátrico só deve sair do lado da criança em duas situações: quando ela melhora ou quando ela morre, especialmente nas cardiopatias complexas ou nos pós-operatórios complicados. Ciente desses fatos, eu me envolvi e mergulhei plenamente nas atividades do programa, dia e noite, participando de tudo o que era proposto.

Vivenciei um programa de residência inteligente, em que, em todos os seguimentos, nós éramos responsáveis pelo paciente e devíamos acompanhá-lo em sua evolução. Assim, após a visita médica da manhã, tínhamos que participar de todas as reuniões clínicas e cirúrgicas e preparar todos os casos internados, pré e pós-operatório. Todos os dias, participávamos de todos os exames, de propedêutica,

acompanhávamos as cirurgias cardíacas, os ecocardiogramas, os eletrocardiogramas e os métodos gráficos, tomografia computadorizada, angiotomografia, ressonância. Mas dentro da cardiologia pediátrica, tenho três casos de amores: clínica cardiológica pediátrica e arritmia, ecocardiografia congênita, pediátrica e fetal, arritmia hemodinâmica e cateterismos cardíacos e congênitos, e intervenções hemodinâmicas. Meu fascínio por tudo isso só aumentou na minha vida e me sentia e me sinto muito feliz por ser cardiologista pediátrica.

Nesse período me dediquei, também, à produção de trabalhos científicos e acompanhei todos os trabalhos de gestão acadêmica desenvolvidos pela professora Dra. Célia Maria Camelo Silva. Participei na organização de listas cirúrgicas, ministrando aulas de cardiologia pediátrica aos colegas que eram admitidos para residência de cardiologia pediátrica.

Deixo registrado aqui meu agradecimento ao professor Dr. Antônio Carlos de Camargo Carvalho, à professora Dra. Célia Maria Camelo da Silva, Dra. Suely Dyógenes e Dr. Victor Manoel Oporto e toda a equipe da Cardiologia Pediátrica pela acolhida, pela amizade, pelo carinho, pelo apoio, pelo incentivo, pelo respeito e pela orientação na tese acadêmica e na vida profissional. Além de tudo isso, eles implantaram em mim a visão do quanto é importante a documentação científica e me conduziram em pesquisas, trabalhos, congressos, capítulos de livros, pós-graduação.

Mais enriquecedor ainda foi descobrir que exercer cardiologia pediátrica requer desprendimento da vida, pois, muitas vezes, é sofrido e me consome muita energia vital. Resumo esses sentimentos na frase que cito aos acadêmicos e aos pais para entenderem a dimensão que é a gravidade de se lidar com o paciente portador de cardiopatia congênita: “Viva intensamente todos os dias de sua vida como se fosse o primeiro e o último, pois a gente anoitece e não amanhece e amanhece e não anoitece. Viva tudo o que tem para ser vivido agora”. Usualmente, quando falo dessa forma, principalmente os pais entendem claramente o que quero dizer.

4.3.5 Especialização em Ecocardiografia Pediátrica, Congênita e Fetal

A seguir, realizei a especialização em Ecocardiografia Pediátrica e Congênita e Fetal no período de 1999 a 2002, após concurso para especialização realizado na Instituição no Setor de Ecocardiografia Pediátrica do Serviço de Ecocardiografia da

Disciplina de Cardiologia da Escola Paulista de Medicina (UNIFESP), no Hospital de São Paulo.

Nesse estágio, foram realizados e interpretados mais de 3.000 exames ecocardiográficos de crianças e adolescentes com cardiopatias congênitas e adquiridas. Agradeço aqui a todos os integrantes do Departamento de Ecocardiografia pelo auxílio na execução dessa especialidade. Aos professores Dr. Antônio Carlos de Camargo Carvalho, Professora Dra. Maria Célia Camelo da Silva, Professor Dr. Orlando Campos Filho e Professor Dr. Valdir Ambrósio Moisés, que viabilizaram e me apoiaram durante a realização da especialização no HSP-EPM-UNIFESP.

A ecocardiografia é fantástica, trata-se de um método de exames muito evoluído até o momento, e acredito que neste milênio não será substituído. É simples, fácil, barato, acessível, reproduzível, não invasivo, podendo ser feito à beira do leito (especialmente na criança e no paciente crítico e neonato) e modificou a história natural das cardiopatias congênitas e da cardiologia pediátrica. Daí sua importância diagnóstica, terapêutica, com seguimentos cardiológicos em todas as idades.

Concomitante à minha participação em todas essas atividades de especialização, ensino e pesquisa com gestão no HSP-EPM-UNIFESP com meus colegas, eu idealizava como seria a construção de uma especialidade tão intensa e exigente como essa. Criar um espaço físico e estabelecer a filosofia das atividades nessa nova especialidade deve ser baseado no tripé de ensino, assistência e pesquisa com formação de profissionais para preparar novos profissionais e gestores. Esses pacientes são de alta complexidade, geralmente podem ter outras comorbidades e necessitam de conhecimento, raciocínio, condução clínica e aplicação de novas tecnologias.

Também, tive oportunidade de realizar vários trabalhos e estudos em ecocardiografia pediátrica e fetal. Participei de vários estudos envolvendo a ecocardiografia que resultaram em tese de mestrado: Dr. Ranulfo Pineiro Matos Neto – Função sistólica do ventrículo esquerdo pela ecocardiografia em crianças e adolescentes com osteosarcoma, tratados com doxorrubicina com e sem dexrazoxane, e doutorado: Dra. Maria Suely Bezerra Diógenes – Avaliação cardiológica em crianças expostas ao vírus da imunodeficiência humana tipo 1 por via perinatal: estudo clínico, eletrocardiográfico e ecocardiográfico Doppler.

Foi com esse espírito que fiz a especialização e nela pude reafirmar todos os meus aprendizados em todas as especialidades realizadas até aqui. Em março de 2009, após aprovação em prova, obtive o título de Especialista em Ecocardiografia Pediátrica, Congênita e fetal pela Sociedade Brasileira de Ecocardiografia, pela SBC e pela Sociedade Brasileira de Cardiologia Pediátrica.

4.3.6 Especialização em Cateterismo Cardíaco e Intervenções Hemodinâmicas em Cardiologia Congênita e Pediátrica

Desde o meu primeiro contato com a cardiologia pediátrica e com o Professor Dr. Antônio Carlos de Camargo de Carvalho me identifiquei e tive muita afinidade, gosto e encanto pela hemodinâmica. Eu iniciei meu contato com a cardiologia pediátrica pela hemodinâmica, quando acompanhava todos os exames e participava das reuniões de revisão dos cates e intervenções em congênita da semana. Assim, ao ingressar na especialização, continuei assídua e me aperfeiçoando em hemodinâmica na Cardiologia Pediátrica e Congênita, sob a coordenação do professor Dr. Antônio Carlos de Camargo Carvalho, Dra. Célia Maria Camelo da Silva no Setor de Hemodinâmica e Intervencionista em Pediatria e Congênita – no HSL-EPMUNIFESP, a partir de 2 de fevereiro de 2003.

O aprendizado em hemodinâmica sempre foi muito rico por ter como orientadora a Professora Dra. Célia Maria Camelo da Silva, que havia chegado de Fellowship, do Hospital for Sick Children (1989) e do National Royal Brompton Hospital (1990-1992), em Londres. Ela é Doutora em Hemodinâmica e foi pioneira em vários estudos e intervenções em cardiologia pediátrica, que se tornaram rotina na abordagem das cardiopatias congênitas tanto diagnósticas, terapêuticas quanto no seguimento clínico. Entre elas se destacaram a implantação da técnica e interpretação dos dados de todas as medidas de resistência vascular pulmonar para avaliação da hipertensão pulmonar pré-operatória nas cardiopatias congênitas de hiperfluxo pulmonar, valvoplastia pulmonar e aórtica com cateter balão, atriosseptostomia com cateter balão nas cardiopatias congênitas com comunicação interatrial restritiva, emprego da técnica rotacional no estudo hemodinâmico em crianças, ultrassom endovascular em cardiopatias congênitas, especialmente em hipertensão pulmonar, fechamento percutâneo de comunicação interatrial, fechamento percutâneo de comunicação interventricular, dilatação e implantação de stent endovascular nas estenoses vasculares, oclusão de fístulas em anomalias de

coronárias e pacientes com fisiologia de coração univentricular, tratamento de obstrução trombótica de dispositivos intravasculares como shunts sistêmico pulmonares nas atrésias pulmonares através da injeção de trombolíticos via cateterismo cardíaco e também estudos abordando proteção radiológica em hemodinâmica e intervenção, estudo das disfunções valvares pulmonares no pós-operatório de tetralogia de Fallot.

Participei de várias pesquisas e estudos e destaco principalmente a atrésia pulmonar com septo interventricular intacto: experiência inicial com a perfuração valvar por radiofrequência em neonatos e lactentes, também pioneira que foi tema da tese de doutorado da professora Dra. Célia Maria Camelo da Silva.

Assim fui descobrindo e ficando fascinada pela hemodinâmica e cirurgia cardiovascular. Continuo com a prática de hemodinâmica e cateterismo cardíaco e intervencionista em crianças e portadores de cardiopatias congênitas em Uberlândia, desde 2004, com o Dr. Vilmar Pereira, hemodinamicista também com prática em cardiologia pediátrica e congênita. Hoje já contamos com mais de 2.400 exames hemodinâmicos e cardiopatias congênitas e um terço deles com intervenção terapêutica. Tenho muito o que aprender e continuo aprendendo, pois não finalizei ainda este capítulo da minha vida.

4.3.7 Mestrado em Pediatria

Empenhada e apaixonada pela área de Cardiologia Pediátrica, no ano de 1992, prestei provas do processo seletivo de ingresso no programa de Mestrado em Pediatria da Escola Paulista de Medicina da UNIFESP, onde fui aprovada. Durante o mestrado, desenvolvi a pesquisa intitulada “Avaliação imunológica de crianças portadoras de Cardiopatias Congênitas”. A dissertação foi orientada pelo professor Dr. Antônio Carlos de Camargo Carvalho com a coorientação da professora Dra. Chloé Camba Musati. Esse estudo foi realizado na UNIFESP dentro das disciplinas Cardiologia e Cirurgia Cardiovascular, Imunologia e Alergia, Anatomia e Patologia Clínica, e contei com bolsa de mestrado concedida pela Coordenação de Aperfeiçoamento de Pessoal de Nível Superior/Ministério da Educação (CAPES/MEC).

Sou extremamente grata ao professor Dr. Antônio Carlos de Camargo Carvalho, pois sua humanidade, sabedoria, competência, firmeza, constância e

confiança depositadas em mim e seus valiosos conhecimentos transmitidos durante o mestrado contribuíram significativamente para o meu crescimento profissional. Agradeço à professora Dra. Chloé, pois seu exemplo, sua disponibilidade e sua força fizeram-me acreditar na seriedade da vida e da ciência, bem como seu estímulo, aprendizado e apoio decisivos foram essenciais na execução desse estudo.

Minha pesquisa de mestrado objetivou avaliar a resposta imunitária de 21 crianças com Cardiopatia Congênita (CC), sendo 10 acianogênicas, e 11 cianogênicas, de 1 a 140 meses. Tetralogia de Fallot e Comunicação Intraventricular foram as anomalias mais frequentes. Dezoito crianças eram desnutridas e seis tiveram infecções recorrentes. Em mais de 50% das crianças com CC, foram observados níveis séricos de imunoglobulinas, IgM e IgA, acima dos limites superiores da normalidade. Os níveis das frações C3 e C4 do sistema complementar mostraram-se adequados, considerando a faixa etária dos pacientes. Participaram da banca examinadora: Professora Dra. Magda Maria Sales Carneiro Sampaio, titular de imunologia do Hospital de Clínicas da Faculdade de Medicina da Universidade de São Paulo, Professor Dr. Dirceu Solé, titular de imunologia do HSP-EPM-UNIFESP, Professora Dra. Maria Suely Bezerra Diógenes.

O mestrado e a defesa foram concluídos no ano de 1995, e obtive o Título de Mestre em Pediatria. Desenvolver essa pesquisa de mestrado foi muito gratificante e reafirmou ainda mais o meu desejo de atuar dentro da área de Cardiologia Pediátrica e Congênita na Pediatria.

4.3.8 Doutorado em Ciências da Saúde

Empenhada no meu crescimento acadêmico e profissional, em fevereiro de 2006, iniciei as atividades na qualidade de aluna do Curso de Pós-Graduação em Cardiologia, nível doutorado, no Departamento de Medicina, disciplina Cardiologia, da UNIFESP, onde desenvolvi a tese de doutorado sob o tema “Ecocardiografia em contraste em cardiopatia congênita”.

Essa pesquisa de doutorado foi realizada com pacientes portadores de Cardiopatia Congênita (CC), cujo objetivo foi: analisar a exequibilidade e segurança da ecocardiografia com contraste em crianças e adolescentes com Cardiopatia Congênita. Participaram da pesquisa 87 crianças e adolescentes com CC e 30 controles normais submetidos a exame ecocardiográfico completo, seguido de

infusão contínua PESDA (Perfluorocarbon Exposed Sonicated Dextrose Albumin). A pesquisa demonstrou que a ecocardiografia com contraste foi exequível e segura em crianças e adolescentes com ou sem Cardiopatia Congênita. Essa pesquisa contou com análise e liberação da CONEP (Comissão Nacional de Ética em Pesquisa), que está diretamente ligada ao Conselho Nacional de Saúde (CNS) em Brasília, que é a instância máxima de avaliação ética em protocolos de pesquisa envolvendo seres humanos.

Para o desenvolvimento dessa pesquisa, tive a orientação do professor Dr. Antônio Carlos de Camargo Carvalho e coorientação da professora Dra. Célia Maria Camelo da Silva e do professor Dr. Valdir Ambrósio Moisés.

A tese foi concluída e defendida em 2012, sob o título “Exequilidade e segurança da ecocardiografia com contraste por microbolhas em crianças e adolescentes com cardiopatia congênita”, Recebi, assim, o título de Doutora em Cardiologia pela Cardiologia e Cirurgia Cardiovascular do HSP-EPM-UNIFESP. A partir dessa tese, vários trabalhos foram realizados, esta fez parte de capítulos de livros e tornou-se um dos trabalhos pioneiros em ecocardiografia com uso de contraste em pacientes portadores de cardiopatias congênitas.

Posteriormente, segui acompanhando a Cardiologia Pediátrica e Congênita e Hemodinâmica e Intervenções, a fim de manter minha capacitação e atualizada. Considero a Cardiologia Pediátrica e Congênita, a Ecocardiografia Pediátrica e Congênita e a Hemodinâmica como minha eterna escola de aprendizado e especialização.

Meu agradecimento especial à professora Dra. Célia Maria Camelo da Silva, pelo tempo disponibilizado em capacitar-se em ecocardiografia com contraste, para esse trabalho acontecer e por estar ao meu lado nesse estudo. Por fim, ao professor Dr. Valdir Ambrósio Moisés, por compartilhar conhecimento, sabedoria, compromisso e, sobretudo, pela arte de fechar o estudo com tanta clareza e fineza.

5 CARREIRA PROFISSIONAL

No início de 1987, fui admitida como professora substituta da especialidade de Pediatria, cargo no qual desenvolvi atividades acadêmicas com alunos do curso de Medicina e residentes da área de Pediatria. Ministrei aulas das disciplinas Semiologia Pediátrica, Puericultura e acompanhei os alunos na graduação com aulas de Hematologia Pediátrica, Semiologia Pediátrica, no internato e na residência médica em Pediatria, nas atividades práticas no Ambulatório de Pediatria Geral, no Pronto Socorro e na Enfermaria de Pediatria.

Motivada pela atuação como Professora Substituta e pela perspectiva da atuação profissional em ensino e assistência de qualidade, prestei concurso para Professora Efetiva e, em 14 de agosto de 1998, fui admitida para o cargo público de docente do curso de Medicina da Faculdade de Medicina da UFU, lotada no Departamento de Pediatria, onde estou até o momento. Desde 1998, ministro aulas para alunos do curso de graduação em Medicina, internato e no Programa de Residência Médica na área de Pediatria Geral, seguindo os programas de ensino determinados pelo Departamento de Pediatria.

Assumi atividades de preceptoria e assistência em todos os cenários da Pediatria: Pronto Socorro, Enfermaria de Pediatria, Neonatologia e Ambulatório de Pediatria e com escala de Plantão de Pediatria. Assim, eu e meu colega Dr. Fernando Jorge permanecemos na preceptoria e como chefe de plantão em todos os cenários da Pediatria. Os demais colegas de residência seguiram suas vidas fora da UFU. O período de preceptoria pelo Departamento de Pediatria como Professora de Pediatria durou de janeiro de 1987 a julho de 1987, quando assumi o contrato de Docente Substituta da Pediatria.

Como substituta, na graduação, fui professora em cenários teóricos e práticos de quase todas as disciplinas de Pediatria, na Residência de Pediatria Geral, nos cenários teóricos e práticos, e ambulatórios de Pediatria Geral, Puericultura. Também atuei como professora em urgências e emergências no Pronto Socorro; como professora diarista com a professora Dra. Valéria Bonett (Nefrologista Pediátrica), o professor Dr. José Martins Borges (Neurologista Pediátrico); como assistente integrante do módulo cardiopulmonar na Enfermaria de Pediatria, com a professora Dra. Maria José Junho Sologuren e o professor Dr. Hélio Lopes Silveira, e no ambulatório de cardiologia pediátrica, com o professor Dr. José Alfredo Cunha.

Posteriormente, com o despertar de meu interesse cada vez mais para os casos de cardiopatia congênita, sob a orientação do Professor Dr. José Alfredo, trabalhei com moléstias infecciosas (com professor Dr. Elísio de Castro), emergência e intensivismo (com professor Dr. Orlando Cesar Mantese), principalmente meningites e meningococcemia, choque e acidentes por animais peçonhentos.

A atividade docente, associada à atividade médica, tem sido muito gratificante, pois possibilita valorosos aprendizados, além de amizades estabelecidas, todas muito presentes em minha vida, algumas distanciadas geograficamente, mas mantendo “coração quente”, pois, de alguma forma, permanecem vivas em minha alma. Eu sigo, também, experiências primorosas com pacientes e familiares, alunos e residentes, equipe de enfermagem, equipes médicas e demais profissionais da área de saúde das duas universidades que frequento.

Dessa forma, minhas formações tanto como docente quanto como pesquisadora são permeadas por saberes que foram construídos ao longo da trajetória de vida de educadora e se mantêm até a atualidade. Na graduação, fui professora em cenários teóricos e práticos em quase todas as disciplinas de Pediatria, na Residência de Pediatria Geral e em diversas áreas de atuação e especialização de Pediatria, como UTI Pediátrica, Neonatologia e Cardiologia Pediátrica e Congênita, Ecocardiografia e Hemodinâmica.

Nesse período, além das atividades de ensino, pesquisa e assistência, sempre mantive atividades médicas desenvolvidas na UTI Pediátrica sob a coordenação do Professor Dr. Orlando César Mantese. Concomitantemente, com o passar do tempo, fui constatando a necessidade de ampliar mais o tratamento da criança com cardiopatia congênita no Hospital de Clínicas na Universidade Federal de Uberlândia, o que coincidiu com a chegada da especialidade de Cirurgia Cardiovascular em adulto (Professor Dr. Hélio Fabry e Professor Dr. Paulo Cesar Santos). Na criança, acompanhei poucos casos de baixa complexidade, como comunicação interatrial, comunicação interventricular e persistência do canal arterial inicialmente itinerante pelo cirurgião professor Dr. João José Carneiro, Professor Titular de Cirurgia Cardiovascular da Faculdade de Medicina de Ribeirão Preto da Universidade de Ribeirão Preto. Posteriormente, foi contratado um cirurgião cardiovascular, Dr. Miguel Arboleda, que ficou no ano de 2004, no Hospital de Clínicas da Faculdade de Medicina da Universidade Federal de Uberlândia. Nesse

período, foram feitas mais de três cirurgias de fechamento de canal arterial, mas, por motivos profissionais e pessoais, ele se mudou para Lima, no Peru.

Como pacientes não paravam de chegar, o Serviço de Cardiologia Pediátrica foi estruturando-se com riqueza de pacientes e diversificados tipos de cardiopatias congênitas. Assim, o início das atividades em Cardiologia Pediátrica e Congênita foi determinado em razão da necessidade de melhorar a assistência médica e multiprofissional das crianças que eram assistidas. Criado esse novo espaço, foi estabelecida a filosofia das atividades da nova unidade no tripé de ensino, assistência e pesquisa.

Dessa forma, em 2005, houve a chegada do novo cirurgião, Dr. Claudio Ribeiro da Cunha, ex-aluno da Faculdade de Medicina da Universidade Federal de Uberlândia, o qual se especializou em Cirurgia Cardiovascular pelo Instituto do Coração da Universidade de São Paulo (INCOR-USP), com formação com estágio em cirurgia cardiovascular com o professor Dr. Miguel Lorenzo Barbero Marcial, reconhecido como um dos melhores cirurgiões em cirurgia cardíaca pediátrica e congênita. Isso possibilitou que, em 2005, déssemos continuidade às cirurgias cardíacas, uma vez por semana, atendendo às demandas tanto das crianças internadas na emergência, neonatos da Unidade de Neonatologia, quanto eletivas provenientes do ambulatório de cardiologia pediátrica. Nossos resultados, desde o início, foram considerados excelentes. Assim, a estrutura foi se qualificando e sendo organizada em cuidados cardiológicos eletivos, com resolução boa, em que as internações por cardiopatia congênita reduziram-se muito. Paralelamente, os neonatos com cardiopatias graves foram atendidos na Unidade Neonatal com bom controle pré-operatório e sendo operados com excelente resultado.

Com o passar do tempo, foram-se capacitando cada vez mais as Unidades de Terapia Intensiva Pediátrica do HC-UFU no cuidado pré-operatório das crianças graves com cardiopatias não operadas e das crianças cardiopatas operadas e na Terapia Intensiva Neonatal nas cardiopatias de apresentação no período neonatal canal dependentes ou não. O crescente movimento de cardiopatia congênita neonatal e pediátrica e das equipes médicas, de enfermagem (representadas pela enfermeira Maria do Carmo) e de multiprofissionais (Fisioterapia, Nutrição, Odontologia, Assistente Social, Escriturários, Manutenção, Humanização) qualificou-se e organizou-se continuamente, transformando–se em um Centro reconhecido pelo Ministério Público da Saúde (MS) e credenciado tanto pelo Ministério da

Educação e Cultura (MEC) quanto pela Sociedade Brasileira de Cardiologia, Sociedades de Cardiologia Pediátrica (SBCP), Sociedade Brasileira de Pediatria (SBP), Sociedade Brasileira de Cardiologia (SBC) e Sociedade Brasileira de Cirurgia Cardiovascular (SBCC), comparando-se aos melhores centros terapêuticos em cardiologia pediátrica do Brasil e de outros países.

Simultaneamente, os outros dois braços da Cardiologia Pediátrica e Congênita desenvolveram-se em toda sua potência, sendo a primeira a Ecocardiografia com mais de 10.000 exames ecocardiográficos em cardiopatia congênita e pediatria até o momento. Já na Hemodinâmica e Intervenções Terapêuticas, o Dr. Vilmar José Pereira, hemodinamicista especializado pela Escola Paulista de Medicina – que tem o programa de hemodinâmica em Cardiologia Pediátrica e Congênita –, e eu, que obtive formação em Cardiologia Pediátrica, Congênita e Hemodinâmica, executamos vários procedimentos, como cateterismos diagnósticos e terapêuticos, como atriosseptostomia com cateter balão, valvoplastia pulmonar e aórtica com cateter balão, fechamento de comunicação interatrial e, por último, interventricular e implantação de endopróteses no tratamento de estenoses vasculares. Hoje, contamos com cerca de próximos de 2.400 estudos hemodinâmicos realizados desde 2004 em pacientes pediátricos portadores de cardiopatia congênita e adquirida. Reitero, ainda, que, em todos os segmentos da Cardiologia Congênita, abrangemos, também, os adultos portadores de cardiopatias congênitas.

Em 2006, iniciamos o curso de Especialização em Cardiologia Pediátrica com o programa de Educação Médica Continuada em Cardiologia Pediátrica (Dra. Bethânia Diniz Ramos e Dra. Fernanda Christiane de M. M. Cisdeli) com duração de dois anos. Após um ano, conseguimos implantar a Residência Médica em Pediatria com atuação em Cardiologia Pediátrica. Após um ano, em 2008, iniciamos o curso de Educação Médica Continuada e Especialização em Ecocardiografia Congênita e Pediátrica em conjunto com a Ecocardiografia em adultos, reconhecida pela FAMED (diretor professor Dr. Ben Hur Braga Taliberti). Nessa época, também iniciamos o programa para residentes de Cardiologia Geral e Ecocardiografia em Pediatria e Cardiologia Congênita e Hemodinâmica e Intervenções tanto em crianças como em adultos portadores de cardiopatia congênita. Além disso, temos o Programa de acompanhamento para o Residente especializando em UTI Pediátrica e Congênita no 1º e 2º ano de Residência. Após um ano, foi iniciada a Residência médica em

Ecocardiografia de adulto. Foi mantida a especialização em Cardiologia Pediátrica e Congênita e iniciamos a especialização em Ecocardiografia Pediátrica, Congênita e Fetal, com duração de dois anos cada uma, seguindo as recomendações da Sociedade Brasileira de Cardiologia, da Sociedade Brasileira de Cardiologia Pediátrica e da Sociedade Brasileira de Ecocardiografia, respectivamente. O Programa de Especialização em Ecocardiografia por Educação Médica continuada mantém-se até a atualidade, no qual o residente realiza o segundo ano de especialização.

Desde 2007, foram formados dez especialistas em Cardiologia Pediátrica e Congênita e dez ecocardiografistas. Nesse período, tivemos mais dois alunos que fizeram apenas um ano de Cardiologia Pediátrica e Congênita por fatores pessoais. Paralelamente, fomos desenvolvendo linhas de pesquisas em insuficiência cardíaca na criança, pré e pós-operatório em cardiopatias congênitas cianóticas e acianóticas, miocardiopatia dilatada, qualidade de vida em pacientes com cardiopatias congênitas (coordenadas pelo professor Dr. Carlos Henrique Martins da Silva), cardiopatias congênitas de apresentação no período neonatal, tanto canal dependentes como canal independentes, ecocardiografia fetal, ecocardiografia em cardiopatias congênitas, ecocardiografia em pacientes com cardiopatias adquiridas, perfil de apresentação epidemiológico e clínico das cardiopatias congênitas em nosso meio, hemodinâmica e intervenções em Cardiologia Pediátrica e Congênita.

Vários estudos têm sido realizados, incluindo o Programa de Residência Multiprofissional de outras áreas, como Odontologia, Enfermagem, Nutrição, iniciação científica, mestrado e doutorado. Hoje, tenho consciência de que esse é um campo amplo e inesgotável para pesquisas e formação de pessoal. Isso fica muito bem documentado com a tese de doutorado realizada entre 2008 e 2012 na Cardiologia Pediátrica e Congênita pelo cirurgião cardiovascular do Distrito Federal e do Hospital de Clínicas da Faculdade de Medicina e Fundação da Assistência Estudo e Pesquisa de Uberlândia da Universidade Federal de Uberlândia, do Dr. Cláudio Ribeiro da Cunha. O estudo foi do “Perfil das citocinas e correlação com a morbidade no período pós-operatório em crianças com diagnóstico de cardiopatias congênitas não cianosantes submetidas à cirurgia corretiva com circulação extracorpórea”, orientada pelo professor Dr. José Roberto Mineo.

No Hospital de Clínicas da Universidade Federal de Uberlândia, por ter uma área de abrangência de cerca de 4 milhões de pessoas carentes de atendimento em

cardiopatias congênitas e cardiologia pediátrica, fomos pioneiros e desenvolvemos o atendimento para pacientes portadores de cardiopatias congênitas, seguimento clínico ambulatorial em cardiologia pediátrica e congênita, ecocardiografia em pacientes com cardiopatia congênita, eletrocardiografia fetal, arritmia cardíaca, métodos gráficos (Dra. Denise Auxiliadora Leite Lasbeck), estudo eletrofisiológico em congênita (iniciado pelo professor Dr. Elias Esber Kanaane e pelo Dr. Petrônio Rangel Salvador Júnior), programa de saúde bucal das crianças e seus familiares com atendimentos de triagem na sala de espera e orientação dos pacientes e pais de como cuidar da boca e dos dentes e sua importância para a saúde (representado pela professora Suzana Ferreira de Paula Silva)

Fomos, também, pioneiros no acolhimento dos pacientes cardiopatas, que hoje se estende a outras áreas. Criamos o programa de humanização no acolhimento das crianças portadoras de cardiopatias congênita e adquirida, que hoje se estende aos outros ambulatórios de pediatria (coordenadora enfermeira Leda Márcia Viana Santos Borges), Projeto Amigos do Coração, em que o ambulatório é a base do desenvolvimento de residentes multiprofissionais com os programas de Educação Multiprofissional em Sala de Espera e Posso Ajudar, Casa de apoio para as mães com crianças portadoras de cardiopatia congênita (ONG coordenada pela enfermeira Juliene Cristine de Oliveira e colaboradores), e a sala do cardiopata, no ambulatório de cardiopatia congênita (que funciona de segunda a sexta-feira com atendimento das 7h às 18h), ao qual os pacientes têm livre acesso para qualquer necessidade relacionada ao tratamento, e atendimento do Paciente Cardiopata (criado e incentivado pelo diretor clínico Cezar Augusto dos Santos), coordenado atualmente pela assistente social Nilma Aparecida Assunção e iniciado com as secretárias Ana, Elena Yara e Vera Lucia.

A equipe de médicos que atuam na Cardiologia Pediátrica e Congênita, em Ecocardiografia Congênita e Pediátrica, Ecocardiografia Fetal, formada na Faculdade de Medicina – Hospital de Clínicas – Universidade Federal de Uberlândia desde 2006, está atuante. Foi composta por médicos procedentes das principais universidades do país, trazendo em seus currículos relevante contribuição à Cardiologia Pediátrica e Congênita, Ecocardiografia em Cardiologia Pediátrica e Congênita e Ecocardiografia Fetal.

Essa é uma parte de minha contribuição para o ensino, a assistência e a gestão acadêmica, como responsável pelas atividades acadêmicas relacionadas à

cardiologia pediátrica na função de coordenadora da Residência de Cardiologia Pediátrica e Ecocardiografia em Cardiologia Pediátrica e Fetal nos últimos 18 anos (desde 2005).

Participei, em 2013 e 2014, com os secretários da saúde de Uberlândia, Exmo. Dr. Almir Fernando Loreiro Fontes e Exma. Dra. Raquel Cazabona, Coordenadora do Programa da Saúde da Criança e do Adolescente da Secretaria Municipal de Saúde de Uberlândia, da elaboração de um Protocolo de Regulação e Priorização de Encaminhamentos de Atenção Básica para ambulatórios de especialidades pediátricas de Uberlândia, relativo às principais cardiopatias e doenças cardíacas em crianças, adolescentes e adultos com cardiopatias congênitas. Durante esse processo, participei de reuniões com os gestores municipais e regionais da área de saúde responsáveis pelo sistema de referência e contrarreferência. Ao final, foi realizada uma proposta de referenciamento de crianças e adolescentes com sopro cardíaco, cansaço aos esforços, arritmias, dor precordial, cianose, lipotimias, síncopes, irmãos com cardiopatias congênitas, vasculites, Kawasaki, neonatos com qualquer sintoma, malformações congênitas familiares. Isso ocorreu de forma compactuada com minha participação como representante no programa de assistência ao cardiopata congênito e pediátrico, adolescente e adulto cardiopata congênito e participação na educação médica continuada em cardiologia pediatria e congênita da Secretaria de Saúde da Prefeitura Municipal de Uberlândia. Ficou, também, estabelecido o HC-FAMED-UFU como Centro de Referência principal da região em atendimento, propedêutica e tratamento clínico e cirúrgico de cardiopatia congênita, cardiologia pediátrica, bem como hemodinâmica e intervenções.

Participei como representante do HC-FAMED-UFU, em 2012 e 2013, do Plano Regional, e Estadual posteriormente (a partir de 2017), que levou, com outras regiões do país, à criação e, após, à ligação com o Plano Nacional de Assistência à Criança com Cardiopatia Congênita (PNACCC) (BRASIL, 2017; CANEO *et al.*, 2012; PINTO JÚNIOR *et al.*, 2004; PINTO JÚNIOR *et al.*, 2013), credenciado pelo SUS em 2013, com o trabalho intitulado “Diagnósticos dos Serviços de Saúde em Cardiologia Pediátrica no Estado de Minas Gerais”. Foram reuniões mensais realizadas às segundas-feiras, em Belo Horizonte, com a Secretaria do Estado de Minas Gerais. Nessas reuniões, ficou determinado que todos os centros que trabalham com cardiopatia congênita têm o dever de aceitar a criança com cardiopatia congênita,

independentemente da regionalização no estado de Minas. Dessa forma, na época, ficamos como Centro de Referência da metade do estado de Minas Gerais, e Belo Horizonte com o restante do estado, além de esta cobrir os tratamentos de cardiopatias congênitas que Uberlândia não conseguisse suprir. Assim, ficamos com esse compromisso em 2013 e também de auxiliar outros centros a se capacitarem. Posteriormente, em Minas Gerais, Passos e Uberaba também iniciaram o Serviço de Cardiologia Pediátrica. Foi admitido que, para fins do PNACCC, a incidência de Cardiopatias Congênitas varia entre 0,8% nos países com alta renda e 1,2% nos países com baixa renda, em que o valor médio de 1% de prevalência é habitualmente aceito para o Brasil e para os demais países da América Latina. Dessa forma, visto que o Brasil registra anualmente 2,8 milhões de nascidos vivos, pode-se estimar o diagnóstico de quase 29 mil novos casos de cardiopatias.

Dessa forma, todos os Serviços de Cirurgia Cardíaca são visitados por essas equipes. A primeira ocorreu em 2017, em que a diretoria do hospital participou da visita com equipe da Cardiologia Pediátrica e Equipe de Cirurgia Cardiovascular. A nossa última visita foi em 2020, imediatamente antes da pandemia, em janeiro.

Atualmente, temos os seguintes perfis no atendimento de Cardiologia Pediátrica e Congênita: em todos os seguimentos, fazemos orientação e ensino para alunos das diversos níveis acadêmicos e de formação (liga de Pediatria, liga de Cardiologia, curso básico de Histologia e Embriologia para familiarização com as doenças congênitas, residência médica em Pediatria, residência médica em Cardiologia Geral, residência médica em Ecocardiografia Geral e especialização em Ecocardiografia, em Cardiologia Pediátrica e Congênita, Medicina Fetal, residência em Cardiologia Pediátrica e Congênita, residência multi em Nutrição, Fisioterapia, Enfermagem; cursos opcionais de várias categorias, como Enfermagem, Medicina, Serviço Social, Recursos Humanos, Psicologia, Humanização, Terapia Intensiva e voluntários no programa de sala de espera do ambulatório.

Chamam a atenção a quantidade de atendimentos efetivados mensalmente:

- Consultas de Cardiologia Pediátrica: 200;
- Exames ecocardiográficos: 400;
- Exames de métodos gráficos e eletrocardiograma: 200;
- Hemodinâmica e intervenções: 12;

- Angiotomografia: 12;
- Ressonância magnética: 4;
- Cirurgias cardiovasculares: 8 a 10.

Desde o início, em 2005, até o momento, contamos com 1.000 cirurgias cardiovasculares e 1.400 estudos hemodinâmicos e cerca de 40% de intervenções.

Por fim, até o momento, conto com 11 alunos especialistas em Cardiologia Pediátrica e Ecocardiografia Pediátrica e Congênita e todos exercem as atividades profissionais. Destaco o Dr. Geórgio, que se especializou e retornou para o Maranhão exercendo Cardiologia Pediátrica e Congênita e Fetal e é referência local para Cardiologia Pediátrica.

Segue a lista dos Médicos Especializados em Cardiologia Pediátrica em ordem crescente de formação de 2007 a 2022:

- Dra. Bethânia Diniz Ramos;
- Dra. Fernanda Cisdelil;
- Dra. Neide Aparecida Faria;
- Dra. Camila Renault Quaresemin;
- Dr. João Ribeiro de Matos Neto;
- Dr. Rodrigo Massini de Melo;
- Dra. Fabiana Lemos de Campos;
- Dra. Viviane Athadeu Gontjo;
- Dra. Cintia Rejane Soares Dupin;
- Dr. Cristiano da Silva Neves;
- Dr. Geórgio Moraes Costa.

5.1 Atividade didática

Sou professora do Curso de Graduação da Faculdade de Medicina da Universidade Federal de Uberlândia, em regime de 40 horas semanais, admitida por concurso público em agosto de 1998.

Desde a minha admissão, ministrei aulas teóricas e práticas na graduação de Medicina, nas seguintes disciplinas:

- Clínica Médica II – com aulas de temas de Cardiologia Pediátrica e Congênita;

- Estágio Supervisionado em Pediatria;
- Semiologia Pediátrica;
- Pediatria Geral;
- Medicina Integrada III;
- FAMED31702 – Saúde Individual VII;
- FAMED901 – Estágio Supervisionado na Área Materno-infantil;
- FAMED31702 – Saúde Individual VII;

Na atividade docente preceptória da Residência Médica, desempenho atividades de:

- Preceptoria da residência Médica em Pediatria Geral com aulas teóricas e práticas em pronto socorro, enfermaria e UTI Pediátrica e UTI Nneonatal;
- Supervisora do Programa de Residência Médica em Clínica e Cardiologia Pediátrica R3 e R4;
- Coordenadora do Programa de Residência Médica em Cardiologia Pediátrica;
- Coordenadora do Programa de Especialização em Ecocardiografia em Pediatria, Congênita e Fetal;
- Coordenadora da Disciplina Semiologia Pediátrica do Curso de Medicina.

5.2 Atividade assistencial

Sou médica no Departamento de Pediatria da UFU desde 1987, tendo atuado nos diversos setores da Pediatria do HCU (Enfermaria, Ambulatório, Pronto Socorro, Unidade de Terapia Intensiva Pediátrica, Neonatologia, Avaliação Cardiológica Clínica, Ecocardiografia, Cardiologia Pediátrica e Hemodinâmica).

Segue abaixo um resumo das principais atividades profissionais assistenciais:

- Ambulatório de pediatria;
- Ambulatório de ecocardiografia pediátrica, congênita e fetal;
- Setor de cardiologia pediátrica, congênita e fetal;
- Enfermaria de pediatria;
- UTI pediátrica;
- Unidade neonatal e UTI neonatal;

- Membro do Comitê Transfusional do Hospital de Clínicas de Uberlândia;
- Chefe do Serviço de Cardiologia Pediátrica do Hospital de Clínicas/UFU, desde agosto de 2014;
- Coordenadora do Programa de Especialização Médica na forma de Educação Médica Continuada em Cardiologia Pediátrica, desde 2006; além do Programa de Educação Médica Continuada igual ao Programa de Residência Médica em Cardiologia Pediátrica e Congênita;
- Programa de Residência em Cardiologia Pediátrica e Congênita desde 2007, no qual disponibilizamos uma vaga, totalizando a formação de 10 alunos, entre eles, oito com tempo integral completados e dois com tempo de um ano;
- Coordenadora do Programa de Educação Médica Continuada em Ecocardiografia Pediátrica, Congênita e Fetal desde 2007;
- Tenho certificação de aprovação dos cursos de reanimação em Suporte Básico de Vida (BLS), reanimação cardiopulmonar em neonatologia pela SBP desde 2004, com revalidação periódica, a última em 2020, antes da pandemia de covid-19. Desde 2016, sou instrutora do PALS pela Sociedade Mineira de Pediatria e reconhecida pelo American Heart Association.

6 PRODUÇÃO CIENTÍFICA

Descrevo a seguir, de modo objetivo e didático, os indicadores bibliométricos de minha produtividade científica, o resumo de minha dissertação e tese de pós-graduação, minha participação efetiva em grupos de pesquisa institucionais, as pesquisas desenvolvidas, os trabalhos publicados, a orientação em programa de pós-graduação, a participação em bancas acadêmicas e a participação em eventos científicos, o que caracteriza minha linha científica de trabalho.

6.1 Indicadores bibliométricos do Sistema de Bibliotecas UFU

Realizou-se revisão dos indicadores bibliométricos pelo Sistema de Bibliotecas da Universidade Federal de Uberlândia - UFU, com auxilio dos bibliotecários do Setor de Biblioteca - UFU e conforme o Sistema de Bibliotecas da Universidade Federal de Uberlândia (2023), os indicadores bibliométricos de produtividade científica disponíveis são:

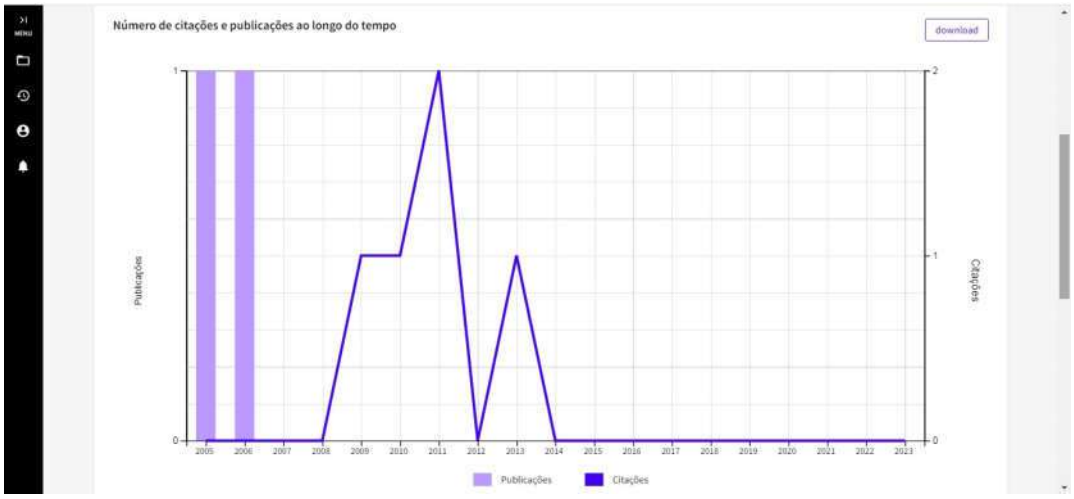
a) H-index: desenvolvido em 2005, como uma ferramenta para combinar quantidade e qualidade da produção acadêmica, sendo definido como o maior número “h” de artigos de um determinado pesquisador que tem, pelo menos, o mesmo número “h” de citações cada um. O índice h pode ser obtido por meio de consulta à base de dados *Web of Science* ou *Scopus* disponíveis no Portal Capes, pesquisando-se pelo nome do autor de quem que se quer conhecer o índice e determinado período em que se quer avaliar.
b) Fator de Impacto: é a média de citações dos artigos de um determinado periódico, calculada a partir do número de citações de artigos desse título em determinado ano, publicados no biênio anterior, dividido pelo número total de artigos publicados nele mesmo, nesse biênio. A base de dados *Journal Citation Report* (JCR), disponível no Portal Capes, fornece o Fator de Impacto dos periódicos indexados pela base de dados *Web of Science*, também disponível no Portal Capes.
c) Qualis (CAPES – Coordenação de Aperfeiçoamento de Pessoal de Nível Superior): trata-se de um aplicativo que permite a classificação e a consulta ao Qualis (classificação dos veículos de divulgação da produção científica,

por área do conhecimento pela CAPES). A classificação está dividida em oito estratos:

Figura 1 — Relatório de Citações

Fonte: Web of Science (2023)

Figura 2 — Relatório de Citações



Fonte: Web of Science (2023)

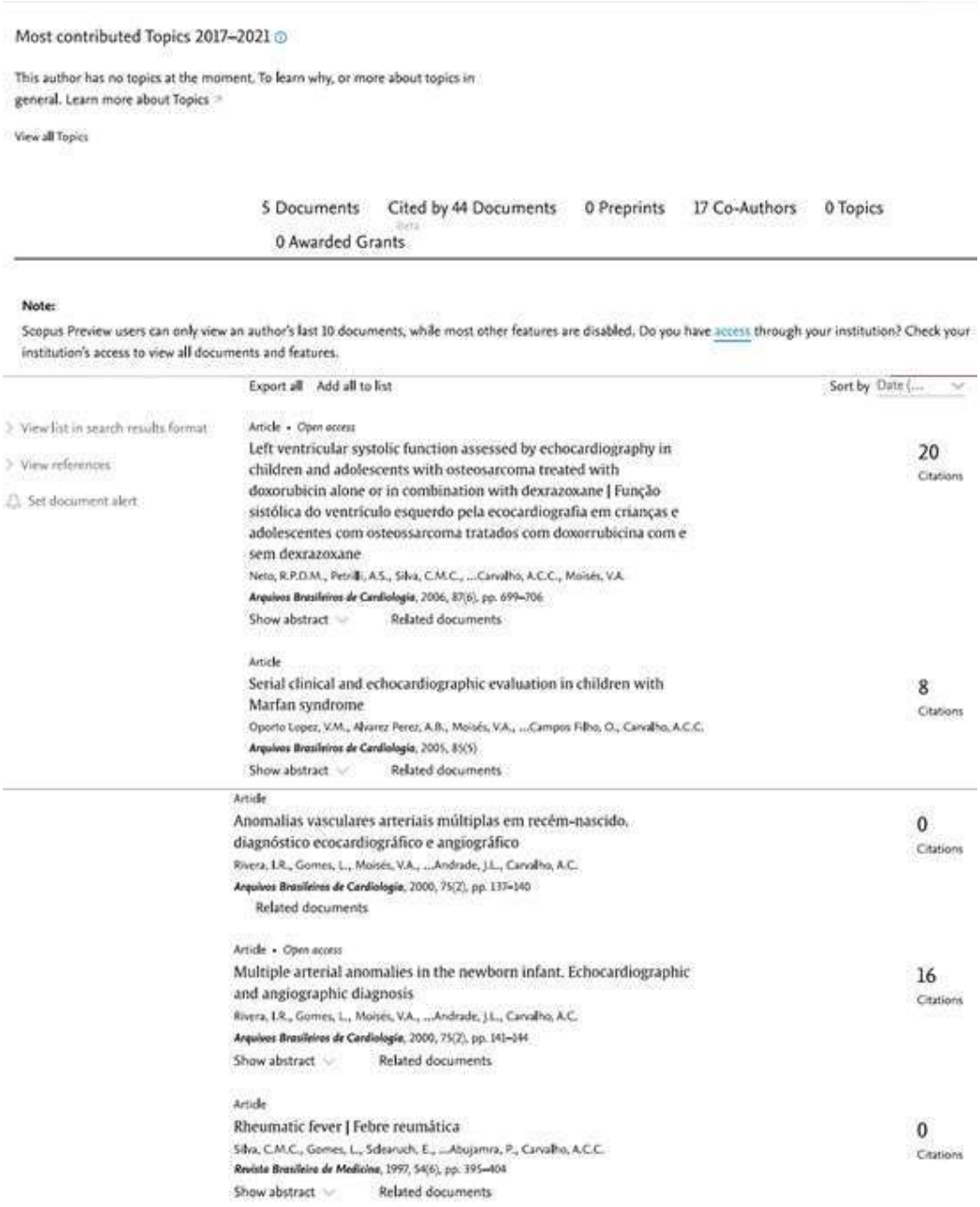
Figura 3 — Citações de Documentos

Fonte: Scopus (2023).

Figura 4 — Gráficos – Tendência de Documentos e Citações

Fonte: Scopus (2023).

Figura 5 — Número de Citações



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Left ventricular systolic function assessed by echocardiography in children and adolescents with osteosarcoma treated with doxorubicin alone or in combination with dexrazoxane | Função sistólica do ventrículo esquerdo pela ecocardiografia em crianças e adolescentes com osteossarcoma tratados com doxorrubicina com e sem dexrazoxane

20 Citations

Neto, R.P.D.M., Petrilli, A.S., Silva, C.M.C., ...Carvalho, A.C.C., Moisés, V.A.

Arquivos Brasileiros de Cardiologia, 2006, 87(6), pp. 699–706

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Serial clinical and echocardiographic evaluation in children with Marfan syndrome

8 Citations

Oporto Lopez, V.M., Alvarez Perez, A.B., Moisés, V.A., ...Campos Filho, O., Carvalho, A.C.C.

Arquivos Brasileiros de Cardiologia, 2005, 85(5)

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0 Citations

Rivera, I.R., Gomes, L., Moisés, V.A., ...Andrade, J.L., Carvalho, A.C.

Arquivos Brasileiros de Cardiologia, 2000, 75(2), pp. 137–140

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Multiple arterial anomalies in the newborn infant. Echocardiographic and angiographic diagnosis

16 Citations

Rivera, I.R., Gomes, L., Moisés, V.A., ...Andrade, J.L., Carvalho, A.C.

Arquivos Brasileiros de Cardiologia, 2000, 75(2), pp. 141–144

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Article

Rheumatic fever | Febre reumática

0 Citations

Silva, C.M.C., Gomes, L., Sdearuch, E., ...Abujamra, P., Carvalho, A.C.C.

Revista Brasileira de Medicina, 1997, 54(6), pp. 395–404

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Fonte: Scopus (20230

6.2. Teses e Dissertações

6.2.1 Mestrado

Título: *Avaliação Imunológica de Crianças com Cardiopatia Congênita*

Dissertação de Mestrado apresentada ao Programa de Mestrado em Pediatria da Escola Paulista de Medicina da Universidade de São Paulo, 1992-1995.

Orientador: Professor. Dr. Antônio Carlos de Camargo Carvalho.

Bolsista: CAPES

Resumo: A autora acompanhou prospectivamente 21 crianças com Cardiopatia Congênita (CC), sendo 10 acianogênicas e 11 cianogênicas, de um a 140 meses. Tetralogia de Fallot e Comunicação Intraventricular foram as anomalias mais frequentes. Dezoito crianças eram desnutridas e seis tiveram infecções recorrentes. Em mais de 50% das crianças com CC, foram observados níveis séricos de imunoglobulinas, IgM e IgA, acima dos limites superiores da normalidade. Os níveis das frações C3 e C4 do sistema complementar mostraram-se adequados, considerando a faixa etária dos pacientes.

6.2.2 Doutorado

Título: *Exequibilidade e segurança da ecocardiografia com contraste por microbolhas em crianças e adolescentes com cardiopatia congênita*.

Tese de Doutorado apresentada ao Programa de doutorado em Pediatria da Escola Paulista de Medicina da Universidade de São Paulo, 1996 a 2012.

Orientador: Professor Dr. Antônio Carlos de Camargo Carvalho.

Coorientação: Professora Dra. Célia Maria Camelo da Silva (UNIFESP), Professor Dr. Valdir Ambrósio Moisés (UNIFESP) e Professor Dr. Wilson Mathias Júnior (INCOR).

Banca examinadora: Professor Dr. José Lázaro de Andrade (INCOR), Dra. Samira Saad Morthy (INCOR), Dra. Solange Bernardes Tatani (UNIFESP), Professora Dra. Maria Suely Bezerra Morthy (UNIFESP).

Resumo: Participaram da pesquisa 87 crianças e adolescentes com CC e 30 controles normais submetidos a exame ecocardiográfico completo, seguido de infusão contínua PESDA (*Perfluorocarbon Exposed Sonicated Dextrose Albumin*). A

pesquisa demonstrou que a ecocardiografia com contraste foi exequível e segura em crianças e adolescentes com ou sem Cardiopatia Congênita.

6.3 Projetos de Pesquisa e/ou Extensão

Título: Ecocardiograma portátil e uso ambulatorial

Descrição: O desenvolvimento de equipamento portátil de ecocardiografia com técnica com definição suficiente para uma avaliação completa, à beira do leito, de forma rápida, permite a exploração de novos horizontes na Cardiologia. Assim como hoje o eletrocardiograma é método de rotina utilizado em todos os consultórios cardiológicos, a exploração do efetivo resultado obtido com o ecocardiograma portátil em comparação com os aparelhos maiores mais sofisticados e as vantagens eventuais de seu uso rotineiro no ambulatório em suspeita de disfunção ventricular, pericárdica e na avaliação de diversos tipos de lesões presumidamente orovalvares vai permitir o cotejo da situação entre benefício potencial futuro e real efetividade presente.

Período de Execução: 2000-2004.

Integrantes: Lourdes de Fátima Gonçalves Gomes (Responsável); Orlando Campos Filho; Lilian Paula de Souza; Valdir Ambrósio Moisés; Valeska Tavares da Silva R. R. Scavonda do Carmo; Manuel Adan Gil.

Título: Cardiopatias e risco gestacional: implicações para o concepto e a mãe

Descrição: PROGRAMA: 33009015007P-9 MEDICINA (CARDIOLOGIA) – UNIFESP.

Período de Execução: 1999-2004.

Integrantes: Lourdes de Fátima Gonçalves Gomes (Responsável); Orlando Campos Filho; Daniel Born; Victor Manuel Oporto Lopez.

Título: Eco Transesofágico

Descrição: O desenvolvimento da ecocardiografia transesofágica provocou grande melhora na avaliação de áreas “cegas” para ecocardiograma transtorácico. Especialmente para lesões de aorta ou lesões valvares com processos infecciosos e nas avaliações intraoperatórias imediatamente após procedimentos cirúrgicos, o ecocardiograma transesofágico passou a ter importância fundamental. Recentemente, também em diversas situações de intervenção na hemodinâmica nos

setores de eletrofisiologia, intervenção percutânea em coronária, terapêutica de aneurismas de aorta ou de Cardiopatia Congênita, o ecotransesofágico passou a ocupar lugar de realce na investigação de lesões residuais ou na indicação de terapêutica intervencionista. São estudados os diversos meios de utilização desta técnica para definição de sua eficiência e acurácia.

Período de Execução: 1994-2000.

Integrantes: Lourdes de Fátima Gonçalves Gomes (Responsável); Orlando Campos Filho; Lilian Paula de Souza; Valdir Ambrósio Moisés; Valeska Tavares da Silva R. R. Scavonda do Carmo.

6.4 Artigos publicados em periódicos

Apresenta-se abaixo as publicações de artigos em periódicos nacionais e internacionais, com as seguintes informações: descrição dos artigos publicados; número de citações; fator de impacto; índices de citação; classificação qualis/CAPES; link de acesso e demais informações pertinentes.

1. MENDONCA, G. S.; CARDOSO, L. M.; PEREIRA, P. G.; LUCIO, M. D.; GOMES, L. F. G.; GUEDES JUNIOR, C. A.; MENDES-RODRIGUES, C.; GIULIANI, C. D. Competências de enfermagem em internações psiquiátricas: recorte temporal da reforma aos dias atuais. International Journal of Current Research, v. 13, p.17.360-17.366, 2021. DOI:10.24941/ijcr.41340.05.2021.]

- Classificação Qualis/CAPES Sucupira (Medicina 1): C
- ISI Impact Factor 2019-2020: 1.532
- SJIF Scientific Impact Factor: 8.132
- Index Copernicus Value (ICV): 72.25
- Disponível em <https://www.journalcra.com/>.

2. COSTA, G. M.; NEVES, C. S.; GOMES, L. F. G.; MATTOS NETO, J. R.; PEREIRA, V. J.; MENDONCA, G. S. Clinical Case-Intracardiac Strange Body Approach in Prematures and Children – Case Report. International Journal of Development Research, v. 10, p. 40.124-40.127, 2020. DOI: 10.37118/ijdr.19832.09.2020.

- Classificação Qualis/CAPES Sucupira (Medicina 1): C
- SJIF Scientific Journal Impact Factor (2023): 8.058
- ORCID: https://orcid.org/0000-0002-9899-807X
- Disponível em: <https://www.journalijdr.com/>.

3. TURATTI, M. F.; MENDONÇA, G. S.; SILVA, C. R.; LÚCIO, M. D.; GOMES, L. F. G.; MARQUES, K. L. S.; OLIVEIRA, F. S. Anthropometric evaluation methods for patients with cerebral palsy. Brazilian Journal of Development, v. 6, p.100864-100880, 2020. DOI:10.34117/bjdv6n12-541.

- Classificação Qualis/CAPES Sucupira (Medicina 1): C
- BJD surpasses 6,000 citations on Google
- H index (2022): 21
- i10 index (2022): 136
- Disponível em <https://ojs.brazilianjournals.com.br/ojs/index.php/BRJD>.

4. MATOS NETO, R. P.; PETRILLI, A. S.; SILVA, C. M. C.; CAMPOS FILHO, O.; OPORTO, V. M.; GOMES, L. F. G.; PAIVA, M. G.; CARVALHO, A. C. C.; MOISÉS, V. A. Função sistólica do ventrículo esquerdo pela ecocardiografia em crianças e adolescentes com osteossarcoma tratados com doxorrubicina com e sem dexrazoxane. Arquivos Brasileiros de Cardiologia, v. 87, p. 763-771, 2006. DOI:10.1590/s0066-782x2006001900013.

- Journal Impact Factor: 2.667
- Journal Citation Indicator (JCI): 0.46
- Histórico do Journal Impact Factor Percentil (JIF): 34,62
- Histórico do Journal Citation Indicator (JCI): 46,88
- Classificação Qualis/CAPES Sucupira (Medicina 1): B2
- Artigo Citações Scopus: 20
- Disponível em: <https://jcr-clarivate.ez34.periodicos.capes.gov.br/jcr-jp/journal-profile?journal=ARQ%20BRAS%20CARDIOL&year=2021&fromPage=%2Fjcr%2Fhome>.

5. LOPEZ, V. M. O.; PEREZ, A. B. A.; MOISES, V. A.; GOMES, L. F. G.; PEDREIRA, P. S.; SILVA, C. C.; CAMPOS FILHO, O.; CARVALHO, A. C. C. Serial Clinical and Echocardiographic Evaluation in Children with Marfan Syndrome. Arquivos Brasileiros de Cardiologia, v. 85, p. 1, 2005.

- Journal Impact Factor: 2.667
- Journal Citation Indicator (JCI): 0.46
- Histórico do Journal Impact Factor Percentil (JIF): 34,62
- Histórico do Journal Citation Indicator (JCI): 46,88
- Classificação Qualis/CAPES Sucupira (Medicina 1): B2
- Artigo Citações Scopus: 08
- Disponível em: <https://jcr-clarivate.ez34.periodicos.capes.gov.br/jcr-jp/journal-profile?journal=ARQ%20BRAS%20CARDIOL&year=2021&fromPage=%2Fjcr%2Fhome>.

6. SILVA, C. M. C.; GOMES, L. F. G. Reconhecimento clínico das Cardiopatias Congênitas. Revista da Sociedade de Cardiologia do Estado de São Paulo (SOCESP), v. 12, p. 717-723, 2002.

- Classificação Qualis/CAPES Sucupira (Medicina 1): C
- Indexada em: LILACS – Literatura Latino-Americana e do Caribe em Ciências da Saúde (www.bireme.br); Latindex – Sistema Regional de Información em Línea para Revistas Científicas de América Latina, El Caribe, España y Portugal
- ISSN 0103-8559
- Disponível em <https://socesp.org.br/revista/>.

7. RIVIERA, I. R.; GOMES, L. F. G.; MOISES, V. A.; SILVA, C. C.; ANDRADE, J. L.; CARVALHO, A. C. Anomalias Vasculares Arteriais Múltiplas em Recém-nascidos. Diagnóstico Ecocardiográfico e Angiográfico. Arquivos Brasileiros de Cardiologia, v. 75, p. 137-140, 2000.

- Journal Impact Factor: 2.667
- Journal Citation Indicator (JCI): 0.46
- Histórico do Journal Impact Factor Percentil (JIF): 34,62

- Histórico do Journal Citation Indicator (JCI): 46,88
- Classificação Qualis/CAPES Sucupira (Medicina 1): B2
- Disponível em: <https://jcr-clarivate.ez34.periodicos.capes.gov.br/jcr-jp/journal-profile?journal=ARQ%20BRAS%20CARDIOL&year=2021&fromPage=%2Fjcr%2Fhome>.

8. SILVA, C. M. C.; GOMES, L. F. G.; LOPEZ, V. M. O.; ANDRADE, J. L.; MATHIAS JR, W. Ecocardiografia com Contraste em Crianças – experiência inicial. Revista da Sociedade de Cardiologia do Estado de São Paulo (SOCESP), v. 9, p. 742-748, 1999.

- Classificação Qualis/CAPES Sucupira (Medicina 1): C
- Indexada em: LILACS – Literatura Latino-Americana e do Caribe em Ciências da Saúde (www.bireme.br); Latindex – Sistema Regional de Información em Línea para Revistas Científicas de América Latina, El Caribe, España y Portugal
- ISSN 0103-8559
- Disponível em: <https://socesp.org.br/revista/>.

9. SILVA, C. M. C.; GOMES, L. F. G.; MATOS, R. Uso de inibidores da enzima conversora de angiotensina em neonatos e lactentes. Revista da Sociedade de Cardiologia do Estado de São Paulo (SOCESP), v. 9, p. 690-741, 1999.

- Classificação Qualis/CAPES Sucupira (Medicina 1): C
- Indexada em: LILACS – Literatura Latino-Americana e do Caribe em Ciências da Saúde (www.bireme.br); Latindex – Sistema Regional de Información em Línea para Revistas Científicas de América Latina, El Caribe, España y Portugal
- ISSN 0103-8559
- Disponível em: <https://socesp.org.br/revista/>.

10. ALVO, M.; SOUZA, R. L.; GOMES, L. F. G.; CASTRO, M. E. C. T.; LUCIA, M. M. S.; NOHRA, A. C.; PALUMBO NETO, M. Abscesso Hepático Ascaridiano na Criança. Pediatria Atual (Rio de Janeiro), v. 10, p. 12-16, 1997.

- Título Abreviado: Pediatr. atual.

- ISSN: 0103-2712
- Editora: Publicações Científicas
- Cidade: São Paulo/SP
- Área temática: Ciências da Saúde
- Assuntos: Medicina
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- Classificação Qualis/CAPES Sucupira (Medicina 1): B1
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- Área temática: Ciências da Saúde
- Assuntos: Medicina
- Título indexado em: Biological Abstracts; Excerpta Medica; Index Medicus; LILACS; Scopus; SES-Saúde Pública
- Scopus – CiteScore measures average (2017): 0,1
- Socpus – SCImago Journal Rank (SJR 2019): 0,108

- Classificação Qualis/CAPES Sucupira (Educação Física e Odontologia): B2
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58. GOMES, L. F. G. Alteração do Ph intramucoso medido através de tonometria gástrica no pós-operatório de cirurgia gástrica em crianças. In: XX Congresso da SOCESP, 1997, Campos do Jordão – São Paulo. Suplemento da Revista da Sociedade de Cardiologia do Estado de São Paulo. São Paulo: SOCESP, 1997. v. 9. p. 104.

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60. GOMES, L. F. G. Balões destacáveis e a oclusão de fístulas coronarianas e pulmonares de grande porte. In: XXX Congresso Brasileiro de Pediatria – Simpósio Internacional de Pediatria, 1997, Rio de Janeiro. Arquivos Brasileiros de Pediatria. Rio de Janeiro: Associação Brasileira de Pediatria, 1997. v. 4. p. S31.

61. GOMES, L. F. G. Cateterismo cardíaco com punção arterial em crianças abaixo de 5 anos. In: XXX Congresso Brasileiro de Pediatria – Simpósio Internacional de Pediatria, 1997, Rio de Janeiro. Arquivos Brasileiros de Pediatria. Rio de Janeiro: Associação Brasileira de Pediatria, 1997. v. 4. p. S36.

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63. GOMES, L. F. G. Coarctação da Aorta: Resultados da Correção Cirúrgica. In: LII Congresso Brasileiro de Cardiologia, 1997, São Paulo. Arquivos Brasileiros de Cardiologia. São Paulo: Sociedade Brasileira de Cardiologia, 1997. v. 70. p.73.

64. GOMES, L. F. G. Diálise peritoneal no pós-operatório imediato (POI) de cirurgia cardíaca pediátrica. In: XXX Congresso Brasileiro de Pediatria – Simpósio Internacional de Pediatria, 1997, Rio de Janeiro. Arquivos Brasileiros de Pediatria. Rio de Janeiro: Associação Brasileira de Pediatria, 1997. v. 4. p. S34.

65. GOMES, L. F. G. Hipercolesterolemia familiar homozigota – relato de caso. In: XXX Congresso de Brasileiro de Pediatria – Simpósio Internacional de Pediatria, 1997, Rio de Janeiro. Arquivos Brasileiros de Pediatria. Rio de Janeiro: Associação Brasileira de Pediatria, 1997. v. 4, p. S30.

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de Janeiro. Arquivos Brasileiros de Pediatria. Rio de Janeiro: Associação Brasileira de Pediatria, 1997. v. 4. p. S34.

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6.7 Apresentação de Trabalho e/ou Palestras Proferidas

1. GOMES, L. F. G. Coarctação de Aorta Atípica e Complexa em criança escolar e a importância da precisão do diagnóstico precoce, 2018. (Simpósio, apresentação de trabalho).
Evento: XXV Semana Científica do Curso de Medicina da UFU: caminhos do Egresso;
Inst. promotora: Faculdade de Medicina da UFU.

2. GOMES, L. F. G. Arritmias cardíacas no neonato – abordagem sistematizada, 2015. (Conferência ou palestra, apresentação de trabalho)
Palestra ministrada no dia 16/7/2015 das 19h30 às 21h30;
Local: Hospital São Luís – Unidade Itaim;
Evento: Centro de Estudos do Hospital São Luís;
Inst. promotora: Hospital São Luís – Unidade Itaim.

3. GOMES, L. F. G. Cardiopatia Congênita no período neonatal, 2015. (Conferência ou palestra, apresentação de trabalho)

Palestra ministrada no dia 12/3/2015, das 19h30 às 21h30;
Local: Hospital São Luís – Unidade Itaim;
Evento: Centro de Estudos do Hospital São Luís;
Inst. promotora: Hospital São Luís – Unidade Itaim.

4. GOMES, L. F. G. Cardiopatias na criança portadora de Síndrome de Down, 2015. (Conferência ou palestra, apresentação de trabalho)
Local: Anfiteatro 8C; Cidade: Uberlândia/MG;
Inst. promotora: Faculdade de Medicina da Universidade Federal de Uberlândia.

5. GOMES, L. F. G. Controle técnico área – dose aumenta a segurança em criança com cardiopatia congênita no Laboratório de Cateterismo e Hemodinâmica, 2014. (Conferência ou palestra, apresentação de trabalho)
Local: Hotel Plaza São Rafael;
Evento: XXIII Congresso Brasileiro de Cardiologia Pediátrica e Cirurgia Cardiovascular Pediátrica;
Inst. promotora: Sociedade Brasileira de Cardiologia Pediátrica e Cirurgia Cardiovascular Pediátrica.

6. GOMES, L. F. G. Controle Técnico da Dose – Área – Produto aumenta a proteção e a segurança em crianças com Cardiopatia Congênita no Laboratório de Hemodinâmica, 2013. (Conferência ou palestra, apresentação de trabalho)
Local: Transamérica Expo Center;
Evento: SOLACI SBHCI 2013 In Partnership Wit TCT;
Inst. promotora: Sociedade Brasileira de Hemodinâmica e Cardiologia Intervencionista.

7. GOMES, L. F. G. Cardiologia Fetal, 2012. (Comunicação, apresentação de trabalho)
Local: Foz do Iguaçu;
Evento: XXII Congresso Brasileiro de Cardiologia Pediátrica;
Inst. promotora: Sociedade Brasileira de Pediatria.

8. GOMES, L. F. G. Controle da dose pela técnica produto dose-aérea aumenta a proteção e segurança em crianças com doença cardíaca congênita no laboratório de cateterismo, 2012. (Comunicação, apresentação de trabalho)
Local: Foz do Iguaçu;
Evento: XXII Congresso Brasileiro de Cardiologia Pediátrica;
Inst. promotora: Sociedade Brasileira de Pediatria.

9. GOMES, L. F. G. Pericardite constritiva – Diagnóstico pouco explorado e tardio em pediatria – relato de dois casos, 2012. (Comunicação, apresentação de trabalho)
Local: Foz do Iguaçu;
Evento: XXII Congresso Brasileiro de Cardiologia Pediátrica;
Inst. promotora: Sociedade Brasileira de Pediatria.

10. GOMES, L. F. G. Cardiopatia Congênita – abordagem clínica, 2011. (Conferência ou palestra, apresentação de trabalho)
Local: Faculdade de Educação Física;
Evento: III Jornada de Fisioterapia da Universidade Federal de Uberlândia;
Inst. promotora: Faculdade de Educação Física / Universidade Federal de Uberlândia.

11. GOMES, L. F. G. Doença de Chagas na Infância – Aspectos Epidemiológicos e Evolução Clínica, 2009. (Comunicação, apresentação de trabalho)
Local: São Paulo/SP;
Evento: XXX Congresso da Sociedade de Cardiologia do Estado de São Paulo;
Inst. promotora: Sociedade de Cardiologia do Estado de São Paulo.

12. GOMES, L. F. G. Transposição de Grandes Vasos, 2009. (Conferência ou palestra, apresentação de trabalho)
Local: Hospital e Maternidade São Luiz;
Evento: Grupo de Estudos Neocor;
Inst. promotora: São Luiz Centro de Estudos.

13. GOMES, L. F. G. Interpretação do Eletrocardiograma em Neonatologia, 2008. (Conferência ou palestra, apresentação de trabalho)

Local: Hospital e Maternidade São Luiz;
Evento: Reunião Científica da Equipe de Neonatologia;
Inst. promotora: São Luiz Centro de Estudos.

14. GOMES, L. F. G. Amparo Maternal, 2007. (Conferência ou palestra, apresentação de trabalho)
Local: Centro de Aprimoramento em Saúde;
Evento: Curso de Pós-Graduação *Lato Sensu* em Fisioterapia Neonatal;
Inst. promotora: Centro de Aprimoramento em Saúde.

15. GOMES, L. F. G. Apresentação não usual da Doença de Kawasaki na Criança – Relato de Caso, 2007. (Comunicação, apresentação de trabalho)
Local: Centro de Convenções de Goiânia;
Evento: IX Congresso Nacional de Pediatria;
Inst. promotora: Sociedade Brasileira de Pediatria.

16. GOMES, L. F. G. Doença de Chagas – Raridade na infância? 2007. (Comunicação, apresentação de trabalho)
Local: Centro de Convenções de Goiânia;
Evento: IX Congresso Nacional de Pediatria;
Inst. promotora: Sociedade Brasileira de Pediatria.

17. GOMES, L. F. G. Avaliação do carvedilol no tratamento de insuficiência cardíaca em cardiopediatria, 2006. (Comunicação, apresentação de trabalho)
Local: Campos do Jordão/SP;
Evento: XXVII Congresso Brasileiro da Sociedade de Cardiologia do Estado de São Paulo;
Inst. promotora: Sociedade de Cardiologia do Estado de São Paulo.

18. GOMES, L. F. G. Transposição dos Grandes Vasos, 2006. (Conferência ou palestra, apresentação de trabalho)
Local: Hospital e Maternidade São Luiz – Centro de Estudos;
Evento: Reunião do Grupo de Estudos de Enfermagem em Cardiologia Neonatal;
Inst. promotora: Hospital e Maternidade São Luiz – Centro de Estudos.

19. GOMES, L. F. G. Hemodinâmica, 2005. (Conferência ou palestra, apresentação de Trabalho)
Vários; Local: Minas Centro; Cidade: Belo Horizonte/MG;
Evento: IX Congresso Mineiro de Terapia Intensiva;
Inst. promotora: Sociedade Mineira de Terapia Intensiva.

20. GOMES, L. F. G. Reconhecimento clínico das Cardiopatias Congênitas e abordagem das arritmias no período neonatal, 2005. (Conferência ou palestra, apresentação de trabalho)
Local: Hospital e Maternidade São Luiz;
Evento: Reunião Científica da Equipe de Neonatologia;
Inst. promotora: São Luiz Centro de Estudos.

21. GOMES, L. F. G.; SILVA, CÉLIA MARIA CAMPOS; OPORTO, VICTOR MANUEL; ABUJAMRA, P.; MATOS NETO, RANULFO PINHEIRO DE; BELO, P.; MOISES, V. A.; CAMPOS FILHO, ORLANDO; CARVALHO, A. C. C.; MALUF, M; BUFFOLO, E.; LIMA, W.; PAOLA, A. A. V. A importância dos achados ecocardiográficos no ótimo manejo da Atrésia Pulmonar com Septo Ventricular Integro, 2001. (Comunicação, apresentação de trabalho)
Local: Hotel Meliá;
Evento: XIII Congresso Brasileiro de Ecocardiografia;
Inst. promotora: Sociedade Brasileira de Ecocardiografia.

22. GOMES, L. F. G. Ecocardiografia com Contraste com Microbolhas – Infusão e uso desta técnica no neonato e criança, 2001. (Comunicação, apresentação de trabalho)
Local: Hotel Meliá;
Evento: XIII Congresso Brasileiro de Ecocardiografia;
Inst. promotora: Sociedade Brasileira de Ecocardiografia.

23. GOMES, L. F. G. Acesso Hemodinâmico em Emergências, 2000. (Conferência ou palestra, apresentação de trabalho)
Local: Minas Centro;

Evento: IX Congresso Brasileiro de Terapia Intensiva;
Inst. promotora: Sociedade Brasileira de Terapia Intensiva.

24. GOMES, L. F. G. Avaliação funcional do Ventrículo Direito no Pós-operatório Intermediário de Correção de Tétrade de Fallot, com reconstrução da Valva Pulmonar, 2000. (Comunicação, apresentação de trabalho)
Local: Campos do Jordão/SP;
Evento: XXI Congresso da Sociedade de Cardiologia do Estado de São Paulo;
Inst. promotora: Sociedade de Cardiologia do Estado de São Paulo.

25. GOMES, L. F. G. Crises Hipoxêmicas nas Cardiopatias Congênitas, 1999. (Comunicação, apresentação de trabalho)
Local: Uberlândia/MG;
Evento: II Congresso da Sociedade de Cardiologia do Triângulo Mineiro;
Inst. promotora: Sociedade de Cardiologia do Triângulo Mineiro.

26. GOMES, L. F. G. Papel da perfuração da Valva Pulmonar com Radiofrequência seguida da Dilatação por Balão de Tratamento de Atrésia Pulmonar com Septo Ventricular Íntegro. Experiência da Unifesp/EPM, 1999. (Comunicação, apresentação de trabalho)
Local: Centro de Convenções do Colégio Marista;
Evento: XXI Congresso Brasileiro de Hemodinâmica e Cardiologia Intervencionista;
Inst. promotora: Sociedade Brasileira de Hemodinâmica e Cardiologia Intervencionista.

27. GOMES, L. F. G. Atriosseptostomia por Cateter Balão à Beira do Leito, 1998. (Comunicação, apresentação de trabalho)
Local: Campos do Jordão/SP;
Evento: XIX Congresso da Sociedade de Cardiologia do Estado de São Paulo;
Inst. promotora: Sociedade de Cardiologia do Estado de São Paulo.

28. GOMES, L. F. G. Choque Séptico, 1998. (Conferência ou palestra, apresentação de trabalho)
Local: Centro Médico de Franca;

Evento: Evento do Departamento Científico do Centro Médico de Franca;
Inst. promotora: Centro Médico de Franca.

29. GOMES, L. F. G. Hemodiafiltração venovenosa contínua em 8 pacientes pediátricos portadores de insuficiência de múltiplos órgãos e sistema – evolução clínica e laboratorial, 1998. (Comunicação, apresentação de trabalho)
Local: Salvador/BA;
Evento: VII Congresso Brasileiro de Terapia Intensiva Pediátrica;
Inst. promotora: Sociedade Brasileira de Pediatria.

30. GOMES, L. F. G. Alterações do PH Intravenoso medido através de tonometria gástrica no pós-operatório de cirurgia cardíaca em crianças, 1997. (Comunicação, apresentação de trabalho)
Local: Centro de Convenções Rio Centro;
Evento: XXX Congresso Brasileiro de Pediatria – Simpósio Internacional de Pediatria;
Inst. promotora: Sociedade Brasileira de Pediatria.

31. GOMES, L. F. G. Atriosseptostomia por Cateter Balão à Beira do Leito, 1997. (Comunicação, apresentação de trabalho)
Local: Centro de Convenções Rio Centro;
Evento: XXX Congresso Brasileiro de Pediatria – Simpósio Internacional de Pediatria;
Inst. promotora: Sociedade Brasileira de Pediatria.

32. GOMES, L. F. G. Avaliação do tratamento cirúrgico na coarctação de aorta em Neonatos e Lactentes, 1997. (Comunicação, apresentação de trabalho)
Local: Centro de Convenções Rio Centro;
Evento: XXX Congresso Brasileiro de Pediatria – Simpósio Internacional de Pediatria;
Inst. promotora: Sociedade Brasileira de Pediatria.

33. GOMES, L. F. G.; CARVALHO, A. C. C.; MUSSATI, C.; JULIANO, Y.; NASPITZ, C.; MALUF, M. Análise quantitativa das populações de linfócitos T E B e

Subpopulações de Linfócitos em crianças com cardiopatias, 1992. (Comunicação, apresentação de trabalho)
Local: Centro de Convenções de Pernambuco;
Evento: XXVII Congresso Brasileiro da Sociedade de Cardiologia do Estado de São Paulo;
Inst. promotora: Sociedade Brasileira de Cardiologia.

34. GOMES, L. F. G.; CARVALHO, A. C. C.; MUSSATI, C.; JULIANO, Y.; NASPITZ, C.; MALUF, M. Avaliação da função fagocitária de polimorfonucleares (PMN) em pacientes com cardiopatias congênitas, 1992.
Local: Centro de Convenções de Pernambuco;
Evento: XXVII Congresso Brasileiro da Sociedade de Cardiologia do Estado de São Paulo;
Inst. promotora: Sociedade Brasileira de Cardiologia.

6.8 Produção Técnica

1. GOMES, L. F. G. Debatedora do Painel – Pós-operatório, 2000.

2. GOMES, L. F. G. Coordenadora do Curso Clínico Cardiopatia Congênita, na Unidade de Terapia Intensiva no Hospital de Clínicas da Faculdade de Medicina da Universidade Federal de Uberlândia. 1999.

3. GOMES, L. F. G. Debatedora: Aplicação de um índice de escore na UTI-Pediátrica e Neonatal para avaliação do tratamento cirúrgico de pacientes com cardiopatia congênita. 2006. 1ª Jornada de Integração da SOMITI.

4. GOMES, L. F. G. Participação técnica dos exames ecocardiográficos bidimensionais com Doppler e fluxo colorido e avaliação clínica ambulatorial na realização do (protocolo clínico) de pacientes portadores de Síndrome da Imunodeficiência Adquirida (AIDS) na coleta de dados na tese de doutorado da Professora Dra. Maria Suely Bezerra Diógenes intitulada: Avaliação cardiológica em crianças expostas ao vírus da imunodeficiência humana tipo 1 por via perinatal:

estudo clínico, eletrocardiográfico e ecocardiográfico Doppler HSP-EPM-UNIFESP. 1997-2001.

5. GOMES, L. F. G. Participação técnica dos exames realizados na hemodinâmica, cateterismo e intervenção na coleta de dados e protocolo terapêutico na tese de doutorado da Professora Dra. Célia Camelo da Silva intitulada: Atrésia pulmonar com septo interventricular intacto: Experiência inicial com a perfuração valvar por radiofrequência em neonatos e lactentes. HSP-EPM-UNIFESP. 1995-2000.

6. GOMES, L. F. G. Participação técnica na coleta de sangue e transferência dos exames do laboratório de imunologia para o laboratório de análises clínicas em pesquisa do HSP-EPM-UNIFESP para realização da dosagem de homocisteína da Professora Dra. Alessandra Carla intitulada: Análise de homocisteína e outros marcadores de inflamação e lesão miocárdica em crianças submetidas à circulação extracorpórea. HC-FAMED-UFU. 2002-2006.

7. GOMES. L. F. G. Participação na seleção dos pacientes e na coleta e na viabilização do cuidado das amostras para a tese de doutorado do Dr. Cláudio Ribeiro da Cunha intitulada: Perfil das citocinas e correlação com a morbidade no período pós-operatório em crianças com diagnóstico de cardiopatias congênitas não cianosantes submetidas à cirurgia corretiva com circulação extracorpórea. HC-FAMED-UFU. 2008-2012.

6.9 Participação em eventos científicos

1. Encontro de Desenvolvimento Docente: Metodologias Ativas de Ensino-Aprendizagem, 2019. (Encontro)

2. XXXVIII Congresso da SOCESP, 2017. (Congresso)

3. 8º LATAM Symposium in Pulmonary Hypertension, 2016. (Simpósio)

4. XXIV Congresso Brasileiro de Cardiologia e Cirurgia Cardiovascular Pediátrica, 2016. (Congresso)

5. XXXVI Congresso da Sociedade de Cardiologia do Estado de São Paulo, 2015. (Congresso)

6. 14º Congresso Brasileiro de Ensino e Pesquisa 2014, 2014. (Congresso)

7. XXIII Congresso Brasileiro de Cardiologia Pediátrica e Cirurgia Cardiovascular Pediátrica, 2014. (Congresso)

8. XXXV Congresso da Sociedade de Cardiologia do Estado de São Paulo, 2014. (Congresso)

9. SOLACI SBHCI 2013, 2013. (Congresso)

10. XXXIV Congresso da Sociedade de Cardiologia do Estado de São Paulo, 2013. (Congresso)

11. XXII Congresso Brasileiro de Cardiologia Pediátrica, 2012. (Congresso)

12. XXXI Congresso da SOCESP, 2010. (Congresso)

13. Simpósio SOCESP, 2009. (Simpósio)

14. XXI Congresso Brasileiro de Ecocardiografia, 2009. (Congresso)

15. XX Congresso Brasileiro de Ecocardiografia, 2008. (Congresso)

16. XXIX Congresso da Sociedade de Cardiologia do Estado de São Paulo, 2008. (Congresso)

17. XVIII Congresso Brasileiro de Ecocardiografia, 2006. (Congresso)

18. XXVII Congresso Brasileiro da Sociedade de Cardiologia do Estado de São Paulo, 2006. (Congresso)

19. IX Congresso Mineiro de Terapia Intensiva, 2005. (Congresso)

20. IX Congresso Paulista de Terapia Intensiva, 2005. (Congresso)

21. Pediatric Interventional Cardiac Symposium, 2005. (Simpósio)

22. XXVI Congresso da Sociedade de Cardiologia do Estado de São Paulo, 2005. (Congresso)

23. XVI Congresso Brasileiro de Ecocardiografia, 2004. (Congresso)

24. XIV Congresso Brasileiro de Ecocardiografia, 2002. (Congresso)

25. XXIII Congresso da Sociedade de Cardiologia do Estado de São Paulo, 2002. (Congresso)

26. 2nd International Symposium of the Brazilian Cochrane Collaboration, 2001. (Simpósio)

27. III Congresso da Sociedade de Cardiologia do Triângulo Mineiro, 2001. (Congresso)

28. XIII Congresso Brasileiro de Ecocardiografia, 2001. (Congresso)

29. III Encontro do Grupo de Estudos em Cardiologia Neonatal – NEOCOR, 2000. (Encontro)

30. IX Congresso Brasileiro de Terapia Intensiva, 2000. (Congresso)

31. XXI Congresso da Sociedade de Cardiologia do Estado de São Paulo, 2000. (Congresso)

32. II Congresso da Sociedade de Cardiologia do Triângulo Mineiro, 1999. (Congresso)

33. LIV Congresso da Sociedade Brasileira de Cardiologia, 1999. (Congresso)

34. XI Congresso Brasileiro de Ecocardiografia, 1999. (Congresso)

35. XVI Congresso Brasileiro de Cardiologia Pediátrica, 1999. (Congresso)

36. XX Congresso da Sociedade de Cardiologia do Estado de São Paulo, 1999. (Congresso)

37. Simpósio Internacional Cardiopatia Congênitas, 1998. (Simpósio)

38. II International Symposium of Pediatric Cardiology, 1997. (Simpósio)

39. 1ª Jornada de Emergência em Pediatria, 1994. (Simpósio)

40. 1ª Jornada de Infectologia Pediátrica, 1990. (Simpósio)

6.10 Organização de Eventos

1. GOMES, L. F. G. Assistente de Atualização Curricular – Monitorização Hemodinâmica com Cateteres Pulmonares, 2000.

2. GOMES, L. F. G. Presidente da Conferência Avanços na Terapia da Asma Grave, 2000.

3. GOMES, L. F. G. Assistente de Atualização Curricular – Atualização em Arritmias, 1999.

4. GOMES, L. F. G. Assistente do Curso Cardiologia para Pediatrias, 1999.

7 ORIENTAÇÕES ACADÊMICAS

7.1 Trabalho de Conclusão de Curso

1. Vera Lúcia Santos Silva. Orientação de tese de conclusão de curso da enfermagem sob o tema: Cuidados de enfermagem em pré e pós-operatório de cirurgia cardíaca. Dissertação Faculdade de Enfermagem – Universidade Federal de Uberlândia. 2005.

7.2 Trabalho de Conclusão de Especialização em Enfermagem Neonatal em Unidade de Terapia Intensiva Neontal

1. Maria Elenice Costa. Orientação de tese de conclusão de Curso da Especialização em Enfermagem de Neonatologia sob o tema: Cuidados de enfermagem em pré e pós-operatório de fechamento de canal arterial em recém-nascidos prematuros. Dissertação Faculdade de Enfermagem – Universidade Federal de Uberlândia. 2009.

7.3 Mestrado

1. Bruno Franco Rossi. Perfil epidemiológico e evolução clínica dos portadores de miocardiopatia dilatada acompanhados no Serviço de Cardiologia Pediátrica do Hospital de Clínicas da UFU. 2018. Dissertação (Ciências da Saúde) – Universidade Federal de Uberlândia.
Ano início e término: 03/2016 a 08/2018 (24 meses).

8 PARTICIPAÇÃO EM BANCAS EXAMINADORAS

8.1 Mestrado

1. GOMES, L. F. G.; SILVA, A. C.; RESENDE, E. S. Participação em banca de Bruna Zanforlin Jácome. Tratamento e evolução de crianças com cardiomiopatia dilatada acompanhadas em um serviço público de cardiologia pediátrica de referência regional, 2022. (Ciências da Saúde) Universidade Federal de Uberlândia. Exame de Qualificação realizado em: 29/9/2022.

1. GOMES, L. F. G.; SILVA, A. C.; RESENDE, E. S. Participação em banca de Bruna Zanforlin Jácome. Tratamento e evolução de crianças com cardiomiopatia dilatada acompanhadas em um serviço público de cardiologia pediátrica de referência regional, 2022. (Ciências da Saúde) Universidade Federal de Uberlândia. Defesa realizada em: 3/11/2022.

8.2 Doutorado

1. RESENDE, E. S.; GOMES, L. F. G.; SILVA, A. C. Participação em banca examinadora de Almir Fernando Loureiro Fontes. Emprego da ecocardiografia de strain na identificação de comprometimento do coração em casos clínicos moderados e graves de COVID-19, 2022. (Ciências da Saúde) Universidade Federal de Uberlândia. Exame de Qualificação realizado em: 28/10/2022.

2. PEIXOTO, J. M.; SALEH, M. H.; GOMES, L. F. G.; ARAÚJO, M. A.; RESENDE, E. S. Participação em banca examinadora de Almir Fernando Loureiro Fontes. Injúria miocárdica avaliada pela ecocardiografia de Strain em pacientes após COVID-19, 2022. (Ciências da Saúde) Universidade Federal de Uberlândia. Defesa realizada em: 29/11/2022.

8.3 Membro de Banca de Concursos Públicos

1. Membro da Comissão Permanente de Docentes para o Processo Seletivo de Residência Médica – 2013 Edital 03/2013, Universidade Federal de Uberlândia, Portaria FAMED nº 24/2013 de 15/10/2013.

2. Membro Suplente da Comissão Julgadora do processo seletivo simplificado regido pelo EDITAL PROGEP nº 10/2022, 2022, Universidade Federal de Uberlândia. Portaria de Pessoal UFU Nº 755, de 22/2/2022.

8.4 Membro de Banca de Trabalho de Conclusão de Residência

1. Membro da Banca de Conclusão de Residência Médica (R3) da Especialidade Pediatria – Programa de Residência Médica da Faculdade de Medicina da Universidade Federal de Uberlândia.

- Residente: Arthur Lacerda Mendonça

- Título: Efeitos da musicoterapia em crianças e bebês internados em unidade hospitalar

Mês/ano: fevereiro/2023.

2. Membro da Banca de Conclusão de Residência Médica (R3) da Especialidade Pediatria – Programa de Residência Médica da Faculdade de Medicina da Universidade Federal de Uberlândia.

- Residente: Tarsila Araujo Paiva Molinar

- Título: Internação de pacientes pediátricos com paralisia cerebral em um hospital de referência nos últimos 5 anos: conhecer para melhor cuidar.

Mês/ano: fevereiro/2023.

9 SOCIEDADES CIENTÍFICAS

1 – Sócia efetiva da Sociedade Brasileira de Cardiologia desde 27/12/2004 com matrícula nº 15709.

2 – Sociedade Brasileira de Cardiologia Pediátrica e Congênita desde 2004.

3 – Sociedade Brasileira de Pediatria – em 1988.

4 – Sociedade Brasileira de Terapia Intensiva – em 1990.

5 – Sociedade Brasileira de Ecocardiografia – desde 2009.

6 – Sociedade Paulista de Terapia Intensiva Pediátrica – desde 1990.

7 – Sociedade Paulista de ecocardiografia desde – 2009.

10 DISTINÇÕES E HONRARIAS POR ATUAÇÃO ACADÊMICA

Ano 2003

Professora Homenageada da 50ª turma de formandos do Curso de Medicina – UFU, Universidade Federal de Uberlândia – Novembro de 2003.

Ano 2006

Prêmio Melhor pesquisa aplicada para o trabalho: avaliação do carvedilol no tratamento de insuficiência cardíaca em cardiopediatria.
Local: XXVII Congresso da Sociedade de Cardiologia do Estado de São Paulo.

11 TITULAÇÕES DE ESPECIALISTA

1- Residência Médica em Pediatria: aprovada no concurso de Residência Médica no Hospital de Clínicas da Faculdade de Medicina – Universidade Federal de Uberlândia em: 2/12/1984 – anos de 1985 e 1986.

2- Especialização em Terapia Intensiva Pediátrica: por concurso público na UTI Pediátrica – Escola Paulista de Medicina – Hospital São Paulo – Universidade Federal de São Paulo – HSP-EPM-UNIFESP: 2/2/1989 – período de 1989 a 1990.

3- Especialização em Neonatologia na Unidade Neonatal por seleção interna na Unidade Neonatal e UTI Neonatal – Escola Paulista de Medicina – Hospital São Paulo – Universidade Federal de São Paulo – HSP-EPM-UNIFESP: 2/2/1991 – período 1992 e 1993.

4- Especialização em Cardiologia Pediátrica e Congênita – Concurso público na Cardiologia Pediátrica e Congênita em 20/1/1996 na Cardiologia Pediátrica e Congênita sob a coordenação do Professor Dr. Antônio Carlos de Camargo Carvalho – Escola Paulista de Medicina – Hospital São Paulo – Universidade Federal de São Paulo – HSP-EPM-UNIFESP: 1/2/1996 – período de 1996 a 1999.

5- Especialização em Ecocardiografia Congênita, Fetal e Pediátrica sob a coordenação do Prof. Dr. Antônio Carlos de Camargo Carvalho, Dra. Maria Célia Camelo da Silva e Prof. Dr. Valdir Ambrósio no Setor de Ecocardiografia Pediátrica, Fetal e Congênita – Escola Paulista de Medicina – Hospital São Paulo – Universidade Federal de São Paulo – HSP-EPM-UNIFESP: 2/2/1991 – período 1991 e 1993.

12 AUXILIAR EM PESQUISAS COMO COLETA DE DADOS

Doutorado em Medicina (Cardiologia)
1992-2000
Professora Dra. Celia Maria Camelo da Silva
Universidade Federal de São Paulo
Título: Atrésia pulmonar com septo interventricular intacto: experiência inicial com a perfuração valvar por radiofrequência em neonatos e lactentes
Antônio Carlos de Camargo Carvalho. Bolsista do(a): Coordenação de Aperfeiçoamento de Pessoal de Nível Superior, CAPES, Brasil. Palavras-chave: atrésia pulmonar com septo ventricular íntegro; Atrésia pulmonar com septo interventricular intacto; Perfuração valvar pulmonar com radiofrequência; Cateterismo intervencionista. Grande área: Ciências da Saúde / Área: Medicina / Subárea: Saúde Materno-Infantil / Especialidade: Hemodinâmica e cateterismo cardíaco nas Cardiopatias Congênitas. Setores de atividade: Saúde Humana.

Doutorado em Medicina
Dra. Maria Suely Bezerra Diógenes
1997-2001
Universidade Federal de São Paulo
Título: Avaliação cardiológica em crianças expostas ao vírus da imunodeficiência humana tipo 1 por via perinatal: estudo clínico, eletrocardiográfico e ecocardiográfico Doppler
Antonio Carlos Camargo Carvalho. Coorientador: Regina Célia de Menezes Succi. Bolsista do(a): Coordenação de Aperfeiçoamento de Pessoal de Nível Superior, CAPES, Brasil. Palavras-chave: HIV – Vírus da Imunodeficiência Humana; Síndrome da Imunodeficiência Adquirida (SIDA); Comprometimento cardiológico; Ecocardiografia Doppler; Eletrocardiografia.
Grande área: Ciências da Saúde / Área: Medicina / Subárea: CARDIOLOGIA. Setores de atividade: Saúde humana e serviços sociais; Atividades de atenção à saúde humana.

Universidade Federal de São Paulo

1998-2003

Mestrado em Cardiologia Pediátrica e Congênita

Dr. Ranulfo Pinheiro de Matos Neto

Função sistólica do ventrículo esquerdo pela ecocardiografia em crianças e adolescentes com osteosarcoma tratados com doxorrubicina com e sem dexrazoxane. 2003. Dissertação (Mestrado em Medicina – Cardiologia) – Universidade Federal de São Paulo.

Universidade Federal de Uberlândia

2002-2006

Doutorado em Imunologia e Parasitologia Aplicadas (Conceito CAPES 6).

Dra. Alessandra Carla Ribeiro Universidade Federal de Uberlândia, UFU, Brasil.

Título: Análise de Homocisteína e outros marcadores de inflamação e lesão miocárdica em crianças submetidas à circulação extracorpórea, ano de obtenção: 2006.

Orientador: Prof. Dr. José Roberto Mineo.

Palavras-chave: Homocisteína; proteína C reativa; Inflamação; Lesão miocárdica; cirurgia cardíaca; cardiopatia congênita.

Grande área: Ciências Biológicas / Grande área: Ciências da Saúde / Área: Medicina / Setores de atividade: Saúde e Serviços Sociais.

Universidade Federal de Uberlândia

Claudio Ribeiro da Cunha

Doutorado em Imunologia e Parasitologia Aplicadas

2008-2012

Doutorado pela (Conceito CAPES 6) Universidade Federal de Uberlândia, UFU, Brasil.

Título: Perfil das citocinas e correlação com a morbidade no período pós-operatório em crianças com diagnóstico de Cardiopatias Congênitas não cianosantes submetidas à cirurgia corretiva com circulação extracorpórea.

Ano de obtenção: 2012.

Orientador: Prof. Dr. José Roberto Mineo.

Palavras-chave: Cirurgia cardíaca pediátrica; resposta inflamatória; circulação extracorpórea; citocina. Grande área: Ciências da Saúde Grande Área: Ciências

Biológicas / Área: Imunologia. Setores de atividade: Saúde e Serviços Sociais. Arquivos Brasileiros de Cardiologia, 2012. v. 99. p. 8-8.

13 LINHA DO TEMPO

Graduação em Medicina pela Universidade Federal de Uberlândia
1979 a 1984

Residência Médica em Pediatria Geral Universidade Federal de Uberlândia
1985 a 1987

Professora Substituta da Faculdade de Medicina – Departamento de Pediatria/UFU
1987 a 1998

Título de Especialista em Pediatria – AMB, SBP e CFM
1988

Residência Médica em Terapia Intensiva Pediátrica na Universidade Federal de São Paulo
1989 a 1990

Especialização em Neonatologia e Terapia Intensiva Neonatal na Universidade Federal de São Paulo
1990 a 1992

Mestrado em Pediatria na Universidade Federal de São Paulo
1992 a 1995

Especialização em Cardiologia Pediátrica e Congênita pela Universidade Federal de São Paulo
1996 a 1998

Professora Efetiva da Faculdade de Medicina – Departamento de Pediatria/UFU
1998 até atualmente

Especialização em Ecocardiografia Pediátrica e Congênita e Fetal na Universidade Federal de São Paulo
1999 a 2002

Doutorado em Ciências da Saúde na Universidade Federal de São Paulo
2006 a 2012

14 CONSIDERAÇÕES FINAIS

Após a assimilação e o aprendizado das determinações legais para a Progressão Vertical na Carreira de Professora Associada IV para Professora Titular, o desafio do momento consistiu em como fazer o memorial.

- Como perspectivas futuras próximas: Tese de mestrado – Dra. Neide Faria – em andamento;
- Tese de Mestrado – Dra. Viviane Athadeu Gontijo.
- Publicações em andamento
- Miocardiopatia dilatada – aspectos epidemiológicos;
- Avaliação nutricional em cardiopatas congênitos;
- Evolução clínica de pacientes portadores de coração com fisiologia univentricular;
- Musicoterapia em cardiologia pediátrica;
- Projeto de extensão: Ecocardiografia funcional para Intensivistas Pediátricos e Neonatais.

A reflexão em relação à vida pessoal foi caracterizada por alternância de momentos de falha da memória para alguns fatos e de muita lucidez para outros. Quanto à vida acadêmica e profissional, foi preciso retomar documentos antigos, mas não esquecidos, que vieram à tona durante este mergulho ao passado. Este necessário exercício foi longo, tendo culminado em um trabalho cuidadoso. O resultado aqui e agora resumido reflete mais uma disposição em captar os acontecimentos e interpretá-los com a devida perspectiva, em um processo dinâmico ainda em construção, em vez de uma visão pretenciosa e definitiva dos fatos vividos.

Após os registros neste memorial, desenvolveu-se a revisão técnica do texto no sentido de adequação às normas vigentes da língua portuguesa, bem como da revisão dos indicadores bibliométricos pelo Sistema de Bibliotecas da UFU.

O capítulo final desta jornada está ainda por ser escrito e nele deverá prevalecer a conclusão de que é preciso trabalhar mais para o próximo, aprender sempre com os novos acontecimentos e nunca se esquecer de agradecer.

REFERÊNCIAS

CAVALCANTE, Anderson. **As Coisas Boas da Vida**. São Paulo: Editora Gente, 2002.

CORALINA, Cora. O Professor. *In*: **Vintém de cobre**: meias confissões de Aninha. 6. ed., São Paulo: Global Editora, 1997.

FRANÇA, J. L.; VASCONCELLOS, A. C. **Manual para normalização de publicações técnico-científicas**. 8. ed. Belo Horizonte: UFMG, 2007.

FREIRE, Paulo. **Pedagogia da Autonomia**. São Paulo: Paz e Terra, 1996.

FREIRE, Paulo. **Pedagogia do Oprimido**. São Paulo: Paz e Terra, 2013.

UNIVERSIDADE FEDERAL DE UBERLÂNDIA — UFU –. Conselho Diretor. **Resolução CONDIR 03/2017** – Regulamenta a avaliação docente no que se refere à Progressão, à Promoção e à Aceleração da Promoção na Carreira de Magistérios Superior. 2017.

PEDROSA, A. Toda boa mãe morre. **Lar em reforma**, 26 out. 2017**.** Disponível em: <https://laremreforma.wordpress.com/author/ariellepedrosa/>. Acesso em: 22 nov. 2022.

RODEN, A. O Pensador. *In*: MAGEE E B. **Um convite à filosofia**. História da Filosofia; [Tradutor Marcos Bagno]. 6. ed – São Paulo: Edições Loyola. 2013:6-9. 1980. Foto Capa Final.

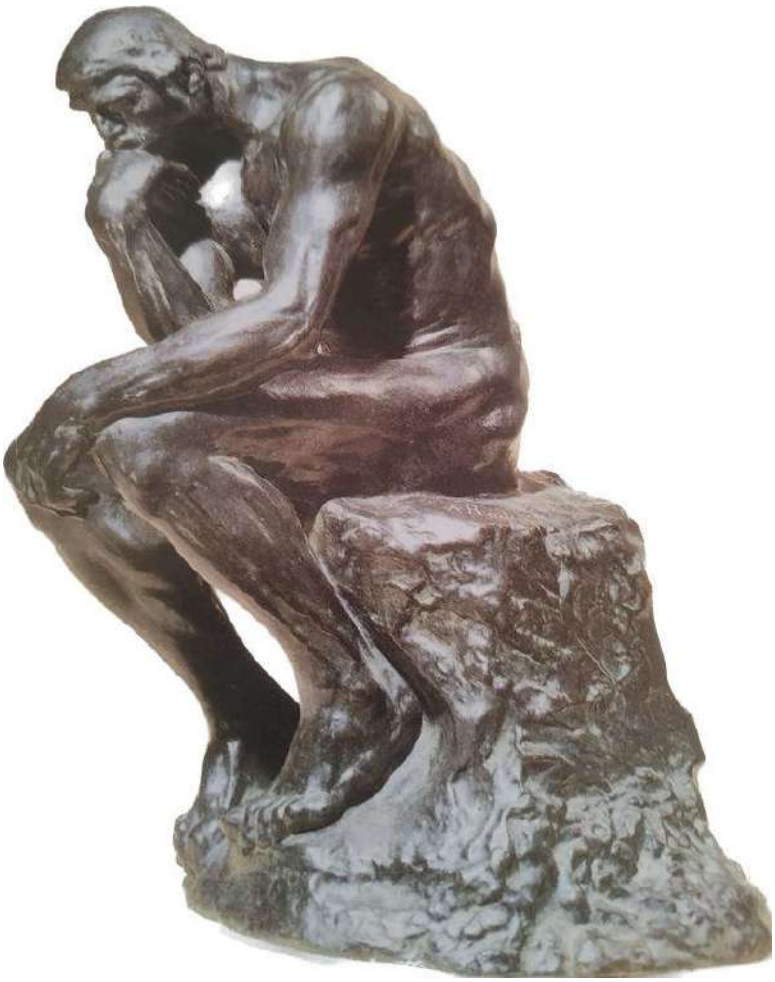
SISTEMA DE BIBLIOTECAS DA UNIVERSIDADE FEDERAL DE UBERLÂNDIA — SISBI . **Indicadores da produção científica**. SISBI-UFU, 20 dez. 2016. Disponível em: <https://bibliotecas.ufu.br/servicos/indicadores-da-producao-cientifica>. Acesso em: 10 mar. 2023.

“Os milionários quiseram comprar a felicidade com seu dinheiro, os políticos quiseram conquistá-la com seu poder, as celebridades quiseram seduzi-la com sua fama, mas ela não se deixou achar balbuciando no ouvido de todos eu me escondo nas coisas simples e anônimas.”

(Augusto Cury)

E quando a gente confia em Deus
Ele nos surpreende.
Quando a gente espera,
Ele nos recompensa.
Quando a gente tem fé,
Ele nos honra
E quando a gente tem esperança
Ele faz tudo dar certo em nossas vidas

Obrigada



“A nudez da famosa estátua de Rodin de um solitário pensador profundamente envolto em reflexão sugere que o homem é de modo peculiar um animal reflexivo e autoconsciente, e que isso é algo fundamental à condição humana.”

(Aguste Rodin)

DOCUMENTOS

COMPROBATÓRIOS

CURSO: MEDICINA

MEC - Universidade Federal de Uberlândia

Diploma r... sob n.° ...35 Livro MEC-/ Fls. ...
em 08 ... Processo n.° 00-0.56 / 85
por delegação de competência do Ministério da Educação e Cultura nos termos da Portaria MEC/DAU nº 71 de 21/12/1977.
Uberlândia, ... / ... / 1985

CONSELHO REGIONAL DE MEDICINA
ESTADO DE SÃO PAULO
Diploma registrado sob n.° 67534
... do Livro n.° ...
com a Lei n.° 3.268 de 30 de setembro de 1957.
P. ... / ... / ...
Chefe da Seção de Registro de Profissionais

REPÚBLICA FEDERATIVA DO BRASIL
MINISTÉRIO DA EDUCAÇÃO E CULTURA
UNIVERSIDADE FEDERAL DE UBERLÂNDIA

CONSELHO REGIONAL DE MEDICINA
DO ESTADO DE MINAS GERAIS
Médico inscrito sob o n.° 18959
a fls. 345 do Livro n° ...
B. H. 09/05/1986
Funcionário: ...
... Vellozo - PRESIDENTE

Colégio Notarial do Brasil - SP
AUTENTICAÇÃO
1013AA744347

O Reitor da Universidade Federal de Uberlândia, no uso de suas atribuições e tendo em vista a Conclusão do Curso de Graduação em Medicina em 21 de dezembro de 1984 confere o título de Médica a Lourdes de Fátima Gonçalves filho de Alcindo Gonçalves Cunha e de Rosana Aparecida Naves Gonçalves nascido a 14 de outubro de 1959 natural de Minas Gerais e outorga-lhe o presente Diploma a fim de que possa gozar de todos os direitos e prerrogativas legais.

Uberlândia, 21 de dezembro de 1984

Reitor [signature]

Pró-Reitor Acadêmico [signature]

Diretor do Centro [signature]

Diplomado Lourdes de Fátima Gonçalves.

REPÚBLICA FEDERATIVA DO BRASIL
ORDEM E PROGRESSO

Universidade Federal de Uberlândia
Faculdade de Medicina
Uberlândia – Minas Gerais

Colegio Notarial do Brasil SP
AUTENTICAÇÃO
1018AA744350

CERTIFICADO

conferido a

Lourdes de Fátima G. Gomes

Certificamos para os devidos fins de direito, que a Dra Lourdes de Fátima Gonçalves Gomes, prestou o Concurso de Residência Médica nesta Instituição em Dezembro de 1984, tendo sido aprovada e cumprindo regularmente o programa de Residência Médica (R1 e R2) na área de *PEDIATRIA* no Hospital de Clínicas da Faculdade de Medicina da Universidade Federal de Uberlândia, no período de 03/01/1985 a 03/01/1987.

Certificamos, outrossim, que o referido *Programa de Residência Médica – SESu/MEC – Brasília – DF – em 25/11/1980, Parecer 033/80,* sendo seu certificado lhe conferindo o Título de Especialista, de acordo com a Lei 6.932/81, publicada no D.D.U., em 09/07/1981.

Uberlândia, 19 de Janeiro de 1988

Prof. Ricardo Custódio Pacheco
Coordenador do Programa de Residência Médica
UFU - FAMED

Associação Médica Brasileira
Sociedade Brasileira de Pediatria
Associação de Medicina Intensiva Brasileira

conferem à

Dra. Lourdes de Fátima Gonçalves Gomes

portador do Título de Especialista em Pediatria o

CERTIFICADO DE ATUAÇÃO NA ÁREA DE MEDICINA INTENSIVA PEDIÁTRICA

São Paulo, 25 de abril de 1990

Dr. Eleuses Vieira de Paiva
Presidente da AMB

Dr. Aldemir Humberto Soares
Secretário Geral da AMB

Dr. Lincoln Marcelo Silveira Freire
Presidente da SBP

Dr. Eduardo da Silva Vaz
Secretário Geral da SBP

Dr. Jairo Constante Bittencourt Othero
Presidente da AMIB

Dr. José Maria da Costa Orlando
Secretário da AMIB

Associação Médica Brasileira

Sociedade Brasileira de Pediatria

conferem o

TÍTULO DE ESPECIALISTA EM PEDIATRIA

à

Dra. Lourdes de Fátima Gonçalves Gomes

por ter obtido aprovação em concurso realizado segundo as normas estabelecidas pela Associação Médica Brasileira e a Sociedade Brasileira de Pediatria.

São Paulo, 31 de maio de 1998

Dr. Eleuses Vieira de Paiva
Presidente da AMB

Dr. Edmund Chada Baracat
Secretário Geral da AMB

Dr. Dioclécio Campos Júnior
Presidente da SBP

Dr. Eduardo da Silva Vaz
Secretário Geral da SBP

SERVIÇO PÚBLICO FEDERAL
Universidade Federal de São Paulo
Escola Paulista de Medicina

PG: 299/98

A T E S T A D O

Atesto para os devidos fins, que **LOURDES DE FATIMA GONÇALVES GOMES**, obteve o Título de Mestre, pelo curso de Pós-Graduação desta Universidade, na Área de **PEDIATRIA.**

São Paulo, 04 de Junho de 1998.

MARCIA MATTOS MARQUES
Diretora da Divisão de Pós-Graduação

Rua Botucatu, 740 - CEP 04023-900 - São Paulo / Brasil
Fone.: (55) (011) 576.4522 - FAX (55-11) 549.2127
E-MAIL: UNIFESP @ EPM.BR

LOURDES DE FÁTIMA GONÇALVES GOMES

AVALIAÇÃO IMUNOLÓGICA DE CRIANÇAS COM CARDIOPATIAS CONGÊNITAS

Tese apresentada à Universidade Federal de São Paulo - Escola Paulista de Medicina, para obtenção de Título de Mestre em Pediatria

SÃO PAULO
1995

UNIFESP

Universidade Federal de São Paulo
Escola Paulista de Medicina
Disciplina de Cardiologia

Setor de Ecocardiografia

Declaração

Declaramos, para os devidos fins, que a Dra. Lourdes Fátima Gonçalvez Gomes frequentou o Setor de Ecocardiografia Pediátrica do Serviço de Ecocardiografia da Disciplina de Cardiologia da Escola Paulista de Medicina – Unifesp, Hospital São Paulo, na qualidade de estagiário voluntário, tendo realizado e interpretados exames ecocardiográficos de crianças e adolescentes com cardiopatias congênitas e adquiridas, no período de março de 1999 a março de 2002.

São Paulo 29 de Outubro de 2004

Prof. Dr. Orlando Campos Filho
Chefe do Serviço de Ecocardiografia EPM-Unifesp

Dr. Valdir Ambrósio Moisés
Coordenador do Setor de Ecocardiografia Pediátrica EPM-Unifesp

Rua Pedro de Toledo, 650 2º andar– São Paulo – SP - CEP 04039-002 Fone: (011) 5576-4230 Fax : 5572-5462

Associação Médica Brasileira

Sociedade Brasileira de Pediatria

conferem à

Dra. Lourdes de Fátima Gonçalves Gomes

portadora do Título de Especialista em Pediatria o

CERTIFICADO DE ATUAÇÃO NA ÁREA DE NEONATOLOGIA

São Paulo, 8 de junho de 2002

Dr. Eleuses Vieira de Paiva
Presidente da AMB

Dr. Edmund Chada Baracat
Secretário Geral da AMB

Dr. Lincoln Marcelo Silveira Ivo
Presidente da SBP

Dr. Eduardo da Silva Vaz
Secretário Geral da SBP

Associação Médica Brasileira
Sociedade Brasileira de Pediatria
Sociedade Brasileira de Cardiologia

AMB
Associação Médica Brasileira

conferem à

Dra. Lourdes de Fátima Gonçalves Gomes

portadora do Título de Especialista em Pediatria o

CERTIFICADO DE ATUAÇÃO NA ÁREA DE CARDIOLOGIA PEDIÁTRICA

São Paulo, 25 de setembro de 2004

Dr. José Luiz Gomes do Amaral
Presidente da AMB

Dr. Aldemir Humberto Soares
Secretário Geral da AMB

Dr. Eduardo da Silva Vaz
Presidente da SBP

Dra. Marilene Augusta R. Crispino Santos
Secretária Geral da SBP

Dr. Jorge Ilha Guimarães
Presidente da SBC

Dr. Carlos Eleverson Lopes Pereira
Diretor Administrativo da SBC

Associação Médica Brasileira

Sociedade Brasileira de Cardiologia

AMB
Associação Médica Brasileira

SOCIEDADE BRASILEIRA DE CARDIOLOGIA
SBC
MCMXLIII

conferem à

Dra. Lourdes de Fatima Gonçalves Gomes

portadora do Título de Especialista em Pediatria o

CERTIFICADO DE ATUAÇÃO NA ÁREA DE ECOCARDIOGRAFIA

São Paulo, 18 de março de 2009

Dr. José Luiz Gomes do Amaral
Presidente da AMB

Dr. Aldemir Humberto Soares
Secretário Geral da AMB

Dr. Jorge Ilha Guimarães
Presidente da SBC

Dr. Carlos Cleverson Lopes Pereira
Diretor Administrativo da SBC

LOURDES DE FÁTIMA GONÇALVES GOMES

EXEQUIBILIDADE E SEGURANÇA DA ECOCARDIOGRAFIA COM CONTRASTE POR MICROBOLHAS EM CRIANÇAS E ADOLESCENTES COM CARDIOPATIA CONGÊNITA.

Tese apresentada à Universidade Federal de São Paulo - Escola Paulista de Medicina, para obtenção do Título de Doutor em Ciências.

SÃO PAULO

2012

UNIFESP

Departamento de Medicina
Disciplina de Cardiologia

DECLARAÇÃO

Declaro para os devidos fins que a Dra. Lourdes de Fátima Gonçalves Gomes, é aluna do Curso de Pós-Graduação em Cardiologia, nível Doutorado nesta Instituição, tendo iniciado suas atividades em fevereiro 1996, desenvolvendo sua Tese sob o tema "Ecocardiografia em Contraste em Cardiopatias Congênitas".

São Paulo, 15 de Junho de 1998

Prof. Dr. Antonio Carlos C. Carvalho
Professor Titular Coordenador do Curso de
Pós-Graduação em Cardiologia

UNIVERSIDADE FEDERAL DE UBERLÂNDIA
FACULDADE DE MEDICINA
COMISSÃO DE RESIDÊNCIA MÉDICA
Av. Pará, 1.720 – Bloco 2H – Sala 13 – Campus Umuarama
CEP 38.405-320 - UBERLÂNDIA – MG

ATESTADO

Atestamos para os devidos fins, que a Dra. ***LOURDES DE FÁTIMA GONÇALVES GOMES***, exerce a função de Supervisora do programa de Residência Médica em ***CARDIOLOGIA PEDIÁTRICA***, no Hospital de Clínicas da Universidade Federal de Uberlândia, desde novembro/2006 até a presente data. x.x.x.x.x.x.x.x.x.x.x.x.x.x.x.x.x.x.x.x.x.x.x.x.x.x.x.x.x.x.x.x.x.x.x.x.x.

Uberlândia, 24 de Maio de 2018.

Profa. Dra. Alessandra Carla de Almeida Ribeiro
Coordenadora Pró-Tempore da Comissão de Residência Médica

UNIVERSIDADE FEDERAL DE UBERLÂNDIA
Coordenação dos Programas de Residência Médica
Avenida Pará, 1720 – Bloco 2H – Sala 13 – Campus Umuarama
Uberlândia-MG - CEP 38405-320 - Telefone: (34) 3225-8626
e-mail: coreme@famed.ufu.br

A T E S T A D O

Atestamos para os devidos fins, que a Dra. ***LOURDES DE FÁTIMA GONÇALVES GOMES***, exerce a função de Preceptora do programa de Residência Médica em ***PEDIATRIA***, no Hospital de Clínicas da Universidade Federal de Uberlândia, no período de 14/08/2020 a 14/08/2022, perfazendo uma carga horária total de 04 horas semanais. x.x.x.x.x.x.

Uberlândia, 14 de Agosto de 2022.

Sinvaldo Gomes Oliveira
Assistente em Administração
Portaria REITO R Nº 618/2006 de 02/06/06

UNIVERSIDADE FEDERAL DE UBERLÂNDIA
Coordenação dos Programas de Residência Médica
Avenida Pará, 1720 – Bloco 2H – Sala 13 – Campus Umuarama
Uberlândia-MG - CEP 38405-320 - Telefone: (34) 3225-8626
e-mail: coreme@famed.ufu.br

A T E S T A D O

Atestamos para os devidos fins, que a Dra. ***LOURDES DE FÁTIMA GONÇALVES GOMES***, exerce a função de Preceptora do programa de Residência Médica em ***MEDICINA INTENSIVA PEDIÁTRICA***, no Hospital de Clínicas da Universidade Federal de Uberlândia, no período de 14/08/2020 a 14/08/2022, perfazendo uma carga horária total de 04 horas semanais. x.x.x.x.x.x.

Uberlândia, 14 de Agosto de 2022.

Sinvaldo Gomes Oliveira
Assistente em Administração
Portaria REITO R Nº 618/2006 de 02/06/06

SERVIÇO PÚBLICO FEDERAL
MINISTÉRIO DA EDUCAÇÃO
UNIVERSIDADE FEDERAL DE UBERLÂNDIA
HOSPITAL DE CLÍNICAS DE UBERLÂNDIA

Hospital de Clínicas
De Uberlândia

DECLARAÇÃO

Declaramos para os devidos fins de comprovação, que a Profª. Drª. LOURDES DE FÁTIMA GONÇALVES GOMES – SIAPE 21234060, docente ligado ao Departamento Acadêmico de Pediatria, desempenha a atividade de Chefe do Cardiologia Pediátrica do Hospital de Clínicas da Universidade Federal de Uberlândia, no período de Agosto/2014 até presente data.

Uberlândia, MG, 07 de JUNHO de 2018.

Prof. Dr. Eduardo Crosara Gustin
Diretor Geral
Hospital de Clínicas
Universidade Federal de Uberlândia

Boletim de Serviço Eletrônico em 30/11/2020

UNIVERSIDADE FEDERAL DE UBERLÂNDIA
Diretoria da Faculdade de Medicina
Av. Pará, 1720, Bloco 2U, Sala 23 - Bairro Umuarama, Uberlândia-MG, CEP 38400-902
Telefone: 34 3225-8604 - famed@ufu.br

PORTARIA DIRFAMED Nº 50, DE 30 DE NOVEMBRO DE 2020

Nomeação supervisores dos Programas de Residência Médica da Faculdade de Medicina da Universidade Federal de Uberlândia.

O DIRETOR DA FACULDADE DE MEDICINA DA UNIVERSIDADE FEDERAL DE UBERLÂNDIA, no uso das atribuições que lhe foram conferidas, e

CONSIDERANDO a RESOLUÇÃO SEI Nº 001/2018, DO DIRETORIA DA FACULDADE DE MEDICINA;

CONSIDERANDO a necessidade de atender à legislação vigente no que tange à Residência Médica no Brasil;

CONSIDERANDO o disposto na Resolução CNRM nº 02/2013, que determina a criação e atribuições da comissão de Residência Médica;

CONSIDERANDO que cada Programa de Residência Médica ficará sob a responsabilidade de um SUPERVISOR, que deve ser médico especialista de cada área de atuação, sendo indicados por seus pares e homologado pela COREME;

CONSIDERANDO os pedidos de substituição de supervisores do Programa de Residência Médica;

CONSIDERANDO o constante dos autos do processo nº 23117.025864/2017-89,

R E S O L V E :

Art. 1º Nomear os supervisores dos Programas de Residência Médica da Faculdade de Medicina da Universidade Federal de Uberlândia:

Programa de Residência Médica Alergia e Imunologia Pediátrica: Prof. Dr. Gesmar Rodrigues Silva Segundo;

Programa de Residência Médica Anestesiologia: Dr. Paulo Ricardo Rabello de Macedo Costa;

Programa de Residência Médica Cardiologia: Prof. Anderson Silveira Duque;

Programa de Residência Médica Cardiologia Pediátrica: Profa. Lourdes de Fátima Gonçalves Gomes;

Programa de Residência Médica Cirurgia Crânio Maxilo Facial: Lucas Gomes Patrocínio;

Programa de Residência Médica Cirurgia de Cabeça e Pescoço: Veruska Tavares Terra Martins da Silva;

Programa de Residência Médica Cirurgia do Aparelho Digestivo: João Bosco Chadú Júnior;

Programa de Residência Médica Cirurgia Geral: Prof. Cezar Augusto dos Santos;

Programa de Residência Médica Cirurgia Pediátrica: Bruna Pires Guerra de Andrade;

Programa de Residência Médica Cirurgia Plástica: Júlio Dante Bonetti;

Programa de Residência Médica Cirurgia Vascular: Profa. Laura de Andrade da Rocha;

Programa de Residência Médica Citopatologia: Olga Maria Lima Aguiar;

Programa de Residência Médica Clínica Médica: Profa. Juliana Markus;

Programa de Residência Médica Clínica Médica R3: Profa. Juliana Markus;

Programa de Residência Médica Coloproctologia: Renato Hugues Atique Cláudio;

Programa de Residência Médica Dermatologia: Profa. Renata Scarabucci Janones;

Programa de Residência Médica Ecocardiografia: Lívia Maria Ambrósio da Silva;

Programa de Residência Médica Endocrinologia e Metabologia: Sandra Regina Xavier Santos;

Programa de Residência Médica Endocrinologia Pediátrica: Débora Cristiane Gomes;

Programa de Residência Médica Gastroenterologia: Prof. Nestor Barbosa de Andrade;

Programa de Residência Médica Geriatria: Prof. Saadallah Azor Fakhouri Filho;

Programa de Residência Médica Infectologia: Prof. Marcelo Simão Ferreira;

Programa de Residência Médica Medicina de Família e Comunidade: Profa. Nicole Geovana Dias;

Programa de Residência Médica Medicina Intensiva: Ricardo Borges de Oliveira;

Programa de Residência Médica Medicina Intensiva Pediátrica: Alan de Paula;

Programa de Residência Médica Nefrologia: Prof. Marcus Vinícius de Pádua Netto;

Programa de Residência Médica Neonatologia: Profa. Daniela Marques de Lima Mota Ferreira;

Programa de Residência Médica Neurocirurgia: Prof. Paulo César Marinho Dias;

Programa de Residência Médica Neurologia: Marcos Campos;

Programa de Residência Médica Neurologia Pediátrica: Nívea de Macedo Oliveira Morales;

Programa de Residência Médica Obstetrícia e Ginecologia: Prof. Welington Ued Naves;

Programa de Residência Médica Oftalmologia: Prof. Flávio Jaime da Rocha;

Programa de Residência Médica Oncologia Clínica: Prof. Rogério Agenor de Araújo;

Programa de Residência Médica Oncologia Pediátrica: Iêda Cristina Cunha Ferreira e Fonseca;

Programa de Residência Médica Ortopedia e Traumatologia: Cleber Jesus Pereira;

Programa de Residência Médica Ortopedia e Traumatologia R4: Cleber Jesus Pereira;

Programa de Residência Médica Otorrinolaringologia: Prof. Lucas Gomes Patrocínio;

Programa de Residência Médica Patologia: Olga Maria Lima Aguiar;

Programa de Residência Médica Pediatria: Profa. Carolina Pirtouscheg;

Programa de Residência Médica Psiquiatria: Prof. Ricardo José Victal de Carvalho;

Programa de Residência Médica Radiologia e Diagnóstico por Imagem: Prof. Túlio Augusto Alves Macedo;

Programa de Residência Médica Radioterapia: Eurípedes Rodrigues Barra;

Programa de Residência Médica Reumatologia: Roberto Ranza;

Programa de Residência Médica Urologia: Prof. Omar Pacheco Simão;

Art. 2º Compete ao supervisor do Programa de Residência Médica:

I. Coordenar, organizar e supervisionar a implantação do Programa de Residência em conformidade com a legislação;

II. Manter atualizadas as fichas dos residentes e todas as normas e resoluções emanadas pelos respectivos Conselhos Nacionais;

III. Zelar pelo bom andamento das atividades práticas e didáticas;

IV. Aplicar a avaliação de cada residente, a partir dos critérios estabelecidos;

V. Participar das reuniões da COREME, sempre que convocado;

VI. Fazer cumprir todas as determinações provenientes dos respectivos Conselhos Nacionais e locais;

VII. Verificar junto aos preceptores o resultado da avaliação individual dos residentes sob sua responsabilidade ao final de cada estágio;

VIII. Elaborar escalas de plantão e férias no início de cada ano do PRM;

IX. Elaborar, anualmente, o Programa de Residência Médica em sua especialidade;

X. Promover a integração dos residentes com a equipe de saúde, usuários (indivíduos, família e grupos) e demais serviços;

XI. Deliberar quanto a licenças e afastamentos solicitados por residentes, que só podem ser concedidos se de acordo com as normas da COREME;

XII. Avaliar e tomar providências cabíveis em relação a eventuais faltas cometidas por residentes ou preceptores, que comprometam o bom funcionamento dos Programas de Residência, resguardados os direitos e as atribuições dos coordenadores dos serviços do hospital;

XIII. Encaminhar a sua respectiva Comissão as faltas de maior gravidade;

XIV. Encaminhar ao Coordenador Geral da COREME:

a. Frequência mensal dos residentes;

b. Os casos de cancelamento da Bolsa de Residência em tempo hábil;

c. A relação anual de residentes com as respectivas férias;

d. A avaliação de aprendizado trimestral individual de cada residente de acordo com a área;

e. As solicitações quanto as questões disciplinares; f. Os pedidos de licença para afastamento dos residentes.

Art. 3º Esta Portaria revoga a PORTARIA DIRFAMED Nº 22, DE 12 DE MARÇO DE 2020 e entra em vigor na data de sua publicação no Boletim Eletrônico.

CARLOS HENRIQUE MARTINS DA SILVA
Diretor da Faculdade de Medicina
Portaria nº 1.464/17

Documento assinado eletronicamente por **Carlos Henrique Martins da Silva**, **Diretor(a)**, em 30/11/2020, às 19:25, conforme horário oficial de Brasília, com fundamento no art. 6º, § 1º, do Decreto nº 8.539, de 8 de outubro de 2015.

A autenticidade deste documento pode ser conferida no site https://www.sei.ufu.br/sei/controlador_externo.php?acao=documento_conferir&id_orgao_acesso_externo=0, informando o código verificador **2422159** e o código CRC **A7EC6A46**.

Referência: Processo nº 23117.025864/2017-89 SEI nº 2422159

Criado por lorena.matos, versão 7 por lorena.matos em 30/11/2020 18:44:42.

Boletim de Serviço Eletrônico em 15/03/2021

UNIVERSIDADE FEDERAL DE UBERLÂNDIA
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Av. Pará, 1720, Bloco 2U, Sala 23 - Bairro Umuarama, Uberlândia-MG, CEP 38400-902
Telefone: 34 3225-8604 - famed@ufu.br

PORTARIA DIRFAMED Nº 18, DE 15 DE MARÇO DE 2021

Nomeação supervisores dos Programas de Residência Médica da Faculdade de Medicina da Universidade Federal de Uberlândia.

A DIRETORA *PRO TEMPORE* DA FACULDADE DE MEDICINA DA UNIVERSIDADE FEDERAL DE UBERLÂNDIA, no uso das atribuições que lhe foram conferidas, e

CONSIDERANDO a RESOLUÇÃO SEI Nº 001/2018, DO DIRETORIA DA FACULDADE DE MEDICINA;

CONSIDERANDO a necessidade de atender à legislação vigente no que tange à Residência Médica no Brasil;

CONSIDERANDO o disposto na Resolução CNRM nº 02/2013, que determina a criação e atribuições da comissão de Residência Médica;

CONSIDERANDO que cada Programa de Residência Médica ficará sob a responsabilidade de um SUPERVISOR, que deve ser médico especialista de cada área de atuação, sendo indicados por seus pares e homologado pela COREME;

CONSIDERANDO os pedidos de substituição de supervisores do Programa de Residência Médica;

CONSIDERANDO o constante dos autos do processo nº 23117.025864/2017-89,

R E S O L V E :

Art. 1º Nomear os supervisores dos Programas de Residência Médica da Faculdade de Medicina da Universidade Federal de Uberlândia:

Programa de Residência Médica Alergia e Imunologia Pediátrica: Prof. Dr. Gesmar Rodrigues Silva Segundo;

Programa de Residência Médica Anestesiologia: Dr. Paulo Ricardo Rabello de Macedo Costa;

Programa de Residência Médica Cardiologia: Prof. Anderson Silveira Duque;

Programa de Residência Médica Cardiologia Pediátrica: Profa. Lourdes de Fátima Gonçalves Gomes;

Programa de Residência Médica Cirurgia Crânio Maxilo Facial: Lucas Gomes Patrocínio;

Programa de Residência Médica Cirurgia de Cabeça e Pescoço: Veruska Tavares Terra Martins da Silva;

Programa de Residência Médica Cirurgia do Aparelho Digestivo: João Bosco Chadú Júnior;

Programa de Residência Médica Cirurgia Geral: Prof. Cezar Augusto dos Santos;

Programa de Residência Médica Cirurgia Pediátrica: Bruna Pires Guerra de Andrade;

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Programa de Residência Médica Clínica Médica: Profa. Juliana Markus;

Programa de Residência Médica Clínica Médica R3: Profa. Juliana Markus;

Programa de Residência Médica Coloproctologia: Renato Hugues Atique Cláudio;

Programa de Residência Médica Dermatologia: Profa. Renata Scarabucci Janones;

Programa de Residência Médica Ecocardiografia: Lívia Maria Ambrósio da Silva;

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Programa de Residência Médica Endocrinologia Pediátrica: Débora Cristiane Gomes;

Programa de Residência Médica Gastroenterologia: Prof. Nestor Barbosa de Andrade;

Programa de Residência Médica Geriatria: Prof. Saadallah Azor Fakhouri Filho;

Programa de Residência Médica Infectologia: Prof. Marcelo Simão Ferreira;

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Programa de Residência Médica Obstetrícia e Ginecologia: Prof. Welington Ued Naves;

Programa de Residência Médica Oftalmologia: Prof. Flávio Jaime da Rocha;

Programa de Residência Médica Oncologia Clínica: Prof. Rogério Agenor de Araújo;

Programa de Residência Médica Oncologia Pediátrica: Iêda Cristina Cunha Ferreira e Fonseca;

Programa de Residência Médica Ortopedia e Traumatologia: Cleber Jesus Pereira;

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Programa de Residência Médica Patologia: Olga Maria Lima Aguiar;

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Programa de Residência Médica Psiquiatria: Prof. Ricardo José Victal de Carvalho;

Programa de Residência Médica Radiologia e Diagnóstico por Imagem: Prof. Túlio Augusto Alves Macedo;

Programa de Residência Médica Radioterapia: Eurípedes Rodrigues Barra;

Programa de Residência Médica Reumatologia: Roberto Ranza;

Programa de Residência Médica Urologia: Prof. Omar Pacheco Simão;

Art. 2º Compete ao supervisor do Programa de Residência Médica:

I. Coordenar, organizar e supervisionar a implantação do Programa de Residência em conformidade com a legislação;

II. Manter atualizadas as fichas dos residentes e todas as normas e resoluções emanadas pelos respectivos Conselhos Nacionais;

III. Zelar pelo bom andamento das atividades práticas e didáticas;

IV. Aplicar a avaliação de cada residente, a partir dos critérios estabelecidos;

V. Participar das reuniões da COREME, sempre que convocado;

VI. Fazer cumprir todas as determinações provenientes dos respectivos Conselhos Nacionais e locais;

VII. Verificar junto aos preceptores o resultado da avaliação individual dos residentes sob sua responsabilidade ao final de cada estágio;

VIII. Elaborar escalas de plantão e férias no início de cada ano do PRM;

IX. Elaborar, anualmente, o Programa de Residência Médica em sua especialidade;

X. Promover a integração dos residentes com a equipe de saúde, usuários (indivíduos, família e grupos) e demais serviços;

XI. Deliberar quanto a licenças e afastamentos solicitados por residentes, que só podem ser concedidos se de acordo com as normas da COREME;

XII. Avaliar e tomar providências cabíveis em relação a eventuais faltas cometidas por residentes ou preceptores, que comprometam o bom funcionamento dos Programas de Residência, resguardados os direitos e as atribuições dos coordenadores dos serviços do hospital;

XIII. Encaminhar a sua respectiva Comissão as faltas de maior gravidade;

XIV. Encaminhar ao Coordenador Geral da COREME:

a. Frequência mensal dos residentes;

b. Os casos de cancelamento da Bolsa de Residência em tempo hábil;

c. A relação anual de residentes com as respectivas férias;

d. A avaliação de aprendizado trimestral individual de cada residente de acordo com a área;

e. As solicitações quanto as questões disciplinares; f. Os pedidos de licença para afastamento dos residentes.

Art. 3º Esta Portaria revoga a PORTARIA DIRFAMED Nº 50, DE 30 DE NOVEMBRO DE 2020 e entra em vigor na data de sua publicação no Boletim Eletrônico.

CATARINA MACHADO AZEREDO
Diretora *Pro Tempore* da Faculdade de Medicina
Portaria de Pessoal UFU Nº 675/2021

Documento assinado eletronicamente por **Catarina Machado Azeredo**, **Diretor(a)**, em 15/03/2021, às 08:24, conforme horário oficial de Brasília, com fundamento no art. 6º, § 1º, do Decreto nº 8.539, de 8 de outubro de 2015.

A autenticidade deste documento pode ser conferida no site

https://www.sei.ufu.br/sei/controlador_externo.php?acao=documento_conferir&id_orgao_acesso_externo=0, informando o código verificador **2637209** e o código CRC **20A6400D**.

Referência: Processo nº 23117.025864/2017-89 SEI nº 2637209

Criado por ana.sena, versão 7 por ana.sena em 15/03/2021 08:17:49.

Boletim de Serviço Eletrônico em 21/10/2021

UNIVERSIDADE FEDERAL DE UBERLÂNDIA
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Telefone: 34 3225-8604 - famed@ufu.br

PORTARIA DIRFAMED Nº 68, DE 20 DE OUTUBRO DE 2021

Nomeação supervisores dos Programas de Residência Médica da Faculdade de Medicina da Universidade Federal de Uberlândia.

A DIRETORA DA FACULDADE DE MEDICINA DA UNIVERSIDADE FEDERAL DE UBERLÂNDIA, no uso das atribuições que lhe foram conferidas, e

CONSIDERANDO a RESOLUÇÃO SEI Nº 001/2018, DO DIRETORIA DA FACULDADE DE MEDICINA;

CONSIDERANDO a necessidade de atender à legislação vigente no que tange à Residência Médica no Brasil;

CONSIDERANDO o disposto na Resolução CNRM nº 02/2013, que determina a criação e atribuições da comissão de Residência Médica;

CONSIDERANDO que cada Programa de Residência Médica ficará sob a responsabilidade de um SUPERVISOR, que deve ser médico especialista de cada área de atuação, sendo indicados por seus pares e homologado pela COREME;

CONSIDERANDO os pedidos de substituição de supervisores do Programa de Residência Médica;

CONSIDERANDO o constante dos autos do processo nº 23117.025864/2017-89,

R E S O L V E :

Art. 1º Nomear os supervisores dos Programas de Residência Médica da Faculdade de Medicina da Universidade Federal de Uberlândia:

Programa de Residência Médica Alergia e Imunologia Pediátrica: Prof. Dr. Gesmar Rodrigues Silva Segundo;

Programa de Residência Médica Anestesiologia: Dr. Paulo Ricardo Rabello de Macedo Costa;

Programa de Residência Médica Cardiologia: Prof. Anderson Silveira Duque;

Programa de Residência Médica Cardiologia Pediátrica: Profa. Lourdes de Fátima Gonçalves Gomes;

Programa de Residência Médica Cirurgia Crânio Maxilo Facial: Lucas Gomes Patrocínio;

Programa de Residência Médica Cirurgia de Cabeça e Pescoço: Veruska Tavares Terra Martins da Silva;

Programa de Residência Médica Cirurgia do Aparelho Digestivo: João Bosco Chadú Júnior;

Programa de Residência Médica Cirurgia Geral: Prof. Cezar Augusto dos Santos;

Programa de Residência Médica Cirurgia Pediátrica: Bruna Pires Guerra de Andrade;

Programa de Residência Médica Cirurgia Plástica: Júlio Dante Bonetti;

Programa de Residência Médica Cirurgia Vascular: Profa. Laura de Andrade da Rocha;

Programa de Residência Médica Citopatologia: Olga Maria Lima Aguiar;

Programa de Residência Médica Clínica Médica: Profa. Juliana Markus;

Programa de Residência Médica Clínica Médica R3: Profa. Juliana Markus;

Programa de Residência Médica Coloproctologia: Renato Hugues Atique Cláudio;

Programa de Residência Médica Dermatologia: Profa. Renata Scarabucci Janones;

Programa de Residência Médica Ecocardiografia: Lívia Maria Ambrósio da Silva;

Programa de Residência Médica Endocrinologia e Metabologia: Sandra Regina Xavier Santos;

Programa de Residência Médica Endocrinologia Pediátrica: Débora Cristiane Gomes;

Programa de Residência Médica Gastroenterologia: Prof. Nestor Barbosa de Andrade;

Programa de Residência Médica Geriatria: Prof. Saadallah Azor Fakhouri Filho;

Programa de Residência Médica Infectologia: Prof. Marcelo Simão Ferreira;

Programa de Residência Médica Medicina de Família e Comunidade: Profa. Nicole Geovana Dias;

Programa de Residência Médica Medicina Intensiva: Ricardo Borges de Oliveira;

Programa de Residência Médica Medicina Intensiva Pediátrica: Alan de Paula;

Programa de Residência Médica Nefrologia: Prof. Marcus Vinícius de Pádua Netto;

Programa de Residência Médica Neonatologia: Profa. Daniela Marques de Lima Mota Ferreira;

Programa de Residência Médica Neurocirurgia: Prof. Paulo César Marinho Dias;

Programa de Residência Médica Neurologia: Marcos Campos;

Programa de Residência Médica Neurologia Pediátrica: Nívea de Macedo Oliveira Morales;

Programa de Residência Médica Obstetrícia e Ginecologia: Prof. Francisco Cyro Reis de Campos Prado Filho;

Programa de Residência Médica Oftalmologia: Prof. Flávio Jaime da Rocha;

Programa de Residência Médica Oncologia Clínica: Prof. Rogério Agenor de Araújo;

Programa de Residência Médica Oncologia Pediátrica: Iêda Cristina Cunha Ferreira e Fonseca;

Programa de Residência Médica Ortopedia e Traumatologia: Cleber Jesus Pereira;

Programa de Residência Médica Ortopedia e Traumatologia R4: Cleber Jesus Pereira;

Programa de Residência Médica Otorrinolaringologia: Valmir Tunala Júnior;

Programa de Residência Médica Patologia: Olga Maria Lima Aguiar;

Programa de Residência Médica Pediatria: Profa. Carolina Pirtouscheg;

Programa de Residência Médica Psiquiatria: Prof. Ricardo José Victal de Carvalho;

Programa de Residência Médica Radiologia e Diagnóstico por Imagem: Prof. Túlio Augusto Alves Macedo;

Programa de Residência Médica Radioterapia: Eurípedes Rodrigues Barra;

Programa de Residência Médica Reumatologia: Roberto Ranza;

Programa de Residência Médica Urologia: Prof. Omar Pacheco Simão;

Art. 2º Compete ao supervisor do Programa de Residência Médica:

I. Coordenar, organizar e supervisionar a implantação do Programa de Residência em conformidade com a legislação;

II. Manter atualizadas as fichas dos residentes e todas as normas e resoluções emanadas pelos respectivos Conselhos Nacionais;

III. Zelar pelo bom andamento das atividades práticas e didáticas;

IV. Aplicar a avaliação de cada residente, a partir dos critérios estabelecidos;

V. Participar das reuniões da COREME, sempre que convocado;

VI. Fazer cumprir todas as determinações provenientes dos respectivos Conselhos Nacionais e locais;

VII. Verificar junto aos preceptores o resultado da avaliação individual dos residentes sob sua responsabilidade ao final de cada estágio;

VIII. Elaborar escalas de plantão e férias no início de cada ano do PRM;

IX. Elaborar, anualmente, o Programa de Residência Médica em sua especialidade;

X. Promover a integração dos residentes com a equipe de saúde, usuários (indivíduos, família e grupos) e demais serviços;

XI. Deliberar quanto a licenças e afastamentos solicitados por residentes, que só podem ser concedidos se de acordo com as normas da COREME;

XII. Avaliar e tomar providências cabíveis em relação a eventuais faltas cometidas por residentes ou preceptores, que comprometam o bom funcionamento dos Programas de Residência, resguardados os direitos e as atribuições dos coordenadores dos serviços do hospital;

XIII. Encaminhar a sua respectiva Comissão as faltas de maior gravidade;

XIV. Encaminhar ao Coordenador Geral da COREME:

a. Frequência mensal dos residentes;

b. Os casos de cancelamento da Bolsa de Residência em tempo hábil;

c. A relação anual de residentes com as respectivas férias;

d. A avaliação de aprendizado trimestral individual de cada residente de acordo com a área;

e. As solicitações quanto as questões disciplinares;

f. Os pedidos de licença para afastamento dos residentes.

Art. 3º Esta Portaria revoga a PORTARIA DIRFAMED Nº 18, DE 15 DE MARÇO DE 2021 e entra em vigor na data de sua publicação no Boletim Eletrônico.

CATARINA MACHADO AZEREDO
Diretora da Faculdade de Medicina
Portaria de Pessoal UFU Nº 3005/2021

Documento assinado eletronicamente por **Catarina Machado Azeredo**, **Diretor(a)**, em 21/10/2021, às 09:10, conforme horário oficial de Brasília, com fundamento no art. 6º, § 1º, do Decreto nº 8.539, de 8 de outubro de 2015.

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Referência: Processo nº 23117.025864/2017-89 SEI nº 3116715

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Boletim de Serviço Eletrônico em 14/02/2022

UNIVERSIDADE FEDERAL DE UBERLÂNDIA
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PORTARIA DE PESSOAL UFU Nº 616, DE 11 DE FEVEREIRO DE 2022

Nomeação supervisores dos Programas de Residência Médica da Faculdade de Medicina da Universidade Federal de Uberlândia.

A DIRETORA DA FACULDADE DE MEDICINA DA UNIVERSIDADE FEDERAL DE UBERLÂNDIA, no uso das atribuições que lhe foram conferidas, e

CONSIDERANDO a RESOLUÇÃO SEI Nº 001/2018, DO DIRETORIA DA FACULDADE DE MEDICINA;

CONSIDERANDO a necessidade de atender à legislação vigente no que tange à Residência Médica no Brasil;

CONSIDERANDO o disposto na Resolução CNRM nº 02/2013, que determina a criação e atribuições da comissão de Residência Médica;

CONSIDERANDO que cada Programa de Residência Médica ficará sob a responsabilidade de um SUPERVISOR, que deve ser médico especialista de cada área de atuação, sendo indicados por seus pares e homologado pela COREME;

CONSIDERANDO os pedidos de substituição de supervisores do Programa de Residência Médica;

CONSIDERANDO o constante dos autos do processo nº 23117.025864/2017-89,

R E S O L V E :

Art. 1º Nomear os supervisores dos Programas de Residência Médica da Faculdade de Medicina da Universidade Federal de Uberlândia:

Programa de Residência Médica Alergia e Imunologia Pediátrica: Prof. Dr. Gesmar Rodrigues Silva Segundo;

Programa de Residência Médica Anestesiologia: Dr. Roberto Araújo Ruzi;

Programa de Residência Médica Cardiologia: Prof. Anderson Silveira Duque;

Programa de Residência Médica Cardiologia Pediátrica: Profa. Lourdes de Fátima Gonçalves Gomes;

Programa de Residência Médica Cirurgia Crânio Maxilo Facial: Lucas Gomes Patrocínio;

Programa de Residência Médica Cirurgia de Cabeça e Pescoço: Veruska Tavares Terra Martins da Silva;

Programa de Residência Médica Cirurgia do Aparelho Digestivo: João Bosco Chadú Júnior;

Programa de Residência Médica Cirurgia Geral: Prof. Cezar Augusto dos Santos;

Programa de Residência Médica Cirurgia Pediátrica: Bruna Pires Guerra de Andrade;

Programa de Residência Médica Cirurgia Plástica: Adriana Santa Cecília Borges;

Programa de Residência Médica Cirurgia Vascular: Profa. Laura de Andrade da Rocha;

Programa de Residência Médica Citopatologia: Fernando Costa Mundim;

Programa de Residência Médica Clínica Médica: Prof. Eduardo Crosara Gustin;

Programa de Residência Médica Clínica Médica R3: Prof. Eduardo Crosara Gustin;

Programa de Residência Médica Coloproctologia: Renato Hugues Atique Cláudio;

Programa de Residência Médica Dermatologia: Profa. Mabel Duarte Alves Gomides;

Programa de Residência Médica Ecocardiografia: Lívia Maria Ambrósio da Silva;

Programa de Residência Médica Endocrinologia e Metabologia: Sandra Regina Xavier Santos;

Programa de Residência Médica Endocrinologia Pediátrica: Débora Cristiane Gomes;

Programa de Residência Médica Gastroenterologia: Prof. Nestor Barbosa de Andrade;

Programa de Residência Médica Gastroenterologia Pediátrica: Profa. Érica Rodrigues Mariano Almeida Rezende;

Programa de Residência Médica Geriatria: Prof. Saadallah Azor Fakhouri Filho;

Programa de Residência Médica Infectologia: Prof. Marcelo Simão Ferreira;

Programa de Residência Médica Medicina de Família e Comunidade: Profa. Nicole Geovana Dias;

Programa de Residência Médica Medicina Intensiva: Ricardo Borges de Oliveira;

Programa de Residência Médica Medicina Intensiva Pediátrica: Alan de Paula;

Programa de Residência Médica Nefrologia: Prof. Marcus Vinícius de Pádua Netto;

Programa de Residência Médica Nefrologia Pediátrica: Yara Aparecida Cunha Ferreira Zuza;

Programa de Residência Médica Neonatologia: Profa. Daniela Marques de Lima Mota Ferreira;

Programa de Residência Médica Neurocirurgia: Prof. Paulo César Marinho Dias;

Programa de Residência Médica Neurologia: Prof. Diogo Fernandes dos Santos;

Programa de Residência Médica Neurologia Pediátrica: Nívea de Macedo Oliveira Morales;

Programa de Residência Médica Obstetrícia e Ginecologia: Prof. Francisco Cyro Reis de Campos Prado Filho;

Programa de Residência Médica Oftalmologia: Prof. Flávio Jaime da Rocha;

Programa de Residência Médica Oncologia Clínica: Prof. Rogério Agenor de Araújo;

Programa de Residência Médica Oncologia Pediátrica: Iêda Cristina Cunha Ferreira e Fonseca;

Programa de Residência Médica Ortopedia e Traumatologia: Cleber Jesus Pereira;

Programa de Residência Médica Ortopedia e Traumatologia R4: Cleber Jesus Pereira;

Programa de Residência Médica Otorrinolaringologia: Valmir Tunala Júnior;

Programa de Residência Médica Patologia: Fernando Costa Mundim;

Programa de Residência Médica Pediatria: Profa. Tatyana Borges da Cunha Kock;

Programa de Residência Médica Psiquiatria: Prof. Ricardo José Victal de Carvalho;

Programa de Residência Médica Radiologia e Diagnóstico por Imagem: Prof. Túlio Augusto Alves Macedo;

Programa de Residência Médica Radioterapia: Eurípedes Rodrigues Barra;

Programa de Residência Médica Reumatologia: Roberto Ranza;

Programa de Residência Médica Urologia: Prof. Omar Pacheco Simão;

Art. 2º Compete ao supervisor do Programa de Residência Médica:

I. Coordenar, organizar e supervisionar a implantação do Programa de Residência em conformidade com a legislação;

II. Manter atualizadas as fichas dos residentes e todas as normas e resoluções emanadas pelos respectivos Conselhos Nacionais;

III. Zelar pelo bom andamento das atividades práticas e didáticas;

IV. Aplicar a avaliação de cada residente, a partir dos critérios estabelecidos;

V. Participar das reuniões da COREME, sempre que convocado;

VI. Fazer cumprir todas as determinações provenientes dos respectivos Conselhos Nacionais e locais;

VII. Verificar junto aos preceptores o resultado da avaliação individual dos residentes sob sua responsabilidade ao final de cada estágio;

VIII. Elaborar escalas de plantão e férias no início de cada ano do PRM;

IX. Elaborar, anualmente, o Programa de Residência Médica em sua especialidade;

X. Promover a integração dos residentes com a equipe de saúde, usuários (indivíduos, família e grupos) e demais serviços;

XI. Deliberar quanto a licenças e afastamentos solicitados por residentes, que só podem ser concedidos se de acordo com as normas da COREME;

XII. Avaliar e tomar providências cabíveis em relação a eventuais faltas cometidas por residentes ou preceptores, que comprometam o bom funcionamento dos Programas de Residência, resguardados os direitos e as atribuições dos coordenadores dos serviços do hospital;

XIII. Encaminhar a sua respectiva Comissão as faltas de maior gravidade;

XIV. Encaminhar ao Coordenador Geral da COREME:

a. Frequência mensal dos residentes;

b. Os casos de cancelamento da Bolsa de Residência em tempo hábil;

c. A relação anual de residentes com as respectivas férias;

d. A avaliação de aprendizado trimestral individual de cada residente de acordo com a área;

e. As solicitações quanto as questões disciplinares;

f. Os pedidos de licença para afastamento dos residentes.

Art. 3º Esta Portaria revoga a PORTARIA DE PESSOAL UFU Nº 588, DE 09 DE FEVEREIRO DE 2022 e entra em vigor na data de sua publicação no Boletim Eletrônico.

CATARINA MACHADO AZEREDO
Diretora da Faculdade de Medicina

Portaria de Pessoal UFU nº 3005/2021

Documento assinado eletronicamente por **Catarina Machado Azeredo**, **Diretor(a)**, em 11/02/2022, às 17:27, conforme horário oficial de Brasília, com fundamento no art. 6º, § 1º, do Decreto nº 8.539, de 8 de outubro de 2015.

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Referência: Processo nº 23117.025864/2017-89 SEI nº 3370428

UNIDADE OFERECE ATENDIMENTO PARA PACIENTES DE DIVERSAS PARTES O PAÍS

HC-UFU é referência em tratamento de cardiopatia congênita

Publicado em 10/06/2021 09h26 Atualizado em 10/06/2021 13h06

O "Dia Nacional de Conscientização da Cardiopatia Congênita", celebrado em 12 de junho, alertar a população e os profissionais da saúde sobre a gravidade da doença e a importância do diagnóstico precoce para o atendimento mais adequado.

De acordo com os especialistas, a cardiopatia congênita é a anormalidade na estrutura ou função do c

O Hospital de Clínicas da Universidade Federal de Uberlândia (HC-UFU/Ebserh) é referência em Cardiologia Pediátrica e tem um Ambulatório de Cardiopatia Congênita que oferece atendimento clínico, exames diagnósticos, cirurgias cardíacas e intervenções hemodinâmicas. O Ambulatório existe há mais de 30 anos e recebe pacientes de diversas localidades do Brasil, o que representa em média mais de três mil pacientes atendidos por ano. “A estrutura terciária para atendimento de cardiopatia congênita com hemodinâmica, exames de imagem e cirurgia cardíaca, em Minas Gerais, está disponível no HC-UFU e em Belo Horizonte”, destaca a cardiologista pediátrica e coordenadora do Ambulatório, Lourdes de Fátima Gonçalves Gomes.

A coordenadora ressalta a importância do trabalho multidisciplinar no atendimento aos pacientes com cardiopatia congênita. “Contamos com o apoio de diversos setores do hospital como o Pronto Socorro e a Enfermaria de Pediatria, a Unidade de Neonatologia, o Serviço de Hemodinâmica, o Serviço de Cirurgia Cardíaca Pediátrico e Adulto, além do Ambulatório de Ginecologia Fetal. É a atuação de toda equipe multidisciplinar que possibilita que o HC-UFU seja referência nesta área”.

Alcides Gil de Souza Neto, 15 anos, foi paciente do Ambulatório de Cardiopatia Congênita por oito anos. Foi encaminhado ao HC-UFU, aos 4 anos de idade, com falta de ar e desmaio. Fez acompanhamento com a equipe e há três anos recebeu alta. “Desde a primeira consulta a assistência foi excelente. Tivemos atendimento médico, alimentação, até brinquedoteca para o meu filho, não nos faltou nada. Só tenho a agradecer”, ressalta José Gilberto Figueiredo Gil, pai do adolescente.

Como forma de agradecimento o pai é voluntário no Programa Amigos do Coração uma iniciativa da gestão de Programas Institucionais de Humanização do HC-UFU, com objetivo de oferecer apoio as crianças e aos familiares, na tentativa de minimizar qualquer desconforto e proporcionar maior aderência ao tratamento. “As crianças esperavam pelo atendimento nos corredores do ambulatório e como tínhamos este espaço externo buscamos parceria e criamos o “Espaço da Criança”, mais conhecido como “Quiosque do Ambulatório de Pediatria” onde, além de brincadeiras, os pacientes podem realizar tarefas escolares com apoio de estagiários”, explica a coordenadora do Programa, Lêda Márcia Viana Santos Borges.

Em parceria com a Pró-Reitoria de Extensão de Cultura da Universidade Federal de Uberlândia (Proex-UFU), o Programa conta com a participação de estudantes de diversos cursos que realizam projetos de extensão. “Meu Dentinho, Meu Coração” é um deles. Os pacientes são avaliados pela equipe da Faculdade de Odontologia e caso precisem de tratamento são encaminhados para o Hospital Odontológico.

Formação

Além da assistência, o HC-UFU forma também especialista na área de cardiopatia congênita com o curso de residência em Cardiologia Pediátrica. A meta, segundo a coordenadora do Ambulatório, Lourdes Gomes, é ampliar a quantidade de cursos, com a criação, por exemplo, da residência em Ecocardiografia em

este problema de saúde pública", destaca.

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DECLARAÇÃO

Processo nº 23117.036866/2018-84

Interessado: Lourdes de Fátima Gonçalves Gomes

Declaramos, para os devidos fins de comprovação e anexação ao relatório de atividades da Progressão Horizontal Associado nível 2 para Associado Nível 3 na carreira docente do magistério superior da UFU, à luz da Resolução 03/2017 do Conselho Diretor, que a Prof.ª Dr.ª Lourdes de Fátima Gonçalves Gomes – SIAPE 2123460, docente ligada ao Departamento Acadêmico de Pediatria, ministrou as cargas horárias nos semestres requisitados, referentes ao interstício de 14/08/2016 a 14/08/2018, junto às disciplinas e estágios de graduação, explicitadas abaixo:

Semestre/ Ano	Data Início e Término	Disciplinas	Hora aula Semanal	Total de Semanas letivas ministradas
2016/2	08/08/2016 a 17/12/2016	FAMED31603 - Medicina Integrada III	04	18 semanas
2017/1	03/04/2017 a 03/08/2017	FAMED31603 - Medicina Integrada III	04	18 semanas
2017/2	21/08/2017 a 22/12/2017	FAMED31603 - Medicina Integrada III	04	18 semanas
2018/1	12/03/2018 a 14/07/2018	FAMED31603 - Medicina Integrada III	04	18 semanas

Semestre	Data Início e Término	Estágio Supervisionado	Hora aula Semanal	Nr. Alunos Orientados	Semanas ministradas
2016/2	04/07/2016 a 25/12/2016 03/10/2016 a 26/03/2017	FAMED31902 – Estágio Supervisionado na área Materno-Infantil	04	10	24 semanas
2017/1	02/01/2017 a 25/06/2017	FAMED31902 – Estágio Supervisionado na área Materno-Infantil	08	10	24 semanas
2017/2	03/07/2017 a 24/12/2017	FAMED31902 – Estágio Supervisionado na área Materno-Infantil FAMED31901 – Estágio Supervisionado na área Materno-Infantil	08	10	24 semanas
2018/1	08/01/2018 a 01/07/2018	FAMED31901 – Estágio Supervisionado na área Materno-Infantil	08	10	24 semanas

Prof.ª Dr.ª Rosângela Martins de Araújo

Coordenadora do Curso de Graduação em Medicina - FAMED/UFU

Portaria R N.º 1461/2017

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Referência: Processo nº 23117.036866/2018-84 SEI nº 0514032

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DECLARAÇÃO

Processo nº 23117.030655/2020-52

Interessado: Lourdes de Fátima Gonçalves Gomes

A COORDENADORA DO CURSO DE GRADUAÇÃO EM MEDICINA DA UNIVERSIDADE FEDERAL DE UBERLÂNDIA, Prof.ª Dr.ª **Rosângela Martins de Araújo**, no uso de suas atribuições legais e regulamentares, **DECLARA,** para fins de comprovação e anexação ao relatório de atividades para a Progressão na carreira docente do magistério superior da UFU, que a **Prof.ª Dr.ª LOURDES DE FÁTIMA GONÇALVES GOMES**, SIAPE 2123460, ligada ao Departamento Acadêmico de Pediatria, ministrou as cargas horárias nos semestres citados abaixo, referentes ao interstício de **14/08/2018 a 14/08/2020**.

Declara, ainda, que a referida professora não foi submetida, formalmente, à avaliação discente e que, até o momento, não consta nesta Coordenação de Curso qualquer registro que a desabone, desempenhando as suas atividades acadêmicas com assiduidade, disciplina, produtividade e responsabilidade.

SEMESTRE	DATA DE INÍCIO E TÉRMINO	COMPONENTES CURRICULARES	HORA-AULA SEMANAL	TOTAL DE SEMANAS LETIVAS MINISTRADAS	NÚMERO DE ESTUDANTES
2018/2	14/08/2018 a 22/12/2018	FAMED31603 - Medicina Integrada III	4,0	18 semanas	54
	14/08/2018 a 30/12/2018	FAMED31901 - Estágio Supervisionado na Área Materno-Infantil: Pediatria	9,3	19 semanas	39
2019/1	11/03/2019 a 13/07/2019	FAMED31603 - Medicina Integrada III	4,0	18 semanas	67
	07/01/2019 a 30/06/2019	FAMED31901 - Estágio Supervisionado na Área Materno-Infantil: Pediatria	9,3	24 semanas	38
2019/2	12/08/2019 a 21/12/2019	FAMED31603 - Medicina Integrada III	4,0	18 semanas	61
	08/07/2019 a 29/12/2019	FAMED31901 - Estágio Supervisionado na Área Materno-Infantil: Pediatria	9,3	24 semanas	46
2020/1	09/03/2020 a 17/03/2020	FAMED31603 - Medicina Integrada III	4,0	01 semana	59
	06/01/2020 a 18/05/2020	FAMED31901 - Estágio Supervisionado na Área Materno-Infantil: Pediatria	9,3	19 semanas	51

PROFA. DRA. ROSÂNGELA MARTINS DE ARAÚJO
Coordenadora do Curso de Graduação em Medicina - FAMED/UFU
Portaria R. Nº 941/2019

Documento assinado eletronicamente por **Rosângela Martins de Araújo**, **Coordenador(a)**, em 20/05/2020, às 15:17, conforme horário oficial de Brasília, com fundamento no art. 6º, § 1º, do Decreto nº 8.539, de 8 de outubro de 2015.

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Referência: Processo nº 23117.030655/2020-52 SEI nº 2035390

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DECLARAÇÃO

Processo nº 23117.057228/2022-83

Interessado: Lourdes de Fatima Gonçalves Gomes

O COORDENADOR DO CURSO DE GRADUAÇÃO EM MEDICINA DA UNIVERSIDADE FEDERAL DE UBERLÂNDIA, Nilton Pereira Júnior, no uso de suas atribuições legais e regulamentares, **DECLARA**, para fins de comprovação e anexação ao relatório de atividades para a Progressão/Promoção na carreira docente do magistério superior da UFU, que a Professora **Lourdes Fátima Gonçalves Gomes**, SIAPE 2123460, ligada ao Departamento de Pediatria, ministrou as cargas horárias nos semestres citados abaixo, referentes ao interstício de**: 14/08/2020 a 14/08/2022**.

Declara, ainda, que a referida professora não foi submetida, formalmente, à avaliação discente e que desempenhou suas atividades acadêmicas com assiduidade, disciplina, capacidade de iniciativa, produtividade, responsabilidade, relacionamento interpessoal e qualidade do trabalho, não tendo, portanto, nada que a desabone.

SEMESTRE	DATA DE INÍCIO E TÉRMINO	COMPONENTES CURRICULARES	HORA-AULA SEMANAL	TOTAL DE SEMANAS LETIVAS MINISTRADA	NÚMERO DE ESTUDANTES
2020/1*	Calendário especial 03/11/2020 a 20/03/2021[1]	FAMED31702 - Saúde Individual VII	6	15 semanas	58
2020/2*	Calendário especial 26/04/2021 a 14/08/2021[2]	FAMED31702 - Saúde Individual VII	6	16 semanas	55
	Calendário internato – 87 06/07/2020 a 27/12/2020[4]	FAMED31901 - Estágio Supervisionado na Área Materno-Infantil	7,3	24 semanas	25
2021/1	Calendário especial 06/09/2021 a 22/12/2021[2]	FAMED31702 - Saúde Individual VII	6	16 semanas	62
	Calendário internato – 88 04/01/2021 a 27/06/2021[4]	FAMED31901 - Estágio Supervisionado na Área Materno-Infantil	7,3	24 semanas	33
2021/2	Calendário especial 02/05/2022 a 20/08/2022[3]	FAMED31702 - Saúde Individual VII	6	15 semanas	65

	Calendário internato – 88 e 89 05/07/2021 a 02/01/2022[4]	FAMED31901 - Estágio Supervisionado na Área Materno-Infantil	7,3	25 semanas	62

RESOLUÇÃO CONGRAD Nº 06, DE 17 DE MARÇO DE 2020 (Calendário 2020 suspenso em 18/03/2020).*
RESOLUÇÃO CONGRAD Nº 16, DE 13 DE NOVEMBRO DE 2020 (7º período 2020/01).[1]
RESOLUÇÃO CONGRAD Nº 07, DE 29 DE MARÇO DE 2021 (1º ao 7º período 2020/02 e 1º ao 8º período 2021/01).[2]
RESOLUÇÃO CONGRAD Nº 35, DE 13 DE DEZEMBRO DE 2021 (1º ao 8º período 2021/02).[3]
http://www.famed.ufu.br/graduacao/medicina/calendarios-especiais.[4]

NILTON PEREIRA JÚNIOR
Coordenador do Curso de Graduação em Medicina
Portaria de Pessoal UFU N.º 3.763/2022

Documento assinado eletronicamente por **Nilton Pereira Junior**, **Coordenador(a)**, em 09/08/2022, às 11:06, conforme horário oficial de Brasília, com fundamento no art. 6º, § 1º, do Decreto nº 8.539, de 8 de outubro de 2015.

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Referência: Processo nº 23117.057228/2022-83 SEI nº 3823692

Available online at http://www.journalcra.com

INTERNATIONAL JOURNAL OF CURRENT RESEARCH

International Journal of Current Research
Vol. 13, Issue, 05, pp.17360-17366, May, 2021

ISSN: 0975-833X

DOI: https://doi.org/10.24941/ijcr.41340.05.2021

RESEARCH ARTICLE **OPEN ACCESS**

COMPETÊNCIAS DE ENFERMAGEM EM INTERNAÇÕES PSIQUIÁTRICAS: RECORTE TEMPORAL DA REFORMA AOS DIAS ATUAIS

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ARTICLE INFO

Article History:
Received 09th February, 2021
Received in revised form
14th March, 2021
Accepted 20th April, 2021
Published online 20th May, 2021

Key Words:
Cuidados de Enfermagem,
Hospital PsiquiÆtrico
Humanizaªo da AssistŒncia

ABSTRACT

Objetivo: Identificar as competŒncias da enfermagem em internaíes psiquiÆtricas, considerando a Reforma PsiquiÆtrica e o processo de dissoluªo do modelo manicomial. **MØtodo:** Revisªo Integrativa de Literatura realizada no perôdo de novembro/2019 a marô/2020. Foram utilizados os descritores cadastrados no DECs – Descritores em Saúde: "Cuidados de enfermagem", "Hospital psiquiátrico" e "Humanização da assistência", e incluídas publicações do perôdo de 2007 a 2018. **Resultados:** Realizada a seleªo da amostra, foram includos na revisªo 14 estudos, dos quais emergiram quatro categorias: a transiªo do modelo assistencial considerando a reforma psiquiÆtrica; as relaíes interpessoais e a comunicaªo terapŒutica como cuidado de enfermagem em internaíes psiquiÆtri cas; o trabalho em equipe multiprofissional; obstÆculos e recursos na prÆtica de enfermagem em saœde mental. **Conclusªo:** As competŒncias de enfermagem nas internaíes psiquiÆtricas assim como as possibilidades de atuaªo tŒm se expandido superando as conteníes diversas e o saber exclusivamente biológico. Reformula -se e reinventam-se os modos de assistŒncia estabelecendo um cuidado psicossocial menos invasivo e mais respeitoso, que usa da disponibilidade e da criatividade do profissional para contribuir na reabilitaªo e no processo de reinserªo do sujeito na sociedade.

Citation: Guilherme Silva de Mendonça, Laíssa Mota Cardoso, Pedro Guimarães Pereira et al. "CompetŒncias de enfermagem em internaíes psiquiÆtricas: recorte temporal da reforma aos dias atuais", *2021. International Journal of Current Research, 13, (05), 17360-17366.*

INTRODUCTION

O cenÆrio da assistŒncia em saœde mental no Brasil passou por importantes transiíes nas œltimas dØcadas, promovidos pela Reforma PsiquiÆtrica com inîcio nos anos de 1970 no país (Amarante, 1998), e se fortaleceu com a redemocratizaªo na dØcada seguinte. Esta mobilizaªo buscou dentre outros interesses a substituiªo do modelo assistŒncial manicomial para um modelo nos contextos de relaªo e (re)inserªo do indivíduo na sociedade (Amarante, 1998), alterando a funªo e ordem do recurso da internaªo psiquiÆtrica no processo terapŒutico, que passou a serum estÆgio neste processo œ œltimo recurso a ser usado no cuidado em saœde mental e deve ser considerada quando os demais dispositivos forem esgotados. Na contemporaneidade, busca-se a adequaªo aos novos moldes de atuaªo interdisciplinar, prestando assistŒncia de forma integral e multiprofissional e se atentando às necessidades do sujeito (Brasil, 2019), indo alØm do trabalho tØcnico e burocrÆtico (Reinaldo, 2007). Valorizando a singularidade humana com toda a sua experiŒncia e circunstîncias de saœde e adoecimento que estÆ inserida. Reconhece-se como abordagem a relaªo interpessoal e a manutenªo da autonomia do indivíduo em sua terapŒutica, evitando o estereótipo assistencial restrito à administraªo de medicaíes, garantia da ordem e vigilîncia (Muniz *et al.,* 2015). Ao encontro do que trouxe a reforma psiquiÆtrica, o Projeto TerapŒutico Singular (PTS) amplamente

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empregado no SUS é uma ferramenta facilitadora para a equipe de saúde no cuidado integral em internações psiquiátricas. Consiste em uma construção multiprofissional e coparticipação do indivíduo no seu plano de cuidado. Porém necessita de atenção e tempo dos profissionais para contínua avaliação e adequação do projeto[10]. Entretanto, sabe-se que historicamente a assistência de enfermagem em saúde mental teve embasamento no cuidado hospitalar e disciplinador. Nesse contexto (Reinaldo, 2007), assim como outras áreas da saúde, ainda sofre interferência do modelo da psiquiatria clássica, com enfoque nos aspectos biológicos, ficando suscetível aos riscos de repetir a determinação do louco como perigoso ou incapaz (Amarante, 2016). O cuidado de enfermagem em saúde mental deve atuar junto ao indivíduo, valorizando seus recursos e encorajando-o ao autocuidado (Lacchini). A determinante estabelecer um vínculo, respaldar sua prática em abordagens teóricas como a comunicação terapêutica, e à luz da teoria das relações interpessoais de Hildegard Peplau. Para empregar a teoria de Peplau composta por três categorias, é necessário autoconhecimento, conhecer ao outro e ao contexto que o indivíduo está inserido, evidencia a importância da escuta, para conhecermos as necessidades do outro, que nem sempre são fisiológicas, mas também subjetivas, envolvendo suas emoções e vontades (Cardoso, 2006). A comunicação como um meio crucial ao cuidado em saúde mental,nesse processo de experiencia gradativa transição, num contato do profissional com uma postura para dar-se de forma terapêutica evitando tão somente o técnico eautomatico (Pontes, 2008). As demandas e funções administrativas, de coordenação da assistência, mais notadamente ligadas ao enfermeiro, podem interferir na disponibilidade para o estabelecimento de vínculo alterando as relações entre enfermeiro e paciente (Duarte, 2011).

A sistematização da assistência em enfermagem coopera com a organização do trabalho que visa o cuidado integral, oferecendo autonomia e respaldo ao profissional. Nas internações psiquiátricas ofoco das ações deve se desprender do diagnóstico, tendo em vista a diversidade e complexidade presente na loucura (Duarte, 2011). A presente revisão integrativa da literatura poderá contribuir no conhecimento e identificação das práticas assistenciais de enfermagem em internações psiquiátricas, considerando as mudanças decorrentes da reforma psiquiátrica e auxiliar na percepção de como esses profissionais têm reinventado as relações de cuidado em saúde mental. Atentando-se à relevância do tema levanta-se o questionamento: Quais as competências de enfermagem em internações psiquiátricas,desde a reforma até os tempos atuais? Este estudo tem como objetivo identificar as competências de enfermagem em internações psiquiátricas, desde a reforma até os dias atuais.

MÉTODOS

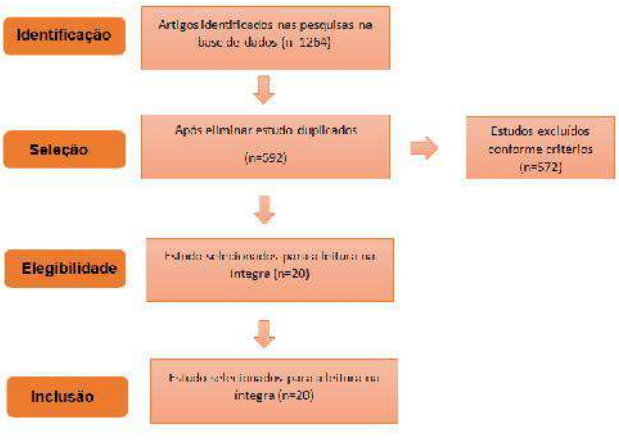
O presente estudo utiliza a Revisão Integrativa da Literatura, que é considerada uma ferramenta singular na área da saúde, pois possibilita a síntese de estudos com um tema em comum, respaldando a assistência no conhecimento científico. Para isso segue critérios cuidadosos de coleta de informações e observação dos dados, sendo um dispositivo pertinente para a Prática Baseada em Evidência(Souza, 2010). Tal metodologia é compostapor seis etapas, sendo a primeira a elaboração da pergunta norteadora, que aqui foi definida: Quais as competências da enfermagem em internações psiquiátricas, considerando a reforma e o processo de dissolução do modelo manicomial? . Seguida então da busca ou amostragem na literatura, foram usados como critério de inclusão na pesquisa: textos online, publicados em periódicos científicos nos últimos dez anos (2009/2018), nos idiomas português e inglês, que envolvam a temática da assistência de enfermagem nas internações em saúdemental, considerando a reforma psiquiátrica. Sendo excluído os editoriais, cartas, relatos de experiência, reflexões teóricas, teses e monografias, assim como os estudos incompletos ou publicados em outros idiomas que não fossem o português ou inglês, e pesquisas publicadas fora do prazo determinado.

O levantamento de dados foi realizado na Biblioteca Virtual de Saúde (BVS). Foram utilizados os termos descritores "cuidados de enfermagem", "hospital psiquiátrico" e "humanização da assistência", que foram cruzados em pares, com o uso do operador booleano "AND", com os resultados descritos na tabela abaixo:

Tabela 1. Descritores pesquisados com o termo booleano *AND*, Uberlândia (MG), Brasil (2020)

Descritores		Número de estudos BVS
Cuidados de enfermagem Hospital psiquiátrico	AND	596
Cuidados de enfermagem Humanização da assistência	AND	645
Hospital psiquiátrico Humanização da assistência	AND	23
Total:		1264

Seguindo os critérios de seleção supracitados foram pré-selecionados 76 estudos das bases de dados LILACS, BDENF, MEDLINE e SCIELO. Após a observação dos títulos e resumos, restaram um total de 20 pesquisas, dos quais foi realizada leitura completa e minuciosa, chegando ao número de 14 estudos que foram explorados nessa revisão 13 em língua portuguesa e 1 em inglês.Concluída a definição dos artigos selecionados no banco de dados virtuais, foi elaborado o fluxograma com a devida representação da seleção dos estudos componentes da amostra final. Para elaboração do presente estudo foi respeitada a Resolução 466/12 do Conselho Nacional da Saúde (CNS), sendo devidamente citados e referenciados todos os autores das obras que foram utilizadas na pesquisa. Foi reservada a identificação das fontes, cumprindo o rigor ético à propriedade intelectual dos estudos explorados, quanto ao emprego de citação nos textos utilizados (Cardoso, 2006). Apresenta-se na Figura 1, o Diagrama de Prisma, (2009), com a evolução da coleta de dados e a quantidade de artigos resultantes, assim como o número dos incluídos seguindo os critérios pré-estabelecidos, que compuseram a amostra do presente estudo.



Fluxograma da seleção dos estudos segundo o *Preferred Reporting Items for Systematic Reviews and Meta-Analyses* (PRISMA 2009)[16]. Uberlândia (MG), Brasil, 2020

RESULTADOS

A busca e análise dos artigos resultaram em 14 (catorze) estudos selecionados, que respeitaram os critérios de inclusão estabelecidos.

Tabela 2. Artigos que compıem o *corpus* da anÆlisepara elaboraªo da revisªo Uberlândia (MG), Brasil (2020)

Artigos	Titulo	Autor/Ano	Objetivo	Metodologia	Perißdico
A1	A complexidade do trabalho de Enfermagem no hospital de custßdia e Tratamento PsiquiÆtrico	VALENTE, Geilsa Soraia Cavalcanti; SANTOS Fernanda Souza[28], 2014	Analisar as formas com que os profissionais de enfermagem lidam com a complexidade existente no ambiente de trabalho confinado do HCTP	Pesquisa descritiva qualitativa.	Revista de pesquisa: Cuidado Ø fundamental. Online
A2	Abordagem da equipe de enfermagem ao usuÆrio na emergŒncia em saœde menta em um pronto atendimento	KONDO, fika Hossar, *et al*. [13], 2011	Conhecer a concepªo da equipe de enfermagem sobre emergŒncias em saœde menta	Pesquisa qualitativa exploratfia	Revista da Escola de Enfermagem da USP
A3	Abordagem existencial do cuidar em enfermagem psiquiÆtrica	FURLAN, Marcela Martins; RIBEIRO, ClØa Regina de Oliveira[11], 2011	Compreender ontologicamente o cuidar em enfermagem na internaªo psiquiÆtrica	Pesquisa qualitativa de abordagem fenomenolßgica	Revista da Escola de Enfermagem da USP
A4	Aßes e cuidados de enfermagem em saœde mental em um hospital-dia psiquiÆtrico: uma revisªo integrativa	JUNIOR, Joªo MÆrio Pessoa, *et al*. [12], 2014	Identificar na literatura evidŒncia dispon◊veis sobre as aßes e cuidados de enfermagem em saœde mental em um Hospital Dia psiquiÆtrico.	Revisªo Integrativa da Literatura	Revista de pesquisa: Cuidado Ø fundamental. Online
A5	Casa de saœdeesperanâ: assistŒncia de enfermagem psiquiÆtricaem um modelo tradicional	RODRIGUES, ´ngela Aparecida Peters, *et al*. [25], 2013	Descrever o contexto da assistŒncia psiquiÆtrica na cidade de Juiz de Fora e sua relaªo com o movimento da Reforma PsiquiÆtrica	Estudo sßcio - histßfico	Revista de enfermagem da UERJ
A6	Construªo de um marco referŒnciapara o cuidado de enfermagem psiquiÆtrica	BORILLE, Dayane Carla; *et al*. [2], 2013	Construir um marco de referŒncia para o cuidado de enfermagem em um hospital psiquiÆtrico.	MØtodo do Arco da Problematizaªo	Revista CiŒncia Cuidado e Saœde
A7	Contenªo fßsica em hospital psiquiÆtrico e a prÆtica da enfermagem	PAES, Marcio Roberto, *et al*[21]., 2009	Investigar como ocorre a contenªo fßsica para paciente em hospital psiquiÆtrico	Pesquisa descritiva	Revista de enfermagem da UERJ
A8	Safety in psychiatric inpatient care: The impact of risk management culture on mental heath nursing practice	SLEMON, Allien; JENKINS, Emily; BUNGAY[26], 2017	Discutir nos ambientes atuais de internaªo psiquiÆtrica, a seguranâ mantida como predominante valor	Revisªo bibliogrÆfica	Revista Nursing Inquiry
A9	Cuidado no hospital psiquiÆtrico sob a ßtica da equipe de enfermagem	DE MELO TAVARES, Claudia Mara; CORTEZ, Elaine Antunes; MUNIZ, Marcela Pimenta[9], 2014	Descrever a percepªo da equipe de enfermagem acerca do cuidado no hospital psiquiÆtrico	Qualitativa, do tipo exploratfia.	Revista Rene
A10	A identidade do cuidado de enfermagem na primeira dØcada do sØculo XXI.	RIBEIRO, Dmaris Kohlbrck de Melo Neu Ribeiro; *et al*. [24], 2013	Buscar evidŒncias cientfficas acerca da identidade do cuidado de enfermagem na prÆtica profissional na primeir de dØcadado sØculoXXI	Revisªo integrativa	Revista Cogitare Enfermagem
A11	Cuidar humanizado: descobrindo as possibilidades na prÆtica da enfermagem em saœde mental	OLIVEIRA, Lucßio Clebson; *et al*.[19] , 2015	Identificar o cuidado humanizado como instrumento da reorganizaªo da prÆtica de enfermagem em saœde mental	Exploratfia de carÆter qualitativo	Revista de Pesquisa: Cuidado Ø fundamental Online
A12	O sentido do cuidado de enfermagem durante a internaªo psiquiÆtrica	OLIVEIRA, Renata Marques; Siqueira, Antßnio Carlos Junior, FUREGATO, Antßnia Regina Ferreira[20], 2017	Identificar o sentido atribußdo aos principais cuidados de enfermagem, prestados durante internaªo psiquiÆtrica	Estudo de campo exploratßrio - descritivo	Revista de Enfermagem UFPE Online
A13	Os cuidados de enfermagem e o exercício dos direitos humanos: Uma anÆlise a partir da realidade de Portugal	MOLL, Marciana Fernandes, *et al*. [17], 2016	Descrever a prestaªo de cuidados de enfermagem em serviços de psiquiatria para adultos de uma cidade de Portugal	Pesquisa qualitativa. Observaªo indireta	Escola Anna Nery
A14	Representaªo social do cuidado de enfermagem em saœde mental: um estudo qualitativo	MACEDO, Jaqueline Queiroz; *et al*. [15],2010	Compreender as representaßes do cuidado de enfermagem em saœde mental	Estudo Qualitativo	Online Brazilian Journal of Nursing

Foram apresentados na tabela seguinte os achados da presente pesquisa, descritos em ordem de código de estudo entre A1 e A14, expondo também os principais dados de cada obra: Autor, ano, título, objetivo, metodologia adotada, e periódico em que foi publicado.

DISCUSSÃO

Realizada a seleção da amostra, com a leitura criteriosa e a análise crítica dos textos incluídos emergiram então quatro categorias: 1-A transição do modelo assistencial considerando a reforma psiquiátrica; 2 – As relações interpessoais e a comunicação terapêutica como um cuidado de enfermagem em internações psiquiátricas; 3 – O trabalho em equipe multiprofissional; 4- Obstáculos e recursos na prática de enfermagem em saúde mental, que favoreceram a interpretação e discussão das informações coletadas.

A transição do modelo assistencial considerando a reforma psiquiátrica A presente categoria é formada pelos artigos A2, A3, A4, A5, A7, A8, A11, A14. Percebe-se que no período que antecede o processo de consolidação da reforma psiquiátrica, o cuidado em internações psiquiátricas consistia em isolar o indivíduo acometido com transtornos mentais, e administrar medicações não apenas como tratamento, mas também como um meio de disciplinar e punir. Competia à enfermagem os papéis de controle, fiscalização e vigilância, assim como as medidas punitivas (A5).

Verifica-se no estudo A8 que embora as práticas em saúde mental exercidas no passado sejam apontadas como cruéis e desumanas, ainda são reproduzidas atualmente no que se trata do isolamento, a disciplinarização, a vigilância e a restrição da autonomia e individualidade como a retirada de objetos pessoais e vestimentas. Muitas dessas ações são justificadas por um discurso de segurança fundamentado no medo, assim como no passado. De acordo com os autores do texto A3, para os pacientes pesquisados o ambiente de internação em saúde mental é descrito como um cenário de violência, onde o que compete ao internado é esperar sua alta, pois ele se sente privado de sua autonomia. Além disso, os entrevistados associaram a figura dos profissionais de enfermagem com a agressividade, por serem esses quem administram medicações de sedação e realizam as contenções. Os autores destacam que a contenção química pode se apresentar tão ou mais restritiva que a física, já que acompanha o indivíduo mesmo após a saída da internação.

O estudo A7 complementa que a contenção física, ou mecânica, que por muito tempo foi praticada de forma inadequada, sofreu mudanças em consequência da reforma psiquiátrica, sendo na atualidade um recurso terapêutico e não mais punitivo. Ao encontro disso no ano de 2012 foi publicada a resolução de número 427 do Conselho Federal de Enfermagem (COFEN), que normatiza os métodos de enfermagem na realização de contenções mecânicas [7]. Conforme esse documento, a contenção mecânica será empregada quando for o único recurso possível para prevenir danos ao atendido e aos demais, sendo proibida quando a finalidade for punir ou disciplinar. Além disso a resolução também descreve os cuidados e monitoramentos necessários ao paciente contido[7]. Na obra A7 os profissionais de enfermagem entrevistados expõem o conhecimento sobre técnicas e cuidados inerentes à contenção física, dentre eles a atenção quanto ao conforto e proteção do paciente e a observação dos sinais vitais e dos membros contidos. Com a mudança na finalidade dessa intervenção, ela passou a requerer uma série de cuidados que anteriormente eram negligenciados.

Em consonância com o que foi recomendado pela Reforma Psiquiátrica no texto A11 os enfermeiros pesquisados recomendam a criação do projeto terapêutico singular, respeitando a individualidade de cada ser, e sugerem também um estreitamento da relação do enfermeiro e da equipe com a família do atendido, o que para os autores expõe o comprometimento em executar o que foi buscado com o movimento reformista. No Brasil a Lei 10.216 de 2001[3], que dispõe sobre a proteção e direitos das pessoas portadoras de transtornos mentais, aponta como função fundamental dos serviços de saúde mental: a reinserção social do atendido no meio em que vive. É importante, de acordo com o estudo A4, que a equipe de enfermagem atue na promoção da autonomia, recuperando a cidadania do indivíduo, o que requer desses profissionais criatividade e disposição para a construção de um cuidado que contemple o exercício das habilidades sociais. O artigo A14 relata uma realidade oposta, dando destaque de forma crítica à falta do comprometimento dos profissionais de enfermagem com o processo de reabilitação e reinserção social dos pacientes em sofrimento mental, que limitam a sua atuação à parte técnica e burocrática, estando ausente em outras atividades. Conforme a pesquisa A4 a atuação da enfermagem em saúde mental passou por transições. Gradativamente, tem se desvinculado da assistência puramente técnica e desprovida de crítica, que se restringia em conter, vigiar e medicar para se aproximar de atividades terapêuticas que prezem pelo vínculo profissional-paciente e o bem-estar do assistido. No entanto embora em alguns aspectos a enfermagem estejam reformulando suas formas de cuidado em internações psiquiátricas, é notável ainda na atualidade a permanência do estigma que persegue o indivíduo com transtorno mental, visto que no estudo A2 profissionais de enfermagem confessam possuir uma resistência em admitir os comportamentos destoantes como algo associado ao processo de adoecimento.

As relações interpessoais e a comunicação terapêutica como um cuidado de enfermagem em internações psiquiátricas Compõem essa categoria os artigos A2, A4, A6, A7, A10, A13. A pesquisa elaborada no estudo A7, relata que os próprios profissionais de enfermagem apontam a necessidade da comunicação e do relacionamento interpessoal para o estabelecimento de vínculo entre a equipe e o cliente. Essa comunicação também foi considerada um instrumento terapêutico nas intervenções com pacientes agressivos. Diante disso, o texto A6 preconiza o respeito mútuo para alcançar a efetividade nessas relações, e para tal é necessário ser capaz de escutar, e aceitar as diferenças e as limitações próprias e as do outro. O estudo também defende que para compreender um ser junto às suas necessidades e possibilidades é fundamental que se constitua o relacionamento interpessoal. No trabalho A4 a aplicação da humanização da assistência se dá justamente nessas relações dos profissionais e pacientes, e interfere de forma relevante no comprometimento do assistido em seu tratamento e em sua qualidade de vida. Em conformidade com os estudos supracitados, o artigo A2 considera a comunicação como uma abordagem eficaz e terapêutica. Também sugere que para alcançar um melhor cuidado, a assistência precisa ir além do corpo físico, contemplando a subjetividade da existência, a nível social, cultural, conhecendo a história e as relações de esse ser humano que está sendo cuidado. Corroborando com o

mesmo pensamento o estudo A10 indica que o cuidar deve estar embasado no convívio e na subjetividade, permitindo uma relação de troca de aprendizados e experiências; e abrangendo o significado do cuidado que compreende dedicação, responsabilização e implicação afetiva. A pesquisa A13 por sua vez compartilha que para alcançarmos o cuidado integral, preconizado por nosso sistema de saúde, é essencial que ocorra o convívio com proximidade entre o atendido, a sua família e a enfermagem, o que coopera para melhores desfechos. Ainda sobre a integralidade, o estudo A6 expõe que é pela relação "pessoa-pessoa" que surge a construção do cuidar integral e humano pela equipe de enfermagem. Os estudos da categoria entraram em concordância quanto aos benefícios que as relações interpessoais e a comunicação terapêutica agregam à assistência, colaborando para o alcance do objetivo do tratamento. Também demandam aos profissionais a responsabilidade de buscar o conhecimento sobre tais instrumentos, e introduzi-los em suas rotinas de trabalho.

O trabalho em equipe multiprofissional: Os estudos A1, A4, A6, A9 e A11 contribuíram para a presente categoria. De acordo com o artigo A4 para a consolidação das mudanças no modelo de assistência em saúde mental e a integralidade do cuidado é essencial que exista a interdisciplinaridade, que se dá nas relações entre as pessoas unindo diferentes saberes que são inerentes na criação do significado da vida. Na pesquisa A6 profissionais de enfermagem de um hospital psiquiátrico definem "equipe" como um conjunto de pessoas com formações diferentes e papéis distintos que dividem uma finalidade em comum. Apontaram a importância de cada profissional exercer o seu papel de forma isolada, porém não foi mencionada a interação entre esses diferentes saberes para a produção de cuidado, apontando um sentido empobrecido do trabalho em equipe, que pode resultar em uma assistência fragmentada. Assim como, no estudo A9 ao serem entrevistados os profissionais de enfermagem admitem uma resistência quanto ao trabalho junto às outras profissões, o que influencia diretamente na qualidade da assistência e sendo essa relação fundamental para a reformulação do cuidado em saúde mental.

A deficiência do trabalho interdisciplinar também pode ser observada no artigo A1, onde os próprios trabalhadores da enfermagem apontaram que um dos fatores que dificulta a assistência é a inexistência do mesmo, além da distância do local de trabalho de cada setor, o que diminuía as possibilidades de comunicação e resultava em um cuidado segmentado. Os entrevistados ainda expuseram uma frustração diante da não participação da enfermagem nas decisões sobre o tratamento dos internados. A legislação da portaria 2.840 de 2014 que cria o programa de desinstitucionalização da saúde mental no âmbito do SUS, traz a necessidade da consolidação do trabalho em equipe multiprofissional, corroborando com o achado nos estudos (Brasil, 2014). Conforme o texto A11, para alcançar o cuidado humanizado que considera a voz e as experiências que fazem do sujeito único, toda a equipe precisa estar alinhada a enxergar o contexto em que o paciente está envolvido. Fazendo-se imprescindível as trocas de saberes entre as diferentes profissões e consolidação das áreas comuns de cuidado que compete a toda equipe, independente da formação.

Obstáculos e recursos na prática de enfermagem em saúde mental: Para discutir essa temática foram utilizados os estudos A2, A4, A9, A13, A10 A11. Diferentes pesquisas apontam as funções burocráticas da enfermagem como um fator que distancia o profissional do paciente. O artigo A11 expõe o conflito entre alcançar o cuidado humanizado dentre as inúmeras tarefas da rotina do enfermeiro, que incluem a gerência, administração, supervisão, além das particularidades institucionais que são de competência desse profissional. Do mesmo modo o artigo A9 afirma que as funções administrativas dadas à enfermagem têm como resultado um afastamento entre o profissional e o paciente, já que essas atividades demandam grande parte do tempo de trabalho. A publicação A4 traz tais atividades burocráticas e a consequente insuficiência de tempo como um desafio a ser superado, assim como a escassez recorrente de recursos materiais e humanos, bem como texto A2 também indica a falta de material como prejudicial à assistência.

Outro obstáculo, exposto pelo trabalho A10, é a posição de poder frequentemente exercida pelo médico, estabelecendo uma relação vertical com a enfermagem, o que limita a sua autonomia frente a assistência prestada. Assim, o estudo A9 a enfermagem psiquiátrica deve participar das decisões em equipe dando amplitude ao cuidado que por vezes se reduz a questões técnicas, desenvolvendo sua criticidade diante da rotina. Em relação às dificuldades encontradas na prática de enfermagem em internações psiquiátricas, foram apontados alguns recursos que podem orientar a organização do trabalho de modo a tentar superar tais obstáculos. A sistematização da assistência de enfermagem aparece como instrumento que beneficia a assistência no estudo A13, assim como o plano assistencial que deve reconhecer a integralidade humana. No entanto, a pesquisa A10 revela a sistematização como parte da identidade da assistência de enfermagem, mas aponta que apesar de sua contribuição, não deve restringir as formas de cuidar e se relacionar entre o profissional e o paciente. O COFEN em sua resolução 358 do ano de 2009, sobre a sistematização da assistência, refere que as etapas do processo de enfermagem precisam ser realizadas em todo serviço em que há atuação da enfermagem, e é composta por: " coleta de dados, diagnósticos de enfermagem, planejamento de enfermagem, implementação e avaliação" (Conselho Federal de Enfermagem, 2009). É possível relacionar as etapas do processo de enfermagem com a construção do PTS do indivíduo assistido, tendo os profissionais de enfermagem muito a contribuir junto à equipe com o plano de cuidado multiprofissional.

CONCLUSÃO

É perceptível que apesar dos estereótipos que cercam a assistência de enfermagem em internações psiquiátricas, com a reforma esse modelo de cuidado vem sofrendo transições. As mudanças se manifestam no modo de se relacionar com o atendido, na disposição em escutar e no cuidado que passa a ser respaldado cientificamente e tem finalidade terapêutica, ultrapassando de forma gradativa o modelo de exclusão e punição vivido no passado. Dessa maneira a enfermagem conquistou uma ampliação dos seus modos de atuação, passando a considerar as subjetividades, o contexto social, familiar, espiritual, e os desejos e recursos do sujeito, para além do ser biológico. Para assistir o outro, passa a ser necessário de fato conhecê-lo, desprender dos estigmas para promoção de vínculo por meio das relações e da comunicação. Os estudos revisados mostraram que os profissionais de enfermagem reconhecem a relação entre a enfermagem e o paciente como

um instrumento terapÊutico, e valorizam a construção e a manutenção do vínculo na internação como um facilitador para que seja alcançado o objetivo do tratamento.

Embora a validação do trabalho interdisciplinar seja encontrada na maioria dos estudos como um meio para se alcançar a integralidade, ainda existe por parte dos profissionais de enfermagem uma resistÊncia e/ou dificuldade de executar o trabalho junto a outras profissões, tornando deficientes as construções coletivas de saberes e intervenções. Acrescenta-se que o enfermeiro por seu papel de gerenciamento da equipe de enfermagem pode estimular e propor a interação com as outras profissões, visando a melhoria da qualidade da assistÊncia com a contribuição de diferentes formações. A atuação da enfermagem em funções burocrÁticas e administrativa surgiu como um desafio, jÁ que toma grande parte do tempo e prejudica a disponibilidade do profissional para se relacionar e se comunicar com o paciente, afetando no estabelecimento do vínculo. A falta de materiais e a quantidade de profissionais adequados, tambØm são apontadas como adversidades. Observando os recursos a serem utilizados, tem-se que a sistematização da assistÊncia e o processo de enfermagem, são indicados como ferramentas que favorecem o trabalho da equipe e organizam o cuidado em etapas interdependentes, desde que não seja visto como uma forma de restringir as relações inerentes à assistÊncia. Contudo, Ø visto que, na atualidade, apesar dos impasses, as competÊncias de enfermagem nas internações psiquiÁtricas e as possibilidades de atuação tem se expandido, indo alØm das contenções diversas e do saber biológico, para um cuidado psicossocial, menos invasivo e mais respeitoso e que usa da disponibilidade, criatividade, disposição e comprometimento para contribuir com a reabilitação e a reinserção do sujeito na sociedade, e que reformula e reinventa os modos de cuidado continuamente. Por fim, ponta-se a necessidade de novos estudos que tratem dos modelos de atuação dos profissionais de saúde nas internações psiquiÁtricas na atualidade. Apesar desses dispositivos não serem prioritÁrios na rede de atenção em saúde mental e sua recomendação hoje ter maior restrição comparada ao passado, ainda cabe redescobri-los, uma vez que esses ainda vivenciam o processo da reforma psiquiÁtrica, estando em constante movimento e readequações, produzindo novas formas de cuidado a serem observadas e compartilhadas.

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Métodos de avaliação antropométrica para pacientes portadores de paralisia cerebral

Anthropometric evaluation methods for patients with cerebral palsy

DOI:10.34117/bjdv6n12-541

Recebimento dos originais: 10/11/2020
Aceitação para publicação: 21/12/2020

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RESUMO
O termo paralisia cerebral (PC) descreve distúrbios do movimento e da postura ocasionados por uma lesão cerebral não progressiva ocorrida durante a infância. Pacientes com esta condição apresentam uma série de comorbidades, entre as quais, desnutrição. Dessa forma o objetivo da presente revisão foi verificar os métodos de avaliação antropométrica que se adequem a esta população. Para tanto utilizaram-se artigos indexados em bases de dados, obtidos através de buscas com diferentes combinações dos descritores "paralisia cerebral", "avaliação nutricional" e "antropometria". Foram utilizadas ainda as listas de referências bibliográficas dos artigos selecionados. Conclui-se que as metas nutricionais dos pacientes com PC devem se basear nos métodos específicos de avaliação para esta população. Contudo, ainda há muitos parâmetros que não foram validados e, neste sentido, mais estudos são necessários para melhorar as intervenções nutricionais.

Palavras-chave: paralisia cerebral, avaliação nutricional, antropometria

ABSTRACT
The term cerebral palsy (CP) describes disorders of movement and posture caused by a non-progressive brain injury ocurring during childhood. Patients with this condition have a number of comorbities, including malnutrition. Thus, the aim of the presente review was to verify the methods of antrhopometric evaluation that suits this population. Thus data base indexed articles were obtained from searches with diferente combinations of descriptors: "cerebral palsy", "nutritional assessment" and "antrhopometry". The list of bibliographic references of the selected articles were also used. Therefore, the nutritional goals of patients with CP should be based on the specific methods of evaluation for this population. However, there are still many parameters that have not been validated and in this sense more studies are needed to improve nutritional interventions.

Keywords: cerebral palsy, nutrition assessment, anthropometry

1 INTRODUÇÃO

A Paralisia Cerebral (PC), também chamada encefalopatia crônica não progressiva da infância, foi descrita pela primeira vez em 1861 e teve diversas definições ao longo dos anos. Atualmente é conceituada como um termo clínico, e não um diagnóstico etiológico, que descreve alterações permanentes do desenvolvimento neuropsicomotor ocasionadas por uma lesão cerebral não progressiva ocorrida durante a fase de desenvolvimento do órgão, ou seja, do período fetal até os primeiros anos de vida, geralmente antes dos dois anos de idade. Manifesta-se como desordens motoras e posturais e comumente vem acompanhada de modificações sensoriais, perceptivas, cognitivas, comportamentais e de comunicação, além de epilepsia e alterações musculoesqueléticas (ROSENBAUM et al., 2007).

No que diz respeito à etiologia, ela é heterogênea e multifatorial, incluindo agentes pré- natais (infecções, parasitoses, intoxicações, radiação, traumatismos, fatores maternos e genéticos), peri-natais (hipóxia, isquemia, prematuridade, parto prolongado) e pós natais (anóxia, traumatismos, distúrbios

metabólicos, infecções, kernicterus, processos vasculares, desnutrição) (ROTTA, 2002; REDDIHOUGH; COLLINS, 2003; FUNAYAMA et al., 2000), sendo as infecções e as hipóxia isquemias as causas mais associadas à encefalopatia (ROTTA, 2002). Quanto a incidência mundial, estima-se que seja de 2 a 2,11 casos a cada mil nascidos vivos (COLVER; FAIRHURST; PHAROAH, 2014; OSKOUI et al., 2013).

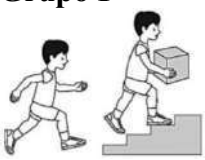
Tão desigual quanto a etiologia são os sinais clínicos e a severidade dos comprometimentos ocasionados ao paciente, dessa forma a PC recebe diversas classificações, sendo as mais comumente utilizadas aquelas que levam em consideração os sinais clínicos e alterações musculares (ROSENBAUM et al., 2007).

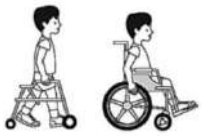
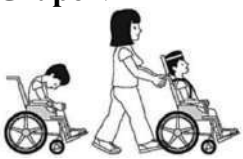
Neste sentido, divide-se, quando considerados os tipos de movimentos predominantes, em espástica – que subdivide-se de acordo com a topografia do comprometimento em unilateral ou bilateral (diplégicas, triplégicas, quadri/tetraplégicas e com dupla hemiplegia) (ROSENBAUM et al., 2007) – discinética, atáxica ou mista (FUNAYAMA et al., 2000); e piramidal, extrapiramidal ou cerebelar em relação a topografia da lesão inicial (WIMALASUNDERA; STEVENSON, 2016; FUNAYAMA et al., 2000).

Ainda com relação às alterações motoras, pode-se classificar, de acordo com o padrão de tonicidade, em hipo ou hipertônica, sendo que os padrões podem variar, num mesmo paciente, em diferentes períodos (GILLES, 2007).

Outra classificação amplamente utilizada para crianças e adolescentes com PC, e de grande importância para a correta avaliação antropométrica, é a com base na capacidade motora grossa (Gross Motor Function Classification System – GMFCS). Nela consideram-se os níveis de mobilidade, discriminando a severidade das disfunções musculares apresentadas pelos indivíduos e classificando-os em cinco níveis, como exposto na figura 1 (WIMALASUNDERA; STEVENSON, 2016; ROSENBAUM et al., 2008; PALISANO et al., 1997).

Figura 1: Descrição e ilustração dos níveis GMFCS

Grupo I 	Anda e sobe escadas sem limitações, corre e pula, mas velocidade, equilíbrio e coordenação podem ser prejudicados
Grupo II 	Anda e sobe escadas com auxílio de corrimão, mas tem dificuldades em se locomover por solos desnivelados ou inclinados e apresenta dificuldade para pular e correr
Grupo III	

	Anda com auxílio de dispositivos como muletas e andadores, pode subir escadas com ajuda de corrimão. Talvez necessite de cadeira de rodas para se locomover por longas distâncias ou em terrenos desnivelados
Grupo IV 	Senta-se em cadeira adaptada, faz transferência com auxílio, anda com andador ou sozinho em curtas distâncias. Consegue tocar cadeira elétrica, mas necessita de ajuda com a tradicional
Grupo V 	Necessita de adaptações para sentar-se, apresenta restrição no controle dos movimentos voluntários e na habilidade de manter a cabeça e o tronco eretos. Totalmente dependente nas atividades diárias e locomoção

Fonte: Adaptado de Wimalasundera; Stevenson, 2016; Palisano et al., 1997. Direito de imagem Copyright© Kerr Graham, Bill Reid e Adrienne Harvey, The Royal Children's Hospital, Melbourne ERC: 070288

No que diz respeito ao prognóstico, embora as alterações encefálicas não sejam progressivas outras doenças neurológicas e funcionais aparecem durante toda a vida e podem evoluir com o tempo, atividades, terapias, envelhecimento, aprendizados e outros fatores (ROSENBAUM et al., 2007). Nesse sentido, não há cura para a encefalopatia não progressiva, sendo o manejo dos sintomas a base do tratamento (WIMALASUNDERA; STEVENSON, 2016).

Com relação às estratégias de meneio nutricional, a avaliação destes pacientes deve incluir histórias médica, nutricional, de crescimento e social completas, além de medidas antropométricas precisas (MARCHAND; MOTIL; NASPGHAN COMMITTEE, 2006), porém diversos estudos demonstraram que os métodos de avaliação utilizados para população geral são inadequados para estes pacientes (MELUNOVIC et al., 2017; WANG et al., 2016; ARAÚJO; SILVA, 2013; CARAM; MORCILLO; COSTA-PINTO, 2008; SOYLU et al., 2008; MARCHAND; MOTIL; NASPGHAN COMMITTEE, 2006) pois superestimam o diagnóstico de desnutrição e subestimam o de obesidade. Sabe-se ainda que esta é uma população de risco para desnutrição, principalmente quando considerados os pacientes com maiores comprometimentos motores (AYDIN, 2018; MELUNOVIC et al., 2017; QUITADAMO et al., 2016; WANG et al., 2016; BROOKS et al., 2011; CALIS et al., 2008; KUPERNIC; STEVENSON, 2008; CAMPANOZZI et al., 2007; SCHWARZ et al., 2001).

Estudo realizado com o objetivo de investigar a ingestão alimentar, avaliação de peso e composição corporal em crianças com deficiência cerebral. demonstrou em seus resultados que a maioria das crianças apresentaram déficit de massa magra, porém não foram encontradas diferenças na composição corporal de acordo com o grau de mobilidade (HOLANDA et. al., 2020).

Com isso, o objetivo da presente revisão foi verificar na literatura os métodos de avaliação antropométrica existentes que se aplicam a esta população e que permitem adequada avaliação nutricional destes pacientes.

2 DESENVOLVIMENTO

O estudo em questão refere-se a uma revisão narrativa da literatura, desenvolvida mediante pesquisa nas bases de dados: Portal Regional de Saúde – Biblioteca virtual de Saúde (BVS) e Biblioteca Nacional de Medicina dos Estados Unidos (PUBMED) e realizada durante o período de outubro a novembro de 2018 utilizando os seguintes descritores (Descritores em Ciências da Saúde – DeCS): paralisia cerebral, avaliação nutricional e antropometria e seus correspondentes nos idiomas espanhol e inglês.

Uma vez que a PC vem sendo discutida e estudada desde o século XIX até os dias de hoje, não se delimitou um período específico para a pesquisa, utilizando todos os artigos que fossem pertinentes para o presente estudo. Para tanto, utilizaram-se como critérios de inclusão artigos científicos disponíveis na integra, nos idiomas português, inglês e espanhol, e que tivessem relação com o tema desta pesquisa, ou seja, que discutissem ou expusessem métodos de avaliação antropométrica para pacientes com PC. Foram excluídos arquivos não disponíveis por completo; em outros formatos que não fossem artigos; publicações repetidas nas bases de dados; e, ainda, aqueles que não tinham relação com o tema.

A seleção dos artigos foi iniciada a partir das buscas nas bases de dados citadas, quando se combinaram os descritores utilizando o booleano AND, de modo que em um primeiro momento buscaram-se por artigos utilizando as combinações: “paralisia cerebral” AND “avaliação nutricional” e “paralisia cerebral” AND “antropometria”, nos três idiomas considerados. Neste momento, foram salvos os artigos encontrados cujos títulos e resumos se encaixavam no tema da pesquisa. Posteriormente, todos os artigos foram lidos na íntegra e selecionaram-se os que respondiam ao objetivo da pesquisa.

Foram utilizadas, ainda, as listas de referências bibliográficas dos artigos selecionados. Assim, durante a leitura destes, quando encontrados artigos de interesse da pesquisa, buscaram-se por eles nos bancos de dados e aqueles disponíveis na íntegra também foram incluídos.

Após extensa leitura dos artigos selecionados, foram destacados nove (Tabela 1) que abordaram técnicas e métodos de avaliação antropométrica específicos ou validados para pacientes com PC.

Tabela 1: Caracterização dos artigos encontrados

Autor	Título	Objetivos
Stevenson, 1995	Uso de medidas segmentares para estimar a estatura em crianças com paralisia cerebral	Testar a utilidade de medidas segmentares para acessar a estatura em crianças com PC.
Krick et. al., 1996	Padrão de crescimento em crianças com paralisia cerebral	Criar uma referência de crescimento para crianças com paralisia cerebral quadriplégica
Stevenson, 1996	Medida do crescimento em crianças com deficiência	Revisão da avaliação do crescimento em crianças com deficiência
Marchand, Motil, NASPGHAN, 2006	Suporte nutricional para crianças com comprometimento neurológico: um relatório clínico da Sociedade Americana de Gastroenterologia Pediátrica, Hepatologia e Nutrição	Diretriz que avaliou o manejo nutricional, incluindo complicações e questões nutricionais de pacientes com alterações neurológicas
Day et. al., 2007	Padrões de crescimento em uma população de crianças e adolescentes com paralisia cerebral	Apresentar curvas de crescimento para pacientes com PC estratificadas pela gravidade da incapacidade, capacidade de alimentação e função motora
Brooks et. al, 2011	Baixo peso, morbidade e mortalidade em crianças com paralisia cerebral: novos gráficos de crescimento	Determinar os percentis de peso-para-idade na PC de acordo com o gênero e o nível GMFCS e identificar os pesos associados a resultados negativos à saúde
Gurka et. al., 2014	Avaliação e correção das equações de espessura das dobras cutâneas na estimativa da gordura corporal em crianças com paralisia cerebral	Avaliar a precisão das equações de dobras cutâneas na estimativa da porcentagem de gordura corporal em crianças com PC em comparação com a avaliação da gordura corporal a partir da absorciometria por raios X de dupla energia
Romano et. al., 2017	Sociedade Europeia de Gastroenterologia Pediátrica, Hepatologia e Diretrizes Nutricionais para Avaliação e Tratamento de Complicações Gastrointestinais e Nutricionais em Crianças com Deficiência Neurológica	Diretriz que objetivou avaliar o manejo nutricional, incluindo complicações e questões nutricionais de pacientes com alterações neurológicas
Minocha et. al., 2018	Avaliação nutricional global subjetiva: uma ferramenta de rastreamento confiável para avaliação nutricional em crianças com PC	Determinar a prevalência de desnutrição em crianças com PC e comparar métodos subjetivos e objetivos de avaliação nutricional.

Avaliação Antropométrica

As Sociedades Europeias (ESPGHAN) e Norte Americana (NASPGHAN) de Gastroenterologia Pediátrica, Hepatologia e Diretrizes Nutricionais recomendam, nas Diretrizes para

Avaliação e Tratamento de Complicações Gastrointestinais e Nutricionais em Crianças com Deficiência Neurológica e no Relatório de Apoio Nutricional para Crianças com Deficiências Neurológicas, que a avaliação antropométrica de pacientes com PC não seja realizada apenas através do peso e da estatura, mas também a partir de pregas cutâneas e circunferências (ROMANO et al., 2017; MARCHAND; MOTIL; NASPGHAN COMMITTEE, 2006).

Isto porque baixos índices de peso para a altura (P/A) ou baixos índice de massa corporal (IMC) não necessariamente indicam depleção dos estoques de gordura, como também podem traduzir-se em níveis de massa magra (MM) menores e altos índices de massa gorda (MG) (SAMSON-FANG; STEVENSON, 2000; KUPERMINC et al., 2010). Ademais, demonstrou-se que o IMC nestes pacientes tem uma correlação moderada com o percentual de gordura corporal (%GC) (KUPERMINC et al., 2013) e, ainda, que as crianças com alterações neurológicas tendem a acumular gordura nas regiões centrais (FRISANCHO, 1981) daí a necessidade de medidas adicionais para correta interpretação dos dados obtidos.

A ESPGHAN sugere também bandeiras vermelhas para o diagnóstico de desnutrição, sendo que a presença de uma delas já é suficiente para caracterizar o paciente com estado nutricional alterado negativamente. São elas: 1) Sinais físicos de desnutrição como úlceras de decúbito, alterações na pele, circulação periférica deficiente; 2) Peso para idade com score Z < 2; 3) Prega cutânea tricipital percentil < P10 para idade e sexo; 4) Área muscular do braço percentil <P10; e 5) Falha no crescimento ou baixo peso. (ROMANO et al., 2017).

Peso e Altura

Quanto ao peso, deve ser aferido em balança calibrada com o paciente usando roupas leves ou nenhuma peça e, naqueles em que a medida não é possível por não se manterem em pé na balança, pode-se pesá-los no colo do acompanhante ou em cadeira de rodas e descontar o peso destes (SAMSON-FANG; BELL, 2013; MARCHAND; MOTIL; NASPGHAN, 2006).

Não foram encontradas fórmulas para estimativa do peso que se apliquem às crianças e adolescentes, nem para os hígidos, nem para os pacientes neurológicos; para adultos sãos existem diversas fórmulas para obter o peso estimado, porém nenhuma delas foi validada para a população com PC, o que faz com que a medida deva ser obtida de forma direta ou que se utilizem estimativas que podem não ser adequadas para estes pacientes.

Em relação à altura, sempre que possível deve ser aferida com o paciente em pé em um estadiômetro ou, ainda, através da altura decumbente quando for possível deitar de forma reta (ROMANO et al., 2017). Entretanto, pacientes com PC podem apresentar uma série de alterações

musculoesqueléticas, bem como deformidades da coluna vertebral (SHIN et al., 2017; ROSENBAUN, 2007) e com isso tornar esta medida um desafio.

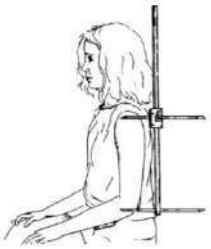
Com isto, Stevenson (1995) desenvolveu fórmulas preditivas (Tabela 2) que possibilitam estimar a altura através de medidas segmentadas (comprimento do braço, da tíbia e altura do joelho) (Figura 2). Importante ressaltar que foi um estudo realizado com crianças e, portanto, as equações se aplicam do nascimento aos 12 anos. Ademais, o estudo demonstrou que a fórmula que utiliza a altura do joelho como indicador para a altura é a que obteve menor erro em relação à altura aferida (R^2=.97) e, portanto, é a mais indicada. Foi demonstrado por Haapala et al. (2015) que esta fórmula apresenta alta confiabilidade entre avaliadores e boa repetibilidade das medidas, o que permite que sejam utilizadas para monitorar o crescimento destas crianças.

Tabela 2: Fórmula para determinar a altura de crianças com PC do nascimento aos 12 anos

Medida Segmentar	**Estimativa da altura (A) em centímetros (cm)**	**Desvio Padrão**
Comprimento do braço (CB)	A= (4,35 x CB) +21,8	±1,7cm
Comprimento da tíbia (CT)	A= (3,26 x CT) + 30,8	±1,4cm
Altura do joelho (AJ)	A= (2,69 x AJ) + 24,2	±1,1cm

Fonte: Traduzido de Stevenson, 1995.

Figura 2: Ilustração das medidas segmentadas propostas para estimativa de altura e a forma de obtê-las

Comprimento do braço		Realizada com um antropômetro através da medida da distância do acrômio à cabeça do rádio. Pode ser realizada com a criança em pé ou sentada e o braço relaxado com o cotovelo fletido a 90º.
Comprimento da tíbia		Realizado a partir da borda superomedial da tíbia até a borda inferior do maléolo medial, através de uma fita flexível.
Altura do joelho		Feita a partir da medida do joelho e o tornozelo dobrados a 90º, com um paquímetro ou antropômetro. Realiza-se a medida da distância do calcanhar à superfície anterior da coxa, sobre os côndilos femorais.

Fonte: Adaptado de Stevenson, 1995.

Uma vez que a fórmula de Stevenson (1995) tem seu uso limitado para crianças até 12 anos, é sugerido que se utilizem as fórmulas propostas por Chumlea, Guo e Steinbaugh (1994) (Tabela 3) naqueles paciente acima desta idade, uma vez que foram validadas para uso até os 19 anos em um pequeno grupo de pacientes com PC (STEVENSON, 1996), ainda que tenha sido observada uma variância entre a estimativa pela fórmula e a altura decumbente (HAAPALA et al., 2015; BELL; DAVIES, 2006).

Tabela 3: Fórmula para determinar a altura de pacientes com PC acima dos 12 anos

Idade (anos)	Sexo e etnia	Estimativa da altura (A) em centímetros (cm)	Desvio Padrão
6 a 18	Meninos brancos	A=40,54 + (2,22 x AJ)	±4,21cm
	Meninos negros	A=39,60 + (2,18 x AJ)	±4,58cm
	Meninas brancos	A=43,21 + (2,15 x AJ)	±3,90cm
	Meninas negros	A=46,59 + (2,02 x AJ)	±4,29cm
19 a 60	Homens brancos	A=71,85 + (1,88 x AJ)	±3,97cm
	Homens negros	A=73,42 + (1,79 x AJ)	±3,60cm
	Mulheres brancas	A=70,25 + (1,87 x AJ) – (0,06 x I)	±3,60cm
	Mulheres negras	A=68,10 + (1,86 x AJ) – (0,06 x I)	±3,80cm

Legenda: AJ- altura do joelho em centímetros; I – idade em anos. Fonte: Traduzido de Chumlea; Guo; Steinbaugh, 1994.

Curvas de Crescimento

Sabe-se que a avaliação antropométrica de crianças e adolescentes envolve as medidas de P/A, peso-para-idade (P/I), altura ou estatura-para-idade (A/I ou E/I) e IMC-para-idade (IMC/I), o que não difere para àquelas com diagnóstico de PC. Entretanto, os padrões de P/I e A/I são menores nas crianças com alterações neurológicas quando comparadas à população geral (SAMSON-FANG; STEVENSON, 2000; DAHL et al., 1996; KRICK et al., 1996; STALLINGS et al., 1995; STALLINGS et al., 1993; STALLINGS et al., 1993b).

Isso porque mesmo quando nutridas estas crianças tendem a ter seu crescimento afetado e uma das consequências é que seu padrão de crescimento pode não ser corrigido mesmo com terapia nutricional (MARCHAND; MOTIL; NASPGHAN, 2006). Talvez isso se justifique pela própria alteração neurológica (ROMANO et al., 2017; ROSEMBAUN et al., 2007) ou ainda pela diminuição da atividade física observada nesses pacientes, pela ausência de força mecânica sobre os ossos, articulações e musculatura e/ou fatores endócrinos, além das altas prevalências de prematuridade e baixo peso ao nascer (TAMEGA et al., 2011; HENDERSON et al., 2007).

Neste sentido, foram desenvolvidas curvas de crescimento apropriadas para os pacientes com alterações neurológicas (Tabela 4), sendo as mais atuais, de Brooks et al. (2011) aquelas recomendadas para a prática clínica, uma vez que abrangem todos os níveis GMFCS de comprometimento, além de levarem em conta se o paciente alimenta-se via oral ou de forma alternativa, por sonda de alimentação.

Tabela 4- Comparação entre as curvas de crescimento desenvolvidas para pacientes com PC

Estudo	Amostra	Características das Curvas Desenvolvidas	Observações e Limitações
Krick et al., 1996	n= 360 crianças com diagnóstico de PC quadriplégica (PCQ)	Meninos e Meninas 0 a 10 anos P/I e A/I Percentis P10, P50 e P90	Comparou com as curvas de crescimento do National Center for Health Statistic (NCHS)* e observou que as crianças com PCQ classificadas com P50 nas curvas para PC, encontraram-se abaixo do P10 das curvas do NCHS; excluiu crianças com PC associadas a outras doenças e síndromes; seu uso limita-se aos pacientes com PCQ; apresenta apenas três percentis de corte
Day et al., 2007	n=24.920 pacientes com PC atendidos no Departamento de Serviços de Desenvolvimento da Califórnia entre 1987 e 2002	Meninos e Meninas 2 a 20 anos P/I, A/I, P/A e IMC/I Percentis P10, P50, P90 GMFCS I ao V	Fornece apenas três percentis de corte; não comtempla crianças menores de 2 anos; apresentaram pontos de corte específicos para avaliação do estado nutricional: P < 10=desnutrição; P ≥ 10 e ≤ 90=eutrofia; P > 90=sobrepeso. **Geralmente é vista com a referência de Steven et al., 2007, pois o nome do autor é Steven M. Day
Brooks et al., 2011	n =25.545 crianças com PC atendidas no Departamento de Serviços de Desenvolvimento da Califórnia atendidas entre os anos de 1988 e 2002.	Meninos e Meninas 2 a 20 anos P/I, A/I, P/A e IMC/I Percentis P5, P10, P25, P50, P75, P 90 e P95 GMFCS I ao V GMFCS V considera via de alimentação	Não comtempla crianças menores de 2 anos; não excluiu do grupo amostral crianças com outras morbidades, o que talvez não reflita o crescimento ideal desta população de forma fidedigna; considera o GMFCS e a via de alimentação (via oral ou por sonda); gráficos de P/I apresentam uma faixa que destaca risco nutricional e de comorbidades associadas ao peso; apresentaram pontos de corte específicos para avaliação do estado nutricional: P < 10=desnutrição; P ≥ 10 e ≤50= eutrofia; P > 50 e ≤ 90=risco de sobrepeso; P > 90= sobrepeso.

*NCHS,1977

No entanto, as curvas indicadas abrangem apenas crianças acima de dois anos, sendo que crianças abaixo dessa idade são contempladas apenas nas curvas propostas por Krick et al. (1996), que, no entanto, é adequada somente para pacientes com PC quadriplégica. Com isso, a ESPGHAN

(ROMANO et al., 2017), sugere que se utilizem as curvas propostas pela Organização Mundial da Saúde (WHO, 2006) para pacientes até os dois anos de idade. O que deve ser feito com extrema cautela.

Estimativa de Composição Corporal

Como citado, faz-se necessário o uso de outras medidas antropométricas além das medidas usuais de peso e altura e as estimativas de composição corporal de crianças podem ser obtidas através de uma série de técnicas (SCARPATO et al., 2017). Nesse sentido, o padrão ouro para obter composição corporal e garantir a adequabilidade da composição corporal é a densitometria com raio-x de dupla energia (DEXA) (KUPERMINC et al., 2008), entretanto este é um método caro e de difícil acesso.

Assim, a ESPGHAN sugere que se utilize, para a estimativa do %GC as correções de Gurka et al. (2010) propostas para população com PC (ROMANO et al., 2017) (Tabela 5), as quais foram feitas a partir da fórmula de Slaugthers et al. (1988). Isto porque tal correções mostraram boa correlação estatística com o a DEXA (ROMANO et al., 2017; SCARPATO et al., 2017).

Tabela 5: Equações originais de Slaughter para determinar percentual de gordura corporal e correção para crianças com PC

População	Equação original de Slaughter
Soma das dobras tricipital e subescapular ≤35mm	
Homem	
Pré-púbere[1] branco - 1.7	%GC=1.21 x (tri+sub) – 0,008 (tri+sub)²
Pré-púbere negro - 3.2	%GC=1.21 x (tri+sub) – 0,008 (tri+sub)²
Púbere branco - 3.4	%GC=1.21 x (tri+sub) – 0,008 (tri+sub)²
Púbere negro - 5.2	%GC=1.21 x (tri+sub) – 0,008 (tri+sub)²
Pós-púbere branco - 5.5	%GC=1.21 x (tri+sub) – 0,008 (tri+sub)²
Pós-púbere negro - 6.8	%GC=1.21 x (tri+sub) – 0,008 (tri+sub)²
Mulher (todas) - 2.5	%GC=1.33 x (tri+sub) – 0,013 (tri+sub)²
Soma das dobras tricipital e subescapular ≥35mm	
Homem (todos)	%GC=0,783 x (tri+sub) + 1.6
Mulher (todos)	%GC=0,546 x (tri+sub) + 9.7
Correções desenvolvidas por Gurka et al.[2]	
Correção geral	+12.2
Correção adicional:	
Homens	-5.0
GMFCS mais severos	+5.1
Negros	-3.1

	Púbere	+2.0
	Pós-púbere	-4.6
	Dobras tri + sub > 35mm	-3.2

Legenda: [1]Pré-adolescente: estágios Tanner 1 e 2; Adolescente: estágio Tanner 3; Pós-pubere: estágio Tanner 4 e 5. [2]Instruções para utilizar as correções para PC: sempre adicionar 12.2 ao resultado da equação de Salughter. Se o indivíduo se encaixar em uma das categorias adicionais, adicionar também a correção respectiva. Fonte: Gurka et al., 2010

Adicionalmente, as pregas cutâneas tricipital e subescapular devem ser utilizadas rotineiramente. Sendo que os valores encontrados devem ser comparados com as tabelas de referência para população saudável, ou seja, OMS (2016) e Frisancho (1981), para identificar o percentil em que o paciente se enquadra. Valores inferiores ao percentil P10 indicam desnutrição (ROMANO et al., 2017; SCARPATO et al., 2017; MARCHAND; MOTIL; NASPGHAN, 2006).

A área muscular do braço, que pode ser calculada a partir das medidas de circunferência do braço e dobra cutânea tricipital (STALLINGS et al., 1993), também é um bom parâmetro para utilizar-se rotineiramente a fim de detectar o estado nutricional dos pacientes, uma vez que esta medida tem uma sensibilidade maior em detectar desnutrição do que o parâmetro de A/I (SAMSON-FANG; STEVENSON, 2000). As tabelas e padrões de referência utilizados para encontras os percentis são os mesmos indicados para população saudável, ou seja, Frisancho 1981 (ROMANO et al., 2017).

A bioimpedância elétrica (BIA) mostrou-se um bom parâmetro de determinação da massa livre de gordura, porém não houve correlação forte na determinação de massa gorda e percentual de gordura corporal (OEFFINGER et al., 2014; RIEKEN et al., 2011; LIU et al., 2005), entretanto, é um método confiável de estimativa de composição corporal quando comparado à DEXA (ROMANO et al., 2017), podendo ser utilizado desde que respeitadas as indicações e limitações do método.

Holanda et al. (2020) destaca que é imprescindível avaliar a composição corporal e alimentar das crianças portadoras de Paralisia Cerebral, com a finalidade de determinar seu estado nutricional e identificar situações de risco viabilizando assim o planejamento de ações de promoção da saúde e prevenção de doenças, deste público específico.

Avaliação Global Subjetiva

Minocha et al. (2018) realizaram um estudo observacional analítico para determinar a prevalência de desnutrição em crianças com PC através de um método subjetivo. Para tanto avaliaram-se objetivamente os dados de 180 crianças, com idades entre 1 e 12 anos, que frequentavam um hospital terciário. Depois de comparar os dados subjetivos com métodos objetivos, concluíram que a Avaliação Global Subjetiva (AGS) pode ser uma ferramenta confiável para avaliar de forma subjetiva o estado nutricional de crianças com PC. Tal instrumento pode ser um bom aliado em triagens e para acompanhamento, uma vez que é não-invasiva, de baixo custo e fácil aplicação.

3 CONCLUSÃO

Os resultados da Revisão Narrativa demonstram que a produção científica acerca dos métodos de avaliação antropométrica específicos para PC é muito escassa. Entretanto, a falta de estudos pode estar associada aos restritos descritores e poucas combinações booleano utilizadas. Talvez novos métodos de busca ampliassem os achados.

Os artigos analisados, demonstram que não se deve estabelecer como meta para os pacientes com PC atingir índices antropométricos definidos para população geral. Estes pacientes constituem um grupo de grande risco nutricional para desnutrição e, por isso, deve-se utilizar os métodos de avaliação nutricional específicos para estes pacientes

Entretanto, muitos dos métodos de avaliação recomendados ainda não são validados para esta população, neste sentido mais estudos podem ser desenvolvidos para melhorar a acurácia e fidedignidade da avaliação nutricional destes pacientes.

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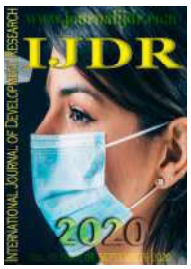
ISSN: 2230-9926

Available online at http://www.journalijdr.com

IJDR

International Journal of Development Research
Vol. 10, Issue, 09, pp. 40124-40127, September, 2020
https://doi.org/10.37118/ijdr.19832.09.2020



RESEARCH ARTICLE OPEN ACCESS

CLINICAL CASE-INTRACARDIAC STRANGE BODY APPROACH IN PREMATURES AND CHILDREN - CASE REPORT

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ARTICLE INFO

Article History:
Received 06th June 2020
Received in revised form
11th July 2020
Accepted 21st August 2020
Published online 29th September 2020

Key Words:

Central venous catheter.
Rupture. Care. Children.

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ABSTRACT

As a universal clinical practice, central venous catheterization involves risks and raises many doubts in the patient's approach to complications, especially in the intravascular and intracardiac rupture of the catheters. Catheter rupture and embolization are the most feared, high-risk adverse events for the patient and stressful for family members and professionals involved. Even following the protocols correctly in handling the catheter, the indication of withdrawal does not prevent these complications. In this way we report the clinical and interventional experience with low weight child with rupture and intracardiac embolization of a large catheter fragment and we discuss the clinical approach in these cases and what is the best moment of withdrawal of these devices.

Citation: ***Georgio Moraes Costa, Cristiano Silva Neves, Lourdes de Fátima Gonçalves Gomes, João Ribeiro de Mattos Neto, Vilmar Jose Pereira, Guilherme Silva de Mendonça.*** **2020.** "Clinical case-intracardiac strange body approach in prematures and children - Case Report*", International Journal of Development Research*, 10, (08), 40124-40127.

INTRODUCTION

Central venous catheterization is a common practice in pediatric, neonatal intensive care units and is increasing in other areas of care from the patient to the emergency room[1]. Central venous catheters provide safe, less invasive, but high-risk vascular access for adverse events[1]. An adverse event is defined as unintentional injury resulting in temporary or permanent disability and even prolonging the time of illness, in the hospital, and death of the patient as a result of the treatment provided[2]. Peripheral Insertion Central Catheter (PICC) facility is indicated by providing safe, fast, effective, improve survival and reduce sequel, mainly venous dissection and thrombosis[3]. Catheter rupture and intravascular embolization account for about 1% of complications associated with central venous access. These events can occur with significant mortality rates[4]. In addition, the technical refinement of the intravascular devices allowed its installation even for children of extremely low weight (less than 1 kg) and stimulates the greater qualification of the professionals involved in the treatment of the child[3]. The decision about insertion of central catheter involves balancing risks and benefits where the benefits must overcome the risks[3]. Thus, continuous monitoring and the search for adverse effects are part of the quality therapeutic indicators and become a fundamental tool because they point out the quality of care and ensure safe care[5]. Once, correctly inserted, and positioned, PICC rupture is the most feared and risky adverse event. Catheter rupture is almost always multifactorial, but inadequate manipulation of the catheter, infusion with great intraluminal pressure, and poor technical quality of the material at the site of the puncture are among the most important determinants[6]. Thus, percutaneous insertion catheter ruptures (PICC) occur most commonly when they are introduced by access into the subclavian vein, resulting from compression of the catheter by the clavicle and the first rib,

known as pinch-off syndrome[7]. Radiography is still the method of excellence in checking the position of the catheter tip as it is accessible, inexpensive, quick, safe and feasible in any hospital unit soon after insertion and during its stay and in the detection of complications. But the immediate approach and follow-up of these patients is what most generates discomforts and doubts for the teams involved. Thus the objective of the report is to document the clinical and interventional experience with low weight child, review of the literature on the events of catheter rupture and to discuss the best moment of withdrawal of these devices.

Case Report

Patient MLGC, twin III, birth weight: 865g, female, 3 months and 21 days, hospitalized for 29 days, due to extreme prematurity (gestational age: 27 weeks), hydrocephalus after intracranial hypertension, was submitted to PICC implantation in the right saphenous vein for infusion of drugs and fluids 06 days before the event. At 29 days of hospitalization and the 6th day of PICC, on 04/28/2019, PICC rupture related to clearing occurred. The radiograph showed the fragment of the catheter with its proximal part intracardiac and the distal part in the right femoral vein. The catheter fragment was large, about 20 cm x 1.9 mm. Because it is of great extent and caliber less than 60% of the lumen of the vessels, the risk of thromboembolic and obstructive complications of the vessel was very small. Therefore, other access for medication and fluids were obtained. The patient remained without abnormalities and was clinically stable.

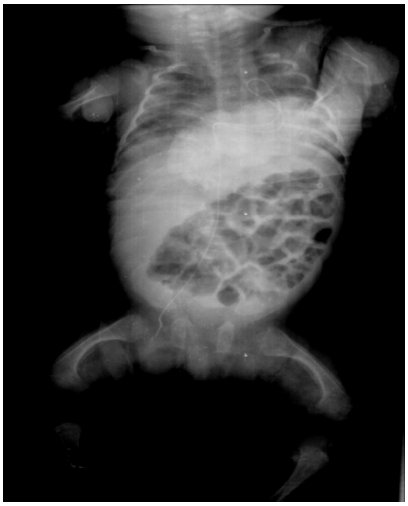

Figure 1. Radiograph

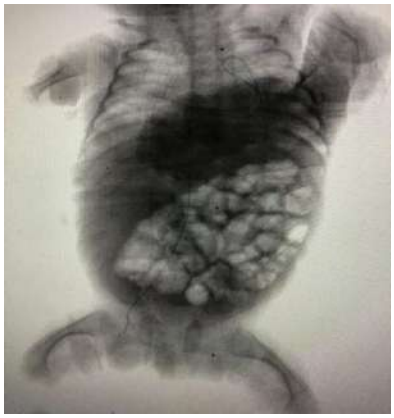

Figure 2. Catheterization

After four days the infant was submitted to interventional catheterization for removal of the foreign body. It was observed by the image that the catheter was in part on the right atrium and part on the right ventricular outflow tract without repercussion. Right femoral vein puncture, 5F introducer passage, insertion of Teflon guidewire was made until the right atrium and with the Judkins catheter the PICC ruptured was moved to the inferior vena cava and the tie catheter was advanced, laced the foreign body and externalized all fragments through by the femoral vein without intercurrences. In the end of the procedure, devices were removed and compressive dressing performed. The procedure was performed with infant intubated with orotracheal cannula number 3 and sedation with midazolam (0.05mg / kg), fentanyl (0.05mg / kg) and ketamine (0.05mg / kg) and mechanical ventilation: respiration rate (45), PEEP (5), inspiration pressure (18), inspiration time (0.45), Fio2 (40%) and peripheral venous access in the right axillary vein.

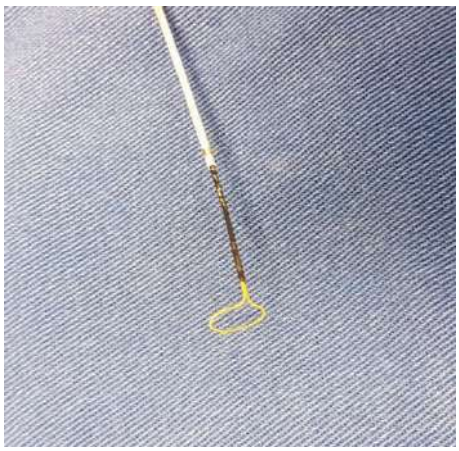

Figure 3. Loop catheter used for extraction

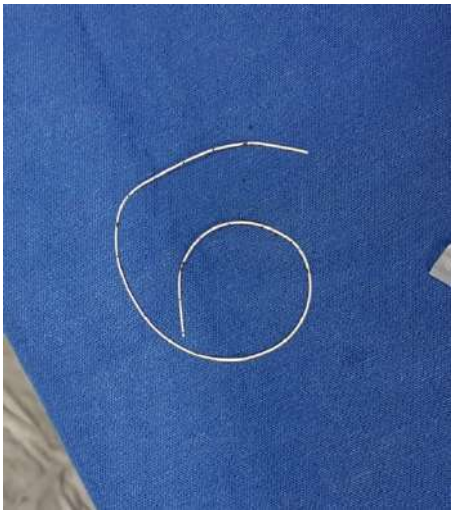

Figure 4. Ruptured PICC catheter (20 cm x 1.9 mm) after extraction

DISCUSSION

It is a worldwide consensus that central venous access devices are essential for the management of critical patients in both intensive care units and inpatient units.[1] Adverse events occur whenever there is inadequate handling. The first care should be made to sure that the tip is properly positioned to avoid perforations of the heart and pericardium avoiding the formation of pleural and pericardial effusions. The second care

is in handling and avoiding extrapolation of the imposed forces, devices of poor technical quality, in addition to the disease and the vascular anatomy of the patient[6-9]. To avoid catheter rupture it is necessary not to use excessive force and syringes with capacity of less than 10ml because they have infusion pressure greater than that supported by the PICC[9-10]. Circuit rupture may or may not be directly related to the length of stay but also to the quality of the infused solutions[11]. The establishment of handling standards, using safe quality catheters, constant checking of catheter position, avoiding infusion of fluids under high pressure are items that should be routine. Furthermore, in preventing catheter rupture, inhibiting the formation of thrombi by means of safe heparinization with maintenance of 1 IU / ml of permeabilizing solution in these devices is indicated by reducing the risk of obstruction and thrombosis or microemboli[12].

Many of these adverse events are asymptomatic, may go unnoticed and undiagnosed. These catheters may undergo incomplete ruptures, with multiple holes in their path, and infusion with multiple leakage similar to a shower[13]. This may be an early, high-risk signal for complete catheter rupture especially during withdrawal. Once ruptured, the possibility of embolization to smaller vessels with obstruction to significant anterograde flow or intracardiac embolization, lung or even arterial circulation by the foramen ovale in the case of neonates and minor children and in patients with intracardiac shunt is great. In these cases, one should avoid handling the patient with unsuccessful attempts at intravascular removal of these foreign bodies. The immediate measure in approaching the patient is not to panic the team of professionals involved in handling the patient, to obtain other access, and to normally maintain the treatment and not attempt to remove the foreign body blindly.Removal of the foreign body should be done electively, planned and scheduled, with the patient clinically stable. Almost always when the foreign body locates in vessels of greater caliber or intracardiac the withdrawal by intervention in hemodynamics is the safest and more successful form. In some cases the surgical approach may be necessary mainly when the foreign body locates in vessels of smaller calibers. In hemodynamics some catheters are specific for removal of foreign bodies from the vascular system.

As the PICC technique popularized much, being universal, it led to the development of devices of smaller caliber more refined. In the absence of the loop catheter for fishing the foreign body there is the possibility of grasping the foreign body with loops made with wire guides of small caliber forming a hook and a loop type in withdrawal[14]. The possibility of serious problems after rupture and embolization of the fragment makes careful observation an excellent tool during the use of the central catheters[15]. Once the adverse event has occurred, monitoring the site of fragment's impaction with radiography is still the best diagnostic and control method these situations. Removal of the foreign body should be planned and scheduled electively and with the patient clinically stable[11]. Desperate measures are more harmful to the patient than the effect of the adverse event itself.In the withdrawal of the embolized fragment, several techniques are used, but those that use the loop-type catheter are currently the most adequate[4]. On the other hand, the hemodynamicist should use the technique that is most familiar and safe. Most interventionalists have the opportunity to encounter a vascular foreign body throughout their clinical practice[13]. Although there is a significant rate of complications in case the catheter fragment remains, the incidental finding in other imaging studies is not uncommon and is reported in 5% to 40% of the cases referred for evaluation[13]. Hence the need to increasingly think about these adverse events and track them continuously. It is very important to implement simple routines, algorithms and good conducts within the hospital units universally among those involved in the treatment. Finding and establishing conduit protocols for such events is essential. Prevent adverse events related to central venous access devices as well as measures to be followed in the occurrence of such adverse events. Remembering that the therapeutic arsenal is in constant modification, procedural increments, new techniques will always arise and reduce the frequency of adverse events is a challenge that should be the goal always.

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Anomalias Vasculares Arteriais Múltiplas em Recém-Nascido. Diagnóstico Ecocardiográfico e Angiográfico

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Múltiplas alterações vasculares arteriais, caracterizadas por enrolamento e torção das artérias pulmonares e aorta foram diagnosticadas mediante ecocardiografia em recém-nascido assintomático com fenótipo sugestivo de síndrome de Ehlers-Danlos. Essas alterações foram posteriormente confirmadas mediante angiografia que apresentava ainda, alterações vasculares periféricas. O eletrocardiograma mostrava provável hemibloqueio do ramo ântero-superior esquerdo e a radiografia de tórax, arco médio escavado com fluxo pulmonar normal.

As anomalias vasculares arteriais múltiplas em crianças são reconhecidas como uma entidade rara e de evolução quase sempre fatal. Poucos casos têm sido descritos desta malformação e a etiologia ainda permanece indefinida[2], sendo referida a sua associação com síndromes como Ehlers-Danlos[3,4] ou cútis laxa congênita[5] ou ainda, secundária a defeitos congênitos do tecido elástico do sistema arterial[6].

Relato do Caso

Recém-nascido de parto normal, a termo, com 3600g, sexo masculino, sem antecedentes gestacionais ou familiares, apresentou com 28 dias de vida, tosse, febre e dispnéia, sendo diagnosticado quadro de bronco-pneumonia e atelectasia em ápice pulmonar direito. Internado foi medicado com ampicilina e amicacina. Na época, foi auscultado sopro sistólico +/4+ na borda esternal esquerda alta, fúrcula, carótida e dorso. Após alta hospitalar, foi encaminhado à nossa instituição para avaliação ambulatorial.

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Recebido para publicação em 13/5/99
Aceito em 13/10/99

O exame físico com 38 dias de vida e 4.370g de peso, revelou alterações gerais caracterizadas por macrocrânia, micrognatia, abdução dos polegares e frouxidão tegumentar e ligamentar (alterações que ficaram mais evidentes durante o acompanhamento clínico ambulatorial); sem edema, sem visceromegalia; freqüência cardíaca de 158bpm e respiratória de 40 ipm; pressão arterial de 110x70mmHg e ausculta cardíaca e pulmonar normais. A avaliação pelo setor de genética reforçou a hipótese diagnóstica clínica de síndrome de Ehlers-Danlos.

O eletrocardiograma mostrou ritmo sinusal, com provável hemibloqueio ântero-superior esquerdo (eixo elétrico a -45°), e a radiografia de tórax levocardia com área cardíaca de tamanho normal e arco médio escavado; área pulmonar normal.

Foi indicado estudo ecocardiográfico que mostrou: *situs solitus*, concordâncias atrioventricular e ventrículo-arterial, septo interatrial com membrana da fossa oval aneurismática, ocluindo defeito tipo *ostium secundum* e sem fluxo interatrial (fig. 1); septo interventricular íntegro e câmaras cardíacas com dimensões e função normais. O tronco arterial pulmonar não apresentava a bifurcação habitual, com a origem das artérias pulmonares em sua região posterior. Havia entrecruzamento no trajeto dos ramos, sendo que o ramo esquerdo se originava na região póstero-direita e, após curto trajeto, dirigia-se para o pulmão esquerdo, enquanto que o ramo direito com origem póstero-esquerda dirigia-se para o pulmão direito (fig. 1). A aorta ascendente apresentava trajeto normal, e o arco aórtico à esquerda, retificado, dirigia-se para a região póstero-esquerda do tórax, não sendo possível visibilizar a aorta descendente (fig. 2). Os vasos da base apresentavam trajeto anômalo com o tronco braquiocefálico fazendo um percurso circular antes da sua bifurcação. Os outros vasos, incluindo a artéria subclávia esquerda apresentavam vários pontos de tortuosidade e entrecruzamento no seu trajeto, em espiral (fig. 1). A aorta abdominal foi bem visibilizada, com tortuosidade evidente ao nível do diafragma (imagem em "sifão") com o trajeto retrocardíaco afastado do átrio esquerdo (fig. 2).

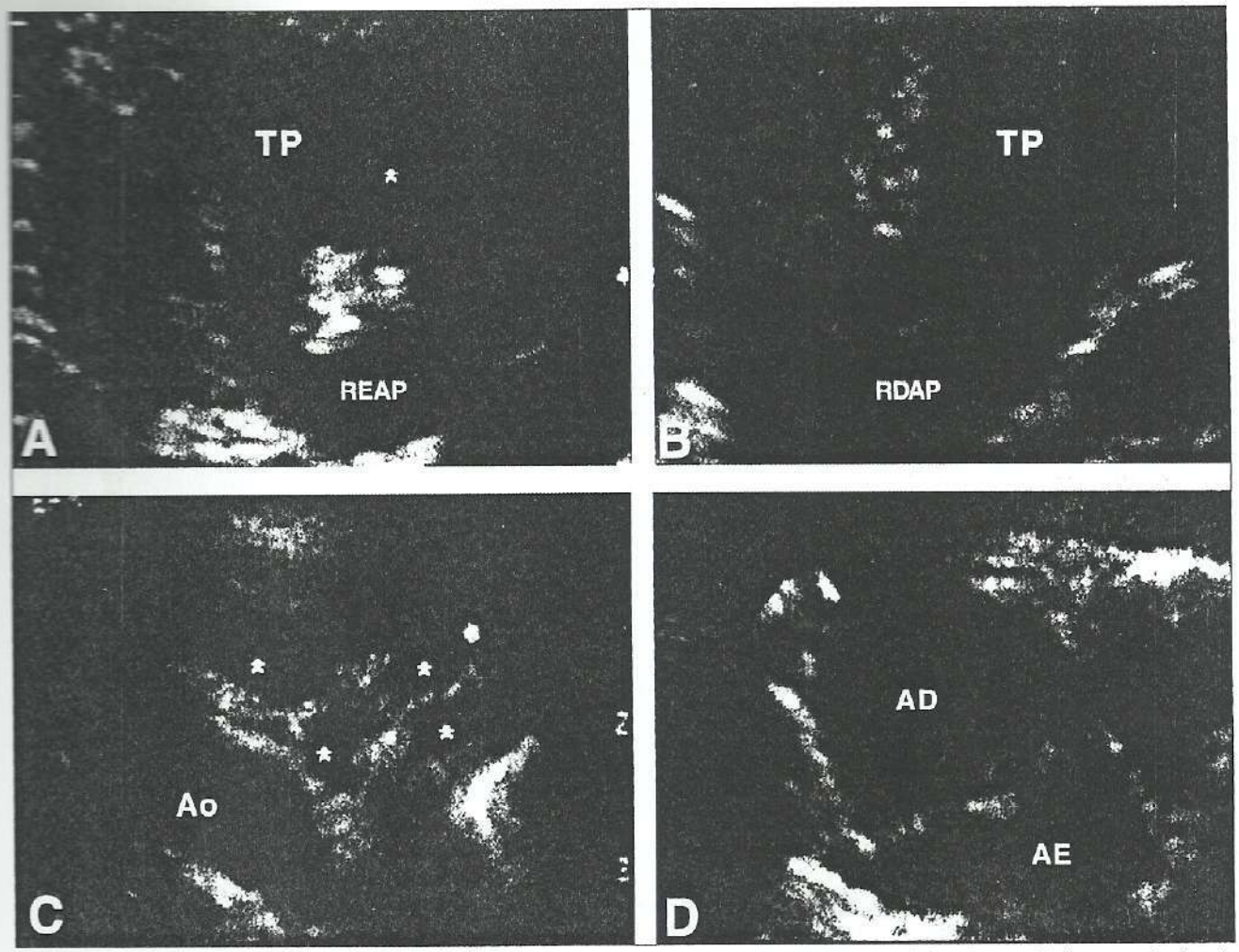

Fig. 1 - Dados ecocardiográficos mostrando: A e B) o entrecruzamento das artérias pulmonares na imagem paraesternal transversal. O asterisco em A indica o ponto de origem do RDAP. Observar a secção transversal do tronco pulmonar e longitudinal das artérias pulmonares; C) vasos da base (*) registrados nos planos longitudinal e transversal na mesma imagem ecocardiográfica; D) septo interatrial. REAP e RDAP- artérias pulmonares esquerda e direita, respectivamente; Ao- aorta; TP- tronco pulmonar; AD e AE- átrios direito e esquerdo, respectivamente.

Com 11 meses de vida o paciente foi submetido a estudo angiográfico que confirmou os achados ecocardiográficos (fig. 3). Após a origem anômala das artérias pulmonares, existia um trajeto espiralar das mesmas no seu terço distal, antes da sua bifurcação. O arco aórtico, à esquerda, dava origem aos vasos da base, que apresentavam tortuosidade e enrolamento em forma helicoidal. Após esse ponto, a aorta se dirigia transversalmente para a região posterior, ficando longe da sua posição habitual, e a sua porção descendente, com vários pontos de tortuosidade, apresentava torção sobre seu próprio eixo, com um curto trajeto ascendente e novamente se dirigia para a região inferior ao nível do diafragma. (imagem em "sifão"). As artérias coronárias apresentavam origem normal, a artéria coronária esquerda, porém, apresentava alguns pontos de tortuosidade e múltiplas irregularidades.

Foi optado por acompanhamento ambulatorial da criança, estando, na última consulta, com dois anos de idade, assintomática e com exame físico normal.

Discussão

Este é um caso extremamente raro de diagnóstico ecocardiográfico de múltiplas anomalias vasculares arteriais. A angiografia confirmou os achados e ainda mostrou outras alterações distais das artérias pulmonares, bem como da aorta descendente. É também, de nosso conhecimento, o primeiro relato de recém-nascido com este tipo de malformação. Em 1858, Coulson[1] descreveu uma tortuosidade da artéria carótida, visível como uma tumoração na região cervical. Relatos de malformações múltiplas, porém, são muito mais raras. Ertugrul[6] relatou em 1967 uma criança de 10 anos de idade, sintomática, com múltiplas alterações da aorta e seus ramos, não sendo encontrada alteração metabólica ou doença sistêmica associada, tendo sido postulado que esta malformação seria decorrente de defeito congênito do tecido elástico do sistema arterial. O entrecruzamento das artérias pulmonares é também referido como uma entidade rara, em alguns casos associada a

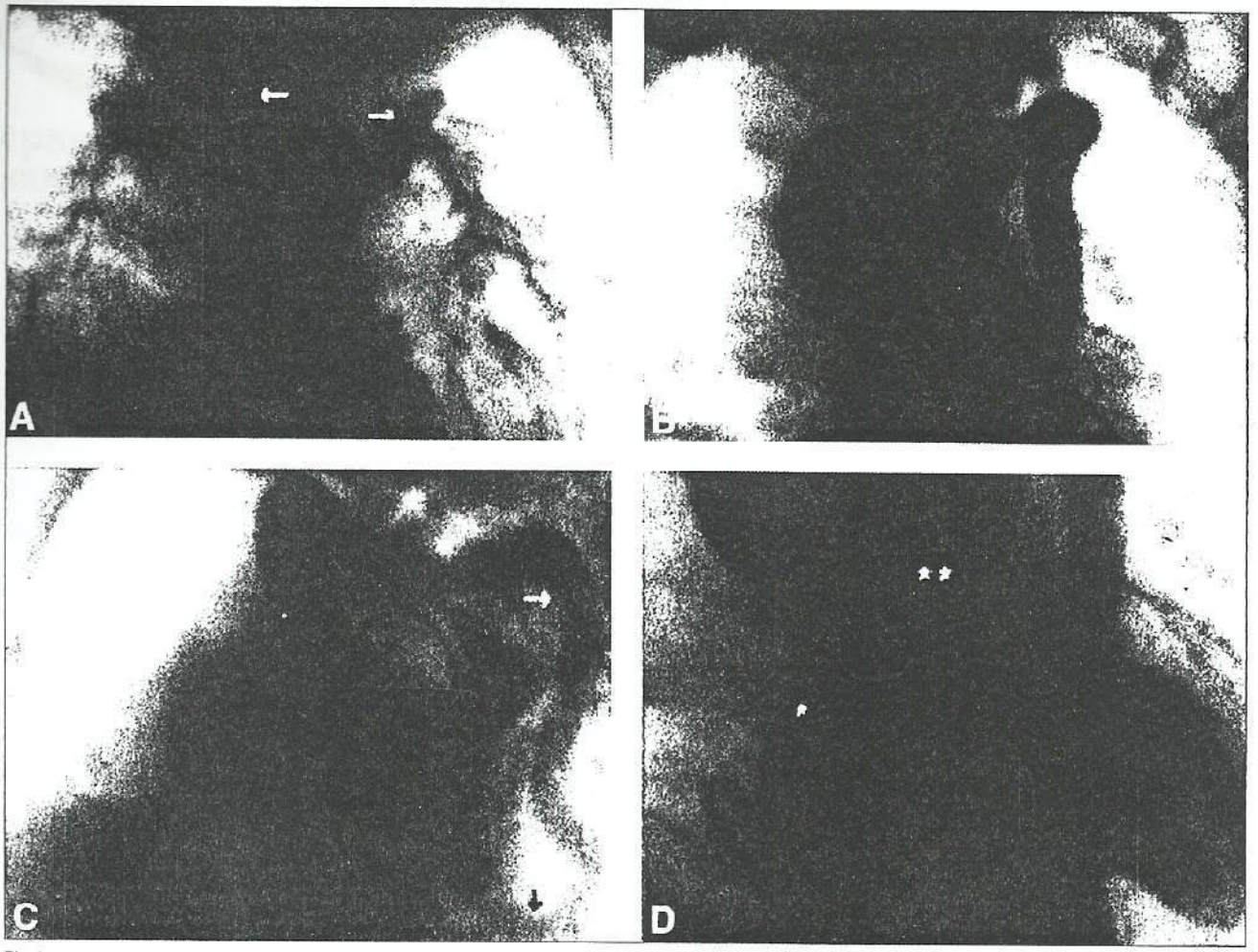

Fig. 3 - Dados angiográficos mostrando: A) origem e trajeto anômalo das artérias pulmonares. As flechas indicam ponto de torção após a sua origem; B) direção anômala do arco aórtico. Os vasos da base apresentam trajeto em espiral; C) aorta descendente com trajeto anômalo e vários pontos de tortuosidade. A flecha indica o ponto de tortuosidade identificado no ecocardiograma (fig. 2e); D) artérias coronárias, direita (*) e esquerda (**).

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Uso de inibidores da enzima conversora de angiotensina em neonatos e lactentes

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O uso de vasodilatadores em crianças, apesar de relativamente recente, tem papel importante no tratamento da insuficiência cardíaca congestiva e baixo débito cardíaco. Nos últimos anos, houve grande progresso no entendimento dos mecanismos celular e molecular envolvidos na regulação do tônus do músculo liso da parede vascular, possibilitando o desenvolvimento de novas classes de drogas, dentre elas os inibidores da enzima conversora de angiotensina.

A terapêutica clássica para o tratamento de crianças com insuficiência cardíaca congestiva secundária a grande "shunt" consiste no uso de digoxina e diuréticos; caso essa terapêutica não seja efetiva, um inibidor da enzima conversora de angiotensina, em geral o captopril, é então adicionado. Apesar de existirem vários estudos quanto a indicação e efeitos hemodinâmicos agudos e tardios em adultos, existem apenas raros estudos sobre seus efeitos em crianças, particularmente em neonatos e lactentes.

Descritores: insuficiência cardíaca congestiva, inibidores da enzima conversora de angiotensina, criança, neonato.

(Rev Soc Cardiol Estado de São Paulo 1999;5:690-4)

RSCESP (72594)-909

Introdução

O uso de vasodilatadores em crianças, apesar de relativamente recente, tem papel importante no tratamento da insuficiência cardíaca congestiva e baixo débito cardíaco. Houve grande desenvolvimento dos estudos nessa área, principalmente no que diz respeito a mecanismos celular e molecular envolvidos na regulação do tônus do músculo liso da parede vascular; como conseqüência, novas classes de drogas foram desenvolvidas.

Apesar de existirem numerosos estudos sobre a indicação do uso de vasodilatadores e de seus efeitos hemodinâmicos agudos e tardios em adultos[1-3], existem apenas raros estudos publicados que descrevem os efeitos da terapia vasodilatadora em crianças, especialmente em neonatos e lactentes.

Em crianças, por apresentarem diferenças fisiopatológicas da função miocárdica relacionadas à idade e da fisiologia cardiovascular em comparação com a população adulta, seria inapropriado transferir a experiência de seu uso em adultos como base para predizer o resultado da terapia vasodilatadora em crianças.

Indicações

Muitas das crianças portadoras de cardiopatia congênita necessitam de tratamento clínico para insuficiência cardíaca congestiva. Classicamente, os digitálicos e diuréticos há muito tempo vêm sendo utilizados, mas recentemente o valor desse tratamento chegou a ser questionado, principalmente, quanto ao uso de digoxina[4]. A terapêutica alternativa, usando vasodilatadores de outro grupo, não havia provado ser completamente satisfatória; todavia, após a disponibilidade dos inibidores da enzima conversora de angiotensina, foi observado aumento das expectativas de melhora do resultado[5,6].

Os inibidores da enzima conversora de angiotensina bloqueiam a conversão do peptídeo inativo angio-

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Ecocardiografia com contraste em crianças: experiência inicial

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A ecocardiografia com contraste foi inicialmente empregada em Cardiologia Pediátrica para obtenção de informações adicionais sobre as anormalidades anatômicas e para otimizar os estudos de fluxo. Alguns pacientes portadores de cardiopatia congênita apresentam potencial para desenvolvimento de isquemia miocárdica, como, por exemplo, portadores de origem anômala de artéria coronária esquerda, e no pós-operatório de cirurgia de Jatene. Já outras cardiopatias, especialmente aquelas hipoxêmicas, são capazes de causar alterações das propriedades intrínsecas da fibra miocárdica, comprometendo sua atividade contrátil, necessitando de avaliação da perfusão miocárdica. Apresentamos nossa experiência inicial com a ecocardiografia de contraste. Nosso estudo teve por objetivo avaliar a perfusão miocárdica em doenças cardíacas congênitas com potencial de isquemia miocárdica. Foram estudados 20 pacientes com idade entre 8 dias e 33 anos de vida, média de 2 anos e 6 meses; 12 desses pacientes eram do sexo masculino e 8, do sexo feminino. O peso variou de 3 kg a 57 kg (mediana, 16,5 kg). A mistura adequada das microbolhas com sangue propiciou ótimas imagens e mapeamento adequado dos ventrículos direito e esquerdo. Com base observacional de escore de perfusão, 15 pacientes demonstraram hipoperfusão global e 12 tinham algum grau de disfunção ventricular. Esses resultados demonstram perfusão miocárdica diminuída em algumas cardiopatias congênitas, sem aparente disfunção ventricular em repouso. Essas evidências podem fornecer esclarecimentos importantes sobre a existência de isquemia miocárdica em crianças e possivelmente prever aquelas que poderão desenvolver disfunção ventricular no futuro.

Descritores: ecocardiografia com contrastes, dobutamina, crianças.

(Rev Soc Cardiol Estado de São Paulo 1999;5:742-8)

RSCESP (72594)-916

Introdução

O uso de contrastes veio complementar as técnicas usuais de ecocardiografia, uma vez que estas, em algumas situações, não são suficientes para avaliar os mecanismos que levam à disfunção ventricular.

Os meios de contraste tornaram-se comercialmente disponíveis a partir da década de 80. Os contrastes desenvolvidos inicialmente tinham meia-vida curta e as bolhas eram muito grandes para ultrapassar o leito capilar pulmonar, tornando, portanto, impossível a opacificação do coração esquerdo.

A ecocardiografia com contrastes foi inicialmente usada em cardiopatia congênita para obtenção de informações adicionais sobre as anormalidades anatômicas cardiocirculatórias, incluindo a análise de morfologia ventricular, e para otimizar os estudos de fluxos, como, por exemplo, regurgitação valvar tricúspide, e, assim, obter a pressão pulmonar ou quantificar melhor as estenoses valvares[1,2].

Acreditamos, porém, que os estudos de perfusão miocárdica são de grande interesse para a Cardiologia Pediátrica. Vários tipos de cardiopatias congênitas apresentam potencial para desenvolvimento de isquemia miocárdica, entre elas origem anômala de artéria coronária esquerda do tronco pulmonar e pós-

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Febre reumática

Rheumatic fever

Unitermos: febre reumática, cardite reumática.
Uniterms: rheumatic fever, rheumatic carditis.

RESUMO

Os critérios para estabelecer o diagnóstico da febre reumática são baseados nas manifestações clínicas e nos exames laboratoriais. Os corticosteróides são indicados para os pacientes com cardite. Aconselha-se a redução da dosagem por volta da segunda semana de tratamento e a suspensão quando todos os sinais de atividade, tanto clínicos quanto laboratoriais tiveram regredido. O incremento de melhores condições sociais e a terapêutica com antibióticos no tratamento das infecções estreptocócicas podem diminuir a incidência da febre reumática.

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MANIFESTAÇÕES CLÍNICAS

A febre reumática é doença que compromete vários sistemas, afetando primariamente o coração, as articulações, o cérebro, o tecido subcutâneo e a pele. Suas manifestações variam de acordo com os órgãos envolvidos e com o grau de comprometimento desses órgãos, sendo o quadro clínico polimorfo.

CARDITE

A cardite é a mais grave e a mais importante manifestação da doença, pois é o único componente da febre reumática que pode levar a sequelas. Ela pode manifestar-se em graus variados, desde a forma discreta, apenas revelada por exames complementares, até a forma fulminante, que pode acarretar a morte em curto período.

A cardite é vista em aproximadamente 50% dos pacientes e pode apresentar-se como única manifestação ou em associação com uma ou mais das outras manifestações da febre reumática. Ocasionalmente, a artrite pode preceder a cardite, e esta geralmente aparece duas semanas após o início da artrite. As evidências de cardite podem ser bem discretas, como nos pacientes portadores de coréia. A taquicardia é um dos sinais clínicos precoces da cardite e a determinação da frequência cardíaca é mais confiável quando o paciente está dormindo. Febre pode elevar a frequência cardíaca e arritmias transitórias podem ocorrer, todavia o bloqueio atrioventricular total (BAVT) não é usualmente visto na cardite reumática.

Em geral, os sinais mais consistentes de cardite reumática incluem a presença de um sopro patológico, particularmente de insuficiência mitral, evidência de cardiomegalia progressiva e de insuficiência cardíaca congestiva (ICC) e sinais de pericardite[1].

A insuficiência mitral é caracterizada por um sopro suave, holossistólico, no ápice, irradiando para a axila, e é mais audível com o paciente em decúbito lateral esquerdo. Na insuficiência mitral grave, um sopro diastólico de enchimento, chamado de Carey-Combs, é gerado pela grande quantidade de sangue que passa através da valva mitral para dentro do ventrículo esquerdo durante a fase de enchimento.

A insuficiência aórtica ocorre em 20% dos pacientes com cardite reumática e pode ser isolada, mas usualmente é associada com insuficiência mitral. Caracteriza-se pela presença de um sopro protodiastólico que se inicia com o componente aórtico da segunda bulha.

A ICC franca ocorre em aproximadamente 5% dos pacientes com febre reumática. Suas manifestações incluem: tosse, dor torácica, taquipnéia, dispnéia, ortopnéia e irritabilidade,

Na evolução, com o controle da atividade reumática há melhora das manifestações clínicas da fase aguda e pela normalização dos exames complementares. As provas laboratoriais mais comumente usadas no acompanhamento são a velocidade de hemossedimentação, a proteína C reativa, a dosagem de antiestreptolisina O e a dosagem de mucoproteínas (a-2 globulina). A dosagem de mucoproteínas parece refletir mais fielmente o controle da doença.

TRATAMENTO DA INSUFICIÊNCIA CARDÍACA

A insuficiência cardíaca deve ser tratada com digital, diuréticos e vasodilatadores. Os digitálicos podem ser usados, mas na vigência de cardite existe sensibilidade maior ao digital e a queda da frequência cardíaca só ocorrerá após o controle da atividade da doença. Os diuréticos estão indicados nos casos com congestão venosa. Os vasodilatadores, como, por exemplo, os inibidores da enzima de conversão da angiotensina, exercem papel importante nos casos com regurgitações valvares.

TRATAMENTO CIRÚRGICO

O tratamento de escolha na fase aguda é o tratamento clínico, mas em alguns raros casos este tratamento se torna insuficiente, devido à grave regurgitação valvar, sendo necessário tratamento cirúrgico de urgência. A insuficiência mitral pode ficar refratária ao tratamento clínico quando há rotura de cordoalhas como complicação da atividade reumática.

SUMMARY

The criteria, for establishing the diagnosis of rheumatic fever are based upon clinical and laboratory datas. The use of corticosteroids is advised in patients with carditis. Usually the corticosteroids can be tapered off by the second week of treatment and discontinued when all signs of activity (clinical manifestations and laboratory test) have fully regressed.

I'm proved social conditions and antibiotic therapy of streptococus infeccion may decrease the incidence of rheumatic fever.

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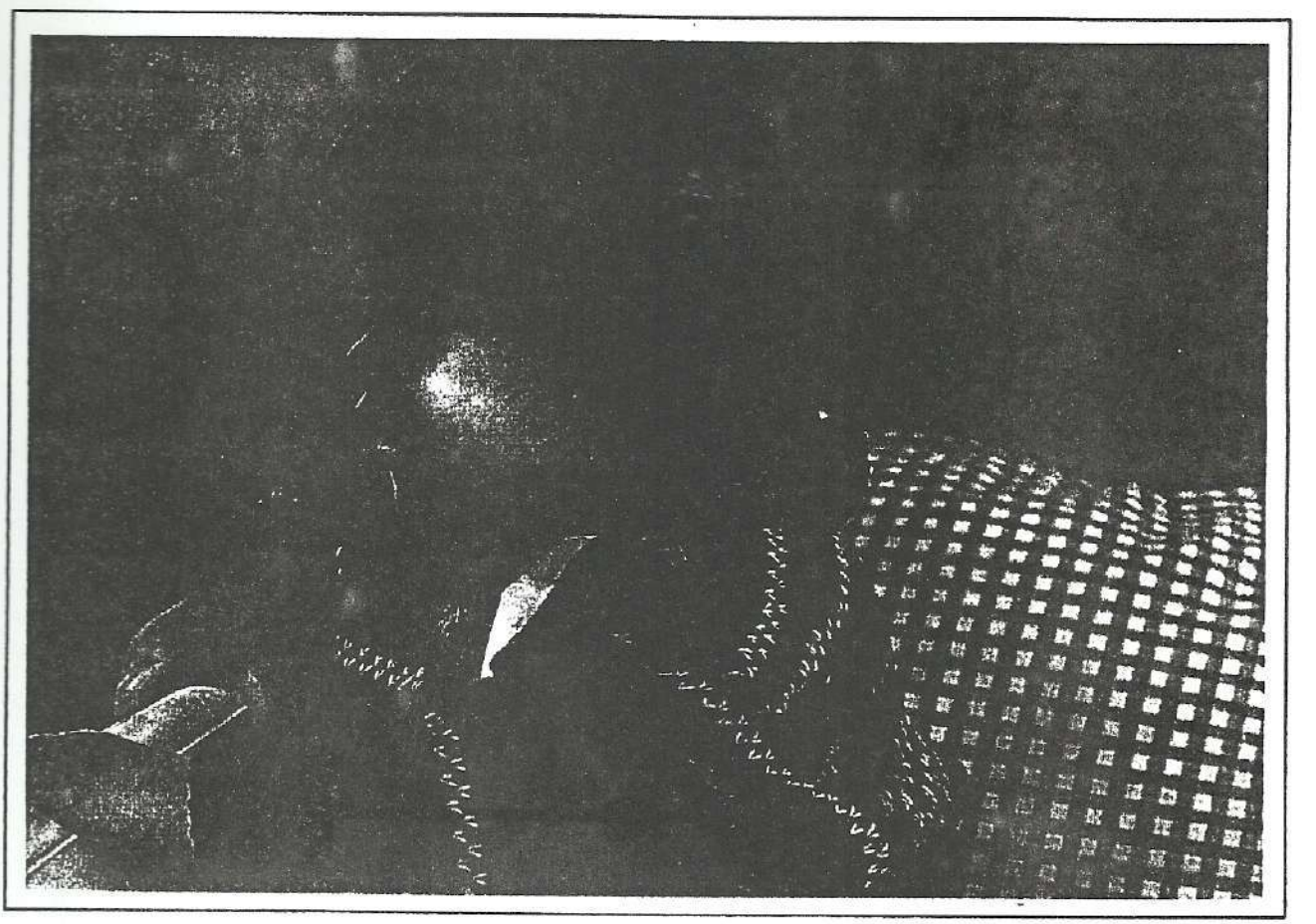
ISSN 0103-2712

PEDIATRIA ATUAL



Neste número:

Vol. 10
JULHO, 1997

7

Abscesso Hepático Ascaridiano na Criança

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Resumo

Os autores relatam um caso de criança de dois anos e cinco meses, do sexo masculino, negra, procedente da cidade de São Paulo, com diarréia aguda inespecífica, complicada com pneumonia, abscesso pulmonar e empiema, acompanhada transitoriamente de eliminação de áscaris no vômito e fezes. Desenvolveu choque séptico e disfunção de múltiplos órgãos e sistemas, com óbito. Na necropsia evidenciou múltiplos abscessos — estendendo-se do fígado, diafragma e pleura até o parênquima pulmonar —, desencadeados pelo parasita.

Aceito para publicação em 10/10/95

Introdução

A infestação por *Ascaris lumbricoides* é a doença helmíntica mais comum. Abrange um quarto da população mundial, incidindo mais na América do Sul, Ásia e África (1).

O ciclo vital do áscaris inicia após a ingestão de ovos com alimentos como frutas e verduras. O ovo eclode e libera larvas rabditóideas, que persistem na luz intestinal ou penetram nas vênulas e linfáticos. Mediante estes vasos as larvas alcançam os pulmões, coração e fígado (15).

ATUALIZAÇÃO TERAPÊUTICA

AT

FELÍCIO CINTRA DO PRADO // JAIRO DE ALMEIDA RAMOS // JOSÉ RIBEIRO DO VALLE

DIAGNÓSTICO E TRATAMENTO

Versão impressa
desta obra: 2018

26ª
EDIÇÃO

artes médicas

2018

A886 Atualização terapêutica de Prado, Ramos e Valle : diagnóstico e tratamento [recurso eletrônico] / Presidente da comissão editorial, Emilia Inoue Sato. – 26. ed. – São Paulo : Artes Médicas, 2018.

Editado como livro impresso em 2018.
ISBN 978-85-367-0269-8

1. Clínica médica. 2. Atualização terapêutica. I. Sato, Emilia Inoue.

CDU 616-07-043.84

Catalogação na publicação: Poliana Sanchez de Araujo – CRB 10/2094

- A cardite subclínica, que se apresenta sem sopro cardíaco, porém com alteração ao ecocardiograma, deve ser considerada e abordada como a cardite clínica tanto no tratamento como na duração da profilaxia.
- Não é recomendável o uso de anti-inflamatórios não hormonais, inclusive o ácido acetilsalicílico, até que se confirme o diagnóstico de FR.
- A prevenção dos episódios iniciais de FR aguda e das recorrências depende do controle das IVAS pelo estreptococo do grupo A; por isso, a importância do tratamento precoce dessas infecções e a adesão à profilaxia secundária.

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68

ABORDAGEM DAS CARDIOPATIAS CONGÊNITAS

CÉLIA MARIA CAMELO SILVA

LOURDES DE FÁTIMA GONÇALVES GOMES

LUCIANA PONSECA DA SILVA

A incidência das cardiopatias congênitas (CCs) varia de 8 a 10 para cada 1.000 nascidos-vivos, abrangendo desde lesões leves e assintomáticas ao longo de toda a vida a cardiopatias muito complexas. A etiologia multifatorial é atribuída a maioria, embora algumas estejam associadas a síndromes genéticas, ação de agentes teratogênicos e doenças maternas, tais como lúpus eritematoso sistêmico (LES) e diabetes melito (DM).

DIAGNÓSTICO

HISTÓRIA CLÍNICA

- História familiar de cardiopatias congênitas.
- História de doenças maternas: diabetes, rubéola, medicações teratogênicas, etc.
- Intercorrências perinatais: prematuridade, aspiração de mecônio, asfixia perinatal.
- Achados de exames pré-natal: malformação cardíaca, arritmia fetal, hidropsia.

EXAME FÍSICO

- Dismorfismo: Trissomia do 21, 18 e 13, etc.
- Cianose central e diferencial (teste do "coraçãozinho positivo"). Ver Quadro 68.1.
- Taquipneia.
- Pulsos – amplitude, simetria, regularidade.
- Sopro cardíaco.
- Hepatomegalia.

QUADRO 68.1 ■ Causas de cianose na criança

CARDIOPATIA CONGÊNITA CIANÓTICA

Obstrução ao fluxo pulmonar
- Atresia pulmonar, estenose pulmonar crítica, tetralogia de Fallot

CONEXÃO VENTRÍCULO ARTERIAL DISCORDANTE

Transposição das grandes artérias
Mistura intracardíaca
- Coração com fisiologia univentricular, tronco arterioso, atresia tricúspide, drenagem anômala total de veias pulmonares

DOENÇAS PULMONARES PRIMÁRIAS

Doenças parenquimatosas
- Síndrome de aspiração de mecônio, síndrome do desconforto respiratório, pneumonia congênita

Doenças extraparenquimatosas
- Pneumotórax, hérnia congênita diafragmática

HIPERTENSÃO PULMONAR DO RECÉM-NASCIDO

Primária
Secundária
- Síndrome de aspiração de mecônio
- Asfixia perinatal, hérnia diafragmática congênita
- Policitemia congênita
- Meta-hemoglobinas

EXAMES COMPLEMENTARES

- **Radiografia torácica:** auxilia na formulação da hipótese diagnóstica, e no diagnóstico diferencial com problemas respiratórios.

- **Eletrocardiograma (ECG)**: pode ser típico em algumas cardiopatias, auxilia no seguimento e é fundamental no diagnóstico de arritmia.
- **Ecocardiograma**: exame fundamental e definitivo para o diagnóstico das CCs – permite definição anatomo-morfológica e funcional.
- **Ressonância magnética (RM) e tomografia computadorizada (TC)**: exames indicados para melhor definição de estruturas não bem visualizadas pelo ecocardiograma, como: ramos periféricos das artérias pulmonares, artérias coronárias, aorta descendente, etc.
- **Cateterismo cardíaco**: complementa o diagnóstico do ecocardiograma e tem papel importante no tratamento (cateterismo terapêutico).
- **Teste da hiperóxia**: consiste em administrar O_2 a 100% por 15 minutos e coletar gasometria da artéria radial direita. Nas casas de cardiopatia cianogênica, a pO_2 será <100 mmHg e a elevação na pO_2 < 20 mmHg.
- **Teste da oximetria de pulso "coraçãozinho"**: consiste em medir a saturação arterial de oxigênio (SaO_2) simultaneamente no membro superior direito e em um dos membros inferiores entre 24 a 48 horas. SaO_2 abaixo de 95% ou diferença entre os membros > 3% requer investigação para afastar cardiopatia.

■ QUADRO CLÍNICO

As formas mais frequentes de apresentação clínica das cardiopatias são: cianose, insuficiência cardíaca (IC), choque cardiogênico ou achado de sopro (ver Quadros 68.1, 68.2 e 68.3).

- **Cianose central**: resultante de hipoxemia arterial, devido a shunt direito-esquerda e/ou hipofluxo pulmonar.
- **Crise de hipóxia**: episódios súbitos de cianose severa, hiperpneia, acidose metabólica, que ocorrem em pacientes com fisiologia de tetralogia de Fallot. O mecanismo provável é o de espasmo da infundíbulo do ventrículo direito (VD). Ocorrem principalmente entre 3 e 6 meses, pela manhã, e pode ser precipitada pelo choro, alimentação e ato de evacuar (ver Quadros 68.4 e 68.5).
- **IC**: é definida como a incapacidade do coração em prover débito cardíaco (DC) adequado para atender à demanda metabólica do organismo. As CCs representam as causas mais frequentes de IC na infância e o seu quadro clínico varia com a idade de apresentação. Nos recém-nascidos e lactentes, caracteriza-se por: dificuldade para alimentação; tempo prolongado e interrupção frequente das mamadas, diaforese, infecções respiratórias recorrentes e hipodesenvolvimento. Nas crianças maiores, predomina a queixa de dispneia e cansaço aos esforços.
- **Choque cardiogênico**: caracteriza-se por hipotensão, cianose, acidose metabólica e oligúria. Pode ser difícil diferenciar entre sepse e distúrbios metabólicos.

■ TRATAMENTO (VER QUADRO 68.4)

Principais medicamentos utilizados no tratamento da IC.

1 | **Diuréticos**
- Furosemida – dose 1 a 6 mg/kg/d, VO ou EV.
- Espironolactona – dose 1 a 3 mg/kg/d.

2 | **Vasodilatadores**
- IECA
- Captopril – 0,5 a 4 mg/kg/d entre duas a quatro tomadas.
- Enalapril – 0,8 a 1 mg/kg/d, uma a duas vezes/dia.

QUADRO 68.2 ■ Causas de insuficiência cardíaca no recém-nascido

I | CARDIOPATIAS CONGÊNITAS (CAUSAS ESTRUTURAIS)

1 | Obstrução à via de saída do VE

Síndrome da hipoplasia do coração esquerdo, EAo crítica, CoAo importante

2 | Regurgitação valvar importante

Tronco arterioso com regurgitação do valve truncal, insuficiência mitral na síndrome de Marfan neonatal, insuficiência tricúspide na anomalia de Ebstein

3 | Grande shunt E – D

PCA, CIV, tronco arterioso, Grandes artérias colateais sistêmico-pulmonares

Obstrução à drenagem venosa pulmonar

Drenagem anômala total das veias pulmonares forma obstrutiva

II | MIOCARDIOPATIAS

- Miocardiopatia (por exemplo: filho de mãe diabética, familiar e idiopática)
- Isquêmica – origem anômala da artéria coronária esquerda e asfixia perinatal
- Miocardites

III | ARRITMIAS

Taquicardia supraventricular, flutter atrial, BAV total

IV | SEPSE/ENDÓCRINA

- Anemia importante
- Tireotoxicose neonatal
- Sepse

VE: ventrículo esquerdo; EAo: estenose aórtica; CoAo: coarctação de aorta; PCA: persistência do canal arterial; CIV: comunicação interventricular; BAV: bloqueio atrioventricular.

QUADRO 68.3 ■ Cardiopatias congênitas que podem se manifestar com quadro de choque cardiogênico

CIRCULAÇÃO SISTÊMICA DEPENDENTE DO CANAL ARTERIAL

CoAo, EAo crítica, síndrome hipoplásicos do coração esquerdo, interrupção do arco aórtico

CIRCULAÇÃO PULMONAR DEPENDENTE DO CANAL ARTERIAL

Atresia pulmonar, estenose pulmonar crítica, atresia tricúspide com atresia pulmonar, ventrículo único com atresia pulmonar

FALTA DE MISTURA

Transposição das grandes artérias

BAIXO DÉBITO SISTÊMICO E EDEMA PULMONAR

Drenagem anômala total das veias pulmonares na forma obstrutiva

1 | **Betabloqueadores**
- Carvedilol – dose de 0,3 a 1 mg/kg/d em duas tomadas.

- Metropolol – 1 a 6 mg/kg/d em duas vezes.

QUADRO 68.4 ■ Crise de hipoxia

4 | **Agentes inotrópicos e vasopressores** – estão indicados na IC grave e choque cardiogênico.

5 | **Digitálicos** – papel controverso, indicado nos casos de IC com taquicardia.

- Digoxina – 5 a 10 µg/kg/d dividido em duas tomadas.
- Agentes inotrópicos EV: dopamina, dobutamina, epinefrina, norepinefrina, milrinone, levosimendan, vasopressina.

TRATAMENTO ESPECÍFICO PARA AS CARDIOPATIAS MAIS COMUNS

O estabelecimento da etiologia definitiva é de importância crucial; assim, o tratamento é direcionado para a causa de base, por exemplo, tratamento cirúrgico e/ou percutâneo para as CCs e implante de marca-passo para BAV.

Cardiopatias acianóticas

1 | **Comunicação interatrial (CIA)**

- **CIAs pequenas < 5 mm:** habitualmente não necessitam de tratamento.
- **CIAs moderadas e grandes:** fechamento eletivo entre os 4 a 6 anos de vida. A forma mais comum é a do tipo *ostium secundum* (80%), sendo a maioria delas elegíveis para fechamento percutâneo pelo cateterismo.

2 | **CIV**

- **CIVs pequenas:** habitualmente não necessitam tratamento e apresentam alta taxa de fechamento espontâneo no decorrer do tempo. Quando subaórtica, existe o risco de prolapso da valva aórtica.
- **CIVs moderadas:** fechamento cirúrgico está indicado quando apresentar:
  a | IC.
  b | Cardiomegalia à radiografia torácica, ou dilatação das cavidades cardíacas ao ecocardiograma.
  c | Prolapso da valva aórtica com ou sem regurgitação.
  d | Ao menos um episódio de endocardite bacteriana.

QUADRO 68.5 ■ Tratamento da crise de hipoxia

e | Infecções pulmonares de repetição.
f | Baixo ganho ponderal sem outras causas.

- **CIVs grandes:** fechamento cirúrgico precoce, preferencialmente a partir do terceiro e antes dos seis meses de vida.
- Cerclagem pulmonar seguida de fechamento de CIV está indicada nos casos com CIVs múltiplas.

3 | **Defeito do septo atrioventricular (DSAV)**

- **DSAV parcial (CIA *ostium primum*):** cirurgia eletiva está indicada por volta dos 4 anos de vida ou mais cedo quando pacientes sintomáticos ou com severa regurgitação da valva atrioventricular.
- **DSAV total (CIA *ostium primum*, CIV do via de entrada, valva atrioventricular única):** correção cirúrgica primária antes do oito meses de vida para prevenção da doença pulmonar obstrutiva crônica (DPOC), especialmente nos portadores de trissomia do cromossomo 21. Nos demais pacientes não sindrômicos, a correção pode ser mais tarde até por volta dos 2 anos de idade.

4 | **PCA**

- **PCAs pequenos:** na ausência de sopro, há controvérsia, se há ou não necessidade de fechamento. Nos pacientes com sopro audível, está indicada, devido ao risco de endocardite e, preferencialmente, por via percutânea.
- **PCAs moderados e grandes:** a época do fechamento depende da gravidade dos sintomas de IC, tamanho do PCA e peso da criança. Nos casos de crianças pequenas com PCA grande, o fechamento cirúrgico é o melhor opção, ao passo que nas crianças maiores, o fechamento pode ser realizado por via percutânea.

Lesões obstrutivas

1 | **EP**

- **EP Leve** – gradiente máximo (pico) < 50 mmHg – não requer tratamento.
- **EP moderada a importante** – gradiente > 50 mmHg. Valvoplastia pulmonar com balão é o tratamento de escolha.

2 | **EP crítica do recém-nascido:** estabilização temporária com prostaglandina E1. Requer tratamento ainda no berçário, nos casos com ventrículo direito bem desenvolvido, está indicada a valvoplastia pulmonar com

balão com ou sem implante de *stent* no canal arterial. Casos com ventrículo direito bi ou unipartite requerem cirurgia para confecção de shunt sistêmico-pulmonar (Blalock-Taussig) e em casos selecionados ampliação da via de saída do ventrículo direito.

3 | **CoAo**

- **Coarctação severa do recém-nascido**: correção cirúrgica precoce, seja para correção da CoA e fechamento da CIV quando presente (correção primária), ou em dois estágios: correção da CoAo com ou sem cerclagem da artéria pulmonar, seguida pelo fechamento posterior da CIV.
- Crianças maiores com coarctação localizada (assintomáticas)
  a | Achado incidental de hipertensão arterial ou de sopro
  b | Opções terapêuticas – aortoplastia primária por balão, implante de *stent* ou correção cirúrgica. A escolha depende da morfologia da CoAo e da idade da apresentação. A opção cirúrgica é a de escolha no primeiro ano de vida.

Cardiopatias cianogênicas

1 | **Tetralogia de Fallot**

A maioria dos casos são elegíveis para correção primária entre os seis meses e 2 anos de vida. Indicações para cirurgia paliativa (*shunt* Blalock-Taussig) são raras. Está indicada em situações especiais:

- Crises de hipóxia ou cianose importante em crianças menores de 6 meses.
- Artérias pulmonares pequenas, no intuito de promover o seu crescimento.
- Trajeto anômalo de artéria coronária cruzando a via de saída do VD.

Após a correção total, os pacientes necessitam acompanhamento, devido ao risco de dilatação tardia do ventrículo direito, arritmias e alguns podem necessitar de troca da valva pulmonar direito à insuficiência pulmonar.

2 | **Tetralogia de Fallot com atresia pulmonar**

Estratégias de tratamentos futuros dependem da anatomia das artérias pulmonares e da presença de artérias colaterais sistêmico-pulmonares e consiste em cirurgia paliativa inicial – *shunt* Blalock-Taussig isolado ou associado à unifocalização. A cirurgia definitiva consiste no fechamento da CIV e implante de tubo VD-TP.

3 | **Atresia pulmonar com septo ventricular íntegro**

Estratégias futuras dependem do grau de hipoplasia do ventrículo direito. Em casos com ventrículo direito bem desenvolvido, está indicada a perfuração da valva pulmonar por radiofrequência seguida por valvoplastia pulmonar com balão com ou sem implante de *stent* no canal arterial. Casos com ventrículo direito displásico, requer cirurgia para confecção de shunt sistêmico-pulmonar e em casos selecionados ampliação da via de saída do ventrículo direito. Nos casos com circulação coronária dependente do ventrículo direito está contraindicada a descompressão de ventrículo direito.

Transposição das grandes artérias

1 | **Transposição dos grandes artérios (TGA) simples (septo ventricular íntegro)**

- Infusão endovenosa de prostaglandina para promover mistura intercirculatória no nível do canal arterial.
- Atrioseptostomia por balão nos casos de CIA restritiva.
- Cirurgia de escolha: cirurgia de Jatene, preferencialmente nas duas primeiras semanas de vida.

2 | **TGA com CIV**

Geralmente não necessita de cirurgia precocemente no período neonatal, é comum desenvolver sinais de IC entre 4 a 8 semanas de vida, e a cirurgia primária deve ser realizada nos primeiros meses de vida.

3 | **TGA com CIV e EP**

- Manifesta-se com quadro de cianose importante.
- Habitualmente requerem *shunt* Blalock-Taussig, seguido por cirurgia de Rastelli ou da translocação da valva pulmonar.

Tronco arterioso

Necessita correção cirúrgica precoce antes dos três meses de vida – fechamento da CIV e tubo VD-TP, quando necessário comissurotomia ou plastia da valva truncal, frequente coinsuficiência.

Coração univentricular

Inclui dupla via de entrada de VE, VD ou ventrículo indeterminado e comumente atrioventricular atrésica ou estenótica – em comum estas cardiopatias apresentam hipoplasia de um ventrículo ou outras associações que impedem a correção biventricular. Necessitam de tratamento estagiado. Pacientes com hiperfluxo pulmonar necessitam cerclagem pulmonar nos primeiros dias de vida, e aqueles com estenose pulmonar importante ou atresia necessitam *shunt* Blalock-Taussig.

No futuro, estes pacientes serão submetidos à cirurgia de Glenn bidirecional, habitualmente a partir do sexto mês de vida, e complementação para Fontan, entre os 2 e 4 anos.

Síndrome hipoplásica do coração esquerdo

Não existe consenso quanto ao melhor tratamento para recém-nascidos com síndrome hipoplásica do coração esquerdo. Intervenções cirúrgicas múltiplas – cirurgia de Norwood e suas variantes ou procedimento híbrido inicialmente, seguida de Glenn bidirecional e após complementação para Fontan e transplante cardíaco são opções disponíveis.

Drenagem anômala total de veias pulmonares

A época do tratamento cirúrgico depende da presença ou não de obstrução ao retorno das veias pulmonares. Quando obstrutiva, comum na forma infracardíaca, a correção cirúrgica deve ser de imediato após estabilização inicial, e na forma não obstrutiva, pode ser realizada nos primeiros meses de vida.

REVISÃO

- A abordagem diagnóstica das cardiopatias congênitas, particularmente no neonato, deve ser realizada de forma sistemática.
- O exame ideal para diagnosticar a cardiopatia congênita é, sem dúvida, a ecocardiografia. Todavia, realizá-la em todo recém-nascido é inviável.
- O teste da oximetria de pulso é simples e apresenta alta especificidade e moderada sensibilidade, características que o habilitam para o rastreamento das cardiopatias.

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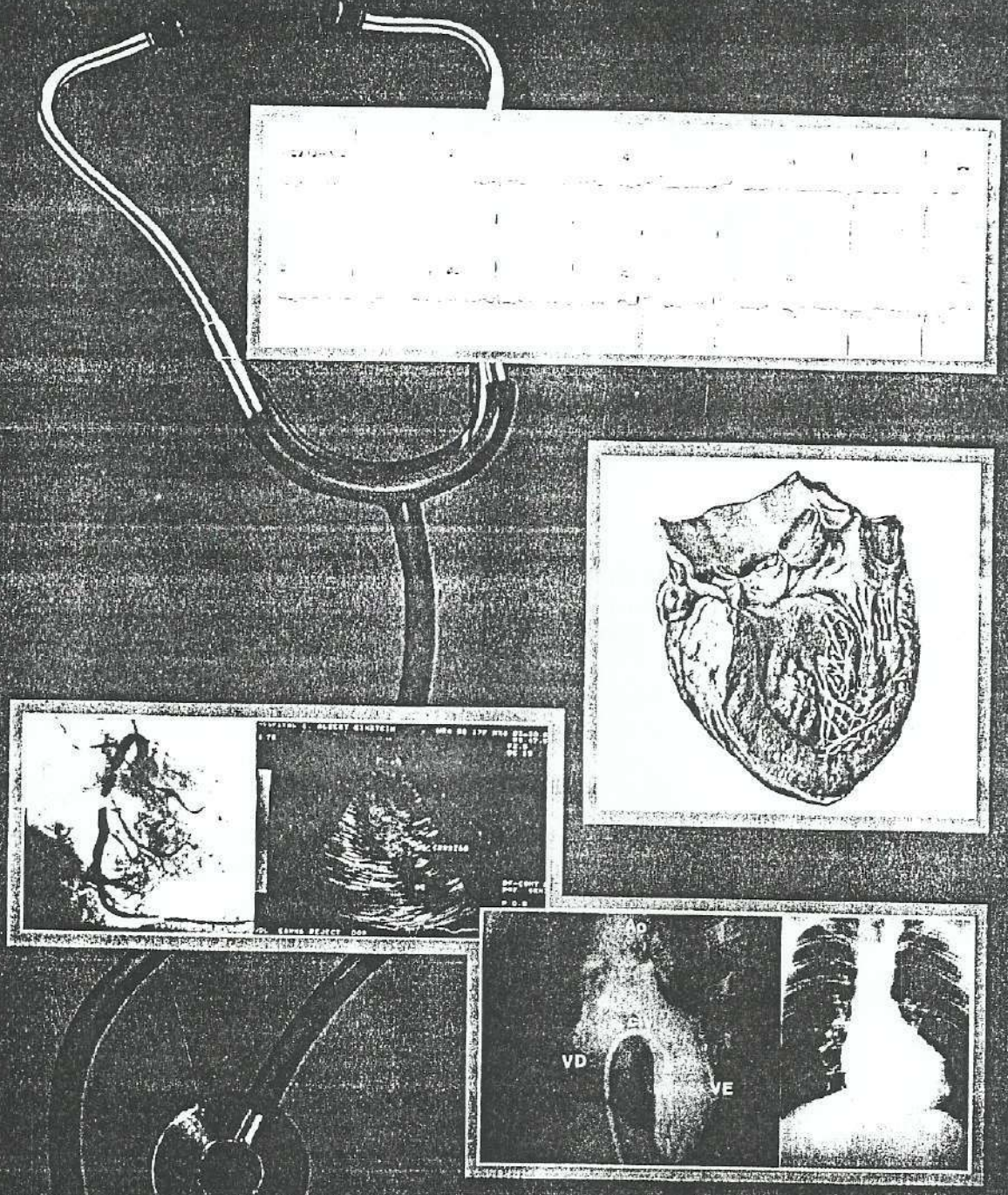
Cardiologia para o Clínico Geral

Celso Ferreira • Rui Póvoa



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Belo Horizonte — Rua Domingos Vieira, 319 — Conj. 1.104

PLANEJAMENTO GRÁFICO — CAPA: Equipe Atheneu

PRODUÇÃO GRÁFICA — O. Prado

Dados Internacionais de Catalogação na Publicação (CIP)
(Câmara Brasileira do Livro, SP, Brasil)

Ferreira , Celso
Cardiologia para o clínico geral/Celso Ferreira, Rui Póvoa
São Paulo: Editora Atheneu, 1999.

Vários colaboradores.

1. Cardiologia 2. Clínica médica 3. Coração — Doenças
I. Póvoa, Rui. II. Título

99-3790

CDD-616.1
NLM-WG

Índices para catálogo sistemático:

1. Cardiologia: Medicina 616.12
2. Coração: Doenças: Medicina 616.12
3. Medicina cardiovascular 616.12

22 Cardiopatias Congênitas

Célia Maria Camelo Silva
Victor Manuel Oporto Lopez
Lourdes de Fátima G. Gomes

INTRODUÇÃO

A incidência de cardiopatia congênita é de seis a oito casos para cada 1.000 nascimentos vivos[1,2,3]. O grau de gravidade varia muito, sendo que apenas um terço destes casos apresenta cardiopatia grave, cuja apresentação no período neonatal pode ser através do quadro de cianose, insuficiência cardíaca, choque cardiogênico ou arritmia, já em outros como nos portadores de valva aórtica bicúspide, a lesão pode permanecer silenciosa por toda a vida. Com os avanços das técnicas diagnósticas e terapêuticas, como o cateterismo intervencionista e os cuidados pós-operatórios um número cada vez maior de portadores de cardiopatia congênita atinge a idade adulta[1-5]. Atualmente, espera-se que a mesma qualidade de tratamento oferecida pelos cardiologistas pediátricos desde o nascimento até a adolescência venha a ser oferecida também na vida adulta. Em resposta a esta necessidade, é que a cardiopatia congênita em adultos tornou-se uma nova área de interesse dentro da cardiologia[1-5].

A maioria das cardiopatias congênitas compatíveis com seis meses de vida intra-uterina permite um nascimento vivo e não devem ser encaradas como um problema estático, e sim como anomalias dinâmicas que se originam no período fetal e que se alteram durante o desenvolvimento pós-natal. No seu curso a cardiopatia congênita pode sofrer modificações, muitas vezes importantes na sua fisiologia, seja pelas alterações dramáticas que ocorrem na transição da circulação fetal para a pós-natal ou mais tardiamente por alterações estruturais e/ou fisiológicas. Por exemplo, uma criança portadora de uma grande comunicação interventricular (CIV) com um grande *shunt* da esquerda para a direita na infância precoce, portanto com sinais de insuficiência cardíaca congestiva (ICC), pode desenvolver progressivamente estenose infundibular e mais tarde apresentar fisiologia e quadro clínico semelhante à tetralogia de Fallot[6,7].

ETIOLOGIA

A etiologia das cardiopatias congênitas parece ser multifatorial, sendo o resultado de uma interação complexa entre fatores genéticos e ambientais. Entre estes determinantes estão hereditariedade, alterações cromossômicas, teratógenos, altitude relativa ao nível do mar por ocasião do nascimento, sexo, idade materna, algumas patologias maternas como diabete, lúpus eritematoso sistêmico etc. Há certa tendência das cardiopatias se repetirem em famílias, como, por exemplo, a comunicação interatrial (CIA) e a persistência do canal arterial (PCA). É também freqüente a associação de CIA nos pacientes portadores da síndrome de Holt-Oram, herança tipo autossômica dominante. Na etiologia da CIA além dos fatores exógenos, em estudo realizado na Escola Paulista de Medicina, ficou evidente a importância do mecanismo genético na recorrência dessa cardiopatia. O risco nessas irmandades com um portador dessa anomalia foi de 25%[8].

Na síndrome de Ellis-van Creveld é comum a presença de átrio único.

Algumas anomalias cromossômicas estão associadas a tipos previstos de cardiopatia congênita: assim, na síndrome de Down (trissomia do 21) é comum a presença de defeito do septo ventricular (DSV) na síndrome de Turner (45 X O) é comum a coarctação de aorta.

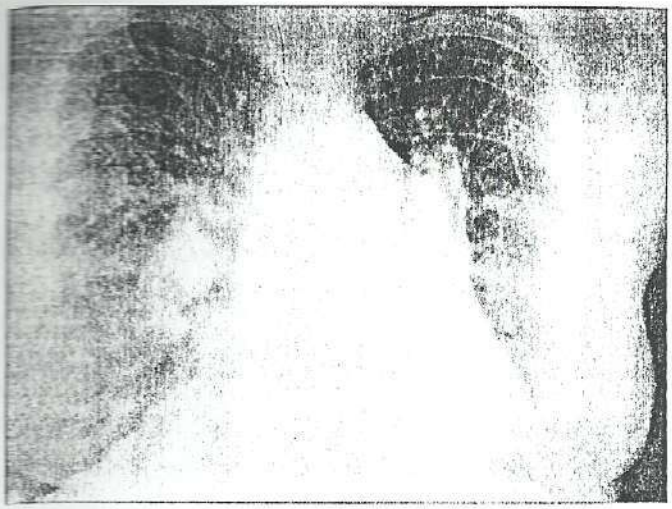

Fig. 22.8 — *Radiografia de tórax de paciente portador de hipertensão pulmonar mostrando dilatação importante do tronco pulmonar e ramos proximais da artéria pulmonar.*

te no cálculo da resistência vascular pulmonar mediante estudo hemodinâmico[5,6,7].

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16-21 JULY, 2017
7th WORLD CONGRESS OF
PEDIATRIC CARDIOLOGY
& CARDIAC SURGERY
Barcelona

Volume 27 • Supplement 4 • Pages S1–S653
doi:10.1017/S104795111700110X
Cardiology in the Young

WORLD CONGRESS OF PEDIATRIC CARDIOLOGY AND CARDIAC SURGERY

7th World Congress of Pediatric Cardiology & Cardiac Surgery Abstracts

ORAL ABSTRACTS

ADMINISTRATIVE

O1289 - IMPROVING TIMELY DISCHARGE BY USING GOAL ORIENTED ROUNDING AND TEAM BASED COMMUNICATION

Alistair Phillips[1], *Nicole Berndsen*[1], *Courtnay Caufield*[2], *Maureen Davis*[2], *Pooja Nawathe*[3], *Stephen Robert*[3], *Vincent Olshove*[1]
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Background: Discharge planning for complex congenital heart patients is challenging for the multi-disciplinary team, especially in a cardiac critical care environment. Delays in discharge are often attributed to poor team planning and communication. Multi-disciplinary collaboration with effective communication ensures a seamless and timely discharge.
Methods: Members of the multi-disciplinary team collaborated to establish a rounding process in the intensive care unit to have patients discharged by noon. The rounding process, implemented in October 2015, had pre-defined goals and involved all members of the team: nursing, physicians, case management and allied health professionals. Patient status, eligibility for discharge and anticipated needs were discussed and planned accordingly by appropriate team members. Rounds were audited for goal completion and efficiency and patient discharge times were tracked.
Results: We identified four crucial multidisciplinary rounds: (1) Operational rounds which are brief overview rounds conducted in a conference room, establishing key goals for the day including preparing for the following days discharges, (2) Work rounds – bedside rounds, (3) 4 pm hand-off rounds – review of key goals days, and reviewing barriers to discharge and (4) Late evening rounds with bedside nurse to make sure goals are on track. Surveys were conducted at 1 month and 3 months after process education and implementation was completed. Responses were from all disciplines. There was an overall increase in process understanding, overall efficiency ($p < 0.1$). Prior to implementation of process, 25% patients were discharged prior to noon. From October 2015-September 2016, 40% of patients were discharged before noon.
Conclusion: Establishing a process for goal-oriented, multi-disciplinary rounding improves team communication and coordination of care in preparation for discharge. Ensuring timely discharge improves patient satisfaction and patient flow within the cardiac critical care unit.

O1660 - A CLINICAL EFFECTIVENESS PROGRAM LEVERAGING AGGREGATE PATIENT DATA AT THE POINT OF CARE IMPROVES VALUE IN PATIENTS UNDERGOING CONGENITAL HEART SURGERY

Andrew Shin, *Ling Loh*, *Joseph Kim*, *Heather Freeman*, *Natalie Pageler*, *Michael-anne Browne*, *Christy Sandborg*, *Paul Sharek*, *Claudia Algaze*
Stanford University, Pediatrics, Palo Alto-United States

Background: Healthcare in the United States is increasingly losing value. Decreasing variation is central to reducing waste and cost. The rapid adoption of information technology is regarded as an important means to promote high value care. We describe a clinical effectiveness program leveraging personalized comparative effectiveness information at the point of care to provide target hospital goals for patients following congenital heart surgery.
Methods: Using an observational pre-post-intervention design, patients undergoing one of the 10 core congenital heart surgeries as defined by the Society of Thoracic Surgeons were included. The setting was a tertiary university-affiliated academic children's hospital between September 6, 2016 and December 19, 2016. Personalized comparative cohorts encompassing 2 years of pre-intervention data were constructed utilizing the electronic health record (EHR). Based on the aggregate data, target hospitalization goals (e.g., target extubation time) were tailored for every patient and made visible peri-operatively for healthcare providers in real-time. Outcome metrics included intensive care unit (ICU), total post-operative length of stay (LOS) and associated cost.
Results: A total of 51 patients were enrolled with 47 (92%) completing the program. Other than the Fontan operation, all surgeries experienced a reduction in LOS. We found an aggregate reduction in LOS and variance for ICU (median 3 [IQR 3-4] vs 5 [IQR 3-7] days, $p < 0.001$; mean $3.6 + 1.9$ vs $6.5 + 6.5$ days, $p = 0.003$), and total postoperative LOS (median 6[IQR 5,8] vs 8[IQR 6,12] days, $p < 0.001$; mean $6.5 + 2.3$ vs $10.9 + 9.9$ days, $p = 0.003$), compared with the pre-intervention period. Mortality, reintubation, ICU and hospital readmission rates were unchanged. The annualized cost savings is estimated to be approximately $2.5 million.

significant gradient reduction occurred. In 2 cases of native CoA (23 and 34 years old man) in early follow-up (6 and 8 months after the procedure) in angio CT small aneurysm formations was observed. Both patients were treated successfully with covered stents. In follow-up in one patient (presented for valve implantation) stent fracture was found and no other complications were observed.
Conclusions: Implantation of new cobalt-chromium AS XL and XXL is a good therapeutical option for the treatment of stenosed great vessels.

P1556 - PATIENTS AGE IMPACT ON THE RESULTS OF THE TRANSCATHETER ATRIAL SEPTAL DEFECT CLOSURE

Anna Kaneva-nencheva[1], Elisaveta Levunlieva[1], Lubomir Dimitrov[1], Kiparisija Nenova[1], Ivan Velkovski[1], Rumen Marinov[1], Stojan Lazarov[2], Dobri Dobrev[1]
National Heart Hospital, Pediatric Cardiology, Sofia-Bulgaria[1]; National Heart Hospital, Congenital Heart Surgery, Sofia-Bulgaria[2]

Transcatheter closure with self-expandable double disc devices (DDD) became a method of choice in the treatment of secundum atrial septal defects (ASD). The complications remain the main concern of the procedure despite the gained experience. The aim of the study is to assess the relationship between the patients age and event-free procedure.
Patients and Methods: We retrospectively reviewed the data base of 169 consecutive patients, 112 children, mean age 9,8 (3,2) years (group 1) and 57 adults, mean age 39 (14,6) years (group 2), with attempted DDD, Amplatzer type, performed in a tertiary heart center.
Results: Event-free course was observed in 148 pts. (88%). There were 6 major complications (1 death due to unrecognized retroperitoneal bleeding; 3 early and 1 late embolizations with surgical removal and 1 acute pulmonary edema in patient on chroniodyalisis). Fifteen minor complications were observed (3 explantations before release due to unstable DDD position; 3 transcatheterly removed embolizations; 1 gastrointestinal bleeding; 1 small pericardial effusion; 7 postprocedural dysrhythmias - atrial fibrillation/flutter). Table. Comparison of the type and rate of complications in both groups
Conclusions: ASD device closure is an effective and relatively safe procedure at any age. Patients age has no impact on the major complications and embolizations. They are related mainly to the preprocedural assessment of the ASD size and morphology. Patients age influences dysrhythmias that need precise pre- and postprocedural estimation and treatment.

Table.

	Group 1(N = 112)	Group 2(N = 57)	p
Closure rate (%)	97	92	ns
Total complications (%)	4,6	28,6	0,000
Major complications (%)	1,9	7,1	ns
Total embolizations (%)	2,8	7,1	ns
Dysrrhythmias (%)	0,9	10,7	0,007
TTE size (mm)	14,3 ± 3,1	18,6 ± 4,4	0,000
TEE size (mm)	14,8 ± 3,3	20 ± 5	0,000
Qp/Qs	1,9 ± 0,5	2,2 ± 1	ns
Device size (mm)	16,5 ± 4	23,3 ± 5,7	0,000
Difference DDD-TTE (mm)	2,1 ± 2,4	4,7 ± 4,2	0,000
Septal aneurism (%)	35,2	41,1	ns

P1559 - RIGHT VENTRICLE DEPENDENT CORONARY CIRCULATION IN A NEWBORN CRITICAL PULMONARY VALVE STENOSIS

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Introduction: Coronary-cavitary connections often associated with pulmonary atresia with intact interventricular septum, may occur in neonates with critical pulmonary stenosis (PS), which adds a worse prognostic.
Objectives: To describe a rare case of critical PS with coronary circulation dependent on the right ventricle (RV) .
Case report: –baby girl, born at term, BW = 3350 g. Maternal history of gestational diabetes and systemic arterial hypertension. A systolic murmur was heard at the first day, being well in hospital time she was discharged home on the third day of life. On her ninth day, she presented to a pediatric cardiologist with tachycardia and weight loss. In her investigation: Chest x-ray showed mild cardiomegaly, slight oligaemic pulmonary fields ; ECG - sinus rhythm, QRS + 60° and incomplete RBBB. Echocardiographic findings: critical PS, PFO with bidirectional shunt, moderate tricuspid regurgitation, a hypertrophied good-sized right ventricle (RV). Estimated RV pressure was 118 mmHg, and a 2mm patent ductus arteriosus. She was admitted to the hospital and started on prostaglandin. At 28 days of life, she was referred to our center for pulmonary balloon valvuloplasty (PVB). Hemodynamics findings: RV-PA gradient of 45 mmHg and RV/AO ratio of 1.39. After RV angiography, PVB with a Power-flex balloon 10 × 2, was performed, followed its deflation, the patient went to complete atrioventricular block, cardiogenic shock and death despite exhaustive CPR manoeuvers. Going back to review her angiographies the left coronary artery were seen arising directly from the right ventricular outflow tract.
Conclusion: decompression of the RV was probably the cause of death. Aortography should be performed prior to PVB in suspected cases

P1569 - LONG TERM RESULTS OF ULTRAHIGH PRESSURE BALLOON ANGIOPLASTY FOR PERIFERAL PULMONARY ARTERY STENOSIS. COMPARISON ANALYSIS WITH STENT PLACEMENT

Mizuhiko Ishigaki[1], Sung-Hae Kim[2], Tanomo Ono[2], Keisuke Sato[2], Jun Yoshimoto[2], Norie Mitsushita[2], Masaki Nii[2], Kisaburo Sakamoto[2], Yasuhiko Tanaka[2]
Shizuoka Children's Hospital, Cardiology, Shizuoka-Japan[1]; Children's Hospital, Cardiology, Shizuoka-Japan[2]

Background and Objectives: Balloon angioplasty for postoperative pulmonary artery stenosis is an important therapeutic option to maintain and facilitate the pulmonary circulation. During the past decade, manykinds of non-compliant balloons which provide excellent trackablity have been widely used. Especially, efficacy of ultra-high-pressure balloons (UHPB) such as CONQUEST@ has been also reported, but their long term benefits are not known. To analyze the long term efficacy of UHPBs against postoperative branch pulmonary artery stenosis compared to stent implantation.
Methods: Retrospective analysis of follow-up catheterization data over three years after angioplasty by UHPBs or stents against the lesions with biventricular corrective surgery. Five cases, 11 branches of UHPB group and 8 cases, 9 branches of stent groups are included.

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PROGRAMA DE PÓS-GRADUAÇÃO EM CIÊNCIAS DA SAÚDE

XIII FÓRUM DO PROGRAMA DE PÓS-GRADUAÇÃO EM CIÊNCIAS DA SAÚDE

PROGRAMA CIENTÍFICO E RESUMOS

UBERLÂNDIA
6 DE OUTUBRO DE 2016

Dados Internacionais de Catalogação na Publicação (CIP)

F745 r Fórum do Programa de Pós-Graduação em Ciências da Saúde (13. : 2016 : Uberlândia, MG)

Resumos / XIII Fórum do Programa de Pós-Graduação em Ciências da Saúde, 6 de outubro de 2016 em Uberlândia, Minas Gerais ; coordenação Angélica Lemos Debs Diniz. - Uberlândia : UFU, 2016.

29 p.

1. Ciências médicas - Congressos. 2. Epidemiologia – Congressos . I. Diniz, Angélica Lemos Debs. II. Universidade Federal de Uberlândia. III. Título.

CDU: 61

Universidade Federal de Uberlândia
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Programa de Pós-graduação em Ciências da Saúde

XIII Fórum do Programa de Pós-Graduação em Ciências da Saúde

PERFIL EPIDEMIOLÓGICO E EVOLUÇÃO CLÍNICA DOS PORTADORES DE MIOCARDIOPATIA DILATADA ACOMPANHADOS NO SERVIÇO DE CARDIOLOGIA PEDIÁTRICA DO HC-UFU

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A miocardiopatia dilatada caracteriza-se pela diminuição da força de contração sistólica miocárdica com consequente dilatação de câmaras cardíacas. É a forma mais comum de miocardiopatia na infância, mas pouco se sabe sobre sua real incidência. A apresentação clinica da doença é de espectro amplo, podendo ser diagnosticada como achado de exame. Pode se apresentar como uma arritmia, cianose generalizada, dor precordial, sincope, e até morte súbita. O diagnóstico de certeza é dado pelo ecocardiograma, que mostra, dentre outras alterações, o ventrículo esquerdo dilatado, com tendência a modificação da sua morfologia e redução da fração de ejeção ventricular esquerda e da fração de encurtamento. O objetivo do estudo é conhecer o perfil epidemiológico e a evolução dos pacientes acompanhados no setor de pediatria do Hospital de Clínicas da Universidade Federal de Uberlândia, avaliar a evolução clínica e o resultado terapêutico nesses pacientes, montar um perfil epidemiológico que possa identificar estes pacientes mais precocemente e abrir novas linhas de pesquisa a serem investigadas em futuros estudos. Serão incluídos pacientes de zero a 13 anos incompletos portadores de miocardiopatia dilatada adquiridas na infância diagnosticadas no período de janeiro de 2005 a setembro de 2015. Será feita uma avaliação dos prontuários, preenchida uma ficha de coleta de dados epidemiológicos e realizado uma análise para a verificação do perfil epidemiológico dos pacientes e avaliação da evolução e do tratamento empregado.

Palavras-chave: Cardiomiopatia dilatada; perfil epidemiológico

Apoio: UFU

Área de Concentração: Epidemiologia da ocorrência de doenças e agravos à saúde

Citação: ROSSI, Bruno Franco; GOMES, Lourdes de Fátima Gonçalves; RESENDE, Elmiro Santos. Perfil epidemiológico e evolução clínica dos portadores de miocardiopatia dilatada acompanhados no serviço de cardiologia pediátrica do HC-UFU. In: Fórum do Programa de Pós-graduação em Ciências da Saúde, 13., 2016, Uberlândia. **Programa Científico e Resumos**. Uberlândia: Horizonte Científico, 2016. v. 10, p. 22.

TL065

SISTÓLICA VENTRICULAR EM CRIANÇAS TRATADAS COM ANTRACICLINAS: UM ESTUDO LONGITUDINAL E PROSPECTIVO COM ECOCARDIOGRAFIA ASSOCIADA A ESFORÇO

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...DE DE MEDICINA DE MARÍLIA, FACULDADE DE MEDICINA DE ...ATU

Introdução: Crianças portadoras de câncer tratadas com baixas doses de antraciclinas ... podem evoluir com cardiotoxidade leve assintomática. O objetivo deste ... foi de avaliar em longo prazo a reserva miocárdica em crianças previamente ... com dose acumulativa de antraciclinas até 100 mg/m². **Métodos:** 27 crianças ... sobreviventes de câncer (25 com leucemia linfoblástica) com remissão ... clínica, sem alterações no ecocardiograma convencional foram avaliadas pela ... ecocardiografia após o exercício 37±15,4 meses (T1) e 101±24 meses (T2) após o ... término do tratamento (grupo ADRIA). Este grupo foi comparado com 25 indivíduos ... saudáveis (grupo controle) com idade e superfície corpórea similares. Todos realizaram ... um protocolo no protocolo de Bruce e o ecocardiograma realizado imediatamente ... após o exercício. Análise estatística: testes Kolmogorov-Smirnov, Mann-Whitney, ... Wilcoxon, com p<0,05. **Resultados:** os grupos foram semelhantes em relação à ... massa cardíaca e função sistólica do ventrículo esquerdo (VE) em repouso em T1 e ... T2. O espessamento da parede posterior do VE corrigido pela superfície corpórea foi ... maior no grupo ADRIA em T2. Após o exercício, menores índices de ejeção do VE e ... maiores incidências na pós-carga no grupo ADRIA foram observadas em T1 e T2.

... índices ecocardiográficos após o exercício

	T1	T1	T1	T2	T2	T2
	Controle	Adria	valor P	Controle	Adria	Valor P
...PPVE	124,9+/-24	82,6+/-25,8	<0,0001	102+/-16,4	90,7+/-21,3	0,042
...	89+/-4	82+/-6	<0,0001	87+/-5	79+/-7	0,0001
...	11,3+/-2,9	8,4+/-2,7	0,0010	11,3+/-2,7	8,6+/-2,0	0,0005
...PVE	29,5+/-9,8	44,2+/-16,8	0,0008	36+/-10,4	49,4+/-16,9	0,0026

...PPVE: espessamento sistólico da parede posterior do VE, FEVE: fração de ejeção ... do VE, IcmVE: índice de contratilidade do VE, ESFPPVE: estresse sistólico final da ... parede posterior do VE. **Conclusão:** A redução da reserva sistólica induzida por baixas ... doses de antraciclinas em crianças e adolescentes assintomáticas permanece alterada ... após de 5 anos, sugerindo que a presença de cardiotoxidade crônica poderia ser ... notada em pacientes com alterações leves após o tratamento com antraciclinas.

TL066

PERFURACAO DA VALVA PULMONAR POR RADIOFREQUENCIA EM CRIANCAS COM ATRESIA PULMONAR E SEPTO VENTRICULAR INTEGRO

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UNIFESP, UNITAU

Introdução: Após a eficácia do tratamento percutâneo da estenose pulmonar crítica do recém-nascido, sua indicação foi estendida para as formas favoráveis de atresia pulmonar com septo ventricular íntegro. **Objetivo:** Avaliar os resultados iniciais e tardios da perfuração da valva pulmonar (VP) por radiofreqüência (RF), seguida de valvoplastia, em portadores de atresia pulmonar com septo íntegro. **Métodos:** Participaram do estudo quinze pacientes consecutivos (idade: 0-8 meses), sendo incluídos treze (86,6%), com ventrículo direito tripartite, de bom tamanho e ausência de sinusóides importantes. **Resultados:** A perfuração valvar foi com sucesso em 9 casos (69,2%). A mediana dos pesos foi 3,9 Kg. Pré-dilatação, a média da pressão sistólica do ventrículo direito (VD) foi de 112 1,6 ± 34,3 mmHg e da aorta (AO) de 71,8 ± 18,5 mmHg, e após a média da pressão sistólica do VD reduziu para 58,6 ± 20,3 mmHg e a da aorta aumentou para 72 ± 4,4 mmHg (p = 0,03). A relação da pressão VD/AO foi de 1,95 ± 0,32 antes e de 0,69 ± 0,09 após (p= 0,0001). O gradiente VD-TP após variou de 5 a 27 mmHg. Entre as complicações graves, observou-se: perfuração cardíaca (3pts). Sete crianças (53,8%) tiveram alta hospitalar. Uma delas necessitou de reintervenção (Shunt Blalock-Taussig). Ocorreram três óbitos (22,6%). O período de internação foi de 15,5 ± 7,8 dias. Após 46,7 ± 39,5 meses, os pacientes sobreviventes encontram-se bem, acianóticos, com fluxo anterógrado adequado. **Conclusão:** A perfuração da valvar pulmonar por radiofreqüência seguida de valvoplastia por balão é uma alternativa segura e eficaz ao tratamento cirúrgico para atresia pulmonar com septo íntegro. Esta técnica pode se tornar o tratamento inicial de escolha no grupo com VD irrigante, infundíbulo amplo e sem sinusóides.

TL067

ASPECTOS CARDIOLOGICOS NA SÍNDROME DE TURNER

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Introdução: a Sd de Turner é uma das aneuploidias mais comuns e afeta 1/2500 ... nascidos, ocorrendo somente em indivíduos do sexo feminino. O distúrbio ... cromossômico característico é a monossomia do cromossomo X (45,X0). O fenótipo ... inclui baixa estatura e perda da função ovariana (hipogonadismo primário) em todos ... os casos, fácies incomum típica, pescoço alado, linha posterior de implantação dos ... cabelos baixa, tórax largo com hipertelorismo mamário e elevada freqüência de ... anomalias renais e cardiovasculares (30% dos casos) com espectro variável além de ... baixa. **Método:** estudo transversal descritivo, de julho de 2010 a outubro de 2011, ... para avaliação do fenótipo cardiológico de 64 pacientes portadoras de Sd de Turner. ... Foi realizado exame físico e ecocardiograma, documentação das lesões cardíacas ... encontradas e necessidade de intervenção cirúrgica bem como evolução. **Resultados:** ... o padrão cromossômico mais comumente encontrado foi 45,X0, embora também ... observamos alguns casos de mosaicismo. A idade das pacientes variou de 2 a 49 ... anos (mediana 15). Em 28 e (43%) foi detectada alguma anormalidade cardiológica, ... sendo a mais comum, prolapso da valva mitral (11e) seguido de valva aórtica ... bicúspide (9e). A Coarctação de aorta ocorreu em 5e (7,8%) e 4e (6,2%) necessitaram ... de correção cirúrgica com bom resultado. Lesões da valva aórtica com repercussão ... hemodinâmica foram observadas em 4e, entretanto nenhum paciente necessitou de ... cirurgia valvar até o momento. **Conclusão:** na sd de Turner, o acometimento cardíaco ... é frequente e apresenta espectro variável, podendo ter caráter evolutivo e geralmente ... bom prognóstico cardiológico favorável. Recomenda-se a investigação cardiológica e ... acompanhamento seriado destes pacientes.

TL068

ASSOCIAÇÃO ENTRE SERVIÇOS DE MEDICINA FETAL E ECOCARDIOGRAFIA FETAL PERMITE MELHOR RASTREAMENTO DE CARDIOPATIAS CONGÊNITAS

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Proposta: O serviço de medicina fetal é um centro de referência obstétrica para detecção de malformações congênitas. Há poucos estudos analisando o impacto da associação entre os serviços de medicina fetal e de ecocardiografia fetal para identificação de fetos de alto risco de cardiopatia complexa. O objetivo deste estudo foi identificar a freqüência e tipos de anormalidades cardíacas em fetos de mães encaminhadas para o serviço de ecocardiografia fetal tanto por especialistas em medicina fetal quanto por obstetras não especialistas. **Métodos:** Estudo prospectivo de uma série de casos consecutivos referidos ao serviço de ecocardiografia fetal no período de janeiro a agosto de 2011. A comparação entre os grupos foi realizada pelo teste exato de Fisher. (nível de significância p<0,05). **Resultados:** O ecocardiograma fetal foi realizado em 36 gestantes, 16 (44%) encaminhadas pelo serviço de medicina fetal e 20 (56%) pelo pré-natal geral. Foi observado que 15 (93,75%) dos ecocardiogramas fetais encaminhados pela medicina fetal apresentaram anormalidade cardíaca comparado a 4 (25%) pelo serviço de pré-natal (p<0,001). Cardiopatia complexa foi encontrada em 6(3,13% das anormalidades cardíacas, arritmia complexa em 15,78%, hipertrofia cardíaca em 15,78% e outros defeitos leves em 5,31%. **Conclusão:** A associação dos serviços de medicina fetal e ecocardiografia fetal permitem um melhor rastreamento e diagnóstico de cardiopatias congênitas fetais.

TEMAS LIVRES DA ÁREA MÉDICA

TL 001

NO SEGUIMENTO PÓS INFARTO AGUDO DO MIOCÁRDIO, A TERAPIA INVASIVA PRECOCE É MENOS EFICAZ NAS MULHERES DO QUE NOS HOMENS

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Fundamentos: Sabidamente há semelhanças e diferenças no comportamento pós infarto agudo do miocárdio (IAM) entre ambos os sexos. Portanto, objetivamos analisar o impacto, na sobrevida a longo prazo, da intervenção coronária percutânea e cirurgia de revascularização miocárdica (terapia invasiva - TI) realizadas na fase precoce do IAM, em ambos os sexos.

Métodos: Foram analisados 923 pacientes (mediana de 64 anos) com IAM, incluídos em banco de dados específico de forma consecutiva e prospectiva entre 1998 e 2004, e seguidos por até 6,4 anos (média de tempo de sobrevida de 4,99 anos). Da população global, 672 (72,8%) eram homens, e os 251 restantes mulheres (27,2%). Durante a fase hospitalar, TI foi utilizada em 70,4% dos homens e em 66,5% das mulheres (p=0,266). Foram construídas diferentes curvas de Kaplan-Meier para cada grupo (homem e mulher). Teste de log-rank foi aplicado nas análises univariadas; análise de regressão "Stepwise Cox", com 23 variáveis incluídas no modelo, foi utilizado nas análises multivariadas.

Resultados: As curvas de Kaplan-Meier demonstraram uma correlação significativa entre TI na fase precoce pós IAM e sobrevida a longo prazo nos homens (p<0,001), porém não nas mulheres (p=0,5). Os modelos ajustados confirmaram as análises univariadas, demonstrando uma correlação significante e independente entre TI e sobrevida nos homens (t-ratio=4,25, P<0,001), e não para as mulheres (t-ratio=1,47, P=0,143).

Conclusão: Terapia invasiva precoce, em pacientes com infarto agudo do miocárdio, é menos eficaz, em seguimento a longo prazo, nas mulheres do que nos homens.

TL 002

INCIDÊNCIA DE CARDIOPATIAS E DOENÇAS ASSOCIADAS EM JOVENS QUE PRATICAM ATIVIDADE FÍSICA E ESPORTIVA

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INSTITUTO DANTE PAZZANESE DE CARDIOLOGIA

INTRODUÇÃO

O número de crianças cardiopatas que praticam atividade física e esportiva tem aumentado. Atualmente a prescrição de atividade física faz parte do tratamento destas crianças.

MÉTODOS

Avaliamos 235 prontuários de jovens cuja idade variou de 7 a 18 anos, atendidos no período de janeiro de 2000 a agosto de 2005, sendo 120 (70%) masculinos. Todas as crianças praticam atividade física organizada e esporte. Registramos a presença de cardiopatias detectadas através de história clínica, exame físico, ECG, ecocardiograma, veloctardiograma, teste ergométrico e exames laboratoriais.

RESULTADOS

Do 180 totais, 139 (77,2%) foram considerados aptos à prática de atividade física. Detectamos cardiopatias em 31 (17,2%), discriminadas na tabela. 5 (2,1%) apresentaram doenças associadas, como Anemia, Asma, Diabetes e Síndrome da Proteinúria do Adeta.

	Total	%		Total	%	%
HA	5	2,7	Est Ao	1	0,5	0,4
S Pré-excit	8	4,4	Ins Ao	1	0,5	
ESV	2	1	Est Pu	2	1	0,9
BRD	2	1	Cor Anom	1	0,5	0,4
Chagas	1	0,5	Displ Arrit	1	0,5	0,4
CIA	1	0,5	Mioc Hipert	2	1	0,9
Marfan	1	0,5	Mio/Pericardite	3	1,6	1,8

CONCLUSÕES:

A incidência de cardiopatias entre jovens que praticam atividade física é significativa. Sabe-se que estes jovens beneficiam-se física e psicológicamente destas atividades. Contudo, alguns deles necessitam afastamento ou restrição da forma a protegê-los de riscos. Neste estudo, 18 (10%) dos jovens foram afastados. Reforçamos a importância da avaliação pré-participação destes indivíduos a fim de reduzir os riscos associados à prática de esportes.

TL 003

REMODELAMENTO POSITIVO EM ARTÉRIAS CORONÁRIAS DE PACIENTES COM SÍNDROMES CORONÁRIAS AGUDAS: COMPARAÇÃO DOS ACHADOS DA TOMOGRAFIA COMPUTADORIZADA DE MÚLTIPLOS DETECTORES E DO ULTRA-SOM INTRACORONÁRIO

IBRAIM MASCIARELLI PINTO; ADIB JATENE; J EDUARDO SOUSA; LUIZ CARLOS SOUZA; ENILTON EGITO; FAUSTO FERES; LUIZ ALBERTO MATTOS; ANDRÉA C ABIZAID; AMANDA SOUSA; ALEXANDRE ABIZAID

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Objetivo: O Remodelamento arterial positivo (RAP) das artérias coronárias é um dos sinais de vulnerabilidade das placas ateroscleróticas em pacientes (P) com síndromes coronárias agudas (SCA). Este fenômeno é bem documentado pelo ultra-som intracoronário (USIC), mas seria interessante que pudesse ser avaliado por um método não-invasivo. O objetivo deste estudo foi analisar a eficácia da tomografia computadorizada de múltiplos detectores (TCMD) na detecção de RAP.

Casuística e Métodos: Entre 01 e 11 de 2005 avaliamos 23 P consecutivos com SCA pela TCMD e pelo USIC. A TCMD incluía aquisições feitas após a injeção de 70 ml de contraste iodado por via endovenosa. As imagens foram transferidas para uma estação de trabalho na qual se mediu, para as finalidades deste trabalho, a área de referência proximal (ARP), a área de referência distal (ARD), a área mínima da luz (AML) e a área do vaso no local da lesão (AV). Todos os P foram submetidos ao USIC nos quais repetiram-se as mesmas quantificações. As análises foram feitas por observadores independentes.

Resultados: A ARP media 9,1± 2,7 mm2 pela TCMD e 10,8± 3,1 mm2 pelo USIC, enquanto que a ARD media 8,2 ± 2,1 mm2 pela TCMD e 9,3 ± 4,3 mm2 pelo USIC. A AML era 0,96 ± 0,3 mm2 pela TCMD e 1,3 ± 0,5 mm2 pelo USIC, enquanto que a AV era 13,4 ± 4,1 pela TCMD e 14,2 ± 5,1 pelo USIC. A comparação de Bland-Altman mostrou que a maior parte dos pontos de comparação encontrava-se dentro do limite de 1 desvio-padrão, sugerindo coerência entre os métodos. A TCMD encontrou RAP em 17 P e o USIC em 19. Nos 2 casos em que houve discordância entre os exames a lesão culpada situava-se na porção distal da artéria coronária relacionada ao evento. Além disso, a TCMD encontrou sinais de placas lipídicas ou mistas em 14 casos e calcificadas em 9, o que foi confirmado pelo USIC.

Conclusão: A TCMD mostra potencial para identificar RAP em coronárias de P com SCA, com resultados que podem ser ainda melhores com o progredir da tecnologia.

TL 004

AVALIAÇÃO DO CARVEDILOL NO TRATAMENTO DE INSUFICIÊNCIA CARDIACA EM CARDIOPEDIATRIA

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Introdução O carvedilol (bloqueador alfa-beta-adrenérgico-não-seletivo) é eficaz anti-falência cardíaca, cujos efeitos a longo prazo no manejo de insuficiência cardíaca (IC) em cardiologia pediátrica necessitam serem melhor avaliados.

Objetivo Avaliar o uso do carvedilol (CVD) associado ao tratamento convencional da IC em crianças e adolescentes em estudo prospectivo de 3 anos.

Métodos Participaram 30 pacientes (pt); idades de 6 meses (m) a 18 anos (medianas: 44 meses); 13 masculinos (43,3%); peso de 4,8 a 48 kg e fração de ejeção (FE) d<50 %. Associou-se o CVD: 0,05 a 0,5 mg/kg/dia (2 a 3x/dia). Avaliou-se: escore de sintomas em IC (ESIC), efeitos adversos, índice cardiotorácico (ICT), eletrocardiograma (ECG), tipo funcional (TF), função sistólica e diastólica (FSD) pelo ecocardiograma (pré-CVD e 3/3 m à 1 ano pós-cura). Utilizou-se: média ± DP, e os testes: *Student t* e Exato de Fisher na análise dos dados.

Resultados Em 17/30 pt (56, 7%) a IC foram por miocardite, cardiopatia congênita em 8 (26,7%), 4/30 (13,3%) com refluxo mitral grave e 1 (3,3%) idiopático. De 29 pt (93,3%) com ESIC de: 12 a 8 pré-CVD. 26 (86%) reduziram: 7 a 4 no 1º mês de CVD; 11/30 (36,7%) tiveram náuseas e vômitos nos 1º 3 dias; tontura em 1 (3,3%). ECG inalterado e redução do ICTa partir de 6 m (0,006 cm/área/m). Inicialmente 22 pt (73,3%%) eram TF II; aos 3 m 14 (46,7%) eram TF I e aos 24 m 28 (93,3%) eram TF I. Tempo médio de normalização da FE: 20,8 m (dosemédia: 0,4 mg/kg/dia). A FE, FSD correlacionou-se ao aumento de CVD (p = 0,0027) e TF (0,003). Cinco pt (16,6%) não recuperaram a FE. Não houve óbitos.

Conclusão O CVD foi seguro, de impacto precoce na melhora clínica, TF e o aumento da dose da droga associou-se à recuperação da função ventricular nestes pacientes.

Volume 11 • Supplement 1 • Pages 1-346

Cardiology in the Young

Abstracts and Posters of the 3rd World Congress of Pediatric Cardiology & Cardiac Surgery: Toronto, Ontario, Canada, 27—31 May, 2001

MAY 28 Time: 11:00–12:30

Session 1: Young Investigator Award

YIR 1

In vivo visualization and quantification of leukocyte endothelial cell interactions during cardiopulmonary bypass with means of intravitalmicroscopy

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Purpose: To visualize and quantify the influence of paediatric cardiopulmonary bypass (CPB) on leukocyte endothelial cell interaction and microcirculation as an early crucial step an inflammatory reactions. Methods: Newborn piglets were subjected to hypothermic CPB (H, n=9). Intravital fluorescence microscopy (IVM)of subcutaneous tissue was performed to analyse leukocyte endothelial cell interactions (grading of rolling-sticking-free flowing leukocytes), capillary perfusion (functional capillary density) and vessel morphology (diameters, length, structure) during CPB and compared to a naïv control group (C, n=7). IVM images were videotaped, digitally processed and analyzed focusing on microvascular alterations as inflammatory signs. Results: IVM pictures were recorded only under stable hemodynamic conditions (H, RR mean=55mmHg, C: RR mean=58mmHg; H: CO=132ml/min/kg, C: CO=96ml/min/kg). After 45 minutes of CPB leukocytes showed obvious signs of activation: Increase in rolling (C: 6.6% H: 19.1%)and sticking of leukocytes (C: 9.0%, H: 47%)on the vascular endothelium. Hematology showed a decline in neutrophil counts at the end of CPB (C: 52.6%, H: 21.5%). Breakdown of functional capillary density (FCD) (C: 317 mm/sqmm; H: 37mm/sqmm) and reduced capillary flow velocities indicated a massive alteration of capillary perfusion. Arteriolar vasoconstriction and venular-capillary vasodilation occured as possible signs of deranged vasoregulation. Conclusion: IVM was shown to be a reliable tool for quantification of inflammatory tissue damage due to CPB. For the first time leukocyte activation, altered microvascular perfusion and changes in vasomotor function were directly visualized. Increased leukocyte adherence, ischemic tissue damage caused by a perfusion deficit and hemodynamic imbalance of the microvascular network are early signs for the onset of an inflammatory reaction caused by CPB.

YIR 2

Tissue engineered vascular autograft (teva): the possibility for developing 'ideal' venous autografts and the influence of cell origin on the outcome

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Tissue engineered vascular autografts (TEVA) were created and the influence of cell origin on the outcome of such grafts was examined. Methods: For Group I (N=4), vascular myofibroblasts were obtained from femoral veins of mongrel dogs. For Group II (N=2), dermal fibroblasts were obtained from subcutaneous tissue of other dogs. In each group, TEVAs were made by seeding these cells onto tube-shaped biodegradable polymer scaffolds composed of PGA non-woven fabrics and co-polymers of l-lactide and ε-caprolactone (PLCL/LA). Next, the IVCs of these same dogs were replaced with TEVAs. After 3, 4, 5 and 6 months in group I, and 1 and 2 months in group II, angiographies were performed, and then the dogs were sacrificed. Implanted TEVAs were examined both grossly and immunohistologically. Results: In group I, the angiographies showed no stenoses or dilatations in each of the TEVAs. Furthermore there was no thrombus inside the graft despite the lack of any anticoagulation therapy. Overall, the gross appearance of all specimens appeared similar to those of native IVCs. Implanted TEVAs contained sufficient amounts of extracellular matrix and immunohistological staining revealed that there was an endothelial cell lining on the luminal surface of each TEVAs. In group II, both dogs showed symptoms of IVC obstruction within several weeks after implantation. Angiographies revealed total occlusion of both TEVAs. Conclusions: These results strongly support the possibility for developing 'ideal' venous autografts with good anti-thrombogenicity and growth potential by using an in vivo tissue engineering technique. The origins of the cells (Group I: mesodermal origin, Group II: ectodermal origin) seeded on the polymer scaffolds affected the outcome of the TEVAs, such that the use of vascular myofibroblasts for creation of TEV

YIR 3

Inflammatory and proliferative response to biodegradable stents produced from iron-based alloys: cellular and molecular studies after incubation with porcine and rabbit fibroblasts, endothelial and vascular smooth muscle cells

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Introduction: In vivo studies after implantation of corrosive degradable stents into porcine coronary arteries and the descending aorta of rabbits have demonstrated minimal inflammatory response and neointimal proliferation. The following study was designed to assess whether differences in gene expression of vascular endothelial derived growth factor (VEGF) and transforming growth factor beta (TGF-beta) could be demonstrated in vitro. Methods: Fibroblasts (FB), vascular smooth muscle (vSMC) and endothelial cells (EC) were isolated from porcine and rabbit aorta and skin. Defined were segments of a biostable 316-L (PUVA, Dreon Medical, Germany) and a corrosive biodegradable iron-stent (NDBK-4, Dreon Medical, Germany) were incubated in cell cultures for up to 7 days. Cultures without stents served as controls. For mRNA-isolation cells were harvested after 0 h, 2 h, 1 day, 3 days and seven days. Proliferation was assessed by cell counts before trypsination. Expression studies of VEGF and TGF-beta were performed after reverse transcription using quantitative real-time PCR (Taqman TM, Applied Biosystems, Germany). PCR cycles to reach the threshold of detection (CT) were determined and normalized to GAPDH (delta CT method). Results: EC and FB were adherent, confluent and increased their cellular density during cultivation with 316-L and degradable iron-stents. In contrast vSMC showed a decreased adherence after 3 days, no vital cells were observed after 7 days in cultures incubated with degradable Fe stents. Incubation with 316-L and control vSMC showed no growth abnormalities. Gene expression demonstrated non-significant differences of less than 1 PCR cycle in all cell lines and incubation periods. Conclusion: Porcine and rabbit EC, FB and vSMC show no different molecular response to incubation with 316-L- and iron-stents. Whether cell death of vSMC may potentially reduce neointimal proliferation requires further investigation.

quantitative PCR

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

YIR 4

Reoperation after the anatomic correction of the transposition of the great arteries

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Technique of the TGA anatomic correction and repair of associated cardiac anomalies in complex cases can lead to morbidity and subsequent reinterventions. This prospective study aims to assess the incidence of reoperation and its results. Between 1982 and January 2000, 1200 pts were operated on at the same center; 102 (8.6%) died and were lost to FU. The 1095 survivors had a mean FU of 58.3 m. One hundred twenty eight reoperations were performed in 110 pts (10%). Reoperation was done for PS in 43 pts, PM implantation and residual shunt in 16 each, AI in 13, cava thrombosis in 11, LVOT obstruction in 9, MR in 7 and for coronary stenosis in 6. Actuarial freedom for reoperation was 90, 83 and 82% at 5, 10 and 15 y. Surgery for PS was not observed after 9 y but operation on AI occurred all along the FU. Coronary obstruction repair was done after 5 y in 3 of the 6 pts. Reoperation incidence was significantly higher in complex TGA (21.4 vs 5.7%, p < .001) as reinterventions on PS and AI (6.6 vs 2.1% and 2.5 vs .4% p < .001). Surgery for LVOT obstruction, PM implantation and shunt closure were only done in complex TGA. After arterial switch operation, reinterventions are more frequent in complex TGA; they occurred more often early in the FU but surgery for PS is seen up to 9 y after the ASO and for AI all along the FU. Need for coronary obstruction relief is rare but seen late.

YIR 5

Activity of paxillin helps drive postnatal pulmonary arterial remodelling

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The pulmonary arterial smooth muscle cell actin cytoskeleton is remodelled immediately after birth and connective tissue deposition increases. We hypothesised that the activity of focal adhesion proteins linking these structures would be enhanced during adaptation. We therefore studied the expression and activity of the focal adhesion protein paxillin in the pulmonary arteries of 48 normal and neonatal chronically hypoxic pulmonary hypertensive piglets from fetal life to adulthood. Results: By immunohistochemistry, paxillin showed site specific and temporal changes in expression after birth being distributed throughout the media in the fetus and newborn and decreasing transiently in the inner media during the first week of life. SDS-PAGE showed two principle paxillin isoforms, 60 and 66 kDa, both decreasing transiently after birth (p < 0.05) and then increasing with age (p < 0.05). 2 – dimensional gel electrophoresis demonstrated multiple isoforms but only one mRNA was evident on Northern blot analysis indicating post translational processing. A postnatal shift in the isoelectric point towards more acidic forms indicated an increase in phosphorylation. The 60 kDa isoform was the more phosphorylated, on both tyrosine and serine. Serine phosphorylation increased transiently after birth (p < 0.01) while both phosphorylations decreased transiently at 14 days of age. In pulmonary hypertension, the transient postnatal reduction in protein expression was abolished and phosphorylation increased (p < 0.01). Conclusion: These findings suggest that the abrupt distension of the vessel wall at birth is associated with changes in the composition and the enhanced activity of focal adhesion proteins, changes which are compromised in pulmonary hypertension. Ultimately, therapeutic control of these focal adhesion signalling pathways would enable us to influence structural remodelling. Supported by The British Heart Foundation.

YIR 6

Trans-ventricular repair of Tetralogy of Fallot in infancy: up to 26 years follow-up

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Objective: To evaluate the need for re-operation, the incidence of arrythmias, the function of the right ventricle and the survival following trans-ventricular repair of tetralogy of Fallot (TOF) in infancy. Patients: Between 1974 and 2000, 91 infants (mean age 200±90days, range 15–364days) underwent a trans-ventricular repair of a simple TOF in our unit. Six patients had a previous aorto-pulmonary shunt. Sixty-six procedures were urgent or emergency. A trans-annular patch (TAP) was inserted in 76 patients (83.6%). Follow-up was 100% complete (mean 14.5±5.2years, range 0–25.8years). Results: There were two operative deaths (2.2%). Thirteen patients underwent re-operations or catheter re-interventions. Freedom from re-operation or catheter re-intervention at 20 years was 87±4%. A re-operation for RVOTO was performed in 3 patients. Twenty-year freedom from re-operation for RVOTO was 97±3%. Six patients required pulmonary valve replacement (PVR) due to severe regurgitation. Twenty-year freedom from PVR was 96±3% (2 patients had PVR more than 20 years after their operation). Use of a trans-annular patch did not significantly affect the need for re-operation or catheter re-intervention. There was one late death. Twenty-year survival was 97±1%. Two patients developed intermittent first or second degree heart block and one developed late recurrent ventricular tachycardia. None had a QRS>180msec. All but one patient are currently in NYHA functional class I having good right and left ventricular function. Conclusion: Trans-ventricular repair of TOF in infancy carries an acceptable operative mortality. Arrythmias are rare and the survival is comparable with that of the general population. Pulmonary regurgitation is mostly well tolerated but its complications may develop even after twenty years postoperatively, and therefore lifelong follow-up is essential.

MODERATED POSTER SESSIONS

Session 2: Cardiac Anesthesia, ICU Care/Neonatal/Respiratory Management

1

Resynchronization pacing is a useful adjunct to the management of acute heart failure after surgery for congenital heart defects

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Acute hemodynamic effect of atrioventricular (AV) and inter/intraventricular (IV) resynchronization accomplished by temporary pacing using epicardial pacing wires was evaluated in 20 children after surgery for congenital heart defects aged 3.4 months to 14.0 years fulfilling the following criteria: 1. presence of AV and/or IV conduction delay and 2. need for inotropic support. AV resynchronization (n = 13) was achieved by AV delay optimization during atrial synchronous RVOT pacing. IV resynchronization (n = 14) was accomplished by atrial synchronous pacing from lateral RV wall in 7 patients with right bundle branch block and normal AV conduction and by atrial synchronous multisite ventricular pacing in 7 patients after AV resynchronization. AV resynchronization resulted in increase in arterial systolic, mean and pulse pressure of 7.2 +/- 8.3 % (p < 0.01), 8.6 +/- 8.1 % (p < 0.005) and 6.9 +/- 15.5 % (p NS), respectively. IV resynchronization used either alone or added to previously performed AV resynchronization led to (further) pressure increase of 7.0 +/- 4.7 %, 5.9 +/- 4.7 % and 9.3 +/- 7.8 %, respectively (p < 0.001 for all). Combined effect of AV and IV resynchronization resulted in systolic, mean and pulse pressure increase of mean (range) 10.2 +/- 5.0 (4.3 – 19.1) %, 8.6 +/- 5.4 (0.8 – 18.8) % and 15.2 +/- 8.5 (6.1 – 33.3) %, respectively (p < 0.001 for all). Increase in systolic arterial pressure after IV resynchronization was positively correlated with both baseline QRS duration (r2 = 0.62, p < 0.05) and extent of QRS shortening (r2 = 0.66, p < 0.05). In conclusion, resynchronization pacing led to a significant increase in arterial pressure and was a useful adjunct to the treatment of acute postoperative heart failure.

2

Somatostatin; a new therapeutic option for the treatment of chylothorax

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The standard treatment of chylothorax in pediatric intensive care today includes conservative therapy with fat free nutrition, total parenteral nutrition and if this is not successful an operative treatment (pleurodesis, ligation of the duct, pleuroperitoneal shunt). We describe four patients who had chylothorax and did not respond to conservative treatment with fat free nutrition or TPN. They were not in the condition for operative treatment and were treated with continuous infusion of somatostatin. Starting dose was 3.5

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

micro/kg/min and infusion was increased every day up to 10 micro/kg/min after three days. In three patients, chylothorax ceased with the continuous somatostatin infusion without severe side effects during the first 9 to 11 days of treatment. One patients with Down syndrome was treated without success and died on cardiac failure. Conclusion: Somatostatin is a therapeutic option for treatment of chylothorax and could reduce surgical intervention, hospitalization time and allows early enteral feeding.

3

Doppler echocardiography during nitric oxide inhalation for neonatal pulmonary hypertension

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To assess the effects of Doppler echocardiography on monitoring pulmonary hypertension during inhalation of nitric oxide (iNO) in the treatment of neonatal hypoxic respiratory failure. 15 neonates (2.44 ± 0.59 kg of body weight) with pneumonia, NRDS or PPHN were admitted into NICU in the age of 1–20 days. All patients had respiratory failure in aggressive mechanical ventilation. Doppler echocardiography combined with Color Flow Mapping was utilized to examine the cardiac function reflected by left ventricular ejection fraction (LVEF) and to quantitate systolic pressure of pulmonary artery (SPAP) based on tricuspid regurgitation (TR), PDA or VSD shunting. Systolic blood pressure (SBP) was read when echocardiography was carried out, and a ratio of SPAP/SBP was calculated. The results showed that all patients had PDA with 5 right-to-left, 5 left-to-right and 5 bi-directional shunt. TR was revealed in 11 cases. One patient had also perimembranous VSD with bi-directional shunt. SPAP increased in 10 patients ranged at the level of 59±17 mmHg prior to iNO and decreased to 42±3 mmHg in 20–120 min, and to 36±10 mmHg in 12–24 hr post initiation of iNO at 3–10 ppm, respectively ($p<0.001$). SPAP/SBP was 0.90±0.12 prior to iNO and decreased to 0.69±0.10 in 30–120 min, and to 0.57±0.17 at 18–24 hr post initiation of iNO ($p<0.001$). However, significant changes were not observed for both LVEF and SBP post initiation of iNO. We concluded that Doppler echocardiography is invaluable in monitoring the efficacy of iNO. The strategy of iNO is effective and safe in the treatment of pulmonary hypertension due to hypoxia.

4

Serum s-100b release after infant cardiac surgery is associated with reduced cerebral blood flow velocity and abnormal electroencephalogram

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BACKGROUND: Serum S-100B has been reported as a marker of cerebral injury after cardiac surgery. The purpose of this study is to explore the relationships between S-100B release and changes in electroencephalogram (EEG) and cerebral blood flow velocity (CBFV) after infant cardiac surgery. METHODS: 16-channel video synchronised, continuous EEG monitoring was performed for a 24-hour-period from 6 to 30 hours after surgery in 27 infants with no preoperative neurological abnormalities. Peak CBFV in the anterior cerebral artery was measured by ultrasonography at 1, 2, 3 and 5 hours post-reperfusion. S-100B was measured by radioimmunoassay at the end of bypass (early S-100B) and 24 hours after reperfusion (late S-100B). RESULTS: Patient age was 57±9 days (range: 4–142 days). Early S-100B level was 1.30±0.48μg/l and late S-100B level was 0.43±0.25μg/l. For the entire study group, high early S-100B correlated with longer bypass time ($r=0.51$, $p<0.0061$) and was associated with the use of circulatory arrest ($p=0.02$), but not the length of circulatory arrest. Higher levels of late S-100B correlated with reduced CBFV at 1, 2 and 3 hours post-reperfusion ($r=-0.45$, $p=0.0195$, $r=-0.44$, $p=0.0221$ and $r=-0.51$, $p=0.008$, respectively). Three of the 27 (11%) patients had an abnormal EEG. An abnormal EEG was associated with high late S-100B levels ($p=0.038$, odds ratio at 0.1μg/L increase of late S-100B, 2.07, 95% confidence interval 1.041–4.115). Neither bypass time nor age was associated with abnormal EEG. CONCLUSION: Higher serum S-100B protein release after infant cardiac surgery is associated with reduced cerebral blood flow velocity. An elevated serum S-100B protein level at 24 hours after open heart surgery in infants may be a marker for post-operative cerebral dysfunction.

5

Recovery of cerebral blood flow velocity in infants following hypothermic cardiopulmonary bypass

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BACKGROUND: Reduced cerebral blood flow velocity (CBFV) in infants after profound hypothermic cardiopulmonary bypass (HCPB) has been reported. Blood haemoglobin level may also affect the recovery of CBFV in infant cardiac surgery. The aim of this study is to evaluate the influence of the degree of both hypothermia and haemodilution on CBFV following infant cardiac surgery. METHODS: Peak CBFV in the anterior cerebral artery was measured by pulsed Doppler intermittently throughout the operation until 5 hours post-reperfusion in 28 infants undergoing cardiac surgery. Arterial haemoglobin was measured at the same time. The patients were divided into three groups according to the degree of intraoperative hypothermia: mild (30–32°C, n=8), moderate (22–25°C, n=16) and deep (17–19°C, n=4). RESULTS: Peak CBFV decreased significantly ($p<0.01$) in all the groups to less than 50% of the preoperative level during HCPB and returned to more than 65% of the prebypass level 1 hour following reperfusion. However, compared to the moderate group, both mild and deep groups had a lower peak CBFV recovery at 1 hour ($p<0.001$), 2 hours ($p<0.01$) and 3 hours ($p<0.05$) after reperfusion with a higher haemoglobin level ($p<0.05$). It was also found that there was a negative correlation between peak CBFV and haemoglobin level at 1 and 2 hours after reperfusion in the whole cohort of patients ($r=-0.56$, $p=0.002$ and $r=-0.37$, $p=0.05$, respectively). CONCLUSION: The recovery of cerebral blood flow velocity in infants with HCPB may be affected by both the degree of hypothermia and haemoglobin level. Reduced CBFV was associated with higher haemoglobin levels following infant cardiac surgery. Attention to postoperative haemoglobin levels may be of importance in optimizing neurological protection.

6

Pulse oximetry enhances accuracy of capnometry in cyanotic heart disease

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We evaluated the relationship between delta-CO2 (the difference between arterial [paCO2] and end-tidal [pET-CO2] carbon dioxide partial pressures) and systemic oxygen saturation (SpO2), to assess whether SpO2 could be used as a correction factor to estimate the paCO2 from the pET-CO2. The anesthetic records of 64 interventions (100 data sets) in 50 children with cyanotic heart disease were evaluated. Interventions included prostaglandin E therapy, interventional catheterization and surgery. pET-CO2 was monitored by a mainstream capnometer attached to the ventilator circuit. Continuous pulse oximetry was performed and arterial blood gas samples were taken during normothermic 'steady state' before and after interventions. The correction formula derived from these retrospective data were validated prospectively in 39 interventions in an additional 34 patients with cyanotic heart disease (61 data sets). Regression analysis revealed a good correlation between SpO2 and delta-CO2 ($r^2=0.89$, $p<0.01$). The resulting regression equation, corrected pET-CO2 = raw pET-CO2 -0.37 × SPO2 + 40, was used to calculate the paCO2 from the pET-CO2 for any spO2 and subsequently validated in the prospective study. The r^2 for the correlation between paCO2 and raw pET-CO2 was 0.12, while the r^2 for correlation between paCO2 and corrected pET-CO2 was 0.95 ($p<0.01$). The raw pET-CO2 bias (+/- 2SD) was 12 (-1.3 and 25.4) mm Hg, while the thus corrected pET-CO2 bias was 0.23 (-4.2 and 4.7) mm Hg. Decision-making with respect to ventilator setting would have been accurately supported in 92% of cases while using corrected pET-CO2 values, compared with 58% of cases when using raw pET-CO2 values alone. Capnometry is enhanced when used in combination with SpO2 monitoring, and correcting pET-CO2 for the degree of hypoxia.

7

Inhaled Nitric Oxide (iNO) in pediatric cardiac surgery: end of the honey-moon?

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Our clinical experience with the use of iNO starts in 1993 and includes 50 pts undergoing corrective surgery, mean age 4 mo (2 wk-9 mo) and mean weight 3.3 Kg (2.7–6.8 Kg). The underlying heart defects were: complete av

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

septal defect 30 | 91% with Down's syndrome), truncus arteriosus 6, total anomalous pulmonary venous drainage 6, single ventricle undergoing modified Fontan operation 2 and 2 pts with LV and RV assisted device for LV and RV failure respectively. Mean dose of iNO was 18±8 ppm, mean duration of treatment was 5±9 d, mean oxygen saturation under treatment was 98±2 %, mean FiO2 0.6. Mean intubation period 8±4 d. There were 3 deaths (6%), 1 for sepsis the other 2 for refractory hypertensive pulmonary crisis. Among our study group, 7 pts, with the longest period of treatment (mean of 8 d), developed hypertensive crisis upon discontinuation of treatment with iNO and required a longer period of endotracheal intubation. Conclusions: iNO is an indispensable therapeutical tool in the post operative management of pediatric cardiac pts but 1) the use of iNO reduces the production of endogenous NO (normally restored after 48 hr from the ECC), 2) during the weaning from NO low saturations should be accepted until the pulmonary endothelium restores its capacity of producing endogenous NO 3) the longer the period of iNO treatment, the more difficult is to wean the patients with subsequent prolungation of their endotracheal intubation this may lead to significant adverse effects 4) together with iNO we suggest the administration of low dose of systemic vasodilators such as nitroprusside, in order to stimulate the endogenous production of cGMP with the same pattern of NO

8

Lessons in nosocomial infection control in pediatric cardiac surgery in a new hospital in a developing country

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Pediatric cardiac-surgical patients are more prone to acquire nosocomial infections compared to adults because of longer duration of ventilation and invasive lines, more number of infusions and lower immunity compared to adult patients. Retrospective analysis of data from 340 pediatric patients who underwent cardiac surgery between July 1,1999 and December 31,1999 in our hospital showed nosocomial infection rate of 12.1%. The measures taken to decrease problem of nosocomial infection included installation of laminar flow in the ICU for preparation and dilution of pharmacological infusions, installation of UV tubes in scrub area, regular check on water quality by water cultures from OT and ICU area, improving quality control and checks on CSSD for sterility, inservice in-house teaching of staff about management of intravascular lines misuse of intravenous and respiratory filters. With these measures, the infection rate in next 6 months(Jan 1 2000 to June 30 2000) decreased to 5.7% in 395 patients. The measures for preventing nosocomial infections in pediatric cardiac surgery patients are relevant in the developing world.

9

Relationship between venous saturation and anaerobic metabolism in neonates following one or two ventricle procedures

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Critical limitations in oxygen delivery result in perioperative organ dysfunction and death. Compared to neonates undergoing an atrial switch procedure (2VP), neonates undergoing single ventricle palliation (1VP) have hemodynamic limitations imposed by the obligate inefficiency of mixing physiology, arterial desaturation, and parallel circulation. In 1VP patients, arterial saturation is sometimes titrated with the intent of improving systemic oxygen delivery, although tissue oxygen levels may then fall below a critical threshold. We hypothesized that the anaerobic threshold would occur at a similar venous saturation in both groups of patients. A prospective perioperative database was maintained on seventy high-risk neonates undergoing either 1VP (n=51) or 2VP (n=19). All patients had continuous SvO2 monitoring for the first 48 postoperative hours. Arterial blood gases were obtained at standard intervals. A standard base excess (BE) < -4 mEq/L or a change in BE exceeding -2 mEq/L/hour were used as indicators of anaerobic metabolism. The relationship between SvO2 and anaerobic risk was tested by logistic regression and the likelihood ratio test. Data from 3000 hours of monitoring were analyzed. The overall anaerobic risk was 4.1% in 1VP and 1.8% in 2VP. In each group the risk of anaerobic metabolism increased as SvO2 approached 30% (p<0.001). There was no significant difference in the risk of anaerobic conditions between 1VP and 2VP until SvO2 <30% (p<0.001). The anaerobic threshold is the same in neonates undergoing 1VP and 2VP. Patients undergoing the Norwood procedure do not have unique tolerance to venous desaturation. Strategies to optimize SvO2 are thus justified in both groups to reduce the risk of sub-lethal organ damage or mortality.

10

Sedation with propofol and remifentanil in pediatric cardiac catheterization

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In pediatric cardiac catheterization sedation and analgesia are necessary. Routinely intravenous sedatives and central-acting analgesics are combined and mainly applied as bolus injection. Because of long half time of commonly used drugs individual control is not satisfactory. Our aim was to establish a secure and well controllable regimen for analgesia and sedation in cardiac catheterizations. We performed cardiac catheterization in 60 pediatric patients, aged 0 – 16 years (mean 3.7 years) using continuous infusion of short-acting agents. Twelve patients (20%) underwent catheter intervention. After premedication with midazolam (0.1mg/kg, i.v.) patients received an initial bolus of propofol (mean 1.5 mg/kg), followed by continuous infusion of propofol and remifentanil. Vital signs, amount of sedatives needed and quality of sedation were monitored. The sedation was carried out by an experienced intensivist. For induction of sleep a mean of 2.5 mg/kg propofol and 0.5µg/kg remifentanil were needed. To maintain deep sedation 0.072 and 0.03 µg/kg/min were required, respectively. Twenty-six patients received temporarily increased infusion rates (mean 0.092 mg/kg/min and 0.025 µg/kg/min) or an additional propofol bolus (0.5 – 1.0 mg/kg) to prevent waking up. There was no significant difference between infants and patients over 1 year of age. All catheterizations could be performed under spontaneous breathing. There were no severe complications related to sedation. In a mean time of 17 min after cessation of drug application patients were awake. Continuous application of propofol and remifentanil provides a safe and feasible method for sedation in

11

Effects of induced hypothermia for treatment of low cardiac output immediately after Fontan operation

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Cardiac output after Fontan operation is dependent on pulmonary vascular resistance because of lack in pumping chamber in pulmonary circulation, and therefore inotropic agents have limited effects on treatment of low cardiac output immediately after Fontan operation caused by acute elevation of pulmonary vascular resistance by cardiopulmonary bypass. Effects of induced hypothermia (rectal temperature of 34°C, under general anesthesia) for treatment of low cardiac output immediately after Fontan procedure (appendage-to-pulmonary artery anastomosis, F) were studied in 16 patients (pts): hypothermia group (H) after F, 30 pts. normothermia group (N) after F, 10) using hemodynamic monitoring by Electromagnetic flowmeter in operating room and Swan-Ganz catheter in ICU. PH was managed in the alpha-stat manner. The results showed 1. Increased pulmonary vascular resistance and reduced ventricular filling immediately after cardiopulmonary bypass. 2. Lower heart rate with comparable cardiac output in group H compared with group N. 3. Higher mixed venous saturation in group H compared with group N. 4. Increased IVC/SVC flow ratio during hypothermia. The results suggest that induced may improve ventricular filling by lowering heart rate, and may exert protective effects in abdominal organs by lowering oxygen consumption and improving blood flow distribution in patients with low cardiac output immediately after Fontan- type procedures.

12

Impact of intraoperative transesophageal echocardiography in surgical repair of congenital heart disease in children

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Background: Intraoperative transesophageal echocardiography (TEE) in surgical repair of congenital heart disease is reported to have a 3–19% surgical impact on pre cardiopulmonary bypass (CPB) attitude, and prompted a return to CPB for further repair of residual defects in 0–13% of cases. Design: We analyzed 465 consecutive intraoperative TEE performed on children ranging from 1 day-16 years old, during congenital heart repair from 1991 to 1999. Material and methods: Intraoperative TEE diagnostic results were reviewed, and their impact on surgical approach were studied both before

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

and after CPB searching for residual defects prompting a return to CPB for further repair. Safety of intraoperative TEE was studied. Total number included failed TEE. Results: TEE had a preCPB diagnostic impact in 109 of 465 examinations (25%), of which 44 (9.5%) had impact on surgical approach. Highest impact was found in anomalous venous returns (57%) ($p = 0.002$), and VSD (17%) ($p = 0.02$). PostCPB TEE revealed major residual defects in 61 patients (13%) of which 28 (6%) prompted further repair under CPB. Highest impact was found in mitral valve repair (45%) ($p = 0.0002$). TEE showed absence of residual defects in 22 of reoperated cases (5%). TEE failure occurred in 5 infants (1.1%) all < 5kg, 2 for missed probe insertion (0.4%), 3 because of ventilation problems (0.6%). Accidental TEE related extubation occurred in 2 infants (0.4%) without consequence. No complications occurred in children > 5kg. Conclusion: Intraoperative TEE in congenital heart repair in children is valuable, with a therapeutic index of 5%. Given the variation of highest impact within diagnostic groups in pre and postCPB periods, and the low complication rate we recommend intraoperative TEE in all children > 5 kg, and cautiously in small infants.

Session 3: Cardiac Imaging: CT, PET, MRI-MRA

13

The quality and usefulness of spiral CT and 3-D images in patients with central airway disease associated with congenital heart disease

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We analyzed the quality and usefulness of the spiral CT and three-dimensional images in 56 patients with central airway stenosis associated with congenital heart disease. Forty-nine patients (88%) were less than 5 years old, including 3 neonates and 33 infants. Their median age was 9.7 months and median body weight was 7.6 kg. Spiral scanning was performed after sedation with chloral hydrate (n=47) and administration of contrast media (n=56) via the pedal route (n=48). It was performed with 1-3 thinner collimation as possible from 1 to 2 mm and overlapped reconstruction of 50–70%. The airway stenoses were located at trachea in 26 and bronchus in 30. Their causes were vascular anomalies (n=8), posteriorly malpositioned aortic arch (n=7) and ascending aorta (n=5), innominate artery compression (n=6), aortic arch aplasia (n=8), absent pulmonary valve (n=6), dilated or dilated cardiovascular structure (n=20), and pulmonary artery sling (n=2). Motion artifact caused mild or negligible image degradation in most patients except 7. The quality was graded as good in 32 and excellent in 15. Non-sedated children with breath-holding showed more severe motion artifact by cardiovascular pulsation. In the evaluation of the airway lesions associated with congenital heart disease, the inspiration artifact caused the image degradation, but diagnostic three-dimensional images could be obtained. The spiral CT and three-dimensional reconstruction may be served as primary diagnostic modality in the uncooperative children with congenital heart disease and suspected airway stenosis.

14

Cardiac response to exercise in patients after atrial correction of transposition of the great arteries assessed by exercise-MRI

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Background: After atrial correction for transposition of the great arteries (TGA), reduced exercise capacity is common. Currently, magnetic resonance imaging (MRI) techniques are available to study biventricular function in response to exercise. The purpose of the present study was to evaluate cardiac function at rest and with exercise in atrially corrected TGA patients using ultra-fast MRI. Methods: In 24 TGA patients after Mustard or Senning correction (20±5 years, NYHA class I or II) and 16 controls (25±5 years) cardiac function was evaluated at rest and with exercise using a Philips MR-scanner and a MR-compatible bicycle ergometer. MR-exercise level was based on 80% of peak oxygen uptake, measured during a preceding graded maximal exercise test. Results: Oxygen uptake at peak exercise was lower in the patients (29±7 vs 42±5ml/min/kg, $p<0.05$), as was peak heart rate (162±23 vs 176±9bpm, $p<0.05$). At rest, 22 (92%) patients had a normal (>=47%) right ventricular (RV) ejection fraction (EF) and all had a normal left ventricular (LV) EF. However, in only 2 (9%) patients, RV EF showed a normal (>=56%) increase with exercise and 5 (20%) patients had a normal LV EF response to exercise. In the patients, RV and LV stroke volume did not increase in response to exercise and RV end-diastolic (EDV) and end-systolic volume (ESV) increased (EDV: 158±57 to 165±59ml; ESV: 70±35 to 76±38ml; both $p<0.05$). LV-EDV and LV-ESV did not change with exercise. Conclusion: Exercise-MRI is feasible in TGA patients after atrial correction, revealing an abnormal response of RV and LV to exercise in 21 of the 23 patients with normal resting RV and LV EF. Not only RV, but also LV function was depressed. These results confirm concerns on long-term ventricular function after atrial correction of TGA.

15

Noninvasive quantification of left-to-right intracardiac shunt volume by magnetic resonance phase-shift techniques in children with normal hearts and with congenital heart disease

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The purpose of the present study was to determine whether velocity-encoded phase-difference magnetic resonance imaging (MRI-PC) can assess the magnitude of intracardiac left-to-right shunting in children with ASD and VSD. The accuracy and precision of MRI-PC and cine gradient echo magnetic MRI were studied in 9 control children following Kawasaki disease without valve regurgitation nor without cardiac shunting. Ten patients with ASD(age range:1 to 13 years) and eleven patients with VSD(age range:1 to 10 years) underwent cine MRI and MRI-PC measurements of flow in the proximal aorta, main pulmonary artery and ASD, following by cardiac catheterization. Stroke volume measured by cine MRI was excellently correlated with the flow in the proximal aorta in the control children($r=0.999$) and in the patients with ASD($r=0.989$). In control children, there was a excellent correlation($r=1.000$) between the flow in the proximal aorta and in the main pulmonary artery. In patients with ASD, the correlation of the pulmonary-to-systemic blood flow ratio(Qp/Qs) measured by MRI-PC and by oxymetry was good($r=0.744$, $p<0.05$). We could measure directly the shunt flow through ASD by MRI-PC for the first time. The shunt flow directly measured by MRI-PC was excellently correlated with the added flow in the proximal aorta and the main pulmonary artery. In patients with VSD, the correlation of the Qp/Qs measured by MRI-PC and by oxymetry($r=0.902$ $p<0.01$) was better than that in patients with ASD. These data show that the clinical value of MRI-PC must be considered within the perspective of the standard imaging modalities usually applied in children with ASD and VSD.

16

Accuracy of 3D contrast-enhanced MR angiography evaluation of aortic abnormalities in children

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To assess the accuracy of 3D contrast-enhanced MR angiography (MRA) in the diagnosis of aortic abnormalities in infants and children, we prospectively studied 72 patients (median age 13 months, range 5 days-15 years) with congenital heart disease. X-ray angiography (X-ray) within a median of 2 days was used as the gold standard for the comparison of results. Diagnoses included three types of aortic pathology: right aortic arch and or aberrant aortic branch vessels (n=27), stenosis or dilatation (n=20), and shunts (n=33, 62 vessels). Gadolinium-enhanced (0.1mmol/kg) MRA was performed on a 1.5 T Signa (GE Medical Systems, Milwaukee, Wisconsin). Independent observers assessed images for aortic anatomy using multiplanar reconstruction (MPR), maximum intensity projection and shaded surface display. The vessel diameters of ascending aorta (AAo, n=50), descending aorta (DAo, n=62), subclavian artery (SCA, n=62) and stenotic segments (n=15) were measured using MPR. MRA was consistent with X-ray in all patients with right aortic arch and or aberrant vessels and with stenosis or dilatation. MRA correctly diagnosed 54 shunts (87%)' 1.6 mm in diameter with X-ray and there was no false positive. Shunt vessels' 1.5 mm in diameter were not identified by MRA. In measuring the vessel diameter of AAo, DAo, SCA and stenosis, mean differences between MRA and X-ray values (MRA–X-ray) were -0.9 (95%CI: -3.1, 1.4), 0.1 (-1.6, 1.7), -0.2 (-1.7, 1.3) and -0.6 (-2.1, 0.9) mm, respectively. Interobserver and intraobserver variability of diameter measurements performed on MRA was' 0.2 (95%CI: -1.0, 1.4) and' 0.6

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

(0.4, 1.6) mm, respectively. MRA is an accurate noninvasive modality to delineate most aortic abnormalities. An additional X-ray may be reserved for selected patients with small short vessels.

17
Evaluation of the pulmonary veins using contrast-enhanced magnetic resonance angiography
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Background: Pulmonary vein (PV) abnormalities are often associated with other cardiac lesions, and their accurate assessment is essential. Transthoracic echocardiography (echo) and angiography have some limitations. Contrast-enhanced magnetic resonance angiography (CE MRA) is a new imaging technique with high spatial resolution, large field of view and short acquisition times. The obtained data can be reconstructed into 3D images and reformatted in unlimited planes. Purpose: To assess the diagnostic value of CE MRA for anatomical evaluation of the PVs. Methods: 26 CE MRA were performed in 25 patients (median age 6yrs, range 2 weeks to 15yrs). Three sets of data were acquired using spoiled gradient echo (SPGR) technique after Gadopentate (0.3 mmol/kg) injection and post-processed for multiplanar and 3D reconstruction with hardcopy software. Each PV was evaluated regarding the site of connection, the course within the angle, the presence of obstruction or hypoplasia and the relationship to the adjacent structures. The MRA findings were compared with echo, angiographic and operative findings, when available. Results: Indications for CE MRA were suspected PAPVC in 8, preoperative evaluation of PVs in 5, intracardiac tumor in 3, confirmation of known PV abnormalities in 3, investigation of pulmonary hypertension in 2 and diaphragmatic hernia in 1 patient. CE MRA defined normal PV in 10, PV obstruction in 8, PAPVC in 5, TAPVC in 2 patients. 99 % (103/104) of all PVs were visualized with CE MRA. 82% (96/104) with echo. CE MRA diagnosis was concordant with echo diagnosis in 79% (68/86), with angiographic diagnosis in 96% (23/24) and with surgical diagnosis in 100% (6/6). Conclusion: CE MRA is a non-invasive fast imaging technique for anatomical evaluation of the PVs, with a high rate of detection and accuracy.

18
Comparision of quantitative pulmonary flow by magnetic resonance imaging and lung perfusion scintigraphy late after Fontan like palliation
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Background: Quantitative evaluation of right/left lung flow ratio using lung perfusion scintigraphy in patients after Fontan or total cavopulmonary connection (TCPC) palliation is difficult. Mainly due to preferential draining of the venae cavae to one lung. Nevertheless scintigraphy is considered the gold standard in determing pulmonary blood flow patterns in individuals with normal cardiac anatomy. We prospectively evaluated the feasibility and accuracy of phase velocity cine (PVC) magnetic resonance imaging (MRI) in combination with flow analysis software (Mass flow ©) to determine the ratio of right to left pulmonary perfusion compared to conventional lung perfusion scintigraphy. Methods: Hemodynamic data of 32 patients 9.4 ± 3.9 years after Fontan like palliation was evaluated. Right and left pulmonary blood flow (RBF/LBF) was quantified with PVC MRI by two observers utilizing Mass flow ©. Right to left flow ratio (FR) was calculated by using following equation: FR = RBF/(RBF+LBF). FR was also cacul by standard lung perfusion scintigraphy after infusion of 99mTc microspheres in an upper limb. Patients after TCPC palliation recieved on two subsequent days an injection into the upper and lower limb, respectively. The right lung flow quota of the two injections were added and set into proportion to the sum totalled left lung flow quotas using the above mentioned equation. Results: Preliminary data for 10 patients are available (figure). Six patients were s/p Fontan palliation (a), four patients s/p TCPC palliation (b). Intra/interobserver variability of FR obtained with PVC MRI was very low. Correlation of FR identified with PVC MRI and scintigraphy was high (r=0.98). In patients with TCPC scintigraphy seems to deliver higher FR than PVC MRI. Conclusion: Flow quantification with PVC MRI is feasible in patients after Fontan like palliation. A potential advantage for flow assessment with PVC MRI against scintigraphy may exist for patients with TCPC. It does not need two injections on subsequent days into the upper and lower limb. This leads to artificial calculation products and increases radiation not only for the patient but also for other patients and staff. Another benefit of MRI may be in the possibility of quantifying and showing the cause of pulmonary blood flow patterns in one session. Neither scintigraphy nor angiography can meet both demands in one examination.

19
Quantitive assessment of ventricular volume, mass, and flow with cardiac magnetic resonance imaging in adults with repaired Tetralogy of Fallot. Relationship with electrocardiographic predictors of sustained ventricular tachycardia, and sudden death
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Background: Pulmonary regurgitation (PR) is a common sequellae of Tetralogy of Fallot (TOF) repair. QRS prolongation predicts sustained ventricular tachycardia and sudden death, the most devastating complications late after TOF repair. We examined the precise relationship of chronic pulmonary regurgitation, ventricular volume, mass and function and QRS duration in adults with repaired TOF. Methods: We used a Picker Edge 1.5T Cardiac Magnetic Resonance scanner to acquire 12-14 contiguous 1 cm cine slices of both ventricles for measurements of volume, mass and function. Outflow curves and regurgitant fractions were measured by phase velocity mapping of flows. QRS was measured manually from standard ECGs. MRI indices were indexed to body surface area (m2). Results: Thus far we studied 37 adults with repaired Tetralogy (mean age 32±10 years, mean time since repair: 22±7 years). Results are presented in the table below. TABLE: Pulmonary regurgitant fraction was 24±17%. The RV Stroke Volume index was significantly higher than that of the LV due to pulmonary regurgitation. Multivariate forward stepwise regression with all the volume, mass and function parameters considered as independent predictors showed that only RVESVi predicts QRS duration (r=0.77 p<0.0001). PR fraction was predictive of RVEDVi (r=0,69 p<0.001) which in turn was predictive of RVESVi (r=0,87 p<0.0001).Furthermore there was a negative correlation between RVEF and PRF (r=-0,44 p=0.006). There was no correlation between QRS duration and left ventricular volume or mass suggesting that QRS prolongation reflects solely RV changes. Conclusions: Pulmonary regurgitation induces changes in RV volume and mass, which in turn lead to QRS prolongation and increased propensity to malignant arrhythmias and sudden death. Cardiac Magnetic Resonance imaging unravels the mechanism of arrhythmogenesis and supports the mechano-electric theory late after repair of TOF.

20
Quantification of mild to moderate pulmonary regurgitation by magnetic resonance (MR) phase velocity mapping in patients after repair of Tetralogy of Fallot (ToF)
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Background: In clinical setting quantification of pulmonary regurgitation (PR) is not feasible. Clinical practice comprises assumptions from Doppler echo or cine-angiography. This study evaluated the feasibility of MR phase velocity mapping in quantifying residual PR in patients after repaired ToF. Healthy volunteers served as controls. Methods: 11 patients (age: 13.9±3.7 yrs) 12.3±4.7 years after ToF repair with echocardiographic signs of mild to moderate PR and 10 healthy volunteers (age: 28.0±6.3 yrs) were studied. Stroke volumes of the right and left ventricle were obtained by flow tracings in the pulmonary artery at the level of the pulmonary valve and midascending aorta. Flow volumes were analyzed by special flow analysis software (MASSFLOW©). PR in the patient group was calculated from negative portions of pulmonary tracings. To identify interobserver variability flow measurements in normal volunteers were carried by two independent investigators. Results: Mean regurgitant volume over the pulmonary valve was 25.5±17.8 ml in the patient group and 2.5±4.9 ml in the control group (p<0.001). This corresponded to 20.7±14.9% and 1.9±3.0% of total forward stroke volume in the patient and volunteer group, respectively. None of the mild regurgitations was missed by MR flow mapping. Net forward stroke volume in the pulmonary artery correlated well with stroke volume in the aorta (r=0.68, p<0.05). Interobserver variability in calculating stroke volumes was very small (r=0.97, p<0.001). Conclusion: MR velocity mapping is a sensitive tool in assessing and quantifying mild to moderate PR with low interobserver variability.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

21

Stability of Amplatzer occluders and image quality during magnetic resonance

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Amplatzer(r) Occluders are new devices undergoing international multicentre trials. Preliminary in vitro studies (AGA Medical Corp., MN) using Siemens 1.5 MRI apparatus found the devices to be stable with minimal artefacts. Between May 1998 and January 2000, 75 Amplatzer(r) Occluders were used to close atrial communications, patent arterial ducts and unwanted extracardiac vascular anomalies in 119 patients, age 0.4 – 77.8 years. Transcatheter closure was attempted and abandoned in two additional patients with atrial septal defects (ASD). The size of the atrial septum was not big enough to accommodate the left atrial disc. There were no undesirable device embolisation, and all device were stable with closure rate comparable to the AGA international registry. Magnetic resonance imaging (MRI) was performed in two patients to investigate transient (3 days) aphasia in one, age 7.9 years, within 6 hours of an Amplatzer(r) Septal Occluder (13 mm ASO) placement, and in another age 0.4 years, to investigate spina bifida 2 months after an Amplatzer(r) Duct Occluder (0.4 mm ADO) to preclude a cardinal vein and a fourth patient to evaluate residual shunt after attempt closure of a large inlet VSD. Three devices were used in the last patient. Imaging was performed on a 1.5-T Magnetom Symphony Imager(Siemens Medical Systems, Iselin, NJ). Multiplanar imaging using spin echo, and HASTE sequences were performed of the body parts of concern, head of 2 patients and spine of one. The position of the devices examined fluoroscopically and radiographically after exposure to MRI and compared to that immediate after placement were found to be stable and did not moved. This clinical study confirmed the in vitro examinations that the Amplatzer(r) Occluders are compatible with magnetic resonance imaging.

22

Noninvasive quantification of left-to-right shunt in 50 pediatric patients by phase-contrast cine magnetic resonance imaging: a comparison with invasive oximetry

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Background: Blood flow can be quantified noninvasively by phase-contrast cine magnetic resonance imaging (PC-MRI) in adults. Little is known about the feasibility of the method in children with congenital heart disease. Methods and Results: In 50 children (mean 6.2 years, range 1.1–17.3) with an atrial or ventricular level shunt, blood flow rate in the great vessels was determined by PC-MRI and the ratio of pulmonary to aortic flow (Qp/Qs) compared with Qp/Qs by oximetry. We found a difference of 7% and a range of –20% to + 20% (limits of agreement, mean ±2SD). In another seven children with congenital heart disease but no cardiac shunting (mean 7.9 years, range 1.3– 17.7), Qp/Qs by PC-MRI was 1.02 (SD ±0.06). No difference between systemic venous and aortic flow volume was found (range –17% to +20%, n=37). Blood flow through a secundum atrial septal defect as assessed by PC-MRI (n=24) overestimated the shunt compared with the difference between pulmonary and aortic flow. The mean difference between three separate PC-MRI measurements in each location was 5.9% (SD 4.0, n=522), demonstrating good precision. The interobserver variability was low. Accuracy of PC-MRI was confirmed by in vitro experiments. Conclusions:

23

MRI Dobutamine stress in adult patients with congenital heart disease: effect on contractility and filling characteristics

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Objective: We examined the role of magnetic resonance imaging (MRI) dobutamine stress in the detection of right ventricular (RV) dysfunction in asymptomatic and slightly symptomatic patients with chronic RV pressure overload. Methods: Thirty asymptomatic or slightly symptomatic patients with chronic RV pressure overload 12 surgically corrected transposition of the great arteries (sTGA)(Mustard or Senning), five congenitally corrected (ccTGA), 13 corrected tetralogy of Fallot and nine age and sex-matched healthy volunteers were included. MRI was applied both at baseline and during dobutamine stress (maximum dose 15_g/kg/min) to determine RV and left ventricular (LV) volumes and ejection fraction (EF). Patients with valvular regurgitation > 10ml/beat were excluded from the study. Results: At baseline RVEF in sTGA was significantly lower than in controls [27(11)% vs 71(9)%, p=0.006]. During dobutamine stress RVEF increased significantly in controls and patient groups except for the Fallot patients. RV stroke volume (SV) increased in controls [22(19)%, p=0.02] and in ccTGA [80(14)%, p=ns]. In Fallot and sTGA, RVSV decreased [-14(12)%, p=0.003 and -10(28)%, p=ns], respectively, accompanied by a significant decrease in RVEDV [-13(15), p=0.004 and -24(15)%, p=0.0001, respectively]. In Controls and ccTGA there was no change in RV end-diastolic volume (EDV) [2(17)%, p=ns] and 4-4(8)%, p=ns], respectively. Conclusion: There is a clear heterogeneity in response to MRI dobutamine stress between different groups of asymptomatic patients with chronic pressure overload. These data suggest impaired filling in sTGA and decreased contractility in Fallot patients. Dobutamine stress MRI may assist in monitoring RV dysfunction.

24

Decreased cardiac reserve in asymptomatic patients with chronic right ventricular pressure overload: evidence for impaired diastolic and systolic function

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Background: In patients with chronic right ventricular (RV) pressure overload RV function parameters at rest are difficult to interpret and deterioration to RV failure is not easily to predict. We examined the role of dobutamine stress in the early detection of RV systolic and diastolic dysfunction in asymptomatic or slightly symptomatic patients with chronic RV pressure overload using magnetic resonance imaging (MRI). Methods: Fourteen patients with chronic RV pressure overload (nine patients with pulmonary artery stenosis, two patients with corrected Tetralogy of Fallot, three patients with pulmonary hypertension)(RVSP>36 mmHg, age 27±7 years, NYHA class I-II) and nine age- and sex matched healthy volunteers were included. Valvular regurgitation >10 ml/beat was an exclusion criterion. MRI was applied both at baseline and during dobutamine stress (maximum dose 15 µg/kg/min) to determine RV volumes, stroke volume (SV) and ejection fraction (EF). Results: At baseline there were no significant differences in RV parameters between the patients and the controls. RV end-diastolic volume (EDV) 115±22 vs 119±34 ml), RV end-systolic volume (ESV) (54±18 vs. 58±19 ml), RVSV (81±15 vs 81±20 ml), RVEF (71±9 vs 69±10%) and cardiac index (CI) (3.2±0.5 vs 3.7±0.7 L/min/m2). During dobutamine stress the increase in CI in patients was significantly lower than in controls (+14±3) vs +89±26%, p<0.01). Patients showed a significant decrease in RVEDV (-11±13%, p<0.01) and RVSV (-34±13%, p<0.01) and no changes in RVESV (+13±25%, p=ns) and RVEF (+12±6% p=ns). Conclusion: In asymptomatic patients with chronic RV pressure overload a decreased cardiac reserve can be demonstrated with dobutamine stress MRI. A decrease in RVSV is accompanied by both impaired RV filling (diastolic dysfunction) and a failure to augment RVEF (systolic dysfunction) during dobutamine stress.

Session 4: Arrhythmias, Electrophysiology, Sudden Cardiac Death

25

Randomized prospective multicenter study on the use of propafenone and sotalol in young patients. Preliminary results

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Efficacy and side effects of propafenone (P) and sotalol (S) were evaluated prospectively in 112 pts (P=55/57 and S=57/63 at inicial randomization/end of follow-up) aged 0–19.6 (median 5.1) yrs. treated for paroxysmal atrioventricular reentrant tachycardia (AVRT) (n=57), persistent junctional reciprocating tachycardia (PJRT) (n=8), focal atrial tachycardia (FAT) (n=18), chronic atrial tachycardia (CHAT)(n=6), intraatrial reentrant tachycardia

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

(JART, treated with S only) (n=11), ventricular arrhythmias (n=5) and others (n=7). 25 pts. (22.3 %) had structural heart disease. 44 pts. (39.3 %) had a mean of 1.1 antiarrhythmic drugs prior to EVS. Follow up ranged from 1 day – 42.5 (mean 10.6±11.9) mos. EVS was effective in 81.1/88.0 % pts. with AVRT, 80.0/66.7 % pts. with PJRT, 63.6/80.0 % pts. with FAT, 0.0/80.0 % pts. with CHAT and 90.9 % pts. with IART (treated with S only). Exclusion from discontinuation for ineffacy/side effects was not different between P (45/57 = 78.9 %) and S (54/63 = 85.7 %). p NS. Electrophysiologic side effects and proarrhythmia occurred in 3/57 (5.3 %) pts. on P (more AVRT in 1, QRS increase of >50 % in 2) and 5/63 (7.9 %) pts. on S (ventricular hegemony in 1, sinus bradycardia in 4) (p NS). Drug dose or increase in QTc were not predictive of proarrhythmia. Systemic side effects were noted in 2/57 (3.5 %) pts. on P and 2/63 (3.2 %) pts. on S (p NS). Conclusions. P and S had comparable efficacy for common types of supraventricular arrhythmias. Significant or symptomatic proarrhythmia and systemic side effects were rare.

26

Clinical profile of idiopathic sustained left ventricular tachycardia

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Idiopathic sustained ventricular tachycardia originating from the left ventricle (ILVT), especially ILVT with a QRS pattern of right branch block and left axis deviation, is a distinct clinical entity and an indication for catheter ablation. The present study evaluated the clinical features, long-term prognosis and indications for treatment in pediatric patients with ILVT. The subjects were 9 patients (5 males, 4 females) with a mean age at onset of 11.2 years (range, 3 to 15 years); the follow-up period was 7.2 years (mean, range, 0.2 to 11.3 years). Their electrocardiograms during ventricular tachycardia (VT) showed right bundle branch block with upward electrical axis of QRS in 8 patients and downward in one. The heart rate during VT was 140–300 beats per minute. Organic heart disease was ruled out in all patients. In 3 patients, VT was found by electrocardiogram performed as part of a routine school examination. Only one patient showed congestive heart failure. Intravenous administration of verapamil eliminated VT in all 7 patients who received this treatment. In electrophysiological studies, VT was induced by programmed stimulation in 6 of 7 patients. Intravenously administered verapamil could effectively prevent tachycardia in 4 of 5 patients. Oral administration of verapamil was effective in 6 of 8 patients. Verapamil with propranolol or procainide was effective in 2 patients who did not respond to verapamil alone. VT disappeared without drugs in 4 patients during the follow-up period, and became non-sustained in another patient. Two of 3 patients with persistent tachycardia underwent catheter ablation. Atrial dysrhythmic sites were effective for ILVT among these patients and ILVT disappeared in half of the cases. Pharmacologic treatment such as with verapamil is still the treatment of choice for ILVT because of good long term prognosis.

27

Effects of adrenergic beta-antagonists on the qt measurements from exercise stress tests in pediatric patients with long QT syndrome

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Background: Adrenergic beta-antagonists(BB) have been successfully used to decrease the incidence of cardiac events in patients with long QT syndrome(LQTS). Published data suggest that BB therapy shortens the QTc interval and QTc dispersion(QTcdi) on resting ECGs in patients with LQTS. This study attempted to determine the effect of BB therapy on QT measurements from exercise stress tests(EST) in pediatric patients with LQTS. Methods: The ESTs of 32 patients (mean age 12.5 yrs; 20 females) with LQTS performed before and after the initiation of BB therapy were evaluated. Measurements were made of the maximum QTc interval and QTcdi during the various segments of the EST. Analysis involved all 32 patients together and a subset of these patients who met Schwartz's criteria for high probability of LQTS. Results: During exercise, the maximum heart rate(HR) pre-BB averaged 191 compared with the maximum HR post-BB which averaged 158 (p value<0.001). There was no statistically significant difference in the pre-BB and post-BB maximum QTc during the supine (0.476 vs. 0.473, p=0.766), exercise (0.490 vs. 0.503, p=0.136), or recovery (0.494 vs. 0.498, p=0.639) segments of the EST. There was also no significant difference in the pre-BB and post-BB QTcd during the supine (0.042 vs. 0.061, p=0.245), exercise (0.064 vs. 0.068, p=0.539), or recovery (0.052 vs. 0.056, p=0.566) segments. Eight patients were classified as high probability of LQTS. When evaluated separately, there was again no significant difference in the pre-BB and post-BB maximum QTc and QTcd during any segment of the EST. Conclusion: In this study, there was no significant reduction in the QTc or QTcd during rest, exercise, or recovery. Therefore, the protection that BB therapy offers may not be secondary to changes in the QTc interval or QTc dispersion.

28

Exercise testing in conjunction with the Schwartz criteria in the diagnosis of long QT syndrome

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Long QT syndrome(LQTS) is a life-threatening condition often difficult to definitively diagnose. Schwartz, et al. developed criteria based on ECG findings, clinical and family history to aid in the diagnosis. Based on this scoring system, patients can be divided into categories (low, intermediate, high probability). Exercise stress testing(EST) is frequently used as a diagnostic tool as well as for risk stratification. The purpose of this study is to evaluate the use of electrocardiographic findings in EST in conjunction with the Schwartz criteria to evaluate pediatric patients. Methods: EST(n=140) of patients referred for evaluation of possible LQTS were reviewed. Based on the Schwartz criteria, patients were divided into group 1(low, 2=intermediate, 3=high probability). In each group, QTc intervals were determined and analyzed in the following conditions: supine, standing, hyperventilation, peak exercise, recovery. Results: Group 1(n=50) had a mean supine QTc of 0.432 ± 0.024. There were no mean values >0.46 for all conditions. In Group 2(n=55), the QTc increased significantly(p<0.01) in all conditions when compared to supine(0.466 ± 0.014). Standing(mean=0.482), hyperventilation(mean=0.480), peak exercise(mean=0.492), recovery(mean=0.493). Of these, the QTc increased to >0.47 in 46% with standing, 53% in hyperventilation, 65% in peak exercise, and 45% in recovery. In Group 3(n=35), the supine QTc(0.48 ± 0.049) did not increase significantly with standing or hyperventilation, but did increase(p<0.001) with peak exercise (mean=0.505) and recovery(mean=0.514). Conclusions: EST can be used as an adjunct to the Schwartz criteria in the diagnosis of LQTS. It appears to be most useful in patients with low or intermediate probability. This may be clinically useful until more definitive diagnostic testing becomes available.

29

Notched T waves on holter recordings enhance detection of patients with IKT2 (HERG) mutations

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Background: The 2 genes, KCNQ1 (LQT1) and HERG (LQT2) encoding cardiac potassium channels are the most common cause of the dominant long-QT syndrome (LQTS). Besides QT interval prolongation, notched T waves have been proposed as a phenotypic marker of LQTS patients. Methods: The T wave morphology of carriers of mutations in KCNQ1 (n=133) or HERG (n=57) and of 100 control subjects (C) was analyzed from Holter ECG recordings. Averaged T wave templates were obtained at different cycle lengths, and potential notched T waves were classified as grade 1 (G1) in case of a bulge at or below the horizontal whatever the amplitude, and as grade 2 (G2) in case of a protuberance above the horizontal. The highest grade obtained from a template defined the notch category of the subject. Results: T wave morphology was normal in the majority of LQT1 and C subjects compared with LQT2 (92%, 96% and 19% respectively, p<0.001). G1 notches were relatively more frequent in LQT2 (18% vs 8% [LQT1] and 4% , p<0.01) and G2 notches were seen exclusively in LQT2 (63%). Predictors for G2 were young age, missense mutations, core domain mutations in HERG. Furthermore, G2 notches were detected at Holter recording in 15/30 patients in whom the ECG was negative for G2 notches. Conclusion : This study provides novel evidence that Holter recording analysis is superior to the 12-lead ECG in detecting G1 and G2 T wave notches. These repolarization abnormalities are more indicative of LQT2 vs. LQT1, with G2 notches being most specific and often reflecting HERG core domain missense mutations.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

30

Exercise testing in evaluation of syncope in children and adolescents

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Aim of the study is to assess role of stress test in evaluation children with recurrent syncope. 324 children (0.5 girls) age 7–19 years (mean 14.8) with recurrent syncope underwent noninvasive procedures: 12-lead electrocardiogram (ECG), echocardiography, Holter, orthostatic test (OT) – modified head up tilt table test and exercise test – modified McMaster ergocycle continuous protocol. All kids had structurally normal heart, with no significant bradyarrhythmias on Holter. Neurological disorders were ruled out by physical examination and electroencephalogram. Follow up was 1.5 – 6 years (mean 2.1). 184/324 (0.57) had positive OT (40 patients (investigation group-IG) with negative single stage OT underwent exercise test immediately after 20 minutes standing period. When (submaximal heart rate had been reached they were positioned in supine position for 20 minutes (min). After that, they were tilted up for next 20 min. Control group (CG) was consisted of 60 healthy, asymptomatic children matched by sex and age. 54/66 patients had positive OT (41/54 vasodilhibitory, 13/54 cardioinhibitory response) after stress test, while only 1/66 in CG ($p<0.01$). 4/56 had exercise induced ventricular tachycardia (EIVT) with presyncope, none in CG. All patients with EIVT had sporadic single uniform premature ventricular contractions on Holter. Exercise test can be safe and useful noninvasive procedure in evaluation children with syncope. It is helpful in excluding exercise related arrhythmias and seems to be reasonable alternative for sphygmomanometer in second stage of tilt table test in children.

31

Predictive factors of late sudden post-operative complete atrio-ventricular block

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Complete atrioventricular block (AVB) rarely occurs late after cardiac surgery but carries a high risk of sudden death. We have reviewed the records of 10 children, to identify predictive factors of late sudden postoperative complete AVB. From 1990 to 1998, 10 patients (pts) were admitted for complete AVB, suddenly occurring 2 months to 8 years after cardiac surgery. Pts had undergone surgery at age 8 days to 12 years. One had a Senning operation and the others had ventricular septal defect closures. All pts had normal pre-operative ECG. After surgery, 9/10 pts had immediate complete AVB, lasting from 2 to 20 days; the last pt had episodes of 2/1 AVB during 7 days. After recovery of permanent 1/1 atrio-ventricular conduction on 24-hour monitoring, ECG still showed abnormalities: infracticular block (5), bifascicular block (1), isolated right bundle branch block (2), long PR interval with left axis deviation (1) and long PR interval with right bundle branch block (1). The diagnosis of complete AVB was made because of symptoms: syncope or cardiovascular collapse in 4 children and on 24-hour monitoring in the 6 others. The level of block was found below the His in all pts who underwent an electrophysiology. All 10 children have had a pacemaker implantation and are doing well. Conclusion: All 10 children with late sudden complete AVB had immediate complete or high degree AVB lasting more than 2 days after surgery. They also had ECG showing different pre and post-operative QRS and/or prolonged PR interval after surgery. These characteristics identify pts who should undergo a postoperative electrophysiology before being discharged, and a PM implantation if the level of block is found below the His.

32

Bidirectional ventricular tachycardia associated with syncope and familial sudden death: another manifestation of channelopathy?

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Bidirectional ventricular tachycardia (BVT) describes tachycardia with beat-to-beat alternation of QRS morphology. To better characterize BVT in pts without other evidence of heart disease, we studied 4 pts, 5–14 yr (QTc 409–452 ms) with ECG evidence of BVT. Symptomatic pts (n=2) had syncope with aborted resuscitation; asymptomatic pts (n=2) had a family (mother and/or sister, all with BVT) history of sudden death. Asymptomatic BVT (150–220 bpm; nonsustained – few beats to few sec) was provoked by exercise in 3 of 4 pts. EP study was performed in 2 of 4 pts; BVT was not inducible by V stimulation. The coding regions of KVLQT1, HERG, KCNE1, KCNE2 and SCN5A were evaluated using bidirectional sequencing. A HERG missense mutation (R1047L) was found in 2 of 4 pts, 2 of 6 asymptomatic family members, and 1 of 50 normal unrelated controls. BVT is controlled by § blocker in 3 of 4 pts and § blocker + propafenone in the other pt. A cardio-defibrillator was implanted in all but the asymptomatic 5 yr old. Conclusions: Nonsustained BVT at a relatively slow rate may appear benign, but it can be associated with syncope and family history of sudden death. BVT usually occurs during exercise and is probably of ectopic mechanism. Additional study is required to determine the significance of HERG mutation, but this finding raises the possibility that BVT is another manifestation of a channelopathy.

33

Catecholamine provoked T wave lability (TWLI): identification of a novel index for risk stratification in congenital long QT syndrome

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Background: Macroscopic T wave alternans is seen infrequently in congenital long QT syndrome (LQTS). Microvoltage T wave alternans (TWA) is a marker of arrhythmic risk in many conditions, but its significance in LQTS is unknown. Methods: 25 genotypically diverse LQTS patients and 16 control subjects were studied during pharmacologic and dobutamine provocation. Genotyping was established by PCR amplification and DNA sequencing of the four most common LQTS genes: KVLQT1 (LQT1), HERG (LQT2) and SCN5A (LQT3). Presence of TWA was determined by Fast Fourier transformation. Aperiodic, beat-to-beat T wave lability during catecholamine provocation was quantified using a newly derived T wave lability index (TWLI) based on a determination of the root-mean-square of the differences in T wave amplitude. Results: Precluded by ectopy, TWA could not be assessed in 9 of 25 patients with LQTS. In the remaining 16 patients, TWA occurred at lower heart rate in LQTS than in controls (117 ± 19 vs 153 ± 37 bpm, $p<0.05$). Dramatic catecholamine provoked T wave lability was observed in a 14-year-old male with F339del-KVLQT1 prior to onset of sustained polymorphic VT. The aperiodic T wave lability was significantly higher in LQTS (TWLI = 0.0945 ± 0.0537 vs 0.0445 ± 0.0127, $p<0.002$). Marked T wave lability (TWLI ≥ 0.075) was detected in all three LQTS genotypes (10/25), but in no control subjects. All high-risk patients having either a history of out-of-hospital cardiac arrest or syncope plus at least one sudden death in the family had TWLI ≥ 0.095 ($p<0.002$). Conclusion: Striking non-alternans T wave lability (TWLI) occurs in patients with LQT1, LQT2 and LQT3 during catecholamine provocation. This novel phenomenon of catecholamine provoked T wave lability may identify patients harboring high-risk genetic substrates.

34

QaT interval-heart rate relation during exercise in patients with KVLAQT1 and HERG mutations

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To elucidate clinical features of long-QT syndrome (LQTS), we analyzed the relationship between QT interval and heart rate during exercise. Patients with LQTS and normal subjects were examined. Subjects were divided into 3 groups: 1) control group with normal QT interval (mean age: 17yrs., n=16; NL), 2) patients with KVLQT1 mutations (mean age: 17yrs., n=9, LQT1), 3) patients with HERG mutations (mean age: 18 yrs., n=8, LQT2). The criterion for QT prolongation was the QTc value above 0.46 seconds at rest. The subjects exercised on treadmill testing using the Bruce protocol until all out. The ECG signals from Holter recordings during the exercise and post-exercise periods were summed every 15 seconds. The R-R intervals and QaT intervals (the interval between Q wave and the apex of T wave) were measured by computer and R-R-QaT plots were obtained. Linear line was drawn between the beginning point and the end point of exercise and slope of the line was determined. QaT dispersion (QaTD) of the patients with KVLAQT1 mutations was also measured during exercise and recovery periods, which was compared with that of normal subjects. The slope of LQT1,

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

0.125±0.045, was significantly smaller than that of NL (0.258±0.046 p<0.01). The slope of LQT2 (0.353±0.080) was significantly larger than LQT1 and NL (p<0.01). QaTD of normal subjects during recovery phase was small, whereas that of LQT1 became larger 3 minutes after the end of exercise. We concluded that LQT1 could be distinguished from LQT2 by QaTD/RR slope from the exercise test and that, in LQT1, large QaTD during recovery phase might contribute to the genesis of torsade de pointes.

35
Endocardial and epicardial steroid-lead pacing in the neonatal and pediatric age-group
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Objective: To compare the performance of steroid-eluting epicardial versus endocardial leads in the pediatric age group. Methods: Evaluation of pacing and sensing characteristics, impedance and longevity of 120 endocardial steroid leads (group A: 54 atrial, 66 ventricular) and 42 epicardial steroid leads (group B: 16 atrial, 26 ventricular) implanted between August 1990 and February 2000. Group A consisted of 73 patients with a mean age at implantation of 12.5 +/- 4.4 years, and group B consisted of 23 patients paced at 5.6 +/- 4.4 years. Priority: Follow-up period in group A: 2.2 +/- 1.8 years and group B: 2.4 +/- 1.9 years (NS). Stimulation threshold at implant for endocardial leads: ventricular 0.65 +/- 0.32 V, atrial 0.72 +/- 0.32 V at 0.5 ms pulse duration. Sensing thresholds ventricular 9.67 +/- 3.96 mV, atrial 3.29 +/- 1.42 mV; lead respectively. Lead impedance: ventricular 657.3 +/- 174.2 Ohms, atrial 508.2 +/- 162.9 Ohms. Epicardial leads at implant: ventricular pacing threshold 1.42 +/- 0.63 V (p < 0.001), atrial 0.95 +/- 0.27 V (p < 0.02). Sensing thresholds for ventricular and atrial leads 11.76 +/- 7.52 mV (p > 0.10) and 3.58 +/- 1.94 mV (p > 0.10) respectively. Lead impedance: ventricular 633.6 +/- 189.0 Ohms (p > 0.10), atrial 598.9 +/- 136.7 Ohms (p > 0.10). At 2 year follow-up, stimulation threshold for endocardial ventricular leads differed significantly from epicardial leads (0.86 +/- 0.43 V vs 1.44 +/- 0.81 V; p < 0.015). Sensing thresholds and lead impedance did not differ significantly. Complications requiring intervention occurred less in group A (n=7 vs n=38). Conclusions: Endocardial steroid leads have better pacing and sensing characteristics at implant and follow-up, with less incidence of complications.

36
Intracardiac echocardiography facilitates transseptal puncture in pediatric ablation
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Background: Transseptal puncture (TS) is commonly used for catheter ablation in pediatric patients but may have serious complications. Intracardiac echocardiography (ICE) has been reported to facilitate TS in adults. This study evaluates the safety and efficacy of ICE for TS in children. Methods: 7 TS sessions were performed in 6 patients (15.3±3.5 y.o., wt 62.7±9.1 kg) with ICE guidance for left-sided accessory pathway ablation. All the patients were referred for TS due to SVT recurrence post-ablation. One had complex heart disease. 8.5 Fr-12.5MHz (2) or 10Fr-12MHz (5) ICE probes were inserted through 8.5 Fr. soft tip femoral venous sheaths into the right atrium. TS was performed using a 6Fr. Mullins sheath and 18 gauge 60 cm needle. Left atrial position was confirmed with pressure monitoring and dye injection. Results: ICE identified the fossa ovalis (FO) in all patients. Tenting of the FO was noted with the transseptal needle prior to TS as seen in the figure (TP identifies the ICE probe). All 7 TS were successful in crossing the thinnest part of the atrial septum upon the first attempt without complication. One TS was repeated due to inadvertent catheter withdrawal. TS was abandoned due to difficulty engaging a very thick FO. Fluoroscopy time was 61.5±41.2 min. for 9.8±7.4 RF lesions (all acutely successful ablations, one late recurrence). One TS would have resulted in posterior RA perforation based on fluoroscopy alone. This was averted with ICE guidance for needle positioning. Conclusion: ICE is safe and effective in guiding TS in pediatric patients. ICE reduces the chances for perforation and facilitates optimal sheath position.

Session 5: Basic Research, Biology/Experimental Teratology, Cellular and Molecular Biology, Vascular Biology

37
Use of a novel anti-factor d monoclonal antibody to inhibit complement, neutrophil, platelet, and cytokine activation in a simulated pediatric cardiopulmonary bypass circuit
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Purpose: Cardiopulmonary bypass (CPB) induces a complex inflammatory response characterized by complement, neutrophil, and platelet activation. This response may be due to several factors, including exposure of blood to nonphysiologic surfaces, surgical trauma, and ischemia-reperfusion of the involved tissues. To investigate the effects of an anti-factor D monoclonal antibody (Mab 166–32) on inhibition of the alternative complement cascade, we undertook the following study using human blood in a simulated pediatric CPB circuit. Methods: We performed 5 paired experiments on Mab 166–32 and irrelevant control Mab at 16 g/dl of human blood. The extracorporeal circuit was primed with 250 ml of human blood and 400 ml of lactated Ringer's solution. During CPB, the hematocrit was maintained at 26–28%, and pump flow remained constant at 500 ml/min. After initiation of CPB, the blood temperature was reduced to 27°C, where it was maintained for 70 minutes, followed by 10 minutes of rewarming and 30 minutes of normothermic CPB. Extracorporeal circulation lasted for 120 minutes, and blood was sampled at 0, 5, 10, 25, 40, 55, 70, 80 and 120 minutes. Activation of complement, neutrophils, and platelets was assayed with immunofluorocytometry and ELISA. Results: After 120 minutes of CPB, the results (mean ± standard error) were as follows: Assay, Control Mab 166–32, P value: C3a (ng/ml) 331±50, 38±33.8, 0.0001; C3a (ng/ml) 237±22.8, 789±10.8, 0.0001; CD62P (platelets) %* 1041.6±191, 480.8±146.5, 0.0001; CD11b (neutrophils)%* 938±269, 395±88.9, 0.0001; IL-8 (pg/ml) 102.5±35.5, 28.7±12.6, 0.0001. *Percentage of baseline value at 0 min (arbitrarily set at 100%) during recirculation. Two-way ANOVA of factor d, randomized block design was used to analyze the data. Conclusion: Anti-factor D antibody significantly inhibits complement activation via the alternative pathway, reducing the inflammatory response associated with CPB.

38
International heart school: trying to help
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In May 1989 The World Forum for Pediatric Cardiology invited Prof. Lucio Parenzan to consider the creation of an International School of Pediatric Cardiology and Pediatric Cardiac Surgery in Bergamo (Italy). The proposal was based on the knowledge that the basic science education of many physicians caring for children with cardiovascular problems in disadvantaged areas of the world are quite good. However, practical experiences and opportunities for sharing pertinent scientific information with leading authorities with special expertise from the more developed areas of the world are frequently non existent or non-existent. A school could overcome these deficiencies by bringing together physicians with specialized expertise in an interactive environment with physicians and health professionals from disadvantaged areas. The teaching of the International Heart School J.W. Kirklin

39
Cell engineered cardiac graft to repair right ventricular outflow tract of the rat
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Objective: The synthetic materials, currently available for the repair of cardiac defects, are nonviable, do not grow as the child develops and do not synchronously with the heart. We developed a beating patch by seeding fetal cardiomyocytes on a biodegradable scaffold in vitro. In this study, we implanted the seeded patch into the right ventricular outflow tract (RVOT) of adult rats. Method: Cultured fetal or adult rat heart cells (1 × 10.6 cells) were seeded into a gelatin sponge and the cell number was expanded in

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

culture for 3 weeks. The free wall of the RVOT in syngeneic adult rats was resected and repaired with either unseeded patches or patches seeded with either fetal or adult cardiomyocytes (n=10 for each group). The patches were examined histologically over a period of 12 weeks. Results: A significant inflammatory reaction was noted in the patch at 4 weeks as the scaffold dissolved. At 12 weeks, the gelatin scaffold had completely dissolved. Seeded cells survived in the patch. Unseeded patches had an ingrowth of fibrous tissue. Gelatin was dissolved at 12th week after implantation. The control patch, but not the cell-seeded patches, was thinner than the normal RVOT. The endocardial surface area of each patch was covered with endothelial cells identified by factor VIII staining. Conclusion: Cell engineered patch was successfully used to replace the RVOT. The seeded cells survived in the RVOT after the scaffold dissolved and the patches remained completely endothelized.

40

Angiogenic factors in patients with congenital heart disease

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BACKGROUND: There is little data available concerning the change of angiogenic factors in patients with congenital heart disease (CHD). PURPOSE: To determine serum concentration of vascular endothelial growth factor (VEGF) and hepatocyte growth factor (HGF) in patients with CHD. METHODS: Fifty five patients with various kinds of CHD (31 boys and 32 girls, SaO2: 57–98%, age: 1 month to 13 years) were enrolled, and were divided into 2 groups according to the SaO2: A (SaO2 >= 92%) and C (SaO2 < 92%). During cardiac catheterization, blood samples were obtained at femoral vein (FV), superior vena cava (SVC), pulmonary artery (PA), pulmonary vein (PV), and femoral artery (FA). Serum HGF and VEGF were determined by ELISA. RESULTS: Among 5 sampling sites, serum HGF at FV was the lowest (0.45 ± 0.31 at FV, 2.20 ± 1.6 at SVC, 3.40 ± 3.15 at PA, 2.55 ± 2.58 at PV and 2.54 ± 2.62 at FA, respectively) and, at FA, serum HGF in C was significantly higher than that in A (3.65 ± 3.04 vs 1.86 ± 2.11, p=.02). On the other hand, among 5 sampling sites, serum VEGF at FV was the highest (382 ± 362 at FV, 225 ± 196 at SVC, 154 ± 145 at PA, 178 ± 209 at PV, and 178 ± 148 at FA, respectively) and, at FV, serum VEGF in C was significantly higher than that in A (534 ± 295 vs 293 ± 196, p= 0004). In A, pulmonary vascular resistance significantly positively correlated with serum HGF at 4 different sites other than FV. CONCLUSIONS: In CHD, serum VEGF and HGF may be produced in different vascular system in response to various stimuli including hypoxemia and vascular resistance.

41

Tumor necrosis factor-alpha and post-ischaemic contractility in an infant model of left ventricular hypertrophy

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OBJECTIVE: Left ventricular hypertrophy is associated with contractile dysfunction, impaired tolerance to ischemia and increased risk during cardiac surgery. Recently, tumor necrosis factor-α (TNF-α) has been implicated in the pathogenesis of both heart failure and ischemia-reperfusion injury. We hypothesized that compensated pressure overload hypertrophy results in increased myocardial TNF-α expression and that it contributes to the increased susceptibility to ischemia-reperfusion seen in hypertrophied infant hearts. METHODS AND RESULTS: Neonatal rabbits underwent banding of the descending thoracic aorta to induce left ventricular (LV) hypertrophy. Myocardial TNF-α protein expression increased progressively with LV hypertrophy. Serum TNF-α was detected only after the onset of heart failure. Prior to the onset of ventricular dilatation and heart failure (determined by serial echocardiograms), hearts from aortic banded and age-matched control rabbits were perfused in the Langendorff mode, and subjected to 45min ischemia and 30min reperfusion. Post-ischemic recovery of developed LV pressure was impaired in hypertrophied hearts as compared with control hearts (59+/-9% vs. 88+/-5%). The addition of neutralizing anti-rabbit TNF-α antibody to cardioplegia and perfusion solutions restored post-ischemic function (82+/-7% recovery). This effect was mimicked by treatment with N-oleoyl ethanolamine, an inhibitor of ceramidase, the key enzyme mediating TNF-α effects on calcium handling. Intracellular calcium was measured by Rhod-2 spectrofluorometry and demonstrated lower diastolic calcium levels and higher systolic calcium transients in anti-TNF-α treated hearts. TNF-α inhibition was also associated with faster post-ischemic recovery of phosphocreatine, ATP, and pH as assessed by 31P NMR spectroscopy. CONCLUSIONS: TNF-α is expressed in infant myocardium during compensated pressure-overload hypertrophy and contributes to the post-ischemic myocardial dysfunction. Inhibition of TNF-α signaling significantly improves post-ischemic contractility, myocardial energetics, and intracellular calcium handling.

42

During pediatric cardiovascular surgery surgical trauma but not the cardiopulmonary bypass (cpb) is responsible for neutrophil activation

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Aim: The activation of neutrophil by the CPB is held responsible for post-operative complications as pericardiotomy or capillary leak syndrome. However, present data do not show evidence of surface expression and serum concentration changes of adhesion molecules are CPB or surgery specific. Furthermore, studies were set in a short time range (from surgery up to 1–2 days after surgery), thus do not include baseline values of the unperturbed immune status of the patients. Methods: 47 surgeries with CPB (patient age: 1–17yrs; CPB group) and 27 surgeries without CPB (age: 1–15yrs; control) were studied. Blood was sampled 24h preoperatively before any in-hospital medication, after anesthesia onset, after connection to CPB at reperfusion, 4h, 1d, 2d after surgery, at discharge and 6 months postoperatively at the ambulance control. Neutrophil antigen expression and serum concentration of soluble adhesion molecules were analysed by flow cytometry and ELISA, respectively. Results: Anesthesia and surgery induced significant and transient decrease of LFA-1 (CD11a/CD18), Mac-1 (CD11b/CD18) and CD 54 surface expression as well as of ICAM-1 and L-selectin serum level below baseline (all p<0.001). Except L-selectin the decrease of all measured parameters was independent of the use of CPB. In the CPB group L-selectin decrease was more pronounced (p=0.006). With CPB CD62L (L-selectin) surface expression increased over baseline and control (p=0.002). Antigen expression did not depend on CPB duration. Preoperative baseline values were reached >2 days to 6 months postoperatively. Conclusions: Cardiovascular surgery leads to suppression of neutrophil adhesiveness that is further reduced by CPB (see CD62L). Activation of neutrophils caused selectively by CPB can not be exclusively responsible for post-operative complications specific for CPB supported cardiovascular surgery. Baseline values are reached days or months postoperatively emphasizing the need to extend the time frame analysed.

43

Corticosteroid treatment does not stimulate interleukin-10 release but reduces neutrophil and monocyte adhesiveness during cardiac surgery

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Aim: Cardiac surgery with cardiopulmonary bypass induces substantial release of the immunosuppressive cytokine IL-10. IL-10 was been found to be a key cytokine in propagating immune paralysis and leading to sepsis, shock and multi-organ dysfunction. A recent study (Surgery 1996; 119: 76) indicates that corticosteroid administration elevates IL-10 release more than 10 fold. Thus corticosteroid could enhance trauma induced immune paralysis and the risk for shock, MODS and infection. Method: A prospective study was performed in children (age: 3 to 16yrs.) who underwent surgery for atrial septum defect (ASDI, n=22, ASDII, n=3). 16 of them received intra-operatively Methylprednisolone (30 mg/kg). Surgical data, medication, intra- and post-operative care were not different between the groups. Blood samples were drawn pre-, peri- and post-operatively. Serum concentration of cytokines and soluble adhesion molecules was determined by ELISA. Expression of leukocyte adhesion molecules (LFA-1, Mac-1) was quantified by flow-cytometry. Results: Patients who underwent surgery showed significant sequestration of IL-10 with maximum values at the end of surgery (peak: 210±540 pg/ml) but no significant difference was found with and without Methylprednisolone. By Methylprednisolone IL-6 release was reduced by >50%, neopterin, TNFα, IL-1β, and histamine release was reduced by >10% (all p<0.05). Post surgical neutrophil and monocyte count, LFA1 and MAC-1 expression and serum levels of ICAM-1 and L-selectin were significantly down regulated (p<0.03). Conclusion: Perioperative administration of methylprednisolone does not elevate IL-10 release but reduces infiltration and adhesiveness of circulating leukocytes. Therefore, intraoperative treatment with corticosteroids seems not to increase the risk for immune paralysis. Migration of monocytes and neutrophils to the site of inflammation in part accounts for post perfusion injury. The beneficial effect of corticosteroids might be the reduction of the migration ability.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

44

31P NMR spectroscopy reveals metabolic abnormalities in asymptomatic patients with hypertrophic cardiomyopathy

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Objectives: Alterations in myocardial metabolism are considered to be causes for contractile dysfunction in patients with hypertrophic cardiomyopathy (HCM). We examined the question of whether metabolic abnormalities precede the manifestation of symptoms in patients with HCM. Methods: Proton-decoupled 31P NMR spectroscopy of the anterosepal region of the heart of 14 young asymptomatic patients with HCM was performed with a 1.5 T whole-body imager. Spectra of the phosphate metabolites were compared with those of normal volunteers. Results: The patients exhibited a significantly reduced ($p < 0.02$) ratio of phosphocreatine (PCr) to ATP of 1.98 +- 0.37, compared with 2.46 +- 0.53 obtained in 11 control subjects. In addition, the group of patients with severe hypertrophy of the interventricular septum ($n = 8$) showed significantly increased ($p < 0.05$) inorganic phosphate (Pi) to PCr ratio with a Pi × HR/PCr of 20.0 +-8.0 versus 9.7 +- 7.2 in control subjects. Both abnormalities are similar to those found in ischemic myocardium. The assumption of deficient oxygen supply is also supported by a significantly increased ($p < 0.01$) phosphomonoester (PME) to PCr ratio with a PME × 100/PCr of 20.7 +- 11.2 compared to 8.4 +- 6.7 in control subjects indicating altered glucose metabolism. Conclusion: 31P NMR spectroscopy detects alterations of myocardial energy metabolism in asymptomatic patients with HCM. These alterations may contribute to the understanding of the pathophysiology and natural history of the disease.

45

Nitric oxide: a vasodilator, and inhibitor of matrix remodelling by suppressing AML1B-elastase cascade

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Nitric oxide (NO), an endogenous vasodilator, inhibits pulmonary vascular remodeling in rats as do inhibitors of vascular elastase. A 20 kD smooth muscle cell (SMC) serine elastase, which is induced by serum-treated elastin (STE), appears critical to the progression of pulmonary vascular disease. In our previous study using differential display to identify transcripts expressed coincident with elastase activation, we identified AML1B, a transcription factor for neutrophil elastase, in SMC. However the direct interaction of NO with AML1B-elastase cascade remains unknown. To uncover the signaling pathway for elastase activation and interaction with nitric oxide (NO), we found STE-induced increase in phosphorylated extracellular signal regulated kinase (ERK). Inhibitions of ERK activation with PD98059 inhibited AML1-DNA binding and elastase. NO donors (SNAP and DETA NONOate) inhibited elastase as did a cGMP mimetic (8-pCPT-cGMP). SNAP inhibition of elastase was reversed by co administration of a PKG inhibitor (Rp-8-pCPT-cGMP). The increase in phospho-ERK was suppressed by NO donors and the cGMP mimetic, and reversed by co administration of the PKG inhibitor, as was nuclear repression and DNA binding of AML1B. Taken together, the present study uniquely links NO/cGMP-generating vasodilators with inhibition of elastase dependent matrix remodeling in vascular disease by influencing AML1B-mediated gene expression.

46

Nitric oxide contributes to the progression of coronary artery lesions in acute Kawasaki disease

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Although nitric oxide (NO) serves many vasoprotective roles including inhibition of platelet aggregation and leukocytes adherence, and stimulation of endothelial cell growth following arterial injury, massive release of NO, particularly synthesized by nitric oxide synthase (iNOS), causes arterial wall degeneration. Conversely, We hypothesized leukocytes as well as endothelial cells actively generate NO, following synthesis of iNOS, and may serve to damage the coronary arterial wall and lead to aneurysmal formation in acute Kawasaki disease (KD). We evaluated the expression of iNOS in leukocytes by flow cytometry, and in coronary aneurysm by immunohistochemical analysis, and we analyzed serum levels of nitrate/nitrite in acute KD. We also studied the number of circulating endothelial cells using a specific anti-endothelial-cell antibody, p1H12, and evaluated the expression of the iNOS in these cells. We studied 55 patients, aged 4 months to 7 years 3 months: those with (n=24) or those without CAL (n=31) in acute KD. The serum nitrate/nitrite levels and iNOS expression in neutrophils were maximal at pretreatment, particularly marked in patients with CAL ($p<0.001$ and $P=0.001$, respectively), and declined rapidly until 2 weeks post onset. While iNOS expression in monocytes and the number of iNOS positive circulating endothelial cells were maximal at 2weeks post onset, when CAL generally develops, especially higher in patients with CAL ($P=0.035$ and $P=0.012$, respectively). The immunohistochemical study showed iNOS immunoreactivity in endothelial cells and monocytes/macrophages in coronary aneurysm in acute KD. The findings in our study suggest that a neutrophil-derived excessive amount of NO, synthesized by iNOS, may play a role in early arterial wall injury, whereas monocytes/macrophages and endothelial cells-derived NO may contribute to the progression of CAL and later vascular remodeling in acute KD, having a possible signaling relation with VEGF.

47

Angiotensin II-induced vascular superoxide production in genetic hypertension

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Studies support a critical regulatory role for angiotensin II (AII) in the development of high blood pressure in genetic hypertension. Furthermore, AII induces increases blood pressure in normotensive animals through activation of specific signaling pathways leading to excess superoxide anion (O_2^-) production in the vasculature. Therefore, we hypothesized that AII-induced vascular O_2^- is increased in genetic hypertension. In addition, we examined whether increased vascular O_2^- production precedes the development of high blood pressure. Systolic blood pressures (SBPs) were recorded by non-invasive tail-cuff in 6- and 12-week-old male spontaneously hypertensive rats (SHR), and normotensive Wistar Kyoto (WKY) rats. Rat thoracic aortas were harvested and incubated in serum-free media containing 0–10 micromolar AII for 12 or 24 hours at 37 C. Following incubation, vascular O_2^- production was measured by lucigenin-derived chemiluminescence and recorded as photon counts/mg tissue weight. Mean SBP was significantly greater in 12-week-old SHR (159±3mmHg) than age-matched WKY rats (135±mmHg, $p=0.001$). In addition, AII incubation significantly increased O_2^- production in 12-week-old SHR at both 12 hours (2.8-fold increase) and 24 hours (1.9-fold increase). In contrast, 12-week-old WKY rats showed no AII-mediated increase in vascular O_2^- production. In 6-week-old rats, mean SBP was not different between SHR (100±12mmHg) and WKY rats (114±3mmHg). However, AII-induced O_2^- production remained significantly increased in prehypertensive SHR (1.8-fold increase), but not WKY. Endothelial denudation, or co-incubation of aortic rings with Cu/Zn superoxide dismutase or diphenyleneiodonium chloride (an NAD(P)H oxidase inhibitor) attenuated the AII-induced increase in O_2^- production in SHR. In conclusion, AII selectively increases vascular O_2^- production in genetically hypertensive rats that precedes the development of high blood pressure. Furthermore, the AII-mediated increase in vascular O_2^- in genetic hypertension is dependent upon an intact endothelium with a specific contribution from NAD(P)H oxidase.

48

A new gene therapy approach in heart failure

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Heart failure is associated with reduced levels of myocardial _ myosin heavy chain (_-MHC) due to increased binding of nuclear factors (PNRB) to a repressor element (PNR) in its gene promoter. We hypothesize that quenching PNRB by PNR decoy will restore myocardium _-MHC levels, resulting in improved contractility. Effect of PNR decoy on _-MHC gene transcription was tested in cardiac cultures of 10- day chick embryos. Cells were transfected with _- MHC promoter/ CAT reporter constructs with PNR (1 Kb/CAT) or without PNR element (670/CAT) by calcium phosphate method. After 24 hrs. PNR decoy oligo or PNR scrambled oligo (PSO) were introduced with lipofectene. Control cells received lipofectene. After additional 48 hrs, cells harvested and CAT activity analyzed by thin layer chromatography. PNR decoy resulted in 3–5 fold increased CAT activity of 1Kb/CAT but not 670/CAT construct. PSO had no effect on CAT activity of either construct. Results show that PNR decoy increases _MHC promoter activity, reducing negative effect of PNR on the transcription process. It offers new avenue for modulating contractility of the failing myocardium.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

MAY 28 Time: 14:00–15:30

Session 6: Coronary Artery Disease/Kawasaki Disease

49

Kawasaki disease in Singapore: incidence and coronary complications

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Data are captured in our Kawasaki Disease (KD) database when patients are referred for echocardiography for or suspected of KD. Echocardiograms to delineate coronary abnormalities are done at 2 weeks, 3 months, 6 months and 6 monthly thereafter from the onset of KD. During the 3 year period under review (June 1997 to June 2000), a total of 249 patients were referred for echocardiography. Twenty-eight patients with normal echocardiograms were excluded as they did not meet the full diagnostic criteria. There were 221 cases of KD during this period, of which 208 were less than 5 years of age, an incidence of at least 32.5 per 100,000 children < 5 years old per year in Singapore. The mean age at diagnosis was 22.4 months. Incidence decreases with age – 93.2% were less than 5 years of age. Infants accounted for 41.6% of cases and 14 (6.3%) were younger than 3 months of age. There was a slight male predominance (p=0.6%). In our patients with KD, 23.1% had coronary dilatation: – 10.9% had coronary aneurysms, while the other 12.7% had generalised ectasia of the coronary arteries. On follow-up echocardiograms, 67.3% had total resolution of the coronary abnormalities – 83.3% of those with ectasia vs. 36.4% of those with aneurysms (p<0.001). Majority (79.3%) of patients with coronary ectasia recovered within 6 months of the illness. No patient with normal echocardiogram at 2 weeks of illness had coronary dilatation at subsequent echocardiography. Seventeen patients (7.7%) had atypical Kawasaki Disease. They tended to be younger (mean 16.2 months, median 6.2 months) compared to the others (mean 23 months, median 15.3 months) (p=0.215). Female sex is associated with higher risk of having atypical KD (OR 3.1, 95%CI 1.1 – 8.7).

50

Coronary artery fistulas: management and follow-up<P>

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Background. Coronary artery fistulas (CAF's) are generally congenital of origin although usually discovered late. This study describes 53 CAF patients. Methods. Retrospective study of 53 patients included: age, gender, mean age at diagnosis, follow-up period of patient, type and characteristics of CAF's, previous history, physical examination, ECG's, exercise tolerance (esist: ET), scintigraphy, echocardiography, management and results of intervention and follow-up. Results: 53 patients had 72 CAF's and a current mean age of 60,21 years(14–89yrs), mean age at diagnosis of 56,01 and a follow-up period of 4,19 years. 58% of the CAF's is of single origin and 59% of single termination, 40% have a corrupted and multiple pathway. 55/72 CAF's originate from the LAD, 24/55 are proximally located and terminate into the PA trunk, the distally located end into the LV. 22/72 CAF's originate from the right coronary artery, 8/72 from the circumflex. 83% were symptomatic at diagnosis. Previous history included myocardial infarctation(MI) in 12. ECG showed repolarisation disturbances in 50%. Fistula related abnormalities in untreated patients were found in ET(80% ischemic changes), in stress MIBI Scintigraphy(11/21 diminished ejection fraction and localized perfusion defects) and on angiography (coronary vessel disease in 47% of which 63% atherosclerotic). 12 patients developped a MI, 8 corresponding to the fistula related artery(FRA). 44 patients (83%) were treated conservatively. 10 patients underwent surgical ligation and 2 transkatheter embolisation (PTE). Conclusion. The origin of LAD-CAF was found to be the most common, if located proximally draining in the main pulmonary artery, if located distally into the right ventricular cavity. Based on the development of associated ischaemic coronary pathology in the FRA of untreated patients we suggest that surgical or interventional therapy is warranted in these patients.

51

Fate of the aortic valve regurgitation after resection of subvalvular aortic stenosis

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Objective: Aortic regurgitation (AR) is a common association with subvalvular aortic stenosis (SAS). The aim was to evaluate the fate of AR after SAS surgical relief. Method: Between 1985 and 1999, 153 SAS patients (pts) underwent fibromuscular repair with resection of SAS. The male/female ratio was 101/52 with a mean age of 13.4 years. Isolated SAS was found in 124 pts (81%) and associated cardiac defects (VSD, mitral valve involvement and double outlet right ventricle) in 29 pts. Gradient across SAS, mean 77 mmHg. Associated AR was found in 109 pts (78%): 35 trivia – 64 mild, 24 moderate and 6 severe. Preoperatively 6 pts had endocarditis, 112 had membranous type stenosis, 24 fibromuscular and 7 tunnel type. Surgery entailed in 140 pts, additional myomectomy in 68. Additional aortic valve surgery was required for 47 of the 119 pts with AR: valve repair 30, replacement 13 and aortic root augmentation 4. Findings: There was one early death and four pts had complete heart block. There was some degree of AR postoperatively in 47 pts. Mean follow up was 4 yrs (range 2 months – 12 years). Relief of SAS improved the degree of AR in 50% of postoperative AR pts. Gradients > 30 mmHg developed in 13 pts, 6 had redo surgery, 4 for relief of Left Ventricular Outflow Tract Obstruction, one for severe AR and one for MR. Conclusions: Resection of SAS has low recurrence. Additional AV surgery was required in 40%. Resection of SAS can improve the degree of AR in over 50% of pts.

52

Epidemiological study of Kawasaki disease in Korea, 1997–1999: comparison with previous studies in 1991–1996

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We retrospectively performed an epidemiological survey on Kawasaki disease(KD) from 1997–1999 in Korea. On behalf of the Korean Pediatric Cardiology Society, we sent a questionnaire to 111 training hospitals, and summarized the data of the survey from 50 hospitals which responded. The total number of patients was 5,862 cases including 1143 in 1997, 1419 in 1998, and 1300 in 1999, which showed no difference in annual incidence and high incidence in May, June, July and August. The male-to-female ratio was 1.31, and their mean age was 29.7 months. The proportions of sibling cases was 0.20%(10/5862), and rate of recurrent cases was 2.3%(90/3862) and the proportion of patients with KD among total hospitalized pediatric patients was 1.15% in average, showing no significant difference according to regions. Echocardiogram was done in 97.4% of KD patients, and coronary arterial(CA) abnormalities occurred in 19.8% of cases(737/3722) including 16.1% of dilatations and 5.1% of aneurysms. Adding 1,709 cases of 1-st survey in 1991–1993 and 2,680 cases of 2-nd study in 1994–1996, in mere cases of 3-rd study, data of the total 8,251 cases in 1990s in Korea showed 28.9 months of mean age, 1.6 of male-to-female ratio, seasonal predilection for summer, 0.24% of sibling cases, 2.3% of recurrent cases, 21.0% of CA abnormalities and 5.2% of CA aneurysms, with statistically decreasing trends of male-to-female ratio and CA abnormalities.

53

Vascular wall morphology and vascular elasticity of coronary aneurysms in long-term after Kawasaki disease: intravascular ultrasound study

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Coronary arterial (CA) lesion in Kawasaki disease (KD) may become a long term coronary risk factor. Seventy KD patients and 10 controls were studied. Consecutive KD patients were followed over more than 10 years from KD onset. Patients comprised 4 groups. Group 1: 18 patients with persistent aneurysms. Group 2: 18 patients with CA stenosis. Group 3: 26 KD patients with regressed CA aneurysms. Group 4: 6 patients with normal coronary findings at the acute stage. CA wall morphology evaluated by IVUS imaging. The % area change of CA lumen was calculated using IVUS imaging to examine the elasticity of the CA wall. IVUS imaging in Groups 1 and 2 showed intima hyperplasia and various degree of calcification at sites of both

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

persistent coronary aneurysms (intima-media complex: 0.71±0.22mm, % calcification area: 55.4±21%) and stenosis (intima-media complex: 0.88±0.44mm, % calcification area: 81.4±20%). However, IVUS imaging in Group 3 showed various degrees of the luminal thickening without calcification (intima-media complex: 0.48±0.12mm, % calcification area: 0%). All IVUS findings in the Group 4 CA wall echo had a single layered appearance were similar to that in the control patients. In Group 1 and 2, coronary artery demonstrated poor elasticity, almost no change in the lumen area (% area change: Group 1: 2.4±1.9%, Group 2: 0.8±1.3%). In Group 3, a significant poorer elasticity was found compare to the control patients (Group 3: 8.1±3.7% vs control: 22.0±13.2%, p<0.05). Group 4 showed no significant difference of elasticity of coronary artery from control (Group 4: 21.2±11.3%). We concluded that long term persistent coronary aneurysm and regressed coronary aneurysms after KD have abnormal vascular wall morphology and poor vascular elasticity. Such follow up should focus on the possible development of premature atherosclerosis and include advice of avoidance atherogenic risk factor.

54

Potentially reversible brachial vasodilatation and impaired flow-mediated reactivity in acute Kawasaki disease

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To elucidate the impact of systemic vasculitis of acute Kawasaki disease (KD) on peripheral vascular dimension and reactivity, vasodilative response to hyperemia by high-resolution ultrasound images of brachial artery was evaluated in 25 children with acute KD (1.5±1.0 months after onset), 10 in recovery phase (10±2.6 mo) and 35 age- and sex- matched controls (C). Brachial basal diameter was significantly enlarged both in acute KD (2.8±.35mm, p<.0001) and in recovery (2.7±.23mm, p<.05) compared with C (2.4± .37mm). Percent of normal value, however, significantly improved in recovery (111±17% vs 124±13% p<.05). Flow-mediated dilatation (% of diameter change) was significantly lower in acute KD (0.8±4.5% vs 16±5.6% in C, p<.0001) and also improved in recovery (12±7.9%, p<.0001). We concluded that systemic vasculitis of acute KD induces pathological peripheral vasodilatation and impairs vascular reactivity which potentially improves in recovery phase.

55

Long-term Outcome of Catheter Intervention in Kawasaki Disease

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Background: Catheter intervention has become one of the promising therapeutic strategies in the treatment of patients with coronary stenosis due to Kawasaki disease. However, long-term follow up data has not been clarified. Patients and Method: We reviewed our single institutional experiences of this procedure. Since 1994 through 2000, 32 patients underwent catheter intervention. The procedures included percutaneous transluminal coronary angioplasty (PTCA, n=18), percutaneous transluminal coronary rotational ablation (PTCRA, n=12), and stent implantation (n=10). The immediate success rate was 60% in the PTCA, 100% in the PTCRA, and 90% in the stent. Age at intervention ranged from 3.9 to 22 years (median 14.5 years) and interval from the onset of disease to intervention was 1.7 to 17 years (median 8.7 years). In all of these patients, follow-up coronary angiography was performed 3 months to 4 years after the procedure. Final follow-up period ranged from 4 months to 6 years (median 3.6 years). Intravascular ultrasound study was also performed if imaging catheter could be approached to the aiming lesion. Two patients complicated with restenosis/aneurysm formation at the site of PTCA. Results: During this follow-up period, neither ischemic symptoms nor myocardial ischemia detecting by myocardial perfusion imaging were appeared in these patients. One patient, whose coronary stenosis could not be dilated by PTCA, have had coronary bypass surgery. Significant restenosis was not found by follow-up coronary angiography. Intravascular ultrasound imaging revealed that good patency of coronary lumen at the site of intervention and thin circumferential intimal layer around the stent. Although progression of neoaneurysm was not observed, resolution of aneurysms was not also confirmed. Conclusion: Long-term result of catheter intervention in this disease is excellent and can postpone or substitute coronary bypass surgery.

56

Myocardial blood flow and coronary flow reserve in patients with "normal" subepicardial coronary arteries after Kawasaki disease assessed by positron emission tomography

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Kawasaki syndrome is an acute childhood vasculitis, affecting the arterial walls in form of a panvasculitis. Aim of the study was to assess myocardial blood flow (MBF) and coronary flow reserve (CFR) in children (12.3 years SD 4.02) with a history of Kawasaki syndrome and angiographically normal subepicardial coronary arteries. Methods: Seven children with Kawasaki syndrome and ECG (resting tracing), echocardiography and positron emission tomography 10 years SD 5.6 after the acute illness, they all were clinical asymptomatic without signs of coronary insufficiency, angiographically the coronary arteries were normal without aneurysm or stenosis. Myocardial perfusion was assessed by N13-PET imaging at rest and after maximal vasodilatation with adenosine; the results were compared with 10 healthy adults (26 years SD 6.3). Results: No patient had signs of myocardial ischaemia; ECG (resp./exercise) was normal without signs of coronary insufficiency or rhythm disturbances, on echocardiography ventricular function was normal without evidence of ischemic areas or signs of enlargement or stenosis of the proximal coronary arteries. There was no statistical significant difference between patients and healthy volunteers in MBF at rest (0.84 SD 0.22 vs. 0.77 SD 0.17 ml/g/min.), whereas MBF after maximal vasodilatation with adenosine (2.56 SD 0.56 vs. 3.18 SD 0.9 ml/g/min p<0.001) and CFR (2.99 SD 0.36 vs. 4.09 SD 1.01 p<0.001) was significantly attenuated in the Kawasaki group. Nine of the patients had small reduced perfusion defects within the myocardium. Conclusion: In children with a history of Kawasaki disease and angiographically normal subepicardial coronary arteries there is a significant accumulation of MBF after maximal vasodilatation and a significant reduction of CFR. Impairment of the vasoreactive ability may indicate a residual damage of the coronary arteries and may be a risk factor for atherosclerosis in adulthood.

57

Histopathological study of neutrophils in coronary arterial lesions in Kawasaki disease

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Conventional therapy for Kawasaki Disease (KD) includes intravenous immunoglobulin (IVIG) and aspirin. This therapy is obviously effective but cannot completely prevent the formation of coronary artery aneurysm. Recently, an elastase inhibitor began to be used for the treatment of KD on purpose to inhibit polymorphonuclear leukocytes (PMNL)-elastase activity. Activated PMNL may damage endothelial cells, resulting in vascular lesions. However, there has been no morphological evidences that PMNL invade vascular walls of coronary arteries in acute phase of KD. Therefore, histopathological investigation was carried out in an attempt to elucidate whether PMNL were present in the coronary arterial lesions, using autopsy cases of KD. The experimental material consisted of eleven autopsy patients who died during acute phase of KD. Duration of the illness ranged from 6 days to 50 days. The tissues were fixed and embedded in paraffin. Hematoxylin and eosin, Elastica van Gieson and azan-Mallory stain were performed for routine histological examination. In addition to densely infiltrating cells in arterial lesion, the antibody CD3, CD20, CD68 and elastase were used for immunohistochemistry. The inflammatory cells appeared in the coronary arterial lesions were mainly composed by macrophages in all patients, however, numerous neutrophils were also identified in non-aneurysmal coronary arterial lesion of the patient who died 10 days after the onset of KD. Neutrophils infiltration marked the peak earlier than those of CD68+ macrophages, CD3+ T lymphocytes and CD20+ B-lymphocytes. It is suggested that neutrophils are strongly involved in the damage of coronary artery at early stage of the illness. As a result of injury of vascular wall caused by neutrophils, vascular dilatation may occur.

58

Echocardiographic videodensity index of the coronary arteries in acute Kawasaki disease: a predictor for subsequent coronary artery abnormalities

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Coronary artery abnormalities developed in 25% of patients with Kawasaki disease. A number of studies tried to analyze indirect markers of systemic inflammatory process for prediction of coronary artery involvement.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

However, inflammation of the walls of these arteries has been demonstrated earlier during the acute phase. Direct analysis on these sites may reflect the ongoing pathological findings. The purpose of our study was to quantitatively analyze the videodensity of the coronary arterial walls in patients with Kawasaki disease indexed to that of myocardium. A total of 72 patients, ranging in age from 2 months to 9 years, were studied. Twenty-eight patients developed coronary artery lesions. An IBP Sonos 4500 echocardiographic system was used to acquire the images and record on videotapes. Optimal still images of the proximal major branches of the coronary arteries were digitally captured and analyzed for videodensity using NIH Image. Three consecutive measurements of each segment of the arterial walls were averaged for analysis. There was significant difference of the videodensity index between those of patients with and without coronary artery abnormalities ($p<0.001$). Videodensity index of greater than 1.32 could predict the lesions with sensitivity of 87% and specificity of 82%. The mean interobserver variability of the measurement was 5%. Echocardiographic videodensity index of the coronary arterial wall may predict for the subsequent occurrence of coronary artery abnormalities in Kawasaki disease.

59

Cardiovascular risk factor reduction in adults with acyanotic congenital heart disease

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Adults with coarctation of aorta, coarctation of aorta with bicuspid valve, or supravalvular aortic stenosis form a subgroup of acyanotic congenital heart disease that is predisposed to premature myocardial infarction. We studied the prevalence of cardiovascular risk factors in this population. The higher cardiovascular risk group (Group A) comprised 52 patients: 17 patients with coarctation of aorta, 28 patients with coarctation of aorta and bicuspid aortic valve, and 3 patients with supravalvular aortic stenosis. There were 25 men (age range 21 to 56 years, mean 34 years) and 17 women (age range 21 to 45 years, mean 32 years). Fifty-six patients with bicuspid valve served as controls (Group B). Cardiovascular risk factors in these patients, i.e. age, sex, hypertension, hypercholesterolemia, diabetes mellitus, obesity, smoking, gout and family history were recorded. An assessment was made of the primary and secondary prevention of the identified cardiovascular risk factors. Group A: 33% were hypertensive ($n=17$), 15% were obese ($n=8$), 28% had hypercholesterolaemia ($n=11$), and 21% were smokers ($n=11$). Group B: 21% were hypertensive ($n=12$), 36% were obese ($n=20$), 4 % had hypercholesterolemia ($n=2$), and 13% were smoking ($n=7$). Physician-directed sedentary lifestyles were identified in the majority. Appropriate advice regarding diet, exercise, smoking cessation, antihypertensive and cholesterol-lowering pharmacological therapy were made and followed-up during subsequent visits. Premature myocardial infarction has been attributed to elevated systolic pressure in the aortic root in patients with coarctation of aorta or supravalvular aortic stenosis in addition to atrial fibrosis in the later. Optimal cardiovascular risk reduction is important in this relatively young population in order to minimize these additional risks.

60

Homocysteine, lipid profile, nitric oxide, vit. B12, and folate values in patients with premature coronary artery disease and their children

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The plasma level of homocysteine (Hcys) and Lipoprotein a (Lipo-a) are independent risk factors for arteriosclerotic vascular disease. Nitric Oxide (NO) and folat values are also important in atherogenesis. We aimed to evaluate these parameters in patients having coronary artery bypass surgery (CABS) prior to 50 years of age and their children. In 53 patients having CABS, 47 children of these patients and 28 normal control subjects, homocysteine, NO, Vit B12, folat, Lipo-a, triglyceride (TG), cholesterole (Chl), LDL cholesterole (LDL), HDL cholesterole (HDL), apolipoprotein-A1 (Apo-A1) and apolipoprotein-B (Apo-B) values were determined. Table: Means of the parameters and p values. Table was send as attachment. Homocysteine values of the patients with premature coronary heart disease and their children are significantly higher than those of controls ($p<0.001$ and $p<0.006$, respectively). Also, NO levels are significantly higher in both groups than controls ($p<0.001$ and $p<0.001$, respectively). B12 values are significantly higher in both groups ($p<0.05$ and $p<0.013$, respectively). Lipo-a levels are higher in both groups but not significant.

Session 7: Cardiomyopathies/Myocarditis/ Heart Failure

61

Preliminary study of evaluation of ventricular function after using carvedilol in children with severe dilated cardiomyopathy

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Background: Little is known about carvedilol in children with dilated cardiomyopathy. Methods: We enrolled 17 consecutive children waiting heart transplantation (age: 2.2 months to 10 years) with chronic heart failure, left ventricular ejection fraction < 35%, in a 6 months follow up double-blind, placebo-controlled study (mean time of evaluation was 304+/-241 days). Patients were randomly assigned to receive either placebo (7 patients) or carvedilol (10 patients) at initial dose of 0.01 mg/kg/day and increased over a period of 60 +/- 30 days to a dose of 0.2 mg/kg/day after which double-blind therapy was maintained for at least an additional 6 months. Results: During the 6 months follow-up and the up-titration of the dosage period in the carvedilol group: 3 pts died (2 in titration period and 1 pt underwent heart transplantation (titration period). In 6 patients evaluated after 6 months with a mean dose of carvedilol of 0.2 mg/kg/day, there was an increase in left ventricular ejection fraction from 15.5 +/- 5.9% to 36.5+/- 12.0% ($p=0.009$), shortening fraction from 15.8+/-3.2% to 22.1+/- 7.6% ($p=0.031$) and the NYHA class improved in 5 patients. The clinical status of 5 patients who were listing was NYHA I in 4 pts and NYHA II in 1 pt and all 5 pts are alive at a mean follow-up period of 591+/- 29 days. In the placebo group, during the 6 months follow-up 1 pt died and 2 pts underwent heart transplantation. In 4 patients evaluated after six months, all were in NYHA IV, there was no change in left ventricular ejection fraction (21.2+/- 4.9% vs 19.5+/- 5.0%, $p=0.68$), or fractional shortening (13.5+/-5.8% vs 14.0+/-3.4%, $p=0.585$). Conclusion: Carvedilol could improve ventricular function in children with severe cardiomyopathy.

62

Left ventricular restrictive dysfunction in children with dilated cardiomyopathy and its clinical implications

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Objectives: This study sought to evaluate the left ventricular diastolic function in children with idiopathic dilated cardiomyopathy (DCM) and its clinical implications. Method: The diastolic function was assessed in 48 children with DCM (26 male, 22 female, aged 3 months-14 yrs, 6.4±4.4 yrs) and 48 age and gender matched normal controls by using Doppler echocardiography. Mitral flow and pulmonary vein flow were recorded and measured at the initial enrollment. All patients were followed serially at 6 months interval. Results: The deceleration time of mitral E wave (DT) in patients with DCM was shorter than normal controls (92±27ms vs 128±40ms, $p<0.01$). E/A ratio of mitral flow increased and the ratio of pulmonary vein peak systolic velocity to peak diastolic velocity (S/D ratio) decreased in children with DCM (both $p<0.05$). These indicated that children with DCM had left ventricular restrictive diastolic dysfunction (RDD). According to our own criteria (data from 675 normal children), 21 (43%) patients showed abnormal left ventricular diastolic function parameters, including 16 (33%) with a shortened DT, 14 (29%) with an increased E/A ratio and 14 (29%) with a decreased pulmonary vein flow S/D ratio. By multinominal logistic regression, RDD was related to the dilation of left atrium and left ventricle, the elevation of pulmonary pressure as estimated by measuring the peak tricuspid regurgitant velocity, and not related to left ventricular ejection fraction. But patients with RDD had higher NY cardiac function scores ($p<0.01$). All patients were followed for 6–57 months (mean 28±16 months, median 22 months). Nine (45%), 2(10%), 9(45%) and 0 of 20 patients with RDD died, deteriorated, unchanged and improved respectively; while 1 (4%), 4(14%), 8(28%) and 15 (54%) died, deteriorated, unchanged and improved, respectively, in 28 patients without RDD ($p>0.05$). Conclusion: Among children with DCM, 42% had RDD, which was related to the NY cardiac function scores and prognosis. The identification of left ventricular RDD by Doppler echocardiography is helpful for assessing the symptoms of disease and predicting their prognosis in children with DCM.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

63

Mitral regurgitation in children with idiopathic dilated cardiomyopathy: its effect on left ventricular thrombus formation and clinical outcome

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Background. Mitral regurgitation (MR) has been shown to have a protective effect on left ventricular thrombus formation in adults with dilated cardiomyopathy. This study sought to determine its effect on left ventricular thrombus formation and clinical outcome in children with idiopathic dilated cardiomyopathy. **Methods.** MR was detected and graded by color Doppler echocardiography in 48 children (mean+/-SD, 6.6+/-4.4 yrs, range, 3 months to 14 yrs; 26 male, 22 female) with idiopathic dilated cardiomyopathy. Presence of left ventricular thrombus and spontaneous echo contrast (SEC) was carefully evaluated at the initial examination. All patients were serially followed. **Results.** MR was identified in 38 (79%) patients at the initial echocardiographic examination. MR was trivial, mild, moderate and severe in 5 (10%), 17 (36%), 13 (27%) and 3 (6%) patients, respectively. MR occurred more frequently in patients less or equal to 6 years. Left ventricular ejection and shortening fraction were significantly lower, and left ventricular dimension was larger in patients with MR. Left ventricular SEC was identified in 20 (42%) patients, and occurred more frequently in children without or with only trivial or mild MR ($p<0.01$). Left ventricular thrombus was found in 4 (8%) patients, all of whom had no or only mild MR. All patients were followed for 6–57 months (mean 28+/-19 months, median 22 months). Fifteen (31%), 17(35%), 6(13%) and 10(21%) patients improved, unchanged, deteriorated and died, respectively. Patients with moderate or severe MR had significantly higher mortality and lower rate of improvement ($p<0.05$). **Conclusions.** Moderate and severe MR in children with idiopathic dilated cardiomyopathy has a protective effect on the formation of left ventricular SEC, which may result in a low rate of left ventricular thrombus. However it is also a marker of poor clinical outcome.

64

ECMO and transcatheter left heart decompression in an infant with acute, severe left heart failure

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We describe a relatively unique technique for support of the heart during an episode of acute and severe heart failure. This is a case report of a 14 month old infant who presented with a 1 week course of poor appetite, low grade fevers, vomiting and progressive respiratory distress. Endotracheal intubation and mechanical ventilation was required during transport from an outlying referral hospital. Severe cardiomegaly with severe LV dilatation and dysfunction was noted at the time of transfer. Inotropic support with epinephrine, dopamine, and milrinone was required. Despite intense inotropic support she developed progressive low cardiac output with several episodes of ventricular tachycardia that initially responded to intravenous lidocaine. On the second post-operative day she had an episode of ventricular tachycardia that deteriorated to ventricular fibrillation and that did not respond to electrical defibrillation. During CPR she was placed on ECMO via cannulation of the right neck. After placement on the ECMO pump she was immediately taken to the catheterization lab. With transesophageal echocardiographic guidance a transeptal puncture was performed and an Amplatz exchange wire anchored in the left ventricle. Progressive static balloon dilatation of the atrial septum was performed with a maximum balloon size of 20mm. The initial LA pressure of 35 mm. Hg was reduced to 9 mm Hg with no gradient across the atrial septum. The pink frothy pulmonary edema fluid coming from the endotracheal tube abated almost immediately after the atrial septal decompression with immediate improvement in lung compliance. An endomyocardial biopsy was performed in the right ventricle with confirmation of acute myocarditis. The patient subsequently required 5 days of ECMO support. At the time of weaning and decannulation from ECMO her left ventricular function had improved markedly with a calculated ejection fraction of 40%. She required hospitalization for 3 weeks after decannulation but during this period of time demonstrated full neurologic recovery and full normalization of left heart function. She was discharged only on a small dose of diuretic and was taken off of the manuplanx for two weeks after discharge. We conclude that very agressive therapy is warranted in infants who present with acute and severe heart failure. We conclude that ECMO cannulation from the neck with transcatheter decompression of the left heart is an effective way of mechanically supporting the failing heart of an infant or child, without the need to perform a sternotomy or thoracotomy. Finally, we once again show the capacity for complete resolution of even the most severe of symptoms and signs associated with myocarditis and acute severe heart failure.

65

Heart failure in children: relationship between functional class, peak VO2 and VE/VCO2 slope

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Standard classification (NYHA) of functional status are subjective and poorly reproducible. The peak VO2 is the best objective index of functional capacity. In adults with chronic congestive heart failure the heightened ventilatory drive may contribute to the limitation of exercise capacity. However, the relationships between these variables have not been studied in children with heart failure. **Methods.** We studied 31 children (17 female), with CHF, age 8.6±2 years, 17 pts in NYHA functional class I, 5 pts in II, 6 pts in III and 3 pts in IV. The mean left ventricular ejection fraction was 27±10% (MUGA). As control group, we used 12 children (4 female), age 9.4±2.2 years. The children underwent maximal treadmill cardiopulmonary exercise testing, using a modified Naughton protocol to determine peak VO2 and VE/VCO2 slope. For analysis, patients were grouped according to functional class: group A (class I-III) and group B (classes III-IV). **Results.** The median peak VO2 (ml/kg/min) was significantly different between the groups, control = 29.3, group A = 22.9 and group B = 14.2 ($p<0.05$) for each comparison. The median of VE/VCO2 slope was significantly different between the groups, control = 33, group A = 39 and group B 47 ($p<0.05$), for each comparison. **Conclusion.** The peak VO2 and VE/VCO2 slope are good parameters to evaluate functional status of children with heart failure. The VE/VCO2 slope may be specially useful to evaluate children who fail to reach maximal exercise.

66

Comparison between New York heart association classification (NYHA) and peak VO2 in the assessment of functional status in children with heart failure secondary to idiopathic dilated cardiomyopathy

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The limitation of exercise capacity is one of most important features of chronic congestive heart failure(CHF). The NYHA functional class scale has been widely used to describe exercise tolerance,but it is subjective and poorly reproducible. The peak VO2 is a objective index of functional capacity in adults with CHF. However, these variables have not been compared in children with CHF. **Methods.** We studied 19 children (12 female), with CHF, age 8.2±2 years. 11 pts were in NYHA functional class I, 4 pts in II, 3 pts in III and 1 pts in IV. The mean left ventricular ejection fraction was 25±9.9% (MUGA). As control group, we used 12 children (4 female), age 9.6±2.2 years. The children underwent maximal treadmill cardiopulmonary exercise testing, using a modified Naughton protocol to determine peak VO2. **Results:** The functional classification (FC) by peak VO2 showed that 9 of the 11 children in FC I would be classified as Weber's class A and 2 would be classified as Weber's class B. Of the four children classified as NYHA FC II, one would be classified as Weber's class A; one, as Weber's class B, one, as Weber's class C and the remaining one, as Weber's class D. Of the three children in FC III, one would be classified as Weber's class B, while the other two would be classified as Weber's class C. The only child in FC IV would be compatible with Weber's class C. **Conclusions.** The peak VO2 may allow more precise grouping of children with heart failure. Further studies are needed to evaluate its prognostic value in this patient population.

67

Daunorubicin-induced cardiotoxicity in cancer children in relation to cardiac function and plasma levels of natriuretic peptide

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We have examined whether plasma levels of atrial natriuretic peptide (ANP) and brain natriuretic peptide (BNP), in addition to echocardiographic evaluation, can be used as specific markers for daunorubicin-induced cardiotoxic

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

effects in children. Consecutively, 34 patients (18 boys and 16 girls) who had previously received doxorubicin-containing chemotherapy were enrolled in this study. Plasma ANP and BNP were assayed simultaneously at the time of first cardiac function evaluation by echocardiography. Of the 34 patients, 8 (23.5%) had left ventricular dysfunction as assessed by echocardiography. Both ANP and BNP plasma levels in these patients were significantly elevated in comparison with healthy IV controls ($p<0.01$) or patients with normal cardiac function ($p>0.05$). BNP plasma levels correlated significantly with cardiac systolic function including EF ($r = -0.43$, $p<0.01$), FS ($r = -0.45$, $p<0.01$), mVcf ($r = -0.42$, $p<0.01$), LVSTI ($r = 0.59$, $p<0.01$). In addition, ANP plasma level correlated significantly with DT ($r = -0.32$, $p<0.05$) and FS ($r = -0.34$, $p<0.05$). There were no significant relationships between levels of natriuretic peptides and diastolic function. These results suggest that plasma ANP and BNP levels could be markers for doxorubicin-induced cardiotoxicity in children. Measurement of natriuretic peptide levels during treatment may allow the identification of some individuals sustaining higher levels of cardiac damage earlier in treatment. Careful and serial evaluation of cardiac function would be needed in patients with normal cardiac function, whose plasma natriuretic peptides are elevated, for the earlier identification of subclinical anthracycline cardiomyopathy prior to the development of congestive heart failure.

68

Multicenter treatment trial for chronic myocarditis in childhood: problems and preliminary results

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Background. A survey in all departments for pediatric cardiology performed in 1994 showed an incidence of dilative cardiomyopathy (DCM) in childhood of 50 cases per year in Germany. According to the results of biopsy performed in 1994, 15 cases of chronic myocarditis (CM) per year are assumed. Study design. Since September 1995, children with newly observed DCM get anticongestive therapy and undergo endomyocardial biopsy after 4–6 weeks. Patients with CM characterized by T-lymphocytes in the biopsy specimen are randomized and depending of the proof of virus genome treated by interferone or immunosuppressive agents or observed. The patients are followed clinically, by echocardiography and by re-biopsy after 6 months. Results. 60 cases were expected, only 31 patients were randomized, 7 under wrong conditions. Out of 24 patients with T-lymphocytes in the biopsy 13 were virus-positive, 11 negative. Only 5 underwent re-biopsy and fulfilled the study protocol completely. The clinical result in the various groups show no significant differences. Conclusion: Until now the model of therapy cannot be assessed finally. The willingness for multicenter studies has to be improved in Germany.

69

Cryopreserved cell transplantation into heart of dilated cardiomyopathic hamsters: a comparison of two cell types

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Cell transplantation may delay or prevent cardiac failure in patients with dilated cardiomyopathy. Cryopreservation of donor cells may permit storage until the time of transplant. We compared the effect of transplanted cryopreserved skeletal muscle cells (SKCs) and cryopreserved vascular smooth muscle cells (VSMCs) on heart function in a hamster model of dilated cardiomyopathy. Methods. Cells from thigh muscle (SKC) and aorta (VSMC) of 4 week old BIO53.58 hamsters were isolated, cultured, and cryopreserved. Cells were thawed and cultured 1 week before transplantation. A total of 4×10^6 cells of each type (SKC, n=30; VSMC, n=18) or culture medium alone (control, n=10) was transplanted into 12 week old recipients. Sham hamsters (sham, n=10) underwent thoracotomy without cell transplantation. Heart function was assessed on a Langendorff perfusion apparatus four weeks after transplant. After fixation, computerized planimetric mapping of the left ventricle determined ventricular size. Results. Both sham and control hearts were dilated with significant LV dysfunction. The cryopreserved SKCs survived, formed muscle-like tissue, preserved systolic function ($p<0.005$), and prevented left ventricular dilatation ($p<0.0001$). Cryopreserved VSMCs survived transplant and prevented heart dilatation ($p<0.001$) but systolic function was not significantly preserved (p= NS). Conclusion: In a hamster model of dilated cardiomyopathy, cryopreserved cells survived transplantation, prevented heart dilatation and preserved heart function. The preservation of heart function was dependent on the type of cell transplanted.

70

Expressional patterns of cytokines in a murine model of acute myocarditis: early expression of cardiotrophin-1

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Although several studies examined the crucial roles of cytokines and the induction of apoptosis in the myocardium, the pathogenic mechanisms of myocarditis remain unclear. To determine the role of cytokines in acute myocarditis, we examined expressional patterns of cardiotrophin-1 (CT-1), tumor necrosis factor (TNF)-alpha and interleukin (IL)-1alpha in a murine model of acute myocarditis. Ten-day-old Institute of Cancer Research mice were injected with Coxsackievirus B3 and killed on days 1, 2, 3, 4, 5, 7, 10, 14 and 28 of injection. TNF-alpha and IL-1alpha expressions were investigated on histological sections from each heart. mRNA Expression of TNF-alpha, IL-1alpha and CT-1 in the heart was examined by reverse transcription polymerase chain reaction and RNase protection assay. Age-matched uninfected mice were used as controls. In addition, to evaluate the pathological role of CT-1 in myocardial damage, we administered an anti-glycoprotein (gp) 130 antibody to CVB3 infected mice since CT-1 transduces its signals via gp-130 dependent signaling pathway. TNF-alpha and IL-1alpha expression was first detected in the cardiomyocytes on day 3, and reached the maximum level on day 7, when inflammatory changes were most prominent. Although an increased expression of TNF-alpha and IL-1alpha mRNAs was also detected on day 3, CT-1 mRNA expression was distinctly augmented on day 2. CT-1 expression preceded TNF-alpha and IL-1alpha expressions in a murine model of acute myocarditis. Interestingly, all CVB3 infected mice with anti-gp-130 antibody treatment died within 8 days. The enhanced expression of CT-1 mRNA might promote cardiac myocyte survival against viral infection and apoptosis by inhibiting the production of proinflammatory cytokines. CT-1 may exert a protective role by modulating cytokine production in CVB3 infected murine hearts.

71

Anomalous left coronary artery from the pulmonary artery: follow-up results after surgical repair

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We assessed the results after surgical repair in anomalous left coronary artery from the pulmonary artery (ALCAPA). Methods. A retrospective analysis was made of seventeen children who presented with ALCAPA between 1987 and 2000. The shortening fraction (SF), the left ventricular end-diastolic diameter corrected for weight and sex (LVEDD$_z$) and mitral regurgitation (MR) were determined. The electrocardiograms were examined. Results. One patient died before surgery. Fifteen patients underwent surgery, direct aortic reimplantation (n = 14) and a subclavian artery anastomosis (n = 1). One patient was treated conservatively. The mean follow-up was six years and seven weeks. There was no late mortality. After surgery, the SF increased by 68% ($p = 0.003$), the LVEDD$_z$ decreased by 44% ($p = 0.002$) and the degree of MR decreased ($p = 0.02$). The electrocardiograms improved in ten patients. Conclusion. This study demonstrates the excellent results after surgical repair in ALCAPA using the direct aorta reimplantation technique.

72

Results and analysis of morbidity and mortality of ventricular assist device (VAD) in children

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Only small series describe VAD as a system to keep children with otherwise unoperable heart failure alive until myocardial recovery or transplantation. Large series are missing. In 40 children, age 6 days – 17 years (median 7y), artificial replacement of heart function with VAD ("Berlin Heart") had been applied for long term support (1–131, mean 20 days) to offer life-saving support in our center between 1990 and 2000. They all were in cardiogenic shock with multiorgan failure, 6 with fulminant myocarditis, 18 cardiomyopathy, 9 chronic stage of congenital heart disease and in 7 weaning from bypass had failed after surgery. Three children with myocarditis were weaned from the system, 16 reached heart transplantation, 20 died and one is still waiting for transplantation. Causes of death were lack of peripheral circulatory resistance, multiorgan failure and shock (14), hemorrhagic complications (5) and one brain death. The treatable problems were thrombosis (17 children), bleeding and rethoracotomy (14), pump exchange (4). There

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

were no severe problems with infection of the system or pump dysfunction. One child has mild cerebral residuals after cerebral infarction, the other survivors are without sequelae.

Session 8: Catheter Interventions

73

Creation of interatrial communications with a new self-expanding shunt-prosthesis: preliminary results in a swine model

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Purpose: to evaluate a new technique for a patent and precise interatrial shunt by using a Nitinol Amplatzer shunt device. Materials and Methods: The self-expanding shunt device is constructed from Nitinol wire with an eccentric hole (4–10 mm), two flat 2–4mm retention discs and a short connecting waist of 2 mm. The device is attached to the delivery cable with a microscrew. Seven minipigs were used in this study. Five had a patent foramen ovale. Two pigs required a septal puncture to enter the left atrium. Placement technique of the device was similar to that of Amplatzer septal occluder. Balloon dilation was used after device placement. Follow up examination was performed at 1 week, 1 month and 3 months. Results: Placement of the device was technically successful in six animals. One animal died from ventricular fibrillation during catheterization. Left atrial angiography showed a precise interatrial shunt in all the 6/7 pigs immediately after placement, and in 4/4 pigs at 3 months' follow up. Pulmonary artery pressure increased at 1–3 months follow up in 4 pigs. One animal presented a significant pulmonary hypertension after the shunt creation. Postmortem examination demonstrated that one shunt with a 4mm device was occluded one month after placement. Four shunts of 10 mm remained patency at its follow up examinations. Neoendothelialization was presented partially or completely at 1 to 3 months. Conclusion: Large, permanent communications between the atria can be accomplished with this new device. It is of easy placement, self-centering, recapturability, self-expansion and proved to be successful to create an interatrial shunt of exact size. This new technique is applicable for the palliation of numerous congenital heart defects and the creation of fenestration in Fontan conduits.

74

Experimental evaluation of a modified Amplatzer duct occluder

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Purpose: To evaluate a new device specifically designed for patent ductus arteriosus (PDA) occlusion based on the PDA anatomy. Material and Method: The prosthesis consists of a cylindrical frame with an 82° angulated and concave aortic retention disc filled with polyester to augment the thrombogenicity. The delivery system of the device consisted of a long, manufactured 7-F thin walled Teflon introducing sheath, an anti-twisting delivery cable. Six mm Gortex grafts were surgically placed in 10 dogs between the descending aorta and pulmonary artery in the location of a patent ductus. Follow up angiograms were made at one week, one month and three months. Result: Percutaneous closure of surgically created PDA grafts was performed in 9 dogs. Complete occlusion of the shunt was obtained in all the animals. Temporary hemolysis occurred in one dog, which subsided following the occlusion of the graft in ten minutes. The aortic ostium of the shunt was completely occluded and covered by smooth glistening neoendothelium at 1–3 months postmortem examination. None of the retention skirts extended into the lumen of the aorta. Conclusion: The superelastic design with a cylindrical frame and convex inclined retension disc fits well the surgically created PDA in animal experiment. It effectively avoids the occurrence of complications, such as protrusion of device and hemolysis after device placement. This device also combines the advantage of small delivery system, easy placement, repositionability and immediate shunt closure.

75

Transcatheter occlusion of patent ductus arteriosus with severe pulmonary hypertension using amplatzer duct occluder and Amplatzer septal occluder

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PURPOSE: To evaluate the immediate and short term results of transcatheter occlusion of patent ductus arteriosus (PDA) with severe pulmonary hypertension using Amplatzer duct occluder (ADO) and Amplatzer septal occluder. METHODS: Between May 1998 to November 2000, among 365 patients with PDA who underwent transcatheter occlusion using ADO, 11 patients (3 male, 8 female) had severe pulmonary hypertension. Patients ranged in age from 1 to 48 years (median 19.5 years) and in weight from 10.5 to 65 kg (median 20kg). ADOs were used in 8 patients. Amplatzer septal occluder was used in 3 adult patients. Pulmonary arterial pressure was measured through the long sheath and aortography was repeated before the release. Chest radiograph and echocardiography were performed 24 hours after device placement; in 9 patients 8 patients completed a 1- month to 1-year follow-up. RESULTS: The device was successfully placed in the PDA in 9 patients, except two patients. There were no complications. Maximal ductus diameter ranged from 5.0 to 10.0 mm (median 6.7 mm). All PDAs were of type A morphology. The pulmonary/systemic blood flow ratio ranged from 1.2 to 2.9 (median 2.0). Angiography showed that 4 patients (44.4%) had complete immediate closure, 2 (22.2%) had a trace shunt, 3 (33.3%) had a small shunt. The mean pulmonary arterial pressure was decreased from 70.8 mmHg to 30.7 mmHg. Echocardiography 24 hours after the procedure revealed complete closure in all patients. 8 patients completed 1 month to 1-year follow up and no ductus recanalization was found. CONCLUSIONS: Transcatheter closure of PDA with severe pulmonary arterial hypertension using ADO is safer than surgery. Amplatzer septal occluder occlusion may be a useful alternative to ADO for a few adult patients with large-sized PDA.

76

Chronic inflammation around intravascular implants in animal models

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Background: There are only few data on local chronic inflammation after implantation of intravascular devices. We studied and compared inflammatory reactions around different implants in tissue specimen from lambs and pigs. Methods: Stainless steel coils and Nitinol coils were employed for interventional closure of a patent ductus arteriosus (n=18) or a ventricular septal defect (n=7). Stainless steel stents (coated and uncoated) were implanted in the inferior caval vein (n=4) or in the ductus arteriosus (n=3). Between 1 and 308 days after implantation the tissue block containing the implant was removed and embedded in Methylmethacrylate. After cutting, grinding, and staining, the sections were evaluated histologically. Results: In specimen from every type of implant but not in every individual specimen there are local lympho-plasmacellular infiltrate and histiocytic reactions with formation of foreign body giant cells. There is an increase of inflammation over time. Nitinol was found to induce a more pronounced inflammatory infiltration compared to stainless steel. No significant difference was found comparing different sites of implantation. Conclusion: Our results indicate that there is an ongoing inflammatory process at the interface tissue/implant in intravascular devices after implantation in lambs and pigs. The significance of our findings for long-term biocompatibility remains uncertain.

77

Interventional closure of patent ductus arteriosus with different morphologies using different occlusion systems

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Over the last years new duct occlusion systems have been developed. Different devices-designs can be used for different duct morphologies. Thus, the number of implants can be reduced, particularly for closure of large ductus arteriosus. The different systems are assigned to investigators according to Krichenko classification. Between 1994 and 2000 112 ductus arteriosus were treated using 118 devices. Detachable EMRIYE (Cook), Tungsten (Balt) coils and non-controlled-release rods (Target) were used. In addition, umbrella systems of Rashkind, Amplatzer and CardioSeal were implanted. 4 patients received more than one device. One coil embolized and was replaced by an umbrella. The mean minimal diameter of 47 ducts, closed by coil devices, was measured with 1.65 millimetre, rang 1.0 – 2.8 millimetre. 40.4% of those ducts were conical, 36% elongated, 13% tubular and few complex or window type ducts. 12 mm Rashkind devices were implanted in 16 ducts with a minimal diameter of 2.1 – 5.6, mean 2.7 millimetres. 75% of those ducts had a conical, 18.7% a window type shape. In 17 cases Rashkind devices were used for closure of 34 ducts with a minimal diameter of 3.6,

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

mean 4.1 millimeter 62% had a conical, 20% an elongated shape. Tubular and window type ducts were noted in 8.8% each. 14 ducts with a minimal diameter of 2.2 – 6, mean 3.9 millimeters were closed by Amplatzer occluders. The majority had a conical shape (64.2%) but 21.5% were complex and 14.3% elongated. One CardioSeal device was implanted in a 7 mm diameter window type duct. Conclusion: To reduce the number of implants for closure of large ducts with conical, elongated or window shape umbrella closure seems to be appropriate. Most small, conical or elongated ducts can be closed by one coil-system sufficiently.

78

Balloon dilation of the right ventricular outflow tract in tetralogy of Fallot

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Balloon dilation of the right ventricular outflow tract was performed in 29 patients with tetralogy of Fallot who required palliative treatment due to severe cyanosis or cyanotic spells. The mean age at dilation was 5.2 years (range 2 11–9 5). Successful dilation with increased systemic oxygen saturation was achieved in 25 patients (92.6%) but failed in 2 patients who had severe infundibular hypertrophy and needed palliative shunt thereafter. Of the 25 successful cases, saturation increased from a mean value of 74 +/- 7% (range 55–84) before dilation to 85 +/- 7% (range 73–94) after dilation (p < 0.001). Sixteen patients still had saturation >85% (mean 89 +/- 4, range 85–97) during the follow-up time of 22 +/- 10 months (range 6–38). By angiocardiography the components of pulmonic stenosis in these 16 patients was mainly at the pulmonic valve with mild infundibular stenosis. Among the remaining 9 patients, one required palliative shunt within 1 month after dilation due to severe spells, eight had an improved oxygenation for a period of time varied from 4 months to 2 years and then worsened thereafter. Pulmonic stenosis in this group was mainly due to infundibular hypertrophy. In conclusion balloon dilation is a safe and satisfactory palliation in tetralogy of Fallot. It is an alternate method to increase oxygen saturation for the patients, in whom enlargement of the pulmonic stenosis is mainly at the pulmonic valve, during waiting for total correction.

79

Percutaneous balloon dilation under transesophageal guidance for neonatal critical aortic stenosis. Medium to long-term results and technical considerations

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Objectives: To report the medium to long-term results of percutaneous transfemoral balloon dilatation (PTBD) with transesophageal echocardiographic guidance (TEEG) for neonatal critical aortic valve stenosis (NCAVS). Patients: Between January 1994 and May 2003, 23 patients (pts), aged 1 to 8 days, underwent PTBD with TEEG for NCAVS. Pts with hypoplastic left ventricle and/or aortic annulus were excluded from the study. Interventions: TEEG was used for ideal positioning of the balloon across the aortic valve and for the assessment of the results of valvuloplasty. The stenotic valve was crossed using the 'Pig-tail' over the wire technique. The balloon (very low profile balloons introduced through a 5F to 6F sheath) diameter was selected to be 70% to 90% of the diameter of the aortic annulus. Results: PTBD was successful (gradient <40 mm Hg) in 22/23 pts (95%). Significant aortic regurgitation (3+/4+) was observed in one pt. Early mortality was 7.4% - 2/23 pts (2 neonates with very poor ventricular functions and shock). Four pts (19%) had pulse loss with restoration after thrombolytic therapy. Four and 3 pts developed transient LBBB and non-sustained ventricular tachycardia, respectively. At a mean follow-up of 42+/-15 months the restenosis rate was 28.5% (6/21 pts) and an intervention was necessary (4 and 2 pts had successful repeat PTBD and surgical valvulotomy, respectively). The over all mortality was 13% (the neonate with the significant AR died during an attempted Ross procedure six months following PTBD). Conclusion: PTBD under TEEG using low profile balloons is an effective and safe alternative to surgical valvotomy for the initial palliation of NCA.

80

Midterm results of interventional management of congenital aortic valve stenosis

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Balloonvalvuloplasty (BVP). Mean age 7.5±7.1 years including 18 pts with age range 1–87d. Five neonates showed congestive heart failure, 4 persistent pulmonary hypertension and 1 had to be resuscitated before the procedure. LF measured by echocardiography was <40% in 6 of them. All the older pts were in stable condition. Aortic valve morphology (AV) Bi was present in 23%. BVP was carried out in general anesthesia, antegrade via femoral vein in 10 neonates, retrograde in the rest. Balloon/valve ring ratio was 1.52±0.11 in single balloon technique. Significant (p<0.001) immediate decrease of the systolic gradient and the Doppler derived gradient after 24 hrs was achieved in all pts (57±26 to 18±14 mmHg, 85±25 to 34±18 mmHg respectively). Re-BVP became necessary within 1 year in 5pts (1 neonate 2x), surgical valvotomy in 3, valve replacement in 5pts. Doppler gradient at last f/u (5.2±3.2y/s, 1-12y) in those treated with BVP alone was 35±14mmHg. AI worsened immediately after the procedure in 2/3 of the pts, was significant in 1 (grade III). During f/u the insufficiency decreased (17x grade I, 13x grade II). In the neonatal group were 2 deaths 1 during the procedure, the other 3 weeks later -due to LV-dysfunction- 4 had CPR in the cathlab, one died from endocarditis after 1y. Occurrance of loss of femoral pulse was significantly higher in the neonatal group (6/10). BVP is the procedure of first choice for patients with congenital AS, including neonatal period, although there is a risk of major complications. Reintervention – BVP or surgery – had to be done in 28%. In the majority BVP is a final palliation.

81

Long-term results after aortic balloon valvuloplasty during the first year of life

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The outcome of 29 patients undergoing consecutive successful aortic balloon valvuloplasty during the first year of life was examined. The mean age at the procedure was 87 days (range 2–365) and the mean weight 4.8 kg (range 2.2–10). Fourteen patients were in critical conditions and 5 had associated aortic coarctation. The valvuloplasty produced a gradient reduction of 70% (range 30–100) with a residual peak to peak gradient more than 30mmHg in 7 patients, mild aortic insufficiency in 11 and moderate in 5. The mean duration of follow up was 6.1 years (range 1–13.3). Seven patients (24%) developed early recurrence: 4 underwent successfull repeated balloon dilation, 2 surgical valvotomy with significant residual gradient associated with severe insufficiency in one case, one patient with recurrence and moderate regurgitation underwent Ross operation 5 years after balloon dilation. Long term follow up showed residual gradient more than 50 mmHg in 9 patients (31%), moderate/severe aortic regurgitation in 7(24%) and mild aortic regurgitation in 13. During the follow up period 4 patients (14%) underwent successfull Ross operation: 3 for stenosis and incompetence of the valve (one of them after surgical valvuloplasty) and 1 for severe incompetence. The other 25 patients (86%) are doing well without any medication. Long term results of balloon dilation of severe aortic stenosis show a good quality of life without any surgical procedure in 83% of patients. In our series there is no late mortality. A second procedure (balloon dilation, surgical valvuloplasty, Ross operation) was necessary only in 7 patients (24%). In our opinion aortic balloon dilation is the more effective and safe palliative procedure for aortic stenosis with excellent long term results and remains the technique of choice for the conservative treatment.

82

Graft and standard 'JOMED' stents for treatment of coarctation

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Management of coarctation has traditionally been surgical repair although balloon angioplasty has benefits and a recognised role. More recently, stents have added to the interventional tools for the management of coarctation especially in adults. We describe our experience with stent implantation in patients with coarctation in whom we used graft or standard stents manufactured by Jomed. Fourteen patients received stents for coarctation, 8 were male and 6 female. The age range was 10–55 years with a mean of 28. Seven had native coarctation. Of the 7 with recoarctation, 2 had previous balloon angioplasty and 5 had had surgery. The stents are balloon expandable and

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

were introduced through a percutaneous sheath (size range 9–11 French). The stent length ranged from 38–58 mm prior to deployment. Four were standard, uncovered stents whereas 10 were graft stents consisting of an expandable PTFE cover within the stent. The procedure was covered with Heparin and antibiotics and Adenosine was used during deployment to prevent displacement during balloon inflation. The mean gradient dropped from 32.4 ± 10.6 mmHg and the angiographic diameter increased from a mean of 8.1 mm ± 15 mm. The balloon size ranged from 10 – 25 mm (mean 15.8 mm). The mean fluoroscopy time was 11.4 minutes. The indications for stents included preferential choice for adults, 1 with an aneurysm following previous balloon dilatation, 1 with Takayasu disease and patent arterial duct and 1 who developed dissection during balloon dilatation. Stent treatment for coarctation is safe, effective and relatively simple. The short and medium term results are very encouraging and it may be the treatment of choice particularly for adults. The covered stents have a role for complicated coarctation and a theoretical advantage with regards to recoarctation.

83

Adenosine cardioplegia during interventional cardiac catheter procedures

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Cardiac interventional procedures are nowadays carried out with great success and with less complications as technology and experience advance. One of the reasons for a sub-optimal result or a complication is the movement of the catheter created by the contracting heart. By using Adenosine to create transient asystole, catheter or balloon movement is abolished during critical moments of the intervention. We used intravenous 0.- Adenosine on 25 patients with congenital heart disease who were undergoing a cardiac interventional procedure. These consisted of 14 patients undergoing balloon valvuloplasty for aortic stenosis and 11 for pulmonary stenosis, 14 undergoing balloon angioplasty for coarctation, 8 receiving a stent for coarctation, 4 with stenosed hemigrafts and one of each with tricuspid stenosis and pulmonary artery stenosis. The dose of Adenosine ranged from 200 – 700 µg/kg given centrally in 50 of the patients. There were no major complications. One patient developed transient atrial fibrillation which ceased spontaneously. Several patients had ventricular premature beats but some of these were associated with balloon inflation within a ventricle. There was no sustained ventricular tachycardia or fibrillation. Adenosine cardioplegia is safe and helps reduce complications associated with catheter movements during cardiac cycle. It also helps to improve the haemodynamic result of the intervention.

Session 9: Surgery-Research and New Techniques, Prosthetic Materials, Surgical Management and Results: Innovative/Experimental Surgery

84

Is transatrial approach adequate for repair of multiple VSDs in Infants?

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The possibility of effective closure of multiple VSD using transatrial approach is still considered dubious. Our experience includes 46 patients with multiple VSDs aged 7.3±1.7 months who were operated on between 01/01/1996 and 31/12/1999, mean weight 6.54 ± 1.6 kg. In 43 (93%) of cases all or some of multiple VSDs were located in the middle or inferior part of the ventricular septum. In the first group – 36 (78%) of patients – only transatrial approach was employed to close all VSDs; in the second group – 7 (15.2%) patients – a combined transatrial-transventricular approach was used. All perimembranous, outlet and infundibular VSDs as well as large trabecular VSDs (n = 2) were closed using patches. The rest of trabecular VSDs were simpled using large pledgets. Total post-operative mortality was 8.7% (4/46), 2.8% (1/36) and 42.8% (3/7) in the first and second group of patients respectively. Significant residual shunt was revealed immediately post-op in 1 patient of the first group (successfully reoperated) and 2 patients of the second group (both died in ICCU). Insignificant residual shunts were visualised using colour Doppler mapping immediately after surgery in 10 (28%) patients of the first and 3 (75%) of the second group. During a mean follow-up period 32.4±9.6 months all 42 discharged patients were assessed. 39 patients had 1 and 3 patients II functional class (NYHA). There was 1 late death due to infection, 1 patient was reoperated 2 years later due to partial detachment of the patch. Some trivial shunts were registered in 3/39 (7.9%) and 1/4 (25%) patients of the first and second group respectively. Our experience suggests that transatrial approach can be adequately used as a method of choice for surgical treatment of multiple VSDs in infants.

85

Modified atrial switch operation (coronary reallocation) for dTGA- Midterm results

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Multi-institutional study showed that units perform less than 20 arterial switch operations (ASO) a year have higher mortality compared to the busy institutions. Coronary translocation is the most important step for achieving the successful outcome in conventional ASO in TGA. We innovated a new technique of ASO, without coronary reimplantation in 1995. We evaluate our mid term results of this new technique. >From September '95 to August '98, 41 cases of TGA variants were treated with the new surgical technique. Age ranged from 15 days to 2.5 years (mean 6 months) and weight ranged from 2–11.5 kgs (mean 4.1 kgs). All patients were operated under cardio pulmonary bypass and were patients had simultaneous repair of coarctation. Overall hospital mortality was 22.5% (9/40). No patient died with coronary insufficiency. All the surviving 32 patients were followed up from 28 months to 66 months. One patient who did not have RVOT patch enlargement in the initial period required relief of RVOT obstruction 2 years after the primary correction. No other patient had significant RVOT/LVOT obstruction or semilunar valve incompetence. Cardiac catheterisation and angiocardiogram was done in 8 patients which showed good growth of neo aorta, neo PA and coronary arteries without any stenosis. Dobutamine stress thallium was done in more 8 patients showed no myocardial perfusion defects and regional wall motion abnormalities. There was no late death. All patients remained in class I without any cardiac medication. To conclude, the new technique of ASO avoids problems related to coronary translocation with excellent mid term results. It is a better alternative for the surgeons who are not well versed with coronary translocation of conventional ASO and with difficult coronary anatomy.

86

New technique of bidirectional Glenn shunt without cardiopulmonary bypass

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Various methods of performing bidirectional Glenn (BDG) shunt without cardiopulmonary bypass (CPB) have been reported earlier, with different techniques of venous drainage. There were no proper guidelines for case selection. We report a novel technique of performing BDG without CPB along with the criteria for case selection. >From July 1998 to Nov 2000, 12 cases of single ventricle and pulmonary venous complex were taken up for BDG without CPB. The age ranged from 9 months to 25 years (median 3 years). The weight ranged from 6–50 Kgs (median 12.5 Kgs). The criteria for case selection were an unrestricted atrial septal defect, no atrioventricular valve regurgitation, and no other intracardiac defects requiring correction. A temporary shunt was established between the SVC or innominate vein to PA for venous drainage during SVC clamping for BDG anastomosis. Central venous pressure (CVP) increased to a mean of 22.4 mm. Hg during SVC clamping, with improvement of oxygen (O2) saturation from 62.4% to 82.4%. After Glenn shunt, CVP and O2 saturation maintained at 13.2 mm Hg and 87.4%, respectively. No patients required blood transfusion. Postoperatively, there were no neurological abnormalities and no hospital mortality. The follow up ranged from 1 month to 28 months. There was no delayed neurological sequele. Our technique provides an excellent venous drainage with improvement of O2 saturation during SVC clamping. We believe the temporary shunt is more physiological and superior to the previously reported methods. It avoids problems related to CPB, blood transfusion and economy. It is easily reproducible with excellent results in a selected group of patients without compromising the completeness of repair.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

87

Anatomical right ventricular exclusion procedure for the pediatric Ebstein and tricuspid regurgitation complex: New surgical approach

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Ebstein's anomaly with severe tricuspid regurgitation(TR) presented in the neonate or infancy is challenging entity. We approached this group of patients with the new procedure with the excessive anatomical exclusion of the dilated, non-functioning RV and RA. Four infant and child were enrolled since 1996, aged 1 to 64 months and weighed 2.8 to 11.8 kg. Pulmonary valve was atresia in 1, critical stenosis in 1 and both stenosis and regurgitation in 2. All 4 had the history of collapse and resuscitation. One infant had the cerebral bleeding from hypoxia. The 64 months-old boy had undergone several surgeries including the prosthetic replacement without clinical benefit. Our approach consisted with (1)the extensive resection and right plication of the RV free wall, (2)closure of TV orifice(PTFE patch in 2, own TV tissue in 1), (3)establishment of the pulmonary flow by Blalock-Taussig shunt in 1, BDG anastomosis in 1 and TCPC in 2(with the lateral tunnel 1 and the extracardiac PTFE graft 1) and (4)the reduction/exclusion of the dilated RA. CPB and aortic cross-clamping times were 102 to 190 and 56 to 82 minutes respectively. All survived the surgery with CVP of 14 to 20mmHg immediately after CPB and 9 to 15 on the discharge from ICU. Clinical improvement was significant in all except one who was the only late mortality from the respiratory failure. As a conclusion, early aggressive anatomical exclusion of this RV and RA, directing to the future Fontan repair, was useful strategy in the challenging entity.

88

Anatomical repair of tricuspid valve for Ebstein's anomaly

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Objective. We propose a new method of tricuspid valvuloplasty for Ebstein's anomaly. Surgical technique. The attachment of the posterior and septal leaflets was incised preserving the antero-septal commissural region. Fibrous bands tethering the leaflets were divided and crimps in the leaflets were stretched. The cords attaching to the apical edge of the leaflets were preserved to prevent leaflet prolapse. After a longitudinal plication of the atrialized right ventricle was performed, the opposing edge of the separately mobilized septal and posterior leaflets were sutured together thereby enlarging the surface of septal leaflet. The mobilized leaflets were attached to the new atrioventricular junction which was a few millimeters below the original one. The tricuspid valve became competent as a result of restoring the coaptation of the three leaflets. Results. Five consecutive patients underwent the technique. Their ages ranged from 2.3 to 13.6 years (median, 6.7 years). The mean CTR on preoperative chest X-rays of 0.70 decreased to 0.55 postoperatively. The tricuspid annular size on echocardiogram was 191 % (range, 128 % to 328 %) of normal preoperatively and reduced to 72 % (range, 61 % to 89 %) of normal postoperatively. The preoperative tricuspid regurgitation, grade 3.5 in average, decreased to 1.0 postoperatively. Angiographic RV ejection fraction significantly increased from 0.36 (range, 0.25 to 0.46) preoperatively to 0.50 (range, 0.37 to 0.66) postoperatively. The RV end-diastolic volume to the normal value significantly decreased from 3.65 (range, 2.85 to 4.46) preoperatively to 1.19 (range, 0.83 to 1.86) postoperatively. Conclusion: Advancement of the septal leaflet and reduction of the tricuspid annular size were effective for restoring the competence of the tricuspid valve in Ebstein's anomaly.

89

Pulmonary autograft (Ross) operation and pericardial collar technique for the right ventricular outflow tract reconstruction

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Technical demands of the Ross operation and two valves at risk have delayed acceptance. The results of 18 patients who underwent Ross procedure and a new pericardial collar technique for the reconstruction of RVOT was documented. Methods: Patients ages ranged from 9 to 37 years (mean 16.2 ± 7.1 years). Three of them had prior open heart operation. Total root replacement technique was used in all patients. Ross / Konno procedure was performed in 3 patients with subaortic stenosis and/or aortic root hypoplasia. We used homografts in 6 patients and stentless bioprosthesis in 12 patients for RVOT reconstructions. A new pericardial collar technique

90

Aortic translocation and biventricular outflow tract reconstruction for d-transposition associated with ventricular septal defect and pulmonary stenosis, a follow-up

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Since 1983, we have participated in 14 aortic translocation operations (AoTLX) for d-transposition of great arteries (d-TGA) associated with ventricular septal defect (VSD) and pulmonary stenosis (PS). This is a clinical summary including a follow-up period of more than 17 years. Ten out of 14 patients were male. Age at operation ranged from 2 years 6 months to 7 years 7 months (median 3 years 5 months). All patients had previous palliative procedures (systemic-pulmonary shunts), except for one. In the majority (10/14), the RV-PA connection was made by a pericardial tunnel. In four patients, a valved homograft was used for this connection. Details of current clinical status were obtained from individual pediatric cardiologists. Postoperative complications include: bleeding (5 patients), ECMO support (3 patients), pneumonia (2 patients), delayed sternal closure (2 patients), hemiparesis (1 patient), and transient cervical blindness/transient atrial failure (1 patient). One patient died hours after separation from ECMO on the 6th postoperative day with sudden circulatory collapse. All other patients (13/14) are alive and well. Eight patients are presently on no cardiac medication. Late re-operation occurred in four patients; one for the obstruction of pulmonary homograft (12 years 10 months after AoTLX), one for RV dysfunction secondary to pulmonary regurgitation (6 years 9 months after AoTLX), one for mitral valve regurgitation secondary to endocarditis (3 years 5 months after AoTLX), and one for left pulmonary artery obstruction (1 year 6 months after AoTLX). In consideration of these follow-up results, we conclude that AoTLX is a valuable surgical option for patients with d-TGA, VSD and PS.

91

The modified Norwood operation for hypoplastic left heart syndrome: using right ventricle-to-pulmonary artery shunt

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Objective. Circulatory collapse due to low diastolic pressure has been a major cause of death after the classic Norwood operation. To prevent this lethal complication, we have constructed a right ventricle to pulmonary artery (RV-PA) shunt in first-stage palliation of hypoplastic left heart syndrome. Methods. Since 1998, 14 infants, weighing 1.6 to 3.7 kg, have undergone a modified Norwood operation. The procedure included a neo-aortic arch reconstruction and a non-valved polytetrafluoroethylene shunt between a small right ventriculotomy and a distal stump of the main pulmonary artery. Neo-aortic arch reconstruction was done by direct anastomosis between ascending aorta, aortic arch, descending aorta and proximal main pulmonary artery. The size of the shunt was 4 mm in 5 patients, 5 mm in 8, and 6 mm in 1. All patients were managed without any particular ventilatory manipulation. Mean value of diastolic blood pressure and Pa CO2 level were retrospectively compared to those obtain from patients (n=12) undergoing the Norwood operation with a modified Blalock Taussig shunt. Result: There were 11 survivors, including 2 patients weighing less than 2 kg. Pulmonary overcirculation did not occur in any of the 14 patients. Patients with the RV-PA shunt had significantly higher mean diastolic blood pressure than those with the B-T shunt(48±10 vs 37±6 mmHg, p<0.01), but mean PaCO2 levels did not differ (45±12 vs 52±15 mmHg, NS). 6 patients underwent bidirectional Glenn with a RV-PA shunt open after a mean interval of 4.5 months, and 2 underwent subsequent Fontan operation. Conclusion. Without delicate perioperative management, the RV-PA shunt in the modified Norwood operation maintains high diastolic blood pressures as well as a stable balance between systemic and pulmonary circulation. This procedure has a possibility to improve the survival of the first stage palliation for Hypoplastic Left Heart Syndrome.

92

Efficacy of additional pulmonary blood flow in the pulmonary circulation of cavopulmonary anastomosis

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Purpose: Pulmonary arteriovenous shunt (PAVS) is a serious complication following cavopulmonary anastomosis (CPA). To prevent the formation of PAVS, the placement of additional pulmonary blood flow (APBF) in CPA is

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

sometimes proposed. However, the role of APBF in the pulmonary circulation of CPA has not been elucidated sufficiently. We developed a rabbit model of CPA with or without APBF and analyzed the physiological characteristics of peripheral pulmonary arteries (PAs), especially hypoxic pulmonary vasoconstriction (an important prognostic for ventilation-perfusion matching). Methods: Twelve Japanese White rabbits (12–16 weeks of age) were used. Under general anesthesia, CPA was established by anastomosing SVC to right PA in an end-to-side fashion. The proximal right PA was completely ligated in 7 rabbits (Atresia group) and partially ligated in 5 rabbits (Stenosis group). Two to six weeks later, the response of peripheral resistance PAs (100–300 mm in internal diameter (ID)) to hypoxia (7% O2 inhalation) and L-NAME, a nitric oxide synthase inhibitor was analyzed by specially designed X-ray TV system. Results: Mean pressure and pulse pressure in right PA were not significantly different between Atresia group and Stenosis group (mean pressure: 8 and 11 mmHg respectively, pulse pressure: less than 2 mmHg in both groups). Baseline ID of resistance PAs was not significantly different between both groups. In Atresia group, resistance PAs did not respond to hypoxia or L-NAME. In contrast, significant constriction of resistance PAs was observed in Stenosis group (hypoxia: -2% vs -24%, p<0.0001, L-NAME: 3% vs -24%, p=0.0066). Conclusions: Receptor constriction of resistance PAs to hypoxia or L-NAME were lost in Atresia group but maintained in Stenosis group. This may support the beneficial effects of APBF in patients with CPA and in part explain the mechanism of BAVS.

93

Autologous reconstruction of the right ventricular outflow tract during ross procedure. Early and mid-term outcome

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OBJECTIVE: The pulmonary autograft remains the best substitute for the aortic valve in children even though different drawbacks remained to be resolved as lack of homograft availability and long-term deterioration. Recently, to address these issues we employed a modification of the Ross procedure(Cleuziou technique). METHODS: Between 1990 and 2000, 39 patients underwent modified Ross operation, in our institutions, for reconstruction of the RVOT utilizing a direct anastomosis between the remaining main pulmonary artery(PA) and ear valve bioprosthesis and creation of a monocusp valve from the anterior PA wall. There were 26(66.7%) males, mean age 20.7 years (range 5 to 42 years). 7(18%) patients underwent concomitant mitral valve repair. RESULTS: The overall hospital mortality was 3(7.7%) (only in Massa series). Preoperatively, 2 of them presented endocarditis and impaired left ventricular function. One of them underwent re-exploration due to important bleeding and the other patient underwent reoperation due to mediastinitis in the 11th postoperative day. Both patients died in the 12th and 24th postoperative day respectively due to progressive congestive heart failure. All survived patients were contacted at follow-up time, mean 27 months. There were no other deaths. All patients resulted to be in NYHA class I or II. In 26 survivors, the echo doppler of the aortic autograft showed none or trivial regurgitation in 33(80%) patients and mild in 8(19%). None or trivial incompetence of the PA monocusp was identified in 17(47.2%) patients, mild in 13(36%), moderate in 5(13.9%) and severe in 1(2.8%). This patient underwent reoperation at 2 years after the first procedure due to right ventricular failure and severe tricuspid regurgitation. CONCLUSION: These acceptable outcomes support the employment of this modification for the Ross procedure especially in children and where the homograft fibrinex does not exist. Other series and longer follow-up should confirm these results.

94

Reduction of platelet adhesion and fibrinous layering by small-diameter polyurethane grafts featuring a very open luminal surface

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INTRODUCTION – Research work demonstrated that the patency and long-term wound healing characteristic of a small-diameter vascular graft (SDVG) is both affected by the chemical nature of the material used for graft fabrication and by the porosity of its structure. To improve the patency of SDVG we developed of a spray phase-inversion

95

The pericardial membrane pulmonary monocusp: surgical technique and early results

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PURPOSE: Long term pulmonary insufficiency resultant from simple transannular patching of the right ventricle outflow tract will ultimately lead to deterioration in right ventricular function. Previously monocusps constructed from xenografts, homografts, fascia lata and autologous pulmonary artery wall have been utilized to maximize pulmonary regurgitation and its deleterious effect on right ventricular function. However, these tend to degenerate in the long term, necessitating re-operation. To circumvent this problem we have utilized a monocusp constructed from 0.1-mm polytetrafluoroethylene (PTFE, pericardial membrane) clinically demonstrated to be resistant to tissue in-growth and degeneration. PATIENTS: Seven children (3 Tetralogy of Fallot, 2 Pulmonary Stenosis) who required division of a small pulmonary annulus underwent monocusp construction utilizing thin PTFE. Three patients had previous corrective surgery. One of these patients had a bovine pericardial monocusp placed 8 years previously, which degenerated. Of the remaining two patients one had a pulmonary valvotomy as a neonate, the other repair of Tetralogy of Fallot with a transannular patch. RESULTS: At a mean (± standard deviation) follow up of 17 ± 8.6 months all patients are alive and are NYHA class 1. Echocardiography demonstrates mild pulmonary insufficiency (PI) in 2 patients, mild to moderate PI in 4 and moderate to severe PI in 1. CONCLUSIONS: The presence of a pericardial membrane monocusp in the pulmonary position may in the long term, preserve against the deleterious effects of transannular patching on right ventricular dysfunction, and be more resistant to degenerative changes characteristic of monocusps constructed of native pericardium or allogeneic tissue.

Session 10: Transplantation

96

Changes in self perception following successful heart or heart-lung transplantation

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Comparison of self perception before and 12 months after heart or heart-lung transplantation has previously demonstrated improvements in perceived physical health, anxiety and body image over time. We now wanted to assess whether there were further changes as the emotional trauma after transplantation. Self and ideal self perception were therefore evaluated in a group of 21 children and adolescents 12 months and 3 years after heart (n=13) or heart-lung (n=8) transplantation. Initial diagnoses of the patients were congenital heart disease (n=9), cardiomyopathy (n=10), primary pulmonary hypertension (n=2) and cystic fibrosis (n=1). A visual analogue scale was devised to provide information about how the child perceived him/herself on a series of different dimensions. Eight constructs representing body image, mood, self image, vocabulary, anxiety, aggression, self esteem and physical health were each rated by the child for two elements – self and ideal-self. At one year post-transplant, self perception scores were significantly (p<0.05) lower (more negative) than ideal self perception scores on all constructs, with children with cardiomyopathy rating themselves as thinner and less anxiety than those with congenital heart disease. Three years after transplantation, self perception scores were still significantly lower than ideal self perception scores on all constructs except for self esteem. There were no significant differences between the different diagnostic categories on any measures of self perception; furthermore, there were no significant changes over time. In conclusion, previously noted early improvements in self perception following successful transplantation are maintained in the medium term. However, self perception remains significantly more negative than ideal self perception, indicating that children one year from transplantation children and adolescents still demonstrate a failure to attain normal status.

97

Development of pre-school children with congenital or acquired heart disease

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Research on intellectual impairment among children with heart disease has focused mainly on older children. The present study aimed to determine whether previous findings are applicable to pre-school children and to assess

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

whether there are any differences between those with acquired or congenital heart disease (CHD). Children under three and a half years old were assessed prior to transplantation (n=26) or open heart surgery (n=24) and compared with a group of healthy children. Development was measured using the Ruth Griffiths Mental Development Scales. Within the transplant group, 11 had CHD and 15 had cardiomyopathy. Of those children awaiting open heart surgery, 10 had cyanotic heart disease and 14 had acyanotic lesions. Whilst the overall mean developmental scores were within the normal range for both the transplant and open heart surgery groups (transplant mean DQ: 95 (+/-18), open heart surgery mean DQ: 102 +/-8), scores were significantly lower than those of the healthy group. Within the transplant group, those with CHD had a significantly lower mean developmental quotient than those with cardiomyopathy (CHD mean DQ: 82 +/-13, cardiomyopathy mean DQ: 104 +/- 16). Furthermore, the CHD patients obtained significantly lower scores than the children with cardiomyopathy on areas of development covering locomotor abilities, speech and hearing, eye hand co-ordination and performance. Contrary to previous findings, there were no significant differences in the open heart surgery group between those with cyanotic and acyanotic lesions. It is concluded that results on intellectual development for older children do not apply to pre-school aged children. Furthermore, for children awaiting heart transplantation, diagnosis is a salient factor in determining outcome in most areas of development.

98

Congenital heart disease and heart transplantation: a single centre experience

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Purpose: To review the outcome of heart transplantation (HTPL) in patients (pts) with congenital heart disease (CHD). Methods: Out of 276 HTPL performed between 1985 and 1999, there were 20 pts (15 male) with CHD (7%). Principal diagnoses were isolated ventricular noncompaction (4), right ventricular dysplasia (3), congenitally corrected transposition of the great arteries (TGA) (3), single ventricle (2), complete TGA after atrial switch procedure (2), double outlet right ventricle (2), tricuspid atresia (2), left isomerism (1), idiopathic atrial dilatation (1). Eight pts had undergone up to 3 palliative procedures or definitive surgery prior to HTPL. All heart transplant recipients were followed prospectively. Results: Mean age at transplantation was 33.2±14.4 yrs (14–51, median 31.4), mean follow-up 6.1±4.0 yrs (0.01–12.4 (median 6.2). An aortic patch was taken from the donor aorta to cover the left pulmonary artery in one patient. This was the only foreign material used for cardiovascular reconstruction. There were 2 perioperative deaths (10%): severe bleeding due to hemostatic abnormalities on the 1st postoperative day and hyperacute humoral rejection on the 17th postoperative day. There was no late death. Postoperative complications included both vermum nasomycelin and renal failure requiring temporary hemodialysis (3 pts), staphylococcus sepsis and postoperative chylothorax in 2 others. Mid-term complications were pneumonia with Actinomyces and Nocardia 3 months postoperatively in 1 and pericutaneous sepsis with temporary multiorgan failure 11 months postoperatively in another pt. Inoperram graft aneurysmectomy was present in 1 pt 9 years after HTPL. All pts enjoy a good quality of life. Conclusions: If the complex anatomy and the technical difficulties secondary to previous palliative procedures/definitive surgery in this heterogeneous population are taken into account, perioperative morbidity and mortality are acceptable. Early and medium-term results are good.

99

Orthotopic cardiac transplantation after Fontan procedure

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The purpose of the study is to demonstrate the outcome of patients who received orthotopic cardiac transplantation (OCT) after a failing Fontan circulation. Twelve patients after Fontan procedure were submitted to OCT. Mean period from Fontan procedure to transplant was 10 years (2–17). Four patients received a total of 6 reoperation after Fontan and before OCT. The indications for cardiac transplantation were protein losing enteropathy (PLE) in 6 cases and heart failure with or without untreatable arrhythmia in 6 patients. There were 2 hospital deaths after OCT respectively due to multiorgan failure (preoperative NYHA class IV) and sudden cardiac arrest due to neurological events. Two late deaths occurred respectively 2 and 7 years after OCT, because of acute and chronic rejection. All survivors are in NYHA class I and one patient delivered a healthy baby. Regression of PLE was observed and documented in all cases, but in the last patient 3 month after OCT the serum protein level has remained unchanged. In conclusion, transplantation is only option for patients with a failed Fontan circulation, emergency status before transplant contributes to peri-operative mortality, regression of PLE does occur after OCT, acute and chronic rejection remain a problem in this group of patients.

100

Pediatric transplantation: normal values for invasive electrophysiology studies

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Diagnosis of chronic transplant rejection (REJECT) after heart transplantation (HTx) still remains a challenge. In children with HTx (pts) so far no data of atrioatrial (AA), atrioatrial (SA) and atrioventricular (AV) conduction measurements have been reported. The purpose of this study was to establish normal values of standard invasive conduction system properties in pts without histological evidence of REJECT. Methods: After informed consent all pts who underwent routine follow-up catheterization for myocardial biopsy and coronary angiograms had EP catheters placed in the HRA and His position. Baseline measurements of old atrial signals [Aold], new atrial signals [Anew] and His-recordings were obtained. SACT and CSNRT were measured and non-atrial incremental pacing [APACE] was performed up to WCL and/or 2/1 AV-block. Then Aold Anew electrical activity was investigated. Pts with biopsy-proven graft rejection (defined ISHLT: A>=1,V>0,CH>0) were excluded. Results: From 4/10/2000 13 pts were enrolled. Mean pts age at HTx was 7.5 mo (range 1 day–48 mo), mean age at EPS was 5.2 (range 1.8–8.2) yrs. All studies were done in conscious sedation, mean fluoroscopy time including angiograms and biopsies was 14.8 min. 12/13 pts showed NSR at baseline, 1 had junctional rhythm, 8/13 of pts had Aold and 2 of these showed AA conduction. Mean Aold-Aold was 449 (520-464) msec, mean Anew-Anew: 588 (422–700) msec. AH was 80 (64–132), HV 46 (range 26–64) and HRA-ESRA 269 (range 14–401) msec. SACT was 110.5/2 (range 50/2–254/2) msec and CSNRT 237.9 (range 136–503) msec. APACE showed mean WCL of 334 (range 280–460) msec and mean 2/1 block at 270 (range 240–360) msec. Conclusion: Normal values for AA, SA and AV conduction for pediatric patients s/p HTx are presented. Their possible clinical impact on graft rejection detection still has to be evaluated.

101

Lessons learned in pediatric cardiac re-transplantation

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Purpose: To examine the ideal indications and outcome of cardiac re-transplantation(re-Tx)in children. Methods: Retrospective review of 364 infants and children who underwent primary Tx between 1985 and 2000. Results: Twenty-seven children have received re-Tx at a median age of 9.1 yrs. Two pts had re-Tx within one day of Tx for primary Tx graft failure, both died. Median interval for re-Tx of the other 25 pts was 6.3 yrs(range 31D-14.2 yrs). Other indications for re-Tx were: Graft vasculopathy(CAV)(19),graft failure(3)and acute rejection(1). Operative mortality for the entire group was 11.1% and 5.3% for the CAV subgroup. Causes of hospital death were: Pulmonary hypertension(2), and sepsis(1). Late deaths were due to rejection(1)and recurrent CAV(1). Post re-Tx, 7 pts had peritoneal dialysis and median GFR was 78(range 33–122). Prior to re-Tx 52% of pts had PRA > 20%, one pt had plasmapheresis before and 3 pts after re-Tx. Rejection events after re-Tx averaged 0.4 episodes/100 pt days. Actuarial survival at 1 and 6 yrs post re-Tx: 85% and 72%. Freedom from CAV after re-Tx at 6 yrs: 57%. Conclusions: Elective re-Tx for CAV offers reasonable palliation. Long-term survival after re-Tx is limited by recurrent CAV and rejection. Renal dysfunction is not uncommon in re-Tx survivors.

102

M-mode tissue Doppler echocardiography may be useful in detecting cellular rejection in pediatric cardiac transplant recipients

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PURPOSE: To investigate whether M-mode tissue Doppler echocardiography (TDE) of the LV posterior wall (LVPW) detects the presence and degree

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

of cellular rejection in pediatric cardiac transplant recipients. METHODS: Thirty-six TDE studies were undertaken in 8 patients, median age at transplantation 9.0yr (range 1.7 to 21.3), median post-transplant interval 2.2mth (range 0.3 to 74.2). 5 males. All TDE studies were performed within 24 hours of an endomyocardial biopsy by an observer blinded to clinical and histological findings. Transmitral maximum mean velocity (MMV) and maximum velocity gradient (MVG) and the time (Ti) to these values from the onset of the QRS complex were measured during ventricular ejection (ve), rapid ventricular filling (rvf) and following atrial contraction (ac). RESULTS: Eleven biopsies were ISHLT histological grade 0, 13 grade 1, 6 grade 2 and 6 grade 3. Univariate analysis revealed MVGrvf to be the only parameter that correlated significantly with rejection grade ($r = -0.39$, $p = 0.02$). Mean MVGrvf differed for rejection grades 0 or 1 vs grade 2 (mean ± SEE 8.3 ± 0.7/s vs 5.5 ± 1.0/s, $p = 0.08$). In multivariate analysis TMVGrvf was the only significant independent predictor of rejection grade ($p = 0.025$). A cut-off level of 8.3/s for MVGrvf yielded a sensitivity of 83% (95% CI: 54, 100), specificity of 40% (95% CI: 25, 61) and negative predictive value of 92% (95% CI: 78, 100). No other TDE parameter nor IVS shortening fraction, LV mass, isovolumic relaxation time, E/A ratio, E deceleration time was significantly associated with rejection grade (all $p > 0.19$). CONCLUSION: MVGrvf and TiMVGrvf of the LVPW by M-mode TDE may be useful for non-invasively determining the need for endomyocardial biopsy in children and adolescents following cardiac transplantation.

103

Fate of infants and children mechanically bridged to heart transplantation, is it worth the effort?

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Objective: Worsening heart failure is a major cause of mortality among pediatric patients awaiting heart transplantation (HTx). We reviewed our 15-year experience of selective use of Mechanical Circulatory Support (MCS) for salvage of these children. Methods: Retrospective chart review and transplant database analysis of all children, up to 18 years of age, placed on MCS for salvage while awaiting HTx. Results: Seventeen of 530 Children (3.2%) listed for heart transplantation between November 1995 and November 2000, were placed on MCS while waiting. Indications for support included cardiomyopathy (n=8), rejection or acute graft failure following primary transplantation (n=6), pericardiotomy (n=2), and complex congenital heart disease (n=1). MCS consisted of extra corporeal membrane oxygenation (ECMO, n=11), HeartMate left ventricular assist device (HTM, n=4), and Sarns centrifugal pump (CP, n=2). The average duration of support was 13.4 ± 15.6 days (range 10 hours to 52 days). Four patients died on MCS (2 ECMO, 1 CP), two patients weaned from support (2 ECMO), and eleven patients were transplanted (6 ECMO, 4 HTM, 1 CP). Actuarial survival following bridge to HTx with MCS is 82% at 11 years. There has been no graft loss beyond 30 days. Complications while on MCS included, bleeding (2 ECMO, 1 CP), ischemic extremity (2 ECMO, 1 CP), sepsis (4 ECMO, 1 HTM), intracranial hemorrhage/infarct (5 ECMO, 1 HTM, 1 CP), and transient renal failure (6 ECMO, 1 CP). Conclusion: MCS prior to pediatric HTx does not negatively impact long-term graft survival and may be effectively used to salvage selected children who deteriorate while awaiting HTx. As miniaturized assist devices become more readily available, perhaps more children awaiting HTx may be salvaged.

104

Cardiac transplantation for multioperated complex congenital heart disease

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Among 142 patients (pts) undergoing heart transplantation (HTx) by the same surgical team between 01/1998 and 11/2000, 15 had complex congenital heart disease (CCHD): TGA 2, common ventricle 5, tricuspid atresia 2, complex DORV 2, TOF with absent pulmonary valve and absent left pulmonary artery 1, ASD and tricuspid malformation 1), with multiple palliative or curative prior operations (on average 2.85/pt). Prior surgical procedures involved pulmonary arteries in 73% of pts, systemic venous return in 46%. Two pts had atrial situs inversus and dextrocardia. Mean age at HTx was 26.4 yrs (13–42 yrs). Mean BSA mismatch between donor and recipient was +12% (+2 to +34%). The technique of sublobal HTx was used in all, allowing anatomic reconstruction of the pulmonary arteries and/or systemic venous return in all but 1 pt, by extensive use of donor tissue. In 1 pt, a Glenn anastomosis had to be kept in place. There was 1 postoperative death 6 days after HTx from sepsis. There were no specific postoperative complications. One patient died suddenly 52 days postop, without any rejection or coronary artery disease at the autopsy. The 14 survivors, including the pt with persistent Glenn anastomosis, are alive and well on average 5.6 yrs after HTx (0.2–10.4 yrs). In conclusion, young pts with end-stage multioperated CCHD may be considered as good candidates for HTx, despite complex anatomy, provided that pulmonary arteriolar are free without major collateral circulation. Operative mortality is similar to other pts and mid-term results are good.

105

Significance of right bundle branch block after pediatric heart transplantation

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Right bundle branch block (RBBB) has been associated with an increase morbidity and mortality after adult heart transplantation. However, its significance has rarely been reported in the pediatric population. Data from 24 heart transplanted patients (mean age 12.1±4.9 years) were analyzed age and sex of the donor, baseline cardiopathy, donors weight related to recipients weight, period of graft ischemia, number of acute rejections, number of endomyocardial biopsies, and right ventricular pressure as well as surface electrocardiograms (ECG) and epicardiary ECG recording obtained at the time of the biopsy. The late delay in the right bundle was defined as preapical right ventricular apical activation (V-RVA) time was delayed more than 30 ms after the onset of the QRS and distal as peripheral if it was less than 30 ms. Complete RBBB, appearing immediately after heart transplant and persisting during follow-up (5.2±3.2 years), was present in 7 (29%), distal in 3 and proximal in 4, while 13 (54%) presented an incomplete RBBB all with a normal V-RVA time. 4 (17%) had a normal ECG. Significant difference was found when comparing the 4 patients with proximal RBBB to the 20 other patients for number of biopsies (11.5±2.9 vs 7.9±4.7, $p=0.03$), number of rejection (2.0±0.8 vs 1.2±0.4, $p=0.015$) and right ventricular pressure (30.8±4.7 vs 22.4±3.6 mmHg, $p=0.03$). There was no significant difference for all other analyzed risk factors. There has been no progression of conduction delay with time. There has been no impact of RBBB on survival rate, functional class nor left ventricular systolic function. In conclusion RBBB is a frequent conduction abnormality following pediatric heart transplantation. It is proximal in about half of the cases, it correlates with number of biopsies, number of acute rejection and increased right ventricular pressure. Further studies are needed to precise the significance of RBBB in pediatric heart transplant.

106

Non compliance and acute rejection after pediatric cardiac transplantation

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Since acute rejection is a major cause of mortality and morbidity after pediatric heart transplantation, we studied the importance of non compliance on its incidence. Forty two patients (28 boys and 14 girls) aged 3 to 19 years (mean 9.8±5 years) were transplanted at our institution because of end-stage myocardial failure secondary to congenital heart disease (33%) or idiopathic cardiomyopathy (67%). Immunosuppressive therapy included cyclosporine, azathioprine and prednisone. Trough blood cyclosporine concentrations were maintained between 200 to 300 ng/ml. Follow-up ranged from 3 months to 15 years (mean 6.4 years). Actuarial survival rate at 3, 5, 7 and 10 years post-op was 85%, 85%, 80% and 65% respectively. Thirty four episodes of acute rejection occurred for an incidence of 0.9 per patient, and 0.5 per 100 patient-days. Freedom from rejection was 61%, 42% and 30% at 1, 5 and 10 years post-op. Among compliant children, the incidence of rejection was 0.24 episodes per patient, while in the group of 9 non compliant patients, the incidence was 1.1 episodes per patient ($p=0.002$). In the compliant group there has been no mortality, while in the non compliant group 4 deaths (44%) occurred secondary to acute rejection ($p=0.009$). Among the analyzed risk factors for non compliance, the post operative period appeared important since non compliance occurred exclusively after 12 months ($p=0.002$). Age had a dominant effect since all non compliant patients were between ages 12 and 19 years. Non compliance was also more frequent in boys ($p=.2$).

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Our results show that, in spite of an excellent survival rate after pediatric heart transplantation, non compliance has a very significant impact on acute rejection and mortality. It occurred more than six months post-op, exclusively during adolescence and more frequently in boys.

107
A modified solution for prolonged myocardial preservation for heart transplantation
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Objective: A new modified solution (SH solution) for prolonged cold storage of the heart has been developed. Method: Using a modified Langendorff model for functional parameter measurements, Wistar rat hearts were subjected to 30 minutes of perfusion with Krebs-Henseleit, 3 minutes of cardioplegic infusion and 10 hours of cold storage (4°C). The hearts were reperfused for 30 minutes and hemodynamic recovery, myocardial high-energy phosphate content and myocardial water content was assessed. The hearts were assigned to four groups (seven hearts per group), according to the cardioplegic solution used: group 1, St Thomas solution; group 2, Stanford cardioplegic solution; group 3, UW (University of Wisconsin) solution; cold storage solution; group 4, SH cold storage solution. Results: After 30 minutes of reperfusion, the LVEDP in group 4 was lower than that in groups 1 and 2 ($p<0.05$). The recovery of the left ventricular developed pressure (LVDP) in group 4 was significantly better compared with group 1 and group 2 (65.4%± 9.1%, 24.6%±6.6%, 27.3%±5.1% respectively, $p<0.05$). The recovery of the left ventricular +dp/dt was significantly better in group 4 compared with groups 1 and 2 (45.4%±21.6%, 27.8%±9.9%, 25.1%±10.8%, respectively, $p<0.05$). For hearts stored for 10 hours in UW cold storage solution or SH cold storage solution, the recovery of heart contractility did not differ significantly. Myocardial high-energy phosphate (ATP) was significantly higher in group 4 compared with group 1 and group 2 ($p<0.05$). The myocardial water content (dry weight/wet weight) in group 4 was lower than in group 1 and group 2 (18.30±0.55, 16.4±0.59, 16.4±0.42, respectively, $p<0.05$). Electron microscopic observation of the myocardium following 10 hours of storage showed damage in all groups, however group 3 and group 4 were significantly worse than group 1 and group 2. Conclusion: SH cold storage solution is as effective as UW solution for storage of the ischemic hearts for up to 10 hours for transplantation. SH solution differs from UW solution in K+ and other components contents, preventing cardiac allograft vasculopathy.

MAY 29 Time: 11:00–12:30

Session 11: Hemodynamics and Physiology, Cardiac Function/Hemodynamics

108
Tissue velocity and strain rate imaging – a new diagnostic approach for congenital valvar aortic stenosis
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The aim of this study was to evaluate cardiac function by myocardial tissue velocities and strain rate (SR) patterns in children with non-corrected, isolated congenital valvar aortic stenosis (AS). The study group consisted of 24 patients, 18 boys and 6 girls, median age 9.0 years (range 0.5–13). Twenty-four age- and gender-matched healthy volunteers were recruited as controls. Echocardiographic examinations were performed with GE Vingmed System Five equipment, using a 3.5 MHz phased array transducer, frame rate >80 Hz. The raw data of three consecutive heart cycles from the parasternal long axis- (LAX) and apical four chamber (4-CV) views were digitally stored and analysed off-line to assess radial and longitudinal velocity vectors of the LV wall movement. Systolic, early- and late diastolic velocity patterns and their duration were studied. Transmyocardial tissue velocity profile (TMVP) was obtained by measuring local velocities across the LV posterior wall (0.5 mm steps). Radial and longitudinal SR of the LV wall was calculated using the velocity gradient method. The systolic and early diastolic velocities in both views as well as the SR values (in 4-CV), were significantly reduced in the patients as compared to their controls ($p<0.0001$). Furthermore, in LAX the duration of the early diastolic wall velocities was significantly longer ($p<0.01$). These changes correlated with the severity of the AS. In early diastole, the TTVP was disturbed, mainly due to decreased endocardial velocities ($p<0.05$). Conclusion: Myocardial wall velocity and in particular strain rate measurements are useful for the assessment of myocardial function in patients with AS. These methods provide new insight in the pathophysiology of this congenital malformation.

109
Clinical significance of early surgical closure of atrial septal defect on neurohumoral factors and cardiac autonomic nervous function
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To investigate potential advantages of surgical closure of an atrial septal defect (ASD) in childhood, cardiopulmonary function, cardiac autonomic nervous activity, and neurohumoral factors were assessed before, 1 month, and 1 year after ASD closure in 26 patients aged from 2 to 32 years (mean 14 years). Results from 14 children (age 2 to 12 years) were compared to those in 7 young adults (age 18 to 32 years). Peak oxygen uptake (VO_2), pulmonary to systemic blood flow ratio, heart rate variability (HRV), arterial baroreflex sensitivity (BRS), heart to mediastinum 123I-metaiodobenzylguanidine activity ratio (H/M), plasma concentrations of norepinephrine (NE), atrial and brain natriuretic peptides (ANP, BNP) were measured and the results compared with those in 46 healthy control subjects aged from 9 to 21 years (mean 16 years). There was no relation between age and pulmonary to systemic blood flow ratio of the ASD patients. Mean capacity peak VO_2, and BRS decreased while both ANP and BNP increased 1 month after ASD closure. The decrease in BRS and increase in ANP and BNP was greater in children than in young adults. 1month after ASD closure ($p<0.05$–0.001). From 1month to 1 years after ASD closure, the decrease in ANP and BNP and increase in HRV and BRS was greater in children than in adult patients ($p<0.05$). One year after ASD closure, BRS improved and ANP decreased significantly compared to the corresponding preoperative values ($p<0.05$). The increase in peak VO_2 correlated inversely with age at operation ($r=-0.67$, $p<0.001$). Our data suggest that the potential capacity for circulatory adaptation to hemodynamic change or surgical stress is greater in children than young adults and that neurohumoral factors, cardiac autonomic nervous function, and exercise capacity improved after ASD closure.

110
Chronic right ventricular pressure overload increases right ventricular contractility but decreases pump function
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Background: In several clinical situations, the right ventricle (RV) is exposed to a chronic pressure overload which may affect both RV and left ventricular (LV) function. We aimed to quantify biventricular function in an animal model of chronic RV pressure overload during baseline and dobutamine infusion. Methods: In young lambs, chronic RV pressure overload at the level of systemic (aortic) pressure was established by adjustable pulmonary artery banding (PAB). Biventricular function was quantified in 5 PAB lambs and 5 age-matched controls by the slope (Ees) of the RV and LV end-systolic pressure-volume relationships (ESPVR), using combined pressure-conductance catheters during inflow reduction. Results: After minimally 8 weeks of PAB, the lambs were clinically in good condition. Chronic PAB resulted in a significant increase in RV end-systolic pressure (Pes) from 12±3 to 64±8 mmHg ($p<0.01$) and significant RV hypertrophy. LV-Pes was unchanged. Cardiac output decreased from 3.6±0.8 to 1.6±0.3 l/min ($p<0.02$) while heart rate remained constant. RV ejection fraction tended to decrease from 61±14 to 45±15 ($p=0.11$), as compared by an increase in RV end-systolic volume (Ves) (15±6 to 22±10 ml/kg) and a decrease in end-diastolic volume (Ved) (38±7 to 31±9 ml/kg). RV Ees increased from 1.1±0.4 to 4.2±1.3 mmHg/ml ($p<0.01$), indicative of increased contractility. In the LV, Ved and Ves decreased significantly (both $p<0.01$), while LV-Ees was unchanged. During dobutamine infusion, the RV but not the LV inotropic response was severely blunted in the PAB group. Conclusions: Medium-term RV pressure overload at systemic (aortic) level results in reduced pump function and contractile reserve despite increased RV contractility. LV volume is reduced but LV contractile function is generally maintained. These findings may characterize a transitional state from compensatory hypertrophy to the onset of ventricular failure.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

111

Diastolic function following heart transplantation in children using biaxial anastomosis

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It has been shown that heart transplantation using biaxial anastomosis (BA) maintains good right heart diastolic function by preserving the donor right atrial geometry However, because the left atrium (LA) anastomosis- in which partial donor and recipient LA cuffs are connected- alters resultant LA geometry, we postulated that left-sided diastolic function might be abnormal. We performed echocardiography 29.3 (SD16.4) months following transplantation on 21 children who underwent BA All patients were free of rejection at the time of study Doppler flow data were compared with 35 age-matched controls RESULTS: No significant difference of the right and left atrial area index was found between BA and controls There was no difference in mitral, pulmonary vein and tricuspid doppler flows data between them Systolic flow/ diastolic flow (S/D) ratio of HV and SVC in BA was significantly lower than in controls CONCLUSIONS (1) Despite altered LA geometry, left sided diastolic function is preserved following BA. (2) Low S/D ratio with low diastolic HV and SVC flow indicates excellent diastolic suction effect into right ventricle.

112

Normal coronary flow reserve and coronary flow response to nitroglycerin in children operated with arterial switch operation for transposition of the great arteries

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Background. Recent studies suggest that coronary flow reserve (CFR) is moderately or severely reduced in children treated with the arterial switch operation (ASO) for transposition of the great arteries (TGA) These studies have been performed with positron emission tomography While these findings are of great concern, they have not been confirmed by other available methods. Methods Ten symptom-free children were examined with echocardiography and heart catheterization 4 to 10 (median 6) years after ASO Selective coronary angiographies were performed A 0.014 inch Doppler FloWire(tm) (Cardiometrics) was used to measure flow velocity in the left anterior descending (LAD) and right coronary arteries (RCA) before and after intracoronary injection of adenosine (0.5mg/kg) and nitroglycerin (5mg/kg) CFR was defined as the ratio of hyperemic to basal average peak velocity (APV). Results. Ventricular function and wall motion were normal in all subjects A total occlusion of the left coronary artery was found on coronary angiography in one child, but all other coronary arteries were without stenosis The median (range) APV at rest was 14.5 (14–21) cm/s in the LAD and 15.5 (9.6–30) cm/s in the RCA The median (range) CFR in the LAD was 3.7 (3.0–4.5) and 3.4 (2.9–4.8) in the RCA. The median (range) increase in APV after intracoronary injection of nitroglycerin was 290% (240–420%) in the LAD and 260%(190–460%) in the RCA. Conclusions The CFR and coronary vasoreactivity as measured with intracoronary Doppler guide wire in children with TGA treated with ASO was within normal limits previously reported for healthy young adults. Coronary angiography should be performed in children with TGA after the ASO, as it can reveal unexpected coronary occlusions. Evaluation of coronary function should preferably be performed with intracoronary Doppler guide wire

113

The use of intra-operative left ventricular pressure volume relations to optimise pulmonary artery band placement during retraining of the subpulmonary left ventricle

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Background. Retraining of the subpulmonary left ventricle (LV) is required before a late arterial switch can be performed in patients with a systemic right ventricle (congenitally corrected transposition (CCTGA), or following Mustard or Senning operation). Pulmonary artery (PA) banding can be injurious to the LV and may not achieve satisfactory retraining. Better methods of maximising efficiency of retraining and minimising LV injury are needed. Methods Intra operative volume and pressure data were recorded with an integrated conductance and micromanometer tipped catheter in the LV in three patients (1 Mustard, 2 CCTGA) undergoing PA banding as part of a plan for staged late arterial switch. Results The response to PA banding shows initial acute adaptation of the LV to PA banding, with a passive linear end systolic pressure volume relation. As the band is tightened further however, a decompensation point (DCP) is reached and thereafter a negative pressure volume relation indicates acute LV failure. The LV pressure at the decompensation point is variable and unpredictable. In one 56-year old patient with CCTGA the decompensation point was determined, and a PA band was placed appropriately. Restudy 2 weeks later showed increased LV contractility, an enhanced adaptive phase, and a DCP higher on the linear adaptive phase which allowed further tightening of the PA band and subsequent benefit to the patient. Such monitoring facilitates accurate monitoring during PA band banding, reducing the acute insult to the LV myocardium by leaving the LV in the adaptive phase of the pressure volume relation. Conclusion Real time LV pressure volume loops allow an improved understanding of the physiology of LV pressure loading and may aid optimal placement of PA banding for retraining of the subpulmonary LV, even in older adult patients

114

Increased angiogenic growth factor levels in cyanotic congenital heart disease

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Background: Previous studies demonstrated that expression of angiogenic growth factors is enhanced in hypoxic models However, little is known about these factors with cyanotic heart disease The purpose of this study was to examine the relationship between plasma levels of angiogenic growth factors (vascular endothelial growth factor (VEGF) and hepatocyte growth factor(@;HGF)) and rate severity of cyanosis Methods: The study included 85 patients with cyanotic heart disease and 87 controls Age ranged from 0 day to 20 years (median 4.2 years) in cyanotic group and from 5 days to 31 years (median 4.6 years) in control group. Mean systemic oxygen saturation was 80.6+17.3% in cyanotic group and 98.1+0.5% in control group. Plasma VEGF and HGF were measured using an enzyme-linked immunoassay. Results: Plasma VEGF level in control is significantly dependent on age ($y=1.12x^{0.27}$ (y=VEGF(x)) $p<0.0001$), and remained as a plateau after 3 months of age. In contrast, such age dependency was not found in HGF. Although VEGF and HGF levels were not different between cyanotic and control group within 1 month after birth, the VEGF level in cyanotic group after 3 months of age was significantly elevated compared to control (149+106 vs 67+12pg/ml, $p<0.0001$) Moreover, VEGF level was significantly negatively correlated with oxygen saturation ($y=-4.40x+7.53$, $R=-0.47$, $p<0.0001$) in cases more than 3 months old. In contrast, no correlation was found between HGF level and oxygen saturation. or VEGF level Conclusion Although physiologically increased VEGF in the neonatal period is rapidly decreased under normal oxygen saturation, a higher VEGF level persists if systemic hypoxia is present. These findings may influence the development of systemic to pulmonary collateral arteries in patients with cyanotic heart

115

Endogenous nitric oxide production in children with congenital heart disease

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Purpose: Endogenous nitric oxide (NO) physiology is altered in many disease states The effect of congenital heart disease (CHD) physiology on endogenous NO production is largely unknown. The objective of this study is assessment of endogenous NO production in children with CHD. Methods. Using chemiluminescence technique, arterial plasma concentration of nitrates and nitrites (NOmet), a marker for endogenous NO production, was measured in 125 children (mean 32–54 mos, range 2d–18y) undergoing hemodynamic assessment during cardiac catheterization for diagnosis and/or treatment of CHD. Results: For all patients, plasma concentration of NOmet was inversely related to PaO2 ($r=-0.54$, $p<0.001$) and arterial oxygen saturation (SaO2) while breathing room air ($r=-0.53$, $p<0.001$). Forty-three patients with cyanotic CHD (room air SaO2 79.7 +/- 9.2%) had elevated NOmet (84.7 +/- 37.3 vs 59.6 +/- 25.5 μM, $p<0.0001$) compared to 82 with acyanotic CHD (room air SaO2 96.4 +/- 3.1%). Eleven patients status post single ventricle palliation with non pulsatile pulmonary blood flow (PBF) had elevated NOmet (90.4 +/- 26.1 vs 62.1 +/- 32.4 μM, $p<0.02$) and lower indexed pulmonary arteriolar resistance (2.19 +/- 0.56 vs 3.2 +/-

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

3.23 units/m2, p<0.01) compared to 114 patients with pulsatile PBF. Conclusions: An inverse relationship exists between endogenous NO and SaO2 and PaO2 in CHD. Cyanotic CHD is associated with increased production of endogenous NO compared to acyanotic CHD. Evidence of increased endogenous NO production in children status-post single ventricle palliation may represent physiologic adaptation to non-pulsatile pulmonary blood flow.

116 ✓

Non-invasive assessment of ventricular filling patterns in patients with univentricular hearts prior to the Fontan operation

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Background: Eligibility of pts with univentricular hearts (UVH) for the Fontan operation (FO) is partially dependent on preserved diastolic function (DF), conventionally invasively assessed by ventricular end-diastolic pressure (VEDP). Objective: to perform non-invasive, pulsed wave Doppler (PWD) assessment of DF in pts with UVH and normal VEDP and compare results to normal children. Methods: PWD interrogation of atrioventricular valve (AV) and pulmonary vein (PV) flow was carried out in 55 pts with UVH immediately prior to FO (mean age 7.3±6 years). All pts were in sinus rhythm with no/mild AV regurgitation. Normal data was derived from 221 normal children (mean age 10.6±3.9 years, range 2–18 years). Age- and heart rate adjusted comparisons of AV and PV flow data were performed using analysis of covariance. Results: Mean VEDP in UVH pts was 9.5±3 mm Hg. PWD data on AV flow in these pts differed significantly from those in normal children with reduced E/A ratio (1.4±0.03 versus 2.2±0.04, p<0.001) and longer deceleration times (175±3ms versus 156±2ms, p<0.001) in the patient group. PV systolic/diastolic TVI ratio was decreased in UVH (0.9±0.04 versus 1.2±0.02, p<0.001) relative to the control group. Conclusions: PWD flow patterns in UVH pts prior to FO were characterized by prolonged ventricular relaxation compared to normal children. However, E/A ratios in UVH were > 1.0 and PV flow was slightly diastolic dominant. This probably reflects a mild degree of diastolic dysfunction (grade II/4 normal) to I/4 dysfunction) and would be consistent with the low VEDP seen in this select group. These data provide a benchmark for PWD assessment of diastolic physiology prior to FO and offer a foundation for further non invasive evaluation of DF in these pts.

117 ✓

Estimation of right ventricular ejection fraction in patients with chronic right ventricular pressure overload using myocardial performance index

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Objective: We examined the value of myocardial performance index (MPI) by means of echocardiography in symptomatic patients with chronic right ventricular (RV) pressure overload. The aim of the study was to obtain from this index a measurement of RV ejection fraction (EF), which is a well-known clinical parameter, but difficult to obtain in patients with RV dysfunction. Methods: RV MPI by the means of Doppler echocardiography and RV ejection fraction by means of magnetic resonance imaging were measured in 10 consecutive (7 male and 3 female) asymptomatic or minimally symptomatic (NYHA I or II) patients (age 28.7 ± 11.6 years) with chronic RV pressure overload. The MPI was obtained through the use of formula (a-b)/b where a is the interval between the cessation and onset of the tricuspid inflow, or the duration of tricuspid regurgitation (if present), and b is the ejection time. Patients with left ventricular dysfunction were excluded from the analysis. Results: The correlation between RVEF and MPI was r = -0.66 p=0.001. For practical reasons the MPI was converted to RVEF-index. The EF-index was calculated by the formula 92-62 × MPI acquired from linear regression analysis. Mean RVEF in the patient group determined by MRI was 53.5 ± 12.3 % and mean RVEF-index was 64.0 ± 14.3 (P=NS). Conclusion: Our study shows a significant inverse correlation between RV MPI and RV EF determined by the means of MR imaging. These data suggest that MPI converted to EF-index can be used in clinical practice as determinant for RV EF in patients with RV chronic pressure overload.

Session 12: Surgical Management and Results: Abnormal Venous Return, Left Ventricular Outflow Obstruction/Aortic Stenosis, Pulmonary Atresia

118

Physiologic repair of anomalous left coronary artery from the pulmonary artery (ALCAPA) by aortic reimplantation in 47 patients: early survival, patterns of ventricular recovery and late functional outcomes

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Objectives: To determine the early and late outcomes of patients with ALCAPA who had repair by aortic reimplantation (AR). Methods: From 1952–2000, 63 patients presented with anomalous coronary artery from the pulmonary artery. 47 (median age 7.7mths, weight 7.7kg) were repaired by AR, and are the subjects of this study. Before repair, 10 patients (21%) presented in extremis and 38 (80%) had a history of heart failure. Results: Hospital survival was 92%. Five patients had perioperative ECMO (median 4d, 2–8) and were significantly more likely to present in critical condition (40% versus 3% if no ECMO, p=0.006) or with ventricular arrhythmias (40% versus 3%; p=0.027), have significantly lower pre-repair EF (10%, range 7–20% n=5 vs 40%, range 9–75%, n=36, p=0.011) or more severe LV dilatation (p=0.027). With up to 15 yr follow-up (mean 4.71 yrs), there were no late deaths. Kaplan-Meier survival was 91% at 1 mth, 1 yr and 5 yrs; freedom from reoperation (for ischemia n=2, PS=1, or severe MR n=1) was 98% at 1 yr and 93% at 10 yrs. At follow up, echocardiography demonstrated improvements in mean EF (64+/-9% vs 33+/-2 % preoperative, p<0.0001); the degree of MR (moderate+ MR 9% vs 38% pre-repair, p< 0.02), wall motion abnormalities (15% vs 62% pre repair p< 0.002). The ratio of measured LVED diameter to the 95th percentile of normal declined from 1.4+/-0.3 to 1.0+/-0.1 (p<0.0006). By repeated measures mixed linear regression analysis, normalization of EF and LV functional parameters occurred within 1 year of repair. Stress testing was normal in 17 of 20 patients in whom it was performed. Conclusions: Physiologic repair of ALCAPA by AR yields excellent early survival and late functional outcomes even in critically ill infants.

119 ✓

Evolving strategies and improving outcomes of the modified Norwood procedure: a 10 year single institution experience

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Purpose: To determine the outcomes of the modified Norwood procedure with a focus on the impact of evolving management strategies and accumulated institutional experience. Methods: From 2/90 to 10/00 171 infants had a modified Norwood operation, and were categorized into 3 operative cohorts (era I: 1990–93, era II: 1994–97, era III: 1998–2000). With consecutive eras, perioperative management, surgical technique and perfusion strategies evolved to promote earlier diagnosis, modified arch reconstruction, avoidance of circulatory arrest, and efforts to balance the circulations with aggressive afterload reduction. Results: There were 117 males (68%) and 54 females with a median age of 6d (1–175d). Median weight was 3.3 kgs (1.7–4.8 kgs). The mean BSA was 0.23 +/- 0.06 m2. HLHS or a variant thereof was present in 118 (70%) infants. Overall 5-yr survival was 43%. Multivariable analysis revealed that preoperative ventilatory support, date of operation, and lower weight were independent predictors of time-related mortality. Morphologic features (non-HLHS diagnosis, ascending aortic size, or noncardiac anomalies) were not associated with increased risk of death. Stage 1 hospital survival for patients in era III was 82% and significantly better than in previous eras (p<0.001). Overall Kaplan-Meier survival at 1 month, 1 year, and 5years was 43%, 31%, and 28% in era I; 60%, 49%, and 45% in era II; and 80% at 1month, 68% at 1 year in era 3 (p<0.001). Interstage I-II attrition was 15% and occurred in 5 children <2 months of age, 9 children 2–3 months, 5 children >3 months. Conclusions: With increasing experience, improvements in perioperative care and surgical technique, good outcomes can be expected for the stage I modified Norwood procedure. Greater monitoring of the interstage period may reduce interval mortality and improve overall survival.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

120
Intraoperative device closure of multiple muscular ventricular septal defects

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Background: Surgical management of muscular VSDs continues to be a challenge, especially when they are multiple, associated with complex cardiac lesions, or require exposure through ventriculotomy. We reviewed the feasibility and outcomes of intraoperative VSD device closure of muscular VSDs.Methods and Results: Intraoperative VSD device closure was performed in 14 patients, 9 of whom had associated complex cardiac lesions, and 10 had multiple VSDs. Two thirds of patients were neonates or small infants. One patient had Swiss cheese septum. Five patients had previous PA banding. There were 2 early deaths, one in a severely ill child with LV failure and another in a patient with hypoplastic left heart. Mean Qp:Qs prior to device insertion was 3.5 to 1.Concomitant PA banding was necessary in 2 patients. Postoperative mean Qp:Qs was 1.5. The device was well seated in all cases on postoperative echocardiography. Residual leaks improved or did not progress except in one patient who needed reoperative PA banding. There was one late death due to aspiration and another patient required heart transplantation for progressive ventricular failure. Conclusion: Intraoperative VSD device placement for multiple muscular VSDs is feasible, avoids ventriculotomy and division of intra-cardiac muscle bands, and can be applied to neonates or small infants.

121
Aortic regurgitation after arterial switch operation

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Aortic regurgitation (AR) after arterial switch operation was studied in 228 patients with a mean follow-up time of 85 +/- 44 months (1–168 months). Mild AR at most was detected in 60 cases (26.5%) and moderate AR or more in 23 cases (10.1%) by echocardiography. Freedom from AR (mild or more) at 10 years after operation was 68.4% and freedom from AR (moderate or more) at 10 years was 84.2%. Progression of AR with time was recognized in some cases, and aortic valve replacement was undergone in 2 cases. Multivariate analysis revealed that older operative age, left ventricular outflow tract obstruction and larger annular diameter of the original pulmonary valve were the significant predictors of AR (moderate or more). In the analysis of AR (mild or more), pulmonary artery banding participated in the significant predictors. In conclusion, aortic regurgitation was detected in considerable part of the patients after arterial switch operation, and close and continual observation in the long term period is important.

122
Late stage results of the Takeuchi procedure for anomalous origin of left coronary artery from pulmonary artery

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From May, 1985 to December, 1999, 30 patients underwent the Takeuchi procedure at our institute. 6 patients were operated on who were any yet of 12 months of age. Post-operative left ventricular function were assessed using cardiac catheterization or echocardiography records retrospectively. Follow ups were performed for 7 patients over a span of 5 years (mean, 8.7+/-4.8 years). Results. There were no early stage or late stage deaths. About 1 month following operations, ejection fractions improved (Number: 9, pre- and post-operative; 42+/-17%, 52+/-20% p=0.06) with reductions of the left ventricular end-diastolic volume (259+/-153%, 165+/-121% of normal value P=0.004). In the late stage period (Number, 5, duration, 2.5 - 14.7 years), the intra-pulmonary tunnels were confirmed to be all patent. Ejection fractions further improved (pre-, post-, late: 31+/-7, 44+/-19, 61+/-4%) with the reduction of the left ventricular end-diastolic volume (249+/-197, 241+/-145, 139+/-22% of N.) Mitral regurgitation decreased to zero or grade I in all patients partially due to our positive indication of mitral annuloplasty. Significant pulmonary artery stenosis occurred in 3 patients due to shrinkage of the equine pericardium used to repair the defects of the main pulmonary trunk. This significant complication could be avoided by using autologous pericardium. The Takeuchi procedure and concomitant mitral annuloplasty provided satisfactory results.

123
Outcome after repair of tetralogy of Fallot with absent left pulmonary artery

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Tetralogy of Fallot (TOF) with absent LPA forms an important subgroup of patients undergoing surgery. Early closure of the PDA in the neonatal period is implicated in perhaps off the LPA origin and causing it to remain hypoplastic or isolated. Thus the repair entails perfusion to only one (usually right) lung after surgery and a homograft is often inserted during repair to provide a competent pulmonary valve. Fourteen pts. with TOF and absent pulmonary LPA, who underwent repair in the last 10 years were included in this study. Their age ranged from 12 –84 months (median 35 mo) and weight 5.25–27 Kg (median 14 Kg). There were 8 males and 6 females. Mean pre-op saturation was 75%, and 8 had a history of spells. Only 6/14 had homografts used for repair and the rest had a monocusp valve placed in the RVOT. The mean CPB time was 122.2+ 25.1 min, cross clamp time 57+14.2 min. Average ICU stay was 3 days and ventilation duration was 28.4 hours. Twelve patients survived, with follow-up available for 3–74 months. Lung perfusion scan done in 6 pts. revealed a mean of 42% perfusion to Rt. and 58% to Lt. lung [via bronchopulmonary circulation]. All pts. were asymptomatic on follow-up. Conclusion: Surgery for TOF with absent LPA has excellent results. Addition of a homograft does not alter outcome or symptoms on follow up.

124
Biventricular repair in patients with double outlet right ventricle with non-committed VSD

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Objective: To determine clinical problems after biventricular repair in patients with DORV with non-committed VSD. Methods: Intraventricular rerouting was carried out in 14 children with this particular feature of malformation. Body weight at operation was 8.2 ± 4.5 (3.0 – 14.0) kg. To construct the channel for the LV outflow tract without obstruction, VSD was enlarged in 10 (71%) and the septal septum was resected in 7 (50%). An external conduit was placed for reconstruction of the RV outflow tract in 2. Results: Three patients, one of whom weighed less than 4kg, died immediately after repair because of low cardiac output. RA pressure after coming off bypass was greater than 10mmHg in these patients. This reflected loss of RV volume produced by the presence of the intraventricular patch. Of the other 11 operative survivors, tricuspid regurgitation has become moderate or severe in 5. Reoperation has been thus far needed in 5 for treating obstruction across the LV outflow tract (in 3), tricuspid regurgitation (in 2), obstruction at the pulmonary conduit (in 2), and residual interventricular communication (in 1). The preoperative presence of the markedly developed outlet septum, seen in 6, was a factor unfavorably associated with operative death (in 2) and reoperation for the obstructed LV outflow tract (in 3). Conclusions: Biventricular repair of DORV with non-committed VSD can be justifiably achieved unless body weight of the patients is less than 4kg and the outlet septum is markedly present. Attention should be paid to postoperative tricuspid regurgitation.

125
Long-term results of aortic valve regurgitation after repair of ruptured sinus of Valsalva aneurysm

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Objectives: We have reviewed our 35 year-experience to investigate the determinants of long-term results of aortic valve regurgitation (AR) after repair of ruptured sinus of Valsalva aneurysm (RSVA). Methods: Thirty three patients aged 7 to 64 underwent surgery for RSVA. The aneurysm ruptured into the right ventricle (n=27), the right atrium (n=5) and the left atrium (n=1). Ventricular septal defect (VSD) was noted in 12 patients. The combined approach through the aortotomy and the involved chamber was used in 24. Either a direct (n=17) or a patch closure (n=16) was performed to close the rupture hole. AR was graded 0–IV by angiographic and/or echographic evaluations. Results: Preoperative AR was noted in 8 (27%) with Grade-I in 6 and Grade-II in 2. In patients with Grade-II, AR remained the same after operation but this later deteriorated to Grade-III resulting in one

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

patient requiring reoperation 10 years later. Newly developed AR was noted in 4 (19%) soon after operation, including Grade I in 3 and Grade-II in 1. A patient with Grade-II AR had the increased AR to Grade-III, requiring valve replacement 22 years later. Late AR was associated with preoperative and early postoperative AR ($p<0.05$) but not with the presence of VSD, location of the fistula, surgical approach, type of repair (direct versus patch). Conclusion: Long-term follow-ups are required since the deterioration of the AR is gradual. Aortic valve repair needs to be considered at primary operation when AR is more than Grade-II. A significant AR should not be remained to prevent late death.

126
Influence of congenital heart disease on survival in patients with congenital diaphragmatic hernia
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Congenital diaphragmatic hernia (CDH) is frequently associated with congenital heart disease and this combination has been considered lethal. The lung-to-head ratio (LHR) is a measure of severity of pulmonary hypoplasia and is a good prenatal predictor of survival for isolated CDH. Good outcome is associated with a LHR>1.4. Our purpose was to assess outcome in CDH pts with heart disease and to see if LHR could help predict outcome in these pts. We reviewed our surgical database for pts referred with a diagnosis of CDH from 4/86-10/00. Of 172 pts, 31(18%) had heart defects. Cardiac lesions included VSD(9), coarctation(4), HLHS(4), TOF(4), AVSD with arch hypoplasia(2), TGA(2), and other(6). Eight had additional anomalies. Of the 31, 7 had fetal demise, 13 had neonatal demise and 6 died after intervention for an overall mortality of 84%(95%CI: 70 %-94 %). The hazard of death from birth to last F/u was 2.9 times higher for those with heart defects($p<0.0001$). Seven of 11 survived hernia repair. The four deaths were attributed to pulmonary hypertension or ECMO complications. Subsequent cardiac repair was undertaken in five. One(HLHS) had ventricular hernia repair and Norwood operation. Another(VSD) underwent hernia repair only. Of these 7, 5 survived(71.4%) with a median f/u of 22 months(range 8-54). LHR was measured in 8/11 pts who had hernia repair. All 3 survivors had LHR>1.2 and the five nonsurvivors had LHR<1.2 (Fisher's exact, $p=0.02$). The pt with HLHS survived Fontan. Conclusion: Heart disease remains a significant risk factor for death in pts with CDH. However, LHR helps predict survival in this high-risk group of pts. In the absence of severe pulmonary hypoplasia, treatment should not be denied in CDH pts with heart defects.

127 ✓
Evaluation of long-term surgical outcome after repair of complete atrio-ventricular septal defect associated with hypoplastic left lateral leaflet and single papillary muscle
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Dealing with left atrio-ventricular valve (LAVV) of common atrio-ventricular canal (CAVC) with hypoplastic left lateral (mural) leaflet, with or without single papillary muscle, is a technical challenge. The closure of the cleft can cause inflow obstruction; and leaving it open may lead to residual or recurrent regurgitation. Method: This retrospective study evaluates the long-term outcome of 36 such patients who underwent complete repair between 1980 and September 2000. Result: Out of 403 CAVC patients who underwent repair, 36(9%, CI: 6.5 to 12.5%) had this anatomy. The median age was 4.4 months (0.1 to 46) and median weight 4.6 (2.9 to 20) Kg. All of them underwent complete repair with the double patch technique. As primary repair, we elected to leave the cleft open. There were 3(8.3%, CI: 2 to 23.%) early deaths. Out of these 3, two died of persistent severe LAVVR despite immediate revision and support of ventricular assist device. In the main series there were 2(5.6%, CI: 1.5 to 18.5%) early deaths ($p=0.13$). Mean early post-operative LAVVR was mild (2.19, CI: 1.9 to 2.5). At median follow-up of 33 months (1 to 169), there was one late death and 11 re-operations in 6 patients (18%, CI: 8 to 36%) including one LAVV replacement. Late LAVVR was more than mild in 20 patients and moderate in 8. Actuarial freedom from death or re-operation was 60 % at 45 months with 12 patients at risk. We conclude that the repair of this anatomy of CAVC is complex. It compares favourably to the regular CAVC anatomy for hospital mortality. However, significant residual or recurrent LAVVR is often present, and it is responsible for a very high rate of re-operation.

128 ✓
Is there a predictive factor for the long-term tolerance of bidirectional cavo-pulmonary connection?
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Some patients seem to tolerate the bidirectional cavo-pulmonary connection (BCPC) circulation longer than others do. Our policy for age of completion of Fontan circulation after BCPC is evolving from elective to symptom related. This retrospective study is to identify markers for a better tolerance of the BCPC circulation. The data of all survivors who had their BCPC between 1986 and 1996, were analyzed, excluding those who had a potential contraindication for the Fontan completion. The series was divided into two groups: those who had their completion earlier than 36 months after the BCPC and the ones completed later than 36 months or are still living with their BCPC. Contingency tables and non-parametric tests were used. Sixty-six patients are included in the series. Thirty-one have been converted before and 35 after 36 months. The groups are of similar age, sex and weight at the time of BCPC creation, and are identical for anatomical data. The lesions analysed included tricuspid atresia, HLHS, isomerism, presence of two ventricles, dominant right or left ventricle, common AV valve, presence of azygos continuation. Physiological data at the time of follow up or Fontan completion also were identical. The following data were analyzed: presence of additional pulmonary flow, presence of cavo-caval collateral circulation, presence of pulmonary AV fistula, degree of AV valve regurgitation and arterial saturations. The creation of BCPC before 1993 was the only significant marker ($p=0.0089$). We conclude that the timing of Fontan is generally subjective and no marker in this study can identify which patients would tolerate long-term BCPC. Moreover, the patients who had their Fontan completed early were comparable at the time of completion to those who are still living under the BCPC regimen.

129
Total Cavo-Pulmonary Connection without cardiopulmonary bypass
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Total Cavo-Pulmonary Connection (TCPC) has been extensively applied to patients with single ventricle physiology. Complications could be related to the effects of cardiopulmonary bypass (CPB). The aim of this study is the realization of TCPC without CPB. Since June 1998, 9 patients were submitted to TCPC without CPB: 7 were males, age ranged from 4 to 17 years (m = 8.1 yrs) and body weight from 15.3 to 37.8 Kg (m = 25.5Kg). Previous operation were bidirectional Glenn anastomosis (2), Glenn and atrisbepticectomy (1). Glenn and PA-banding (1), modified Blalock-Taussig shunt (2) and PA-banding (1). After total heparinization, 2 cannulas were placed between each Vena Cava and the right atrium. A Glenn anastomosis was performed in those patients without previous one and then, an extracardiac conduit was placed between IVC and R.P.A (all patients). The conduit size ranged from 14 to 22 mm (m = 17.1mm). There was one hospital death due to multiple organ failure (11.1%). Early postoperative complications were pleural effusion (2), pericardial effusion (2), embolic cerebral vascular accident (1) and pulmonary infection (1). The median ICU stay was 2.5 days and median hospitalization time was 10.7 days. Follow up ranged from 1 to 25 months (m = 7 months) and all survivors are in functional class I, without signs of venous obstruction on bidimensional echocardiograms. Despite the small number of patients, TCPC without CPB is clinically possible with results comparable to those with CPB.

Session 13: Cardiomyopathies/Myocarditis/Heart Failure

130
Treatment for cytomegaloviral carditis
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In order to improve the prognosis of cardiac, caused by Cytomegaloviral (CMV) we investigated efficacy of interferon therapy. 19 children aged from 2 till 14 years old with CMV cardiac (everybody had diagnostic titer of CMV IgG and IgM and hadn't such titer of any others viral antibodies) and

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

increased Resistant Index of Celiac trunk, that was one of subtests of Low Output heart failure, were treated by well-known actions, Gancidovir in age doses and interferon in order to increase cytotoxic immunity and intensify CMV elimination from cells. We used α2b-interferon 'Viferon' in rectal suppositories and Cicloferon in intramuscular injection by determined scheme. Course of Viferon lasted 2 months and Cicloferon – 3,5 months with intervals. Results: 15 children (79%) had not recidivation of carditis during 2 years, 4 children (21%) had 1 or 2 recidivations this period. Conclusion: interferon therapy has good efficacy in treatment for carditis caused by persistence Cytomegalovirus infection.

131

Dobutamine stress echocardiography in the evaluation of cardiac function

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Dobutamine stress echocardiography has been performed in cancer children treated with anthracyclines chemotherapy. The study was designed to examine whether dobutamine stress echocardiography may make earlier and more accurate detection of cardiac dysfunction possible than standard cardiac function tests in doxorubicin-treated patients. Consecutively, 38 patients (24 boys and 14 girls ; who had previously received doxorubicin containing chemotherapy were enrolled in this study. In addition, age-matched, 13 healthy controls were also recruited. Dobutamine was infused at rates of 2.5 to 5 μg/kg/min and echocardiographic measurements were obtained at rest and at the end of dobutamine infusion. Of the 38 patients, 2 patients had left ventricular dysfunction as assessed by echocardiography at rest. In addition, 2 of 34 patients with normal cardiac function at rest showed an abnormal response to dobutamine, compared with healthy controls. Left ventricular ejection fraction and fractional shortening in doxorubicin treated patients were not different from those in control subjects at rest but demonstrated significantly lower values ($p<0.05$) after 5 μg/kg/min dobutamine infusion. Left ventricular end-systolic interventricular wall stress was significantly elevated ($p<0.01$) after both 2.5 and 5 μg/kg/min dobutamine infusion in doxorubicin-treated patients compared with the mean values in control subjects. Moreover, dobutamine stress echocardiography revealed abnormal correlation between rate corrected mean velocity of circumferential fiber shortening and end-systolic wall stress in doxorubicin treated patients. No significant differences were found in diastolic function between control subjects and doxorubicin treated patients. In conclusion, Dobutamine stress echocardiography is a very sensitive method to detect subclinical cardiac dysfunction in patients receiving anthracyclines.

132

Influence of left ventricular hypertrophy on qt dispersion and rate-corrected qt interval in children with hypertrophic cardiomyopathy

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The aim of the study was to assess the influence of left ventricular hypertrophy (LVH) on QT dispersion (QTd) and rate-corrected QT interval (QTc) in 21 children (age: 14.2±4.6 years) with hypertrophic cardiomyopathy (HC). The control group consisted of 24 healthy children (14.3±4.4 years). All patients underwent standard ECG (50 mm/s speed), 48-h ambulatory ECG monitoring and echocardiography. Left ventricular mass (LVM) was calculated using the area length formula and LVM was related to body surface (LVMI). Results: Ventricular arrhythmias (VA) were recorded in 6 (28%) patients with HC: 4 patients (19%) had ventricular pairs or runs of ventricular tachycardia (VT); QTc interval (439±4 ms) and QTd (40±16 ms) were significantly greater in children with HC than in controls (384±3 ms, 24±8 ms, respectively) ($p<0.005$). QTc > 440 ms was found in 6 (28%) patients with HC. No patients in the control group had QTc > 440 ms. No significant association was found between QTc and VA and LVH. There was a significant positive correlation between QTd and LVM ($r=0.465$, $p<0.0001$), LVMI ($r=0.536$, $p<0.0005$) and left ventricular posterior wall thickness (LVPW, $r=0.729$, $p<0.00001$). A marked increase in QTd was recorded in children with HC and VA compared with those without VA (55±19 ms vs 30±10 ms, $p<0.04$). QTd > 60 ms was observed only in children with HC and hazardous VA (VT and ventricular pairs). Conclusions: Children with HC have prolonged QTc interval and increased QTd. Increase of LVM and LVPW thickness is associated with increasing of QTd in children with HC. Increase of QTd may be one of the predisposing conditions for development of VA in these patients.

133

Discriminative ability of standard echocardiography and tissue doppler imaging techniques for the detection of effects of treatment with anthracyclines

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The purpose of this study was to determine retrospectively the best set of standard echocardiographic- and/or tissue Doppler imaging (TDI) parameters needed for discrimination of survivors of childhood cancer treated with anthracyclines from healthy controls. Previous studies have shown that TDI has the ability to detect regional effects on myocardial velocities. The study comprised 159 subjects (60 patients, 99 volunteers); age range 8.5–17.6 yr. The survivors received 50–420 mg/m2 cumulative dose of anthracyclines, with a mean follow-up of 7.05 (±2.3) yr. All underwent standard echocardiographic studies of blood flow velocities and ventricular dimensions, followed by measurements of systolic and diastolic peak myocardial velocities using TDI technique from long-axis and apical 4-chamber views. The parameters used in the multivariate discriminant score (S-score) were selected from a large set of 51 parameters using stepwise selection (significance level $p=0.05$). The S-score and receiver classification probability (C-index) were used to measure the overall discriminative performance of conventional and TDI techniques separately and in combination. The overall discriminative performance of the standard echo-Doppler parameters (C = 0.77) was lower than that of the TDI (C=0.84). The highest C-index was obtained using both techniques (C=0.89). The best set of parameters includes: LV fractional shortening and MV flow velocity, 2 long-axis and 5 apical 4-CV TDI wall velocities (systolic and diastolic). In the patients group, the index score S was positively associated with cumulative dose of anthracyclines ($p=0.004$). However, there was no association with age at start of chemotherapy or with duration of follow-up. In conclusion, standard echocardiography should be used in combination with the new TDI technique for the detection of effects of anthracyclines on the myocard.

134

Cardiomyopathy in children with mitochondrial disease

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Mutations within the genes of the respiratory chain may result in defects of oxidative phosphorylation. Energy-dependent organs e.g. the brain, heart and skeletal muscle are particularly vulnerable to defects in energy metabolism. Encephalomyopathy and cardiomyopathy therefore represent common manifestations of mitochondrial disease. AIMS: To make an inventory of the occurrence of cardiomyopathy in children with mitochondrial disease and to describe their clinical course and cardiological manifestations. METHODS: 300 children with CNS and neuromuscular disease combined with hyperlactatemia were referred to our institution between 1984 and 1999. 106 had mitochondrial myopathy based on morphological and biochemical investigations of skeletal muscle biopsies. Diagnosis of cardiomyopathy was based on ECG and echodoppler. RESULTS: Seventeen patients had cardiomyopathy, all hypertrophic non-obstructive. Onset of symptomatic mitochondrial disease ranged from birth to 10 years of age. 8/17 children had cytochrome c oxidase (COX) deficiency, while the remaining 9 had various defects. The diagnosis of cardiomyopathy was made from birth to 27 years. Left ventricular (LV) posterior wall and septal thickness were both increased with z-scores of +4.6±2.6 and +4.3±1.6 (mean±SD) respectively. LV diastolic diameter z-score, +1.3±3.4, and fractional shortening (FS), 24±13% displayed marked variation. 7 children had mitral regurgitation. 6 of these had FS<17%. 9/17 children had abnormal ECG with pre excitation in 3, supraventricular arrythmia in 4 and right bundle branch block in 5. Nine patients developed heart failure (age: 3 days–27 years). 12/17 children with cardiomyopathy died or underwent heart transplantation including all 8 children with COX deficiency. Mortality was higher than in children without cardiomyopathy (71 vs 25%, $p<0.001$). CONCLUSIONS: In children with mitochondrial disease, cardiomyopathy was common (17%) and associated with increased mortality. The prognosis for children with COX deficiency and cardiomyopathy seemed particularly unfavourable.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

135

Idebenone reduces cardiac hypertrophy in friedreich's ataxia

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Background Friedreich ataxia (FRDA) is an autosomal recessive neurodegenerative disease causing limb and gait ataxia and hypertrophic cardiomyopathy. The disease gene encodes a protein of unknown function, frataxin. The loss of frataxin is due to a large GAA trinucleotide expansion in the first intron of the gene and causes an oxidative stress with the combined deficiency of a Krebs cycle enzyme, aconitase, and three mitochondrial respiratory chain complexes (I-III). Idebenone, a short-chain quinone, may act as a potent free radical scavenger and protect heart muscle against oxidative stress. We have carried out an open trial of idebenone in a large series of FRDA patients and followed their left ventricular mass and function. Methods A series of 28 FRDA patients aged 4–22 years (20 boys, 18 girls) were given idebenone orally for six months (5 mg/kg/day). Heart ultrasound parameters were recorded on the same scanner prior to and after six months of oral idebenone. Findings After six months, heart ultrasound revealed a more than 25% reduction of left ventricular mass in 50% of patients ($p<0.001$). Heart hypertrophy was largely stabilized in the other half patients. A reduced shortening fraction (11–26%) was originally observed in 6/28 subjects and improved in 5/6 individuals. In one patient, the shortening fraction only responded to 10 mg/kg/day of idebenone. No correlation between responsiveness to idebenone and age, sex, initial ultrasound parameters or number of GAA repeats in the frataxin gene could be found. Interpretation This study demonstrates the efficacy of idebenone in controlling heart hypertrophy in FRDA. Owing to the absence of side effects of the drug, we suggest giving idebenone (5–10 mg/kg/day) continuously to FRDA patients at onset of hypertrophic cardiomyopathy and even preventively prior to heart involvement.

136

Effective and safe therapy with coenzyme Q10 for idiopathic dilated cardiomyopathy

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It has been reported that myocardial mitochondrial failure can be improved by the administration of coenzyme Q10(CoQ10) in adults, but there is no report about it's therapeutic effect in children. CoQ10 has been shown to be deficient in myocardial tissue biopsies taken from dilated cardiomyopathy (DCM) hearts compared to normal hearts. CoQ10 (ubiquinone) is a mitochondrial utilized by mitochondria in electron transport within the respiratory chain and thereby has a significant effect upon oxidative phosphorylation as well as intramyocardial energy provision. Therefore, we evaluated the clinical and functional responses to CoQ10 therapy in children with DCM. We report three cases aged 4, 6 and 12 months each with a clinical picture of congestive heart failure. Echocardiographic examination revealed left ventricular dilatation and decreased systolic function. Metabolic, biochemical and viral investigations were found to be negative, and were diagnosed as idiopathic DCM. After 1, 1, and 8 months respectively of conservative therapy with digoxin, diuretics and ACE inhibitors, they were given CoQ10 (30mg/die) orally. The clinical symptomatology improved with improvements of NYHA class from class III to I in all cases. Between the time point of stabilization on conventional medications and the initiation of CoQ10 therapy, there was no significant change in fractional shortening (0.14, 0.21, 0.13 vs 0.11, 0.20, 0.20) or ejection fraction (0.31, 0.49, 0.30 vs 0.25, 0.48, 0.48). However, after CoQ10 therapy for 6 months there was a significant increase in fractional shortening (0.30, 0.37, 0.36) and ejection fraction (0.62, 0.74, 0.75). The improved cardiac function show that therapy with CoQ10 is remarkably beneficial due to correction of CoQ10 deficiency in DCM. It seems to be safe and and effective for our three cases of DCM. Our study is going on to evaluate CoQ10 therapy in more number of cases to prove its beneficial effect.

137

Risk factors for disease-related death in childhood hypertrophic cardiomyopathy

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Objectives. To establish clinical features associated with increased risk of disease-related death in childhood hypertrophic cardiomyopathy (HCM). Methods. Retrospective cohort study from six regional centres of paediatric cardiology with a geographical basis for referral. There were 122 patients with HCM presenting at age <19, mean follow-up 9.9 years, with 27 deaths. Multivariate analysis was used to correlate ECG- and echocardiographic measures with risk of disease-related death. Results. Echocardiographic features at presentation showing positive correlation with risk of disease-related death were LV wall-to-cavity ratio ($p=0.0008$) and presence of LV outflow tract gradient ($p=0.002$). Rate of increase in septal thickness ($p=0.0002$) and LV wall thickness ($p=0.0002$) correlated positively. Consequently, low septum-to-cavity ratio ($p=0.002$), last LV wall-to-cavity ratio ($p=0.006$), low septal thickness ($p=0.007$), last LV wall thickness ($p=0.008$) and late left atrial enlargement ($p=0.008$) all correlated with risk of death. ECG features that correlated at presentation were Sokolow-Lyon index ($p=0.0008$) and total R–S sum in limb leads ($p=0.007$). Subsequent changes in ECG voltages were also correlated as was presence of frequent ventricular ectopics ($p=0.002$). Verapamil-dose and myectomy did not affect risk of death. Propranolol was protective with an inverse relation between propranolol dose and risk of death ($p=0.001$). Risk of sudden death was correlated with septal thickness, both at presentation ($p=0.001$) and at late follow up ($p=0.0004$), late LV wall thickness ($p=0.004$) and ECG voltages (R-S sum $p=0.0003$, Sokolow-Lyon index $p=0.0002$). The best predictors for heart-failure related death were the severity of relative hypertrophy at presentation expressed as septum-to-cavity ratio ($p=0.001$) and LV wall-to-cavity ratio ($p=0.0003$); ECG voltages did not correlate. Conclusion. ECG and echocardiographic features can be used for risk stratification of childhood patients with HCM. One treatment agents, high-dose propranolol (>4.5 mg/kg), reduces the risk of death.

138

Alpha-Dystrobrevin mutations in nonisolated left ventricular noncompaction and evidence for genetic heterogeneity

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Left ventricular noncompaction (LVNC), a form of cardiomyopathy presents in infancy with a hypertrophic and dilated left ventricle with deep trabeculations and commonly with reduced systolic function. Deficient of the FK binding protein 12 (FKBP12) gene result in non-isolated LVNC associated with congenital heart disease in mice. Mutations in the gene G4.5, which maps to chromosome Xq28, have been described in patients with isolated LVNC, suggesting that LVNC and Barth syndrome (X linked disorder also associated with dilated cardiomyopathy) are allelic. Female patients with LVNC, however, have been also reported suggesting that X-linked inheritance in some instances (Ichida et al. J Am Coll Cardiol 1999). We analyzed the patients with non-isolated LVNC for a series of candidate genes selected using the final common pathway hypothesis.

139

Efficacy of carvedilol in management of congestive heart failure in infants with dilated cardiomyopathy

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Carvedilol is a non selective beta blocker with alpha receptor blocking properties and has been shown to be efficacious in the management of congestive heart failure (CHF) in adults with reduced ventricular function. Experience in children and especially infants is limited. The purpose of this study was to assess the efficacy and safety of carvedilol in symptomatic infants with dilated cardiomyopathy (DCM) not responding to conventional anti-congestive therapy and afterload reduction. METHODS: From 12/98 to present infants < 2 years presenting with DCM were assessed. Those who showed clinical signs of CHF on adequate doses of digoxin, diuretics and ACE inhibitors and had echocardiographic estimation of ejection fraction (EF) < 30% were included in the study. Echocardiographic assessments of EF, fractional shortening (FS), mitral regurgitation, electrocardiograms (ECG), and blood chemistries were performed prior to starting carvedilol and at last follow up. Holter monitoring was performed on 7 patients. A symptom score was also given to each patient to assess functional status. Carvedilol was initiated in the hospital at 0.1 mg/kg/day orally divided in two doses and titrated up to 1 mg/kg/day with continuation of conventional anti-CHF therapy. RESULTS: 10 patients (5 males), age 13+/-6.8 months, weight 6.9+/-1.8 kg were identified. Over a follow up period of 10 +/- 2.5 months, EF (%) increased from 24+/-5% to 40+/-11% ($p<0.05$), SF (%) increased from

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

12.24+/-6.6% to 18+/-7% (p=NS). Seven patients had an average weight gain of 8% (range: 1–20%). One patient had significant decrease in ventricular ectopy. Seven patients became completely asymptomatic. There was one mortality. There were no complications related to carvedilol therapy. CONCLUSION: Carvedilol is well tolerated in referred infants with DCM resulting in significant improvement in functional status and EF (%).

140
Malignant mutations in hypertrophic cardiomyopathy: a rare find indeed

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Background: Hypertrophic cardiomyopathy (HCM) is a genotypically and phenotypically diverse disease involving the cardiac sarcomere. Previous genotype-phenotype studies have identified three mutations (R403Q, R453C, and R719W/Q) as highly malignant defects within the most common HCM-predisposing gene, beta myosin heavy chain. Routine clinical screening for these malignant mutations has been suggested to identify high-risk families. Methods: We screened 249 near-consecutive, unrelated pts (118 female) of HCM seen at Mayo Medical Center's HCM Clinic during a 3 year period from April 1997 to April 2000. DNA was obtained following informed consent. PCR amplification of exons encoding R403Q (exon 17), R453C (14), and R719W/Q (19) was performed, and the mutations were detected using mutation specific restriction enzyme assays and denaturing high performance liquid chromatography. Results: The mean age at diagnosis of HCM was 41 years with 53 subjects diagnosed before age 25. 127 pts (51%) had evidence for obstruction with a mean peak resting gradient of 67 mm Hg. The mean maximal wall thickness was 22 mm. 83 pts (33%) were familial. There was a family history of sudden death in 54 (22%). Only 2 of the 249 (0.8%) had a malignant mutation. Conclusion: This finding underscores the profound genetic heterogeneity in HCM. Less than 1% of unrelated individuals seen at a tertiary referral center for HCM possessed a malignant

141
Nitric oxide inhibits apoptotic enzyme activity in a genetic model of cardiomyopathy: Implications for reversal of ventricular remodeling in heart failure

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We have been involved with several aspects of von Willebrand factor (vWF) in the context of pulmonary hypertension. Since finding have potential pathophysiological and clinical implications we planned this report as an attempt to summarize observations. Studies involved 52 patients (37 female) aged 3.2 to 50 (median 20) years with primary (PPH, 12 pts) or secondary (SPH, 40 pts) precapillary pulmonary hypertension including 25 associations with congenital heart disease (CHD-PH). Analysis of vWF included measurements of plasma antigenic (vWF:Ag) and biological (ristocetin cofactor) activities and assessment of multimeric as well as subunit structure (Western blotting). vWF:Ag was increased in patients vs controls ($p<0.001$) with exceedingly high levels in PPH ($p=0.003$ vs. SPH) but unrelated to age or gender group. In contrast, biological activity was decreased in patients as a result of defects in the multimeric structure ($p=0.004$). Besides, hypoxia was associated with heightened vWF:Ag ($p=0.014$). Improvement of arterial oxygen saturation was followed by a decrease in vWF:Ag levels and partial correction of multimeric defects. Also, bleeding of hematomas by means of hemodialysis in patients with Eisenmenger syndrome (CHD-PH) had no effect on arterial oxygen saturation, but did provoke a significant reduction in vWF:Ag levels ($p=0.021$), albeit structural abnormalities persisted. Subunit analysis showed that altered multimeric structure was mainly due to increased proteolytic degradation of vWF in vivo ($p=0.032$) probably associated with decreased sialic acid content of carbohydrate components ($p<0.05$). Finally, high vWF:Ag levels ($p=0.0087$) and the multimeric abnormalities of vWF ($p=0.0206$) were significantly correlated with decreased one-year survival in both PPH and CHD-PH groups. Thus, vWF is involved in a complex network of pathophysiological phenomena in pulmonary hypertension, including endothelial dysfunction, hypoxia, hyperviscosity and proteolysis. Abnormalities which have impact on short-term prognosis may be used as indexes for therapeutical decisions.

Session 14: Epidemiology/Outcomes Research

142
Evaluation of precursors of atherosclerosis in children aged ten to fifteen years – Yugoslav study

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The aim of this study was to determine serum lipides level and other non lipid risk factors for atherosclerosis. Method: This 5 years cohort study began in 1995 and follows risk for development of coronary heart disease (CHD) on 5249 Yugoslav children (2675 girls and 2574 boys) – YUSAD study. We measured total serum cholesterol level (TSHC), LDL-cholesterol levels (LDL-HL), HDL-cholesterol levels (HDL-HL), serum triglyceride levels (STL), apo-A1, apo-A11, apo-B, glucose and fibrinogen level, blood pressure (BP) and calculated atherosclerosis index and body mass index (BMI). Conclusion: Girls have average systolic BP of 106.00 +/- 0.24 mmHg, average diastolic BP of 68.69 +/- 0.12 mmHg and higher serum LDL-HL, STL and apo-B levels than boys. Boys have average systolic BP of 109.85 +/- 0.26 mmHg, average diastolic BP of 69.89 +/- 0.44 mmHg and significantly higher percent of preferable LDL-HL comparing to girls: 68.9% vs. 57.8%, same as preferable apo-B serum levels: 92.4% to 87.9%.

143
Delayed neuro-developmental outcome in infants with transposition of the great arteries (TGA) after arterial switch operation (ASO) observed by localized in vivo spectroscopy (1h-mrs) and bayley scales of infant development II (BSID II)

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Introduction: Abnormal neuro-developmental outcome has often been reported in children with TGA after corrective surgery. Underlying mechanism of these abnormal results is thought to be multifactorial. The purpose of this study was to evaluate the cerebral metabolism of infants with TGA before and after ASO by 1H-MRS and the neuro-developmental testing at 1 year by BSID II. Methods: 1H-MRS was done before ASO on 30 full-term neonates with TGA 2–18 days after birth. Follow up MRS and BSID II were performed at 11–13 months after ASO. Localized 1H-MRS was performed on the parietal white matter (PWM) and occipital gray matter (OGM) of the brain to calculate the [NAA/Cr], [Cho/Cr], [mI/Cr] and [NAA/Cho] metabolites ratios. The age-matched normal full-term neonates (N= 15) and infants (N= 30) were included for comparison. Results: The [Cho/Cr] in PWM before ASO (1.44 ± 0.14 vs. 1.24 ± 0.12) and after the surgery (1.08 ± 0.15 vs. 0.83 ± 0.10) were higher and the [NAA/Cho] (0.61 ± 0.08 vs. 0.79 ± 0.10) in OGM was lower before surgery for the TGA infants than for the normals. The results of BSID II showed that 7 showed mild or severe mental and moderate language developmental delays, and 4 showed moderate motor developmental delay. Conclusion: The low NAA and high Choline levels, observed within a few days after birth and at 1 year indicate that the cerebral metabolism has already been damaged prior to surgery and was not normalized by 1 year. The results of BSID II were in concordance with those of 1H-MRS, suggesting that the abnormal hemodynamics of TGA in the fetal life might well have an effect on neuro development.

144
Evaluation of cerebral metabolism for children undergoing open heart surgery for closure of atrial septal defect (ASD) under cardiopulmonary bypass by localized in vivo 1h mr spectroscopy (1h-mrs)

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Introduction: The adverse effects of open heart surgery (OHS) under deep hypothermia and circulatory arrest in neonates and infants have been reported. A recent study reported an abnormal developmental outcome after surgical closure of atrial septal defect (ASD) under CPB as compared to non-surgical device closure of ASD. In this study, we investigated whether a brief CPB under mild hypothermia and normal flow would have any deleterious effects on the brain metabolism. Methods: Seven children (age = 18

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

= 47 months) undergoing surgical closure of ASD had 1H-MRS examinations shortly before and 2 months after the operation. Results from ASD patients were compared with those from the age-matched normal children. Localized 1H-MR Spectroscopy was performed on the parietal white matter (PWM) and occipital gray matter (OGM) of the brain to calculate the values of the [NAA/Cr], [Cho/Cr], [mI/Cr] and [NAA/Cho] metabolites ratios. Results: The metabolites ratios measured by 1H-MRS for patients with ASD before and after surgery were not significantly different although significantly different from those for normal children: the elevated [Cho/Cr] ratio was observed for ASD children (1.05 ± 0.16 vs. 0.81 ± 0.12, p<0.05). Data from two patients who also had MRS examination 3 days after surgery were not different when compared with preoperative data. Conclusion: The results of this study indicate that CHS under mild hypothermia and normal flow did not affect the cerebral metabolism. An additional observation from 2 patients that cerebral metabolites measured 3 days after CHS were almost the same as preoperative values further indicates that CHS performed under this condition did not significantly alter brain metabolites.

145

Long-term survival in patients with repair of tetralogy of Fallot: a multivariate analysis of risk factors for late death

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BACKGROUND: The risk of sudden death after surgical repair for tetralogy of Fallot is 25 to 100 times greater than in an age matched normal population but risk factors of cardiac and sudden death are not well identified. OBJECTIVE: This study sought to analyze risk factors of late cardiac and sudden death after surgical repair for tetralogy of Fallot. METHOD: We reviewed our experience from 1971 to 1995 with 306 patients retrospectively. Cox regression was used to investigate potential risk factors. The variables included age and weight at operation, with or without pulmonary atresia, palliative operation, transannular patch, aortic cross clamp time, postoperative right ventricular systolic pressure, main pulmonary artery systolic pressure, pressure gradient of right ventricular outflow tract, ratio of systolic pressure in the right ventricle to that in the left ventricle (RV/LV), complete right bundle branch block (CRBBB) and duration of QRS complex. RESULT: There were 16 late deaths (5.4%). The 25-year survival rate was 94.9%. Cardiac death occurred in 6 patients (2.0%) including 4 sudden deaths (1.3%). The risk factor for cardiac death was high RV/LV (Odds ratio: 20.1, p=0.0008) especially, RV/LV>=0.7 (11.5, p=0.0257), while that for sudden death was pulmonary atresia (17.5, p=0.0069). CRBBB and duration of QRS complex showed no influence. No significant risk factor of sudden death was detected in cases excluding pulmonary atresia (n=272). CONCLUSION: Reducing RV/LV is necessary for prevention of late cardiac death. Association of pulmonary atresia is a risk factor for sudden death.

146

Congenital heart defects, maternal febrile illness, and multivitamin use: a population-based study

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The relation between febrile illness during pregnancy and cardiac defects in the offspring was assessed in a population-based case-control study in metropolitan Atlanta. Case-infants (905) with cardiac defects were actively ascertained from multiple sources. The control-infants (3,029) were infants without birth defects who were selected from birth certificates by stratified random sampling. Reported maternal febrile illness, from one month before pregnancy through the third month of pregnancy, was compared with no fever or infection during the same period. Maternal febrile illness was associated with an increased risk for heart defects in the offspring (odds ratio [OR] 1.8; 95% confidence interval [CI] 1.4–2.4). When influenza-like illness was the source of the fever, the relative risk was 2.1 (95% CI 0.8–5.5). The risk associated with febrile illness was strongest for tricuspid atresia (OR 5.2), left obstructive defects (OR 2.7), transposition of the great arteries (OR 2.9), and ventricular septal defects (OR 1.8). These risks were generally lower among mothers who used multivitamins during the periconceptional period than among mothers who did not use multivitamins during the same period. For example, compared with no fever or infection and no multivitamin use, fever without multivitamin use was associated with a 2.3-fold increased risk for heart defects (95% CI, 1.5–3.5). However, fever with multivitamin use was associated with a risk that was similar to the reference group (OR 1.1, 95% CI, 0.6–2.2). If confirmed and causal, these findings suggest that febrile illness in human may be a significant and perhaps preventable cardiac teratogen in humans.

147

Does isolated pulmonary insufficiency impair exercise performance

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The clinical impact of pulmonary insufficiency (PI) remains unknown. While the impact of PI on exercise performance has been studied in patients with repaired Tetralogy of Fallot (ToF), results are often difficult to interpret because of residual RVobstruction or branch pulmonary artery abnormalities. We performed cardiopulmonary stress testing using cycle ergometer in 2 groups of children with PI. 17 children following isolated surgical pulmonary valvectomy and a matched group of 17 children with primary repair of ToF with transannular patch. All patients had free PI on echocardiogram and no discernible RV obstruction. Exercise variables were compared to age matched normal controls. There was no difference between the 2 surgical groups in age (12.3+/-2.8 [ToF] vs 12.1+/-2.9 years, p=.89), weight (47.5+/-10.1 [ToF] vs 51.4+/-12.0 kg, p=.37), BSA (1.4+/-.16 [ToF] vs 1.4+/-.19 m2, p=.31), age at surgery (1.9+/-1.5 [ToF] vs 1.4+/-2 years, p=.37) or follow-up interval (16.6 [7.2–18.5] [ToF] vs 16.5 [7.7–21.6] years, p=.14). Paired t-tests demonstrated that patients with isolated surgical pulmonary valvectomy had a lower peak oxygen consumption (VO2) (mean difference -3.1, p=.036) and a lower anaerobic threshold (AT) (mean difference -3.0, p=.009) when compared to matched patients who repaired ToF. There was no difference in heart rate, arrhythmia, or oxygen pulse. When compared to age & sex matched normal controls, both groups had a significant reduction in VO2 (mean differences -5.4, p=.003 [ToF] and -6.4, p=.0003 [valvectomy]) and AT (mean difference -3.0, p=.007 [ToF] and -6.5, p<.0001 [valvectomy]). Isolated pulmonary insufficiency has a detrimental effect on exercise performance. While both repaired ToF and valvectomy groups are limited patients with ToF have better aerobic capacity. Further study with assessment of RV compliance may prove to be of value.

148

Do households close to hazardous waste sites have a greater risk of congenital heart disease?

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Previous studies have shown that children born to mothers living close to hazardous waste sites have a greater risk for selected congenital malformations. The purpose of this study was to determine if there was an increased risk of congenital heart disease (CHD) associated with proximity to hazardous waste sites in Dallas County. Using data from a population-based study, we looked at 1030 CHD cases born to Dallas County from 1975–1988, and 3093 matched controls randomly selected from 1980 Dallas census data. A case control study design and Chi square analysis were used. Cases and controls were similar with respect to the sex of the child, maternal age and ethnicity. We used Environmental Protection Agency data to identify the locations of 440 hazardous waste sites and 2 National Priority List sites that were functional throughout the study period. Seventy four percent of all residential addresses and 68% of all hazardous waste sites were mapped to geographical coordinates using ArcView geographical software. We found an increased risk for congenital heart defects in association with maternal residence within 1 mile of hazardous waste sites [odds ratio (OR)=1.3, 95 % confidence interval (CI)=1.1–1.5, p<0.01]. There was also an increased risk with maternal residence within 1 mile of a National Priority List site [OR=3.1, 95 % CI=1.8–4.9, p<0.05]. We conclude that maternal residential proximity to hazardous waste sites was associated with an increased risk for heart defects in their offspring in Dallas County during our study period. These results have important implications regarding the relationship between environmental exposures and CHD. Future prospective studies are needed to identify the specific factors involved in the induction of CHD.

149

Developing a research tool to assess quality of life in children and adolescents with heart disease: preliminary data

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Each year approximately 1 million children are born worldwide with

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

congenital heart disease. Despite increased survival in this population there is no validated tool to assess the quality of life (QOL) of this group. The purpose of this project was to develop an instrument to assess the QOL of children and adolescents with congenital and acquired heart disease. The first step in the development process was to assemble focus groups (n=14) composed of 3 groups of children (age 8–12, n=26), 3 groups of adolescents (age 13–18, n=28), 3 groups of parents of children (n=29), 3 groups of parents of adolescents (n=25), 1 group of physicians (n=8), and 1 group of nurses (n=12) to ascertain what specific items were deemed most important to include in a QOL instrument. The most commonly identified items from children and parents of children groups were physical limitations and/or restrictions, receiving special treatment at school and/or at home, missing school for medical therapy and medication burden. In contrast, physicians and nurses identified physical limitations and/or restrictions, emotional distress from parental separation, pain from procedures, and difficulty for children to understand why they are ill. The most commonly identified items from adolescents and parents of adolescents groups were physical limitations and/or restrictions, feeling different from peers, overprotective parents, fear of dying, and medication burden. In contrast, physicians and nurses identified physical limitation or restriction, feeling different from peers, poor body image, loss of control/privacy, and social limitations. In summary, the focus group data revealed similarities in QOL items between patients and parents in both age categories. In contrast, physicians and nurses identify a different set of items than the patients and the parents of the patients. Caregivers should be aware of these differences when discussing QOL issues with patients and their families.

150

Hypertrophic cardiomyopathy in Noonan-syndrome: reevaluation, long-term follow-up and prognosis

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Objectives: Hypertrophic cardiomyopathy (HCM) is well known in Noonan syndrome(NS). The aim of this study was to determine the incidence, the time of onset, and long term course of HCM in NS. Methods: A uniform study protocol was established to confirm the diagnosis of NS including clinical criteria condensed from a metaanalysis of relevant papers. Due to the autosomal dominant genetics of NS the index patients were invited for clinical and cardial examination together with their families. Data of patients meanwhile deceased were included only in cases of doubtless diagnosis of NS. Under these conditions 124 patients were reevaluated (97 index patients, 30 mothers, 29 fathers, 11 of 26 brothers, 8 of 24 sisters, and data of 8 deceased). Results: Diagnosis of NS was made in 82 of 124 patients (47 male, 35 female). The follow-up time was 18.4 ± 8.2 years, the age at diagnosis 6.9 ± 5.8 years and the age at death 7.1 ± 5.5 years. At least one major cardiac lesion was found in 62 patients (76%). Congenital heart defects were: pulmonary stenosis in 35 (43%) patients (22 valvular, 7 supravalvular, 6 peripher), septal defects in 25 (28%) patients (10 ASD II, 8 perimembranous VSD, 5 AVSD), and HCM in 14 (17%) patients. Obstructive type of HCM (HOCM) was present in 9, noncobstructive type (HNCM) in 5 patients. Clinical symptoms due to HCM manifested in 5 patients during infancy. Development of HCM in NS later than 12 years of age was not found. Death occurred in 8 (10%) patients with NS, in 6 of them (75%) death was related to HCM. Conclusions: HCM is a frequent cardial lesion in NS. HCM in NS occurs usually in the first years of life and is associated with high mortality.

151

Influenza and parainfluenza type 3 virus infection in neonates with the cardiac arrhythmia

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The aim of this study was to assess the occurrence of influenza and parainfluenza type 3 virus (PIV 3) infection in neonates with cardiac arrhythmia. 70 neonates admitted to our department (1998 – 2000), due to heart arrhythmia, participated in this study. The level of antibodies in hemagglutination inhibition test against influenza A (subtypes H1N1 and H3N2), influenza B and PIV 3 was tested. In 25 cases immunofluorescence technique and embryonated egg culture (only for influenza) from the pharynx swabs were used. In 11(16%) of 70 neonates PIV 3 and in 26(37%) influenza viruses were found: in 8(11%) influenza A (H3N2), in 2 influenza A (H1N1), in 5(7%) influenza B. Two neonates were co-infected with A (H3N2) and B, 1 with all types influenza viruses and 1 with influenza B and PIV3. Among 11 neonates infected with PIV 3 ventricular arrhythmia (Ho Lown's scale) was observed in 5 and mild periodical bradycardia in 6 cases. Among 8 children infected only with influenza A (H3N2): in 2 ventricular arrhythmia (I and IIa Lowns scale), in 2 sinus tachycardia, in 1 supraventricular tachycardia and in 3 periodical mild bradycardia was diagnosed. In 2 children co-infected of influenza A(H3N2) and B supraventricular tachycardia was observed. 5 neonates infected with influenza B, 1 co-infected additionally with PIV 3 and 2 infected with A (H1N1) virus mild periodical bradycardia was diagnosed. In one neonate with co-infection of all 3 subtypes of influenza viruses ventricular arrhythmia (Ho Lown,s scale) was found. Conclusions. 1. Neonatal cardiac arrhythmia could be the consequence of influenza and parainfluenza type 3 viruses infections. 2. Infection with influenza A (H3N2) and co-infection with influenza A (H3N2) and B could be a risk factor of serious cardiac arrhythmia in neonates.

152

Length of stay for cardiac procedures from a multi-center pediatric cardiology consortium; a fifteen year experience

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Introduction: Pediatric Cardiac Care Consortium consist of 45 cardiac centers that submit data to a central registry at the University of Minnesota. Length of stay (LOS) has become an area of focused attention in the past several years due to managed care, capitated payments and competition among centers. We have previously reported a progressive decrease in surgical mortality for children over one month and one year of age. We hypothesize that LOS is decreasing over time, in a fashion similar to the mortality decrease. Methods: LOS is investigated for the PCCC patients enrolled from years 1982–1997 inclusive. 98,017 admissions for catheterization, surgery or both are considered. The study period is considered as early (1982–1986), mid (1987–1991) and late (1992–1997). The patients are grouped as less than one month of age, one month to one year, one year to 21 years. Results: LOS has decreased for children 1 – 12 month and 1 – 21 year through the 15 years of PCCC data studied. LOS for infants less than one month has been relatively stable. This was also previously seen for 30 day surgical mortality. The adjusted average length of stay in days for the various age groups for PCCC. The number in parentheses is the number of admissions averaged. Conclusion: LOS has improved over time. Neonatal LOS increases the overall average. Investigators to improve cardiac care should continue to focus on improving early solitary outcomes.

Session 15: Fetal Cardiology

153

Ventricular volume evaluation in the fetus with congenital heart disease using 3d echocardiography

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We examine the feasibility and appearance of 3D fetal echo assessment of ventricular volumes in 29 normal fetuses and 22 fetuses with congenital heart disease (gestation 18–35 weeks). Gated volume sets could be obtained in 51 of 57 cases. Normal right and left ventricular volume, stroke volume data and RV/LV ratios were obtained, together with normal changes with gestation. In patients with congenital heart disease, those with a hypoplastic heart (index lesion serology of Fallot) demonstrated normal combined ventricular volumes and right/left volume ratios for gestational age. Fetuses with single ventricular heart with intact septum (hypoplastic left or right heart) demonstrated a small but significant reduction in combined ventricular size and stroke volume for gestational age.

154

Flow in accurate 3d models of the early embryonic human heart – computational fluid dynamic models

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https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Background: Questions remain regarding the influence of fluid dynamics on the developing embryonic heart. Computational fluid dynamic (CFD) experimentation provides a unique medium for detailed examination of flow through these complex structures. The purpose of this investigation was to demonstrate: 1) streaming blood flow patterns exist in the early embryonic heart; 2) fluid surface stresses change significantly with anomalous alterations in fetal heart lumen shape. Methods: Stage 10 & 11 early human embryo hearts (AFIP collection) were digitized as calibrated 2D cross-sectional sequential images. These images were aligned and the cardiac lumen was delineated in each. A 3D surface was constructed from the stacking of these 2D images. This surface was meshed for input into a finite volume CFD flow solver. CFD flow solutions were obtained (steady and pulsatile flow). Particle traces were placed in the inlet and outlet portions of these two stages. Finally, sections of the embryonic heart were artificially enlarged with transformation of the computational grid. CFD flow solutions were obtained and surface stress changes analyzed. Results: Streaming was shown to exist with particles released on one of the other side of the cardiac lumen tending not to cross over and mix with particles released from the opposite side of the cardiac lumen. Figures show the result of shear stress changes (stage 10) in the altered lumens (A-narrowed;B-widened). Summary: We describe a technique used in developing sophisticated computational fluid dynamic models of embryonic hearts. Streaming exists in steady and pulsatile flow scenarios in the developing embryonic heart. Differences exist in local shear stress distributions with surface shape anomalies of the fetal heart lumen. These observations may shed light on the potential role of fluid dynamic factors in partially determining patterns of abnormal heart development.

155

Abnormal cardiac axis on early transvaginal ultrasonography: key finding of major cardiac malformation

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Introduction: First-trimester diagnosis of cardiac anomalies has become possible with high-frequency high resolution ultrasound probes. However, small size of fetal heart, difficulties in spatial orientation, unfavorable fetal position, and limited angles of insonation may impede adequate examination. The axis of fetal heart is independent of fetal age and usually can be assessed even when it is difficult to see detailed cardiac anatomy. We analysed the cases with abnormal cardiac axis on early transvaginal ultrasonography with special attention to the intracardiac and extracardiac abnormalities. Materials and Methods: Routine transvaginal ultrasonography is carried out at around 11–13 weeks' gestation in our hospital. There were 4 cases of abnormal cardiac axis during Dec. 1998 – Nov. 2000. Results: Four cases of abnormal cardiac axis were diagnosed at first trimester: one dextrocardia, two dextrocardia, and one levocardia with transverse axis. Second trimester fetal echocardiography revealed that the case of dextrocardia also had complete atrioventricular septal defect and severe pulmonary stenosis in left isomerism. Two cases of mesocardia were one functional single ventricle with pulmonary stenosis in right isomerism and one corrected transposition of the great arteries with absent right atrioventricular connection. The case of levocardia with transverse axis was confirmed to be tetralogy of Fallot. Conclusion: While in early transvaginal obstetric scanning, it is not possible to adopt a segmental approach to the fetal cardiovascular system, it is possible to establish left or right inversion of the cardiac apex, the stomach, and the liver. Abnormalities of cardiac position and/or axis are key findings in diagnosis of developmental cardiac defects by early transvaginal ultrasonography. So, we suggest that the transvaginal ultrasonography could be used for early detection of cardiac malformation.

156

Long QT syndrome (LQTS) during the perinatal period

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In a family with 2 different mutations in KCNQ1, A525T (maternal) and R518X (paternal), we investigated the heart rhythm during the last trimester of pregnancy and the neonatal period. Of 5 children, 2 had only the A525T mutation and 3 children carried both mutations. During the third trimester, measurements of fetal heartrate in the 3 fetuses with 2 mutations, showed low heartrate variability and concurrent bradycardia. During the 36th pregnancy the fetus had a pericardial effusion. In week 36 later cardiotachygraphic tracings demonstrated periods of alternating bradycardia (100 beats/min) and short periods of tachycardia (200 beats/min) suggesting periods of torsade de pointe tachycardia. The mother had low serum potassium and the arrhythmias disappeared after correction. In the 3 children with 2 mutations the post partum QTcs were > 600ms and in 1 child frequent episodes of T-wave alternans were seen. In the children with 1 mutation the neonatal QTcs were 500ms and 420ms. Congenital LQTS was symptomatic prior to birth. The presence of two mutations was associated with low in-utero heartrate variability and bradycardia. Maternal hypokalemia may induce in-utero tachyarrhythmia in fetus with LQTS caused by mutations in KCNQ1.

157

Fetal left atrial supraventricular tachyarrhythmia model in lambs

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To evaluate hemodynamic effect of rapid LA pacing on fetal circulation, we made a fetal supraventricular tachyarrhythmia model and measured the Ao pressure, CVP, and LV and RV outputs in 10 fetal lambs. Under maternal and fetal anesthesia, catheters were inserted into the fetal SVC and ascending Ao through a fetal neck incision. Pacing leads were sutured onto the fetal LA appendage via left thoracotomy. Ventricular output was estimated using a echo-device by a transcutine approach. Fetal hemodynamics were observed without pacing (control), and at the atrial pacing rates of 200, 300, 350, and 400/min. The Ao pressure decreased when LA was paced at 300/min or more and CVP increased when LA was paced at 350/min or more. The LV and RV output decreased when LA was paced at 350/min or more. The LV output was 215 +| 54 ml/kg/min at control, 205 +| 60 ml/kg/min at 200/min, 178 +| 56 ml/kg/min at 300/min, but decreased to 164 +| 44 ml/kg/min at 350/min and to 144 +| 37 ml/kg/min at 400/min. The RV output was 336 +| 56 ml/kg/min at control, 336 +| 95 ml/kg/min at 200/min, 273 +| 91 ml/kg/min at 300/min, but decreased to 256 +| 80 ml/kg/min at 350/min and to 202 +| 76 ml/kg/min at 400/min. Fetal circulatory failure could be mutually confirmed when LA was paced at 350/min or more in LA supraventricular arrhythmia model.

158

Cardiac wall motion velocities in normal fetuses evaluated by pulsed doppler tissue imaging technique

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The purpose of this study is to determine regional myocardial velocities in normal fetuses using Doppler tissue imaging technique (DTI), and to compare them with those in healthy children (HC). Fifteen fetuses, gestational age 28–31 weeks (mean 30±2.9 weeks), and 16 HC (mean age 2.5±1.5 yrs) were examined. DTI sample volume was placed in the middle segment of left ventricular lateral wall (LVLW), inter-ventricular septum (IVS), and right ventricular wall (RVW) in the four chamber views, to review longitudinal systolic and diastolic velocities. Peak velocities of ventricular ejection (VE), rapid ventricular filling (E), and late ventricular filling (A) were measured, and E/A ratio were calculated. In fetuses/HC mean values of peak ventricular ejection velocities for LVLW, IVS and RVW ranged from 4.6–7.8/7.5–11.8 cm/sec, for rapid ventricular filling 4.1–5.5/12.9–17.4 cm/sec, for late ventricular filling 5.6–9.4/5.5–7.6 cm/sec and for E/A ratio 0.65–0.75/2.2–3.2. RVW and IVS had significantly lower systolic velocities in fetuses in comparison with HC ($p<0.001$), but there was no difference in systolic velocities for LVLW. During diastole all fetal walls moved significantly slower during E phase than in HC ($p<0.0001$), and E/A ratios were significantly lower ($p<0.0001$). There was no difference in A velocities between fetuses and HC. Pulse DMI is a new promising ultrasound technique for determining regional myocardial velocities and that can be used to assess cardiac functions in fetuses. Diastolic myocardial velocities indicate that fetuses have lower ventricular compliance than children. Their systolic velocities measured by DMI are significantly lower for RVW and IVS, but similar for the LVLW.

159

Prenatal diagnosties of congenital heart diseases – ten-year experience

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To assess the reliability and accuracy of fetal echo in prenatal detection of cardiac abnormalities and to analyse the outcomes of fetuses with these anomalies. During 10 yrs period, 1622 fts have been scanned by pediatric cardiologists. Gestational age was 15–39 weeks (mean 25.6±9.2 wks). Fetal echo results were compared with postnatal echocardiography or autopsy

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

findings. Cardiac abnormalities were recognized in 94/1622 (5.8%) fe: 73 had structural heart defects, 13 had cardiac arrhythmias and 8 had both (3 of them cardiac tumors and arrhythmias). Among the 86 with structural heart defects, more than 60% had critical obstructive lesions or complex CHD. About one half (10/21) of the fetuses with arrhythmias had persistent, severe rhythm disturbances (5 with complete AV block, 4 with supraventricular tachycardia, and one with atrial flutter), while other had premature atrial contractions. Of the 54 fetuses with cardiac diseases, 27 (28.7%) had echocardiographic criteria for heart failure, and 11 of them had fetal hydrops as well. Overall outcome in study group was poor: 39 (41.5%) pregnancies were terminated, two in (2.1%) died in utero, 20 (21.3%) infants died in neonatal period, while 33 fetuses (35.1%) with cardiac anomalies survived during the follow-up (6 months to 9 years). There were 8 fetuses with false-negative diagnosis, and two with false-positive findings (the sensitivity of the method was 91.5% and specificity 99.8%). Fetal echocardiography is a highly sensitive and specific diagnostic method. Fetuses with critical obstructive lesions or complex heart defects had poor prognosis, particularly in cases with associated heart failure or cardiac arrhythmias.

160

Magnetocardiography in the detection of fetal arrhythmias

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Background: Fetal magnetocardiograms (FMCG) are recordings of the magnetic field generated by the fetal heart. Whereas other methods provide a measure for the mechanical performance of the fetal heart, fetal magnetocardiography reflects the electrophysiological phenomena directly. FMCGs can be reliably obtained from 20 weeks gestational age. Methods: FMCGs were obtained in 36 normals, 3 fetal complete AV-blocks, 2 fetal flutters and 2 with premature atrial contractions (PAC). The FMCGs were obtained using a 19-channel SQUID Magnetometer system, cooled by liquid Helium. Results: Normal fetal heart rates could be reliably obtained from 20 weeks gestational age and varied from 110 to 160 bpm with a beat-to-beat variability of 5–25 bpm. P wave duration 46±11 ms, PR interval 99±16 ms, QRS-width 52±9 ms, T-wave and QT-segments were discernable in 50%. In the 3 complete AV blocks measurements between 25 and 35 week gestational age. P-P interval varied from 395±36 ms to 460±46 ms, R-R interval ranged from 784±22 ms to 1060±4 ms, while QRS-width was not significantly different from normals. In case A an abrupt progression from a 2nd to 3rd degree AV-block was observed, from 784±33 to 960±62 ms for the R-R interval. Case B and C showed R-R intervals of 1063±4 ms and 1001±32 ms. In two patients periods of fetal flutter were observed with atrial rates of respectively 438bpm and 480bpm and ventricular rates of 219bpm and 240bpm, showing a 2 : 1 atrioventricular block during flutter periods. Fetal echography and postnatal ECG confirmed all rates and measurements. Conclusions: FMCG is able to register fetal heart rate reliably from 20 weeks gestational age on and can be used to classify arrhythmias. It is possible to determine the atrial and ventricular rates and the duration of P-waves, PR-intervals and QRS-complex.

161

The morphology of the human fetal heart from 7 to 41 weeks' gestation

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Background: Currently, there is a trend to examine the fetal heart earlier in gestation. Detailed anatomic studies, with respect to prenatal diagnosis, have not been performed, but compared to appearances seen later in gestation. Objectives: To describe the morphology of the fetal heart from 7 to 41 week's gestation, by a single methodology, and with respect to ultrasonic views. Methods: 241 fetal hearts were examined by dissecting microscopy. External features were documented, and sections made in specific anatomic planes relevant to ultrasonic examination. The size, morphology and relative positions of structures was documented in relation to gestational age. Results: Between 7 and 11 weeks the long axis of the heart shifts leftwards. Overall cardiac dimensions increase in curvilinear fashion, but the relative increase in size actually falls with age. Prior to 15 weeks, the arterial trunks are similar in size at valvar level, but the ascending aorta is larger. The Eustachian valve is a prominent right atrial structure prior to 12 weeks, and there is rapid expansion of the pulmonary venous orifices between 7 and 12 weeks. Offsetting of the atrioventricular valves is present from 7 weeks, and increases in proportion to heart size. Sections of crossing outflow tracts could be obtained from 8 weeks, as could a three-vessel view and the views of the converging aortic and ductal arches. Conclusions: In the first trimester of pregnancy there is rapid, relative increase in heart size. There are differences in the position and morphology of the heart between early and later gestation. The main features necessary for the exclusion of some major cardiac defects, nonetheless, are present from as early as 7 weeks. Diagnosis, at this stage, will require ultrasonic resolutions of the order of 0.25 to 0.7mm.

162

Detection of cardiac hypertrophy in the fetus using magnetocardiography

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To determine the developmental changes in the myocardial currents during fetal life, to evaluate the clinical usefulness of fetal magnetocardiography (fMCG) for prenatal diagnosis of cardiac hypertrophy, we approximated the magnitude of the one-current dipole of the fetal heart using fMCG. Eighty-eight fetuses without fetal or maternal complications and 7 fetuses with cardiomegaly on fetal echocardiograms were included in this study. The gestational age (GA) ranged from 20 to 40 weeks. Before recording fMCG, position and depth (D) of the fetal heart were determined by ultrasound. The normal component of fMCG was recorded using a 9-channel SQUID system (Hitachi) in a magnetically shielded room. The total current activity of the heart was estimated as a one current dipole (Q). Apart from the peak magnetic field (B, Tesla) among 9 waveforms using the following equation: $B=(\mu/4\pi)(Q/4\pi D^2)$, where μ is a constant of the magnetic permeability. The Q value of the normal subjects ranged from 41 to 650 nAm, and showed positive correlation with GA, reflecting an increase in the amount of myocardial current, i.e., myocardial mass. Three of the fetuses with cardiomegaly caused by various cardiovascular abnormalities ranged from 291 to 1,330 nAm. In 6 fetuses, the Q value was higher than the mean + 2SD of the control for each corresponding GA. Moreover, right ST segment depression was demonstrated on fMCG tracing in a fetus with marked cardiomegaly due to Ebstein malformation. Although fetal orientation might influence the magnitude of the dipole, making it smaller, fMCG recorded by a multi-channel SQUID system is a clinically useful tool for non-invasive prenatal, and electrical evaluation of fetal cardiac hypertrophy. Observation of all three components of fMCG is desirable to estimate the dipole strength more precisely.

163

Outcome of 412 congenital heart disease diagnosed with fetal echocardiography

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Purpose: To evaluate the prenatal outcome in patients with fetal echocardiographic diagnosis of congenital heart disease (CHD). Methods: Between January 1992-October 2000, 682 pregnant women (age 28±2, 5years) between the 18--24- week of the gestation were included in the study. Inclusion criteria were: suspected CHD in a previous echocardiogram (379pz 55,6%), extracardiac malformations (118pz-17,3%), chromosomal anomalies (62pz-9,1%), maternal diabetes (7pz 1%), familiarity for CHD (14pz 2%), abnormal results of screening tests (108pz-15%). Pediatric cardiologic and gynecologic per formed the echocardiography together. Results: Echocardiographic evaluation was normal in 270 pz(39,6%) and abnormal in 412 pz(60,4%), of which 242(58,7%) with isolated CHD and 170(41,3%) with associated malformations. Among the CHD, 172(42%) were neonatal emergencies. The outcome of the 412 fetus with abnormal exams was evaluated among the 242 fetus with isolated CHD, 73(30,1%) were born and still alive, 7(2,9%) dead in the utero(IUD), 25(10,3%) died in the first days of life, 30(12,4%) were still in utero, 59(24,4%) were lost during the follow-up(LOST) and 48(19,9%) underwent to an abortion(AB). Among the 170 with associated malformations, 28(16,5%) were still alive, 12(7%) were IUD, 34(20%) died in the first days of life, 17(10%) were still in utero, 32(18,6%) were LOST and 43(25%) AB. Among the 265 surgical interventions performed on newborn younger than 1 year old, 10% had been planned before using the fetal echocardiogram, with the result of a better outcome: survival percentage of prenatal diagnosis of severe pulmonary stenosis or atresia was 67%vs41% of no prenatal diagnosis($p<0.05$), the same for the transposition of the great arteries(80%vs53%,$p<0.05$). Conclusions: The outcome of fetuses with CHD is strongly related to associate extracardiac and/or chromosomal anomalies. In some selected CHD (TGA, pulmonary stenosis with intact ventricular septum) the outcome is better if the diagnosis was performed prenatally.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

164
Fetal tachyarrhythmia – management and outcome
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INTRODUCTION: This study was designed to evaluate a new classification of fetal tachyarrhythmias (FT) using M-Mode echocardiography, and to test a new therapeutic approach based on this classification. METHODS: Between 1987 to 2000, we studied a group of 65 fetuses with tachyarrhythmia of 200bpm and over. Observing the fetal heart during 30-45 minutes we classified the FT in two groups: 1) Non sustained (NS): when the sinus rhythm was predominant and lasted more than 50% of the time of observation; 2) Sustained (S), when the tachycardia lasted more than 50% of the time of observation. For the NS group we used oral digoxin to the mother. For the S group we used two types of treatment: 2.1) Direct treatment with desbenonide (Cedilanid-D) associated with oral digoxin and 2.2) Oral digoxin as a control group. Second drugs were used when conversion was not achieved (amiodarone and flecainide). RESULTS: Twenty-five of these fetuses had NSFT (supraventricular tachycardia n=22 and atrial flutter n=3) and 40 had SFT (supraventricular tachycardia n=31, atrial flutter n=8 and ventricular tachycardia n=1). In 22 cases of NSFT treated, we achieved 95.4% (n=20) of conversion to sinus rhythm. In 37 cases of SFT treated, 17 were treated directly with 82.3% of conversion (n=14). Conversion immediately after the cardiocentesis was achieved in 42.8% of the cases. Twenty fetuses with SFT were treated orally with only 30% of total conversion. In the latter group the rate of no conversion, the number of days of conversion and the number of drugs associated were higher (amiodarone in 12 cases and flecainide in 4 cases). We had also 4 cases of maternal digoxilis intoxication. All treated fetuses with NSFT survived besides 2 (triplet uterine, rabdomyomas) and 5 fetuses died with SFT (severe hydrops in 3, rabdomyomas in 1 and complications of the cardiocentesis in 2). Structural abnormalities were found in 5 cases. CONCLUSIONS: The results showed that fetal echocardiography was accurate in diagnose the type of tachyarrhythmia and the classification in sustained and not sustained was effective in orient the therapeutic approach. We believe that using this classification we can separate the more severe form of this disease with greater potential to develop fetal hydrops (SFT) and the direct treatment in this group is justified. Since we can achieve the conversion so fast prenatal therapy may prevent unnecessary operative or premature deliveries and should always be considered.

MAY 29 Time: 14:00–15:30

Session 16: The Adult with Congenital Heart Disease, Pregnancy/Delivery for the Women with Congenital Heart Disease

165
The impact of cardiac surgery scars on well-being of young adult patients with congenital heart disease
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Background: Post thoracotomy scarring is generally considered to have a negative impact on adult patients' well-being and have spurred development of transvascular treatments for congenital heart disease. Methods: A preliminary questionnaire asked 10 cardiac patients 'to describe personal consequences (if any) of having a cardiac surgery scar.' The results provided basis to design the second questionnaire which asked specific questions and rated the impact of scars on identified areas of concern. Responses were tested against the scar location/size and patients' demographic and clinical data. Results: 13 patients declined the study. 100 patients (53 men) aged 18 to 50 (mean 27 years) participated. 5.8% of our patients reported that the scar affected them less now than in adolescence. The body was perceived as disfigured by 58%. The scar was concealed occasionally or every day by 48% of patients. Attention to scar made 19% of patients feel negative, 58% neutral and 23% positive. Effect of the scar on: Very negative Negative Neutral Positive Very positive. Recreation and Sport 2% 11% 78% 5% 3% Self-Esteem 4% 16% 66% 12% 2% Self-Confidence 2% 10% 72% 9% 3% Career 1% 4% 91% 3% 1% Success in life 1% 5% 85% 6% 3% Health appreciation – 4% 35% 40% 21% Friendships 1% 6% 86% 3% 4% Sexual relationship 5% 8% 70% 10% 2%. Conclusion: A sternotomy or thoracotomy scar left a neutral/negative impact on patients' well-being. We noted small variations relative to the location and size of the scar, and patients' characteristics.

166
Aortic valve problems after coarctation repair
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Background: Although the association between aortic coarctation (coarc) and aortic valve (AoV) and aortic arch anomalies is well known, not much is reported on the incidence of these sequelae long-term after surgical repair of aortic coarctation. Methods: From 180 patients, who had undergone coarc repair at our institution and were >16 years of age at the time of the study, 124 had adequate follow up. Echo data, cardiologic charts and surgical records were studied retrospectively. Results: The mean age at coarc repair was 11 years and the mean follow-up 18 years. Sequelae: AoS 18 pts (14.5%), 16 pts intervention, 2 gradient >30 mmHg; Ao regurgitation 46 pts (36.7%): 5 pts intervention, 40 pts AoR (23 pts 1+, 13 pts 2+, 5 pts 3+, 2 pts 4+); AoS/AoR 3 pts (2.4%), 3 intervention. Ascending Ao dilatation > 40mm: 35 pts (28.2%). Ao arch pathology: 29 pts (23.3%). Cardiac related death: 3 pts (2.4%). Conclusion: In the long term follow up of patients after coarc repair, AoV and aortic arch problems occur in 46% of the patients. Close life-long surveillance of coarc patients is mandatory not only because of re coarc and hypertension but also because their aortic valve and ascending aorta are at risk.

167
Long-term follow-up of Senning operation : comparison of mri and radionuclide angiography in the evaluation of right ventricular function
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Right ventricular function is an important determinant of late morbidity and mortality after the Senning operation. The value of noninvasive MRI in the assessment of right ventricular function 15 to 25 years (mean 17.6 years) after this procedure has been investigated and compared with findings on radionuclide ventriculography in 40 adult patients (mean age : 18 years). The age at the operation was 2 months (± 2). The mean resting right ventricular ejection fraction by first pass radionuclide angiography was 57.1 (± 16.5). Right ventricular ejection fractions measured by MRI were slightly higher : 60.9 % (± 7.6). The resting right ventricular function evaluated by the two methods was normal in all patients. Correlation between the two methods was poor but significant ($r = 0.66$, $p = 0.001$). The anatomy of great arteries and residual atrial defect (3 patients) were clearly assessed using MRI. The intraatrial baffle and systemic venous connection were easily visualized by MRI. 3 patients had stenosis of the intraatrial baffle. Tricuspid regurgitation was easier to detect by Doppler echocardiography than MRI. MRI and radionuclide angiography are useful non invasive methods of investigating right ventricular function. MRI provides also a complete anatomical assessment, which can be repeated as often as necessary after the Senning operation.

168
A surgical correction of the patients with eisenmenger syndrome
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Till the present time the problem of patients with Eisenmenger syndrome has not been solved. Nowadays there are three directions in its treatment: conservative – with cardioglycosides, radical – lungs transplantation; palliative – with prosthesis of the main pulmonary artery – pulmonary artery narrowing. Actually ventricular septal defect (VSD) combined with heart lung preparation (Lillehei Hit-Edwards) are uncurable. In Saint-Petersburg Pediatric State Medical University two stage method of surgical correction of Eisenmenger syndrome was invented. The method was developed on autopsy specimens and approved experimentally on the animals. The first step during the artificial circulation: VSD closing and formation of the shunt between the pulmonary artery trunk and left auricle or between the pulmonary artery (left or right) and the same vein was made on the open heart. The first shunt is better on two reasons: there is no danger of vessel stenosis or kinking formation and independent occlusion of

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

the shunt has happened rapidly. The changing of the level of veno-lateral blood shunting from intracardiac to extracardiac allows to adapt the heart-lung system to new hemodynamic conditions during the postoperative period, because the external shunt works as a safe-valve. Then with the pressure decreasing in the pulmonary artery system independent occlusion of the artificial shunt happens. However there is no necessity to wait for independent occlusion. With the pressure decreasing to 40–50 mm Hg the endovascular occlusion of the shunt is possible.

169

Transition from pediatric to adult health care among a cohort of young adults with congenital heart defects

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Purpose: To examine the transition from pediatric to adult care for patients with complex congenital heart defects (CHD). Method: From a cohort of children seen at the Hospital for Sick Children in Toronto we selected 422 patients with the most serious defects who are now age 19–21. The consensus recommendation is that this group of patients should be seen annually at a specialized CHD center. Questionnaire and/or interview data were obtained from 233 of 365 patients that we were able to contact and who consented to participate. Data on clinic visits for all eligible patients were obtained from all 15 Canadian Adult Congenital Heart Network (CACH) centers. Results: Of the eligible patients, 41% had been seen at least once at a CACH centre and 2.5% of patients were registered but had not been seen (i.e., referral received, appointment pending, cancelled appointment, or no-show). Of the patients who had attended at least one appointment at a CACH centre, 74% had been seen within the past year, 13% within the past 2 years, and 5% had not been seen in over 3 years. Demographic (e.g., distance to centre) and psychosocial predictors (e.g., health behaviours, health status, psychological adjustment) of successful transition will be examined. Conclusions: The majority of 19–21 year old CHD patients who should be seen annually at a specialized CHD centre have not made a successful transition to adult care. Once patients have successfully made the transition to adult care, the vast majority of patients are seen on a regular basis. This is the first study to document the transition from pediatric to adult care in a large cohort of CHD patients.

170

Mortality and risk factors for late deaths in tetralogy of Fallot long after repair - Japanese nationwide multicenter survey

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Purpose: This study aims to analyze mortality and risk factors for late deaths in adult patients after repair of tetralogy of Fallot (TOF). A Japanese multicenter study. Background: Although favorable outcome of surgery for TOF has been documented, late deaths from cardiac causes are still known to occur. Mortality and features associated with late deaths in a large series of these patients are rather sparse. Methods: Twelve major cardiovascular centers in Japan followed two postoperative hospital survivor groups: Group A (n=121) received repair of TOF in 1972. Group B (n=181) received repair of TOF in 1982. Surgical techniques are thought to be largely different between these 2 years. We analyzed mortality, demographic data, surgical history and risk factors for late mortality in these patients. Results: Significant differences were observed in cardiac anomalies (13 % v 34 %, p<.001), outflow patch repair (49 % v 93 %, p<.001), transventricular patch repair (15 % v 61 %, p<.001) and age at repair (9.4 years v 7.6 years, P=.04). Late deaths from cardiac causes was similar in number (7 pts v 2 pts). Actuarial 15- and 25-year survival rates were similar (96% and 93%) in Group A v 98% in Group B, (P=.94). Re-intervention (reoperation) free 15-year survival rates were 93% v 94% (P=.93). Risk factors for late deaths were age at repair (p=.04), history of reoperation (p=.001), sick sinus syndrome (P=.002) and residual ventricular septal defect (p=.02). Transventricular patch repair was associated with reduced long term survival in Group A (p=.01). Conclusions: In spite of significant differences of demographic and surgical history between patients with TOF repaired in 1972 and in 1982, late survivals were both excellent. High risk subsets of patients for late mortality can be identified.

171

Aortic root dilation in adults with repaired TOF

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OBJECTIVES: We aimed to evaluate aortic root size and possible factors associated with its dilation in adults with repaired tetralogy of Fallot (TOF). BACKGROUND: Aortic valve and/or aortic root replacement are sometimes required due to progressive aortic root dilation and aortic regurgitation (AR) in repaired TOF. METHODS: We analyzed serial demographic and echocardiographic data in 51 patients with repaired TOF (17 dilators with aortic root ≥44 mm (age: 32.6 ± 7.4 years), 17 intermediates with aortic root between 35mm and 44mm (age: 36.6 +/- 11 years) and 17 non-dilators with aortic root <35mm (age: 30.9 ± 7.7 years)) and 17 non-TOF repaired controls (age: 35.9 +/- 8.4 years). RESULTS: Aortic root size (mm) at the study end was 49 +/- 5.5, 39 +/- 2.4, 31 +/- 2.3 respectively (p=.0001). Over a mean period of 5.2 +/- 3.8 (REPAIRED 3.8 years), annual increase of aortic root (mm/year) was 2.1 +/- 2.8, 0.22 +/- 0.8, 0.2 +/- 0.3, 0.4 +/- 1.6 (p=.001). Amongst the subgroup of dilators a longer shunt-to-repair period: 12 +/- 9 years (p=.08), and a higher incidence of pulmonary stenosis: 6/17 (p=.04), right aortic arch: 3/17 (p=.01), AR: (moderate): 4/17 (p=.005), aortic valve replacement: 2/17 (p=.10), greater cardiothoracic ratio: 59 +/- 4 (p=.0001) and larger left ventricular end-diastolic dimension (mm): 52 +/- 5.9 (p=.001) were observed compared to all other subgroups. CONCLUSIONS: Aortic root dilation with left ventricular enlargement is not uncommon in repaired TOF. Long-standing volume overload on aortic root and intrinsic properties of aortic root appear to have a cause-effect relationship with this dilation. Adults with TOF may be at risk of aortic root rupture. Meticulous follow-up of aortic root is thus recorded.

172

Clinical profiles of adult patients with unoperated isolated secundum atrial septal defect in Korea

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To evaluate the natural history and clinical characteristics of adult patients with isolated secundum atrial septal defect(ASD), we reviewed 206 patients with unoperated ASD who had registered at the Grown-up Congenital Heart clinic, Severance Medical Center from Nov 1994 to July 2000. Results : The subjects ranged in age from 15 to 71 and consisted of 57 men and 149 women. 2. 127 patients(61.6%) had symptoms at presentation. Dyspnea on exertion(78), chest pain/discomfort(23), palpitation(16) etc. And remaining 79(38.4%) were asymptomatic who were noted to have ASD during medical screening. cardiomegaly(53), heart murmur(15), abnormal EKG(6) etc. 3. Twenty-two (10.6%) had atrial fibrillation(AF) at presentation. The mean age of the patients with AF(50.2 +/- 7.6 years) was higher than that of the patients without AF(39.2 +/- 14.6 year). Defect diameter was larger in the patients with AF than without AF(p<0.001). And in 3 patients who were operated after 50-year-old, AF developed and persisted during follow up period after operation. 4. Pulmonary hypertension(PHT)(defined as mean pulmonary artery pressure at rest > 25mmHg) was found in 21 patients. Defect diameter of PHT patients was over 15mm in 19 patients. Frequencies of PHT in patients with large defect(>15mm), according to different age group were : 16~34 years old – 2/58(3.4%), 35~54 years old – 9/54(16.7%), over 55 years old – 7/40(17.5%)(p<0.05). Two patients aged 26 and 47 had PHT even with small defect(<15mm) suggesting coexisting primary PHT. 5. Frequency of mitral regurgitation(>II/IV) was 6% and mitral valve prolapse was 18%. Conclusion : Although ASD patients have good natural history, quite a number of ASD patients showed problems such as AF and PHT if they were not operated on over 40 years. So early closure is recommended even without symptoms.

173

22q Deletion syndrome in adults with tetralogy of Fallot: cardiovascular and other clinical features

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Objective: To determine the cardiovascular phenotype of 22q Deletion Syndrome (22qDS) in adults with tetralogy of Fallot (TOF). Background: The association between 22qDS and TOF is well established in paediatric populations (~20%) but not in adult cohorts. Furthermore, limited data exist

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

regarding late onset features in that population. Methods: Consecutive adult patients with TOF attending the University of Toronto Congenital Cardiac Centre for Adults were systematically screened for clinical features of 22qDS (learning difficulties, dysmorphic facial features, hypernasal speech, other birth defects, hypocalcemia). Comprehensive reviews of pediatric and adult case notes provided data on the cardiovascular phenotype. 22qDS was confirmed in subjects meeting clinical screening criteria using fluorescence in-situ hybridization (FISH) methods and a standard probe. Results: Seventeen (9.8%) of 174 subjects had confirmed 22qDS. Subjects with 22qDS had higher rate of aberrant subclavian artery (P=0.003) and reoperations following repair (p=0.04) than other TOF subjects; rates of other cardiovascular features did not differ between the two groups. Learning difficulties ranged from very mild to severe and six (35%) 22qDS subjects had a major psychiatric illness (e.g., major depression or schizophrenia). Conclusions: 22qDS is common in adult patients with TOF and may be associated with extra-cardiac features including late onset psychiatric disease. The results suggest that adults with 22qDS usually have a similar cardiovascular phenotype to others with TOF but their course may be more complicated.

174

Patterns of in-patient care for adults with congenital heart defects

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To determine patterns of in-patient care for adults with congenital heart defects (CHD), we analyzed data from all discharges in New York and Massachusetts in 1996. Cases more than 16 years of age with ICD-9-CM codes indicating CHD were selected. Admissions were grouped as cardiac surgery, cardiac catheterization, non-cardiac surgery, and medical. Cardiac surgical procedures were further grouped as follows: pacemaker insertion, septal defect or patent ductus closure, valvotomy or replacement, cardiac revascularization, or other cardiac surgical procedure. Ischemic cardiac disease, type of insurance, and in-hospital death were also tabulated. Among 2,728,499 discharges, only 3017 (0.1%) had CHD (age range 16–98 years, median 40). In-hospital death occurred in 109 (3.6%). Concomitant ischemic disease was present in 368 (12%). Most admitted patients had insurance: commercial 797 (26%), HMO/managed care 678 (22%), government 1419 (45%); other 34 (1%); none/self pay 189 (6%). Half of admissions included procedure: cardiac surgery 919 (29%), catheterization 559 (18%), non-cardiac surgery 293 (9%), medical 1576 (51%). Among cardiac surgical admissions, 38 were pacemakers (4%), 276 septal defect or patent ductus closure (30%), 226 valvotomy or replacement (25%), 130 coronary revascularization (14%), and 233 other cardiac surgery (26%). Admissions occurred at 270 institutions. Most centers that admitted adults with CHD did so rarely. The majority (204 centers, 76%) admitted <10 patients in 1996; 55 admitted 10–49 (20%); only 13 (5%) admitted 50+ patients. Death during admission was higher at institutions admitting fewer patients (3.9% at <10 admits/year, 2.6% 10–49, 2.6% 50+, p= 012). Of 60 institutions that performed cardiac surgery, over half (55%) performed <10 cases/year in patients with CHD. In-patient care for adults with CHD is not centralized. Many institutions occasionally admit and perform procedures in these patients.

175

A controlled trial of exercise training in adult patients with repaired tetralogy of Fallot

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Background: Positive effects of physical training in adults with acquired heart disease have been reported. The role of exercise training in adult with congenital heart disease however is less well defined. Furthermore, there have been concerns over the safety of exercise training in such patients. We assessed the safety and effect of exercise training in adults with repaired Tetralogy of Fallot (TOF). Methods: Eighteen adult patients with repaired TOF were randomized to participate in a three month supervised exercise program (exercise group, 9 patients) or to continue their usual exercise routine (control group, 9 patients). Each patient in the exercise group received an individualized exercise program to be performed 3 times a week, whereas the control group was told to continue living their life as usual. Cardiopulmonary testing was performed in all patients at baseline (before randomization) and at the end of the study. Results: No death or incidents occurred during the study period. There was a significant increase in peak oxygen consumption in the exercise group by the study end (22.1 ml/kg-1/min-1 vs 24.3 ml/kg-1/min-1, p=0.049), whereas it remained unchanged in the control group (21.8 ml/kg-1/min-1 vs 22.1 ml/kg-1/min-1, p=0.825). There was also a trend in the exercise group toward an increase in exercise duration (720 sec vs 780 sec, p=0.084), while a decrease was present in the control group (776 sec vs 724 sec, p=0.062). Conclusion: In clinically stable adult patients with repaired TOF, a moderate level of exercise training appears safe and improves aerobic capacity. Exercise training should thus be encouraged in these patients.

176

Pregnancy outcomes after atrial repair for transposition of the great arteries (TGA)

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We report on pregnancy outcomes after atrial repair of TGA from 12 centers in a national registry. After public announcement, clinicians reported pregnancies using a standard format. Fifty pregnancies were reported in 31 women. The majority (n=29) had Mustard procedure, 2 had Senning. Time between atrial repair and pregnancy ranged from 9 to 27 yrs (m. 19.2 yrs). Age at pregnancy was at 25 yrs (range 16–38). At pregnancy 42 women were NYHA Class I, 6 were Class II and 2 were Class III. 71% of the infants were delivered prematurely and weighed 2809 g. Mean gestational age was 35 wks. 10% were delivered by Cesarean section, 3 for cardiac indications. Maternal complications included arrhythmia in 5, and hemoptysis in 2. Heart failure occurred in 25% of the pregnancies, developing during the second and third trimester and in 2 at 5–6 days post-partum. Severe right ventricular failure led to cardiac transplantation 5 months after delivery in one Amadine who developed heart failure required hemodialysis post-partum then died suddenly one month after delivery. There is one late death. We conclude that pregnancy after atrial repair carries a moderated degree of risk and should be undertaken with caution.

Session 17: Surgical Management and Results: Valves/Conduits

177

Functional outcome of Ross procedure in children

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OBJECTIVE: The aim of our study was to analyze functional effects of the replacement of aortic root with pulmonary valve autograft (Ross procedure) in children with congenital anomaly of the aortic valve. METHODS AND DATA: Between December 1997 and December 2000, a total of 30 patients underwent the Ross procedure. Indications for surgery was severe aortic regurgitation (21 patients) or a combination of aortic regurgitation and stenosis (8 patients). Our patient with severe subaortic stenosis underwent Ross-Konno procedure. Average age at operation was 12.9 +- 4.1 years (from 4.5 to 17 years). RESULTS: Survival on average follow up period of 15 months is 100%. 16 patients (54%) had no or trace neo-aortic regurgitation, trivial aortic regurgitation was discovered in 13 patients (43%). One patient with structural anomaly of the pulmonary valve had mild neo-aortic regurgitation. Within the follow up period there has been a significant (p< 0.001) reduction in the diastolic diameter of the left ventricle (LVDd) both in absolute and index terms. Other echocardiographic parameters (FS, EF) were within the normal range. Dysrhythmia was not noted in any of the patients, while T-wave inversion on ECG was recorded in 4 patients. 28% of all operated patients are without medication, while the rest are on medication for a period of six to twelve months after surgery in correlation with the patients' clinical state. CONCLUSION: Replacement of aortic root with pulmonary valve autograft is a surgical method of choice in children with congenital or acquired anomaly of the aortic valve. With the technical aspects of this procedure well accomplished, mortality nears zero. The functional outcome of operation is encouraging, but follow-up is too short to make any firm conclusion.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

178
Long-term fate of aorto- and homografts in the reconstruction of the right ventricular outflow tract
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Allografts are considered the conduit of choice for the reconstruction of the right ventricular outflow tract (RVOT), yet their availability is limited and therefore xenografts are implanted as well. We compared the long term durability of both grafts in the RVOT. Patients: From 1/1974 to 6/2000 401 patients (survival more than 30 days) with pulmonary stenosis (n = 153), tetralogy of Fallot (n = 96); DORV (n = 21) or TGA (n = 46) patients with Rastelli type correction; patients with truncus arteriosus communis (n = 105) were studied. Results: 20 year survival analysis showed a significantly (p = 0.01) better survival of patients with TOF/pulmonary atresia (81 ± 5%) and Rastelli-type surgery (81 ± 8%) compared to TAC patients (59 ± 8%). Conduit related (allograft vs xenograft as initial graft) patient survival analysis showed no significant (p = 0.51) difference, survival being 84 ± 4 % for allograft and 77 ± 5% for xenograft patients. Comparing allo- vs xenograft, the conduit exchange rate was not significantly different (p = 0.2) for conduit diameters ≤ 15mm, being 41 ± 9% for allografts and 30 ± 6% for xenografts. For allografts with diameter greater than 15mm, exchange free survival rate was significantly (p = 0.02) higher, being 87 ± 8% compared to 51 ± 10% for xenograft conduits. Conclusion: Conduits of either or gin with diameters less than 15mm exhibit a high probability for replacement. The reason for conduit exchange is not always structural deterioration but also outgrowth of the small diameter. For conduit diameters > 15mm allograft conduits performed significantly better as compared to xenografts. Allografts remain an important tool in the reconstruction of the RVOT with excellent 20 year graft and patient survival.

179
Pericardial tissue valve and gortex conduit: an excellent alternative for right ventricular reconstruction in children
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Objectives: There is still no perfect conduit for reconstruction of the right ventricular outflow tract (RVOT) in children. Homografts are not always available in the appropriate size, and degenerate in a few years. This study evaluates the pericardial valve with Gortex conduit as an alternative for RVOT construction. Methods: From 1/1993–6/30/99 a pericardial tissue valve was inserted in all patients undergoing RVOT reconstruction or pulmonary valve replacement (PVR) who were large enough to accommodate a sizer valve. In patients without a native main pulmonary artery a new technique was used to construct an RV-PA conduit out of a flap sheet of Gortex, since ductus frequently leads to stenosis. Data was collected by retrospective review, follow-up echocardiogram and assessment by a single cardiologist. Results: There were 48 patients, 22 undergoing a PVR alone and 26 a RV-PA valved Gortex conduit. Diagnosis included TOF (n=25), truncus arteriosus (n=9), VSD with PA (n=5); DORV (n=4), D-TGA with PS (n=2), and 1 each IAA with sub AS VSD with PI and PS s/p Ross procedure. Patient age ranged from 3–33 years, and all surgeries were reoperations. The valve sizes ranged from 19–33mm, and the median hospital length of stay was 4 days. There were 2 (4.2%) perioperative and 1 (2.1%) late deaths, none related to the valve or Gortex conduit. At a follow-up of 9–80 months (37 ± 16 months), all remaining 45 patients are NYHA class I, all valves are functional, and no patient (0/45) has required valve or conduit replacement or revision; more importantly echocardiogram revealed no significant valve or conduit stenosis (mean gradient 10±8 mmHg), as well as no evidence of regurgitation or truncal degeneration. Conclusions: A pericardial tissue valve and Gortex conduit provides a reliable alternative for RVOT reconstruction in pediatric patients. It is readily available in all sizes, molds to the limited retrosternal space, and has outstanding intermediate results with no evidence of failure or deterioration up to 7 years after insertion.

180
Medium term follow up after percutaneous transseptal mitral commissurotomy in children below 12 years
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Follow up results of percutaneous transseptal mitral commissurotomy (PTMC) in the young children are not known. Records of 60 children, mean age 10.6±1.2 years (range 7–12 years), who underwent PTMC before Dec 1999, were analyzed for their symptom status and mitral valve area (MVA) following PTMC. Forty two (70%) patients have completed more than one year follow-up and were included in the present study. Before PTMC their NYHA class was III in 52% (n=22), IV in 14.3% (n=6) and MVA 0.59±0.15 cm2 (range 0.5 to 0.9 cm2). Two patients had severe MR and 2 had subpulmonal stenosis. Of the remaining 38 patients who had successful PTMC during mean follow up of 33.4±15.2 months (range 12 to 82 months), all were in either NYHA class I (74%) or class II (26%). Mean MVA at follow up was 1.49±0.5 cm2 (range 0.9–2.9 cm2) as compared to area just after PTMC 1.54±0.29 cm2 (range 1.2 to 2.7 cm2). Restenosis as defined by valve area <1.1 cm2 after successful PTMC was present in 5 out of 38 (13%). That did not correlate with MVA before or after PTMC and age of the patient. Conclusion: PTMC in children <12 years is an effective palliation on medium term follow up.

181
Evaluation of risk factors for homograft calcification
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Homograft (HG) conduits are routinely used to establish continuity between the RV and PA. HG in children are noted to calcify very early in the postop period. The retrospective study aims to evaluate risk factors for HG calcification and correlate it with other HG related complications. Of 222 patients (129 m, 93 f) with a median age 5.5 years (1mo-44yrs) who underwent surgery using HG during July 1991-Jan 2000, 63.5% were children. The primary diagnosis were TOF/DORV (63%), TGA VSD (17%), TOF/PA (10%), Truncus (3%) and others. A total of 236 HG were implanted in 222 patients. 91.3% aortic and 48.2% pulmonary HG. The sizes ranged from 8–28mm. 146 had no gradient, 32 mild (< 30mm), 4 moderate (30-50mm) and 4 severe obstruction (>50mm). 154 pts had a follow up of at least 1 mo available (3–66 mo). Calcification was detected in 43 (32%) of which 75% were aortic HG (p<0.01). Median time for detection of earliest calcification was 17mo (3–40mo). 42% of these had no obstruction, while 35% had mild, 14% moderate and 9% severe HG obstruction. Rapid wt gain after surgery (>5kg/year) was associated with a higher incidence of calcification (p<0.01). This correlated with times of growth spurt - age groups (1–4 and peripuberty) (p<0.04). There was no correlation between diagnosis, position, HG size, ABO compatibility, donor age, postop fever, HG gradient/ regurgitation and calcification. Conclusion: Aortic HG and rapid weight gain postop were associated with a higher risk of HG calcification.

182
Surgical mitral valvuloplasty in paediatric patients
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Introduction: Valve surgery in children, is aimed at restoring correct hemicyanotics with limited resort to prostheses, which would imply early deterioration or definitive hypercoagulation. Objective: To report a serie of paediatric pts with acquired mitral pathology for whom it was possible, in all cases but one to repair the damaged valve. Material and Methods: Between Oct.98 and Jul.00, 25 children with heart valve disease, 22(88%) rheumatic and 3(12%) post-endocarditic, were operated upon. Sixteen female, mean age age 11.6±3.94 yrs (5 to 17) and mean weight 29.4±9.02kg. All pts had mitral pathology: 16 had pure mitral regurgitation(reg) and 9 had reg+ stenosis. Four had associated severe aortic reg and 3 significant tricuspid reg. Results: In all operations the intention was to repair the mitral valve. Only one pt had a mechanical prosthesis implanted. In 24 cases complex valvuloplasties were performed: (expended commissurotomy, shortening of chordae, reconstruction of valve leaflets-patching or extension of the posterior leaflet and restuping of the annulus). In 4 pts homografts were implanted in aortic position. Average CPB time was 76.4±33.99mins and Ao clamping time was 56.08±32.29mins. One pt died in hospital from sepsis, there were no other mortality or morbidity. Surgical results were accessed by intra-operative TEE, and only one pt had a significant residual defect (mitral reg grade II/III and mitral gradient 10mm Hg). Follow up (Max. 2 yrs). Mean reduction in LVd was 18.7%±7% at LA 21,6%±12% and in PAP was 29,6%±17%; two pts have mitral re-stenosis but only one needs to be re-up. Conclusions: Mitral valve repair in children can be achieved in the majority of but only one cases, although it might be palliative.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

183

Reconstruction of the right ventricular outflow tract in children undergoing the Ross procedure

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Objective To determine whether surgical options for reconstruction of the RV outflow tract have problems after the Ross procedure in children. Methods Since 1992, 30 children have undergone the Ross procedure, including the Ross-Konno procedure in 6. Of these, a pulmonary homograft was used for reconstruction of the RV outflow tract in 4, and a tailored heterologous pericardial roll in 4. In the other 22, autologous tissue was exclusively used as a posterior wall of the channel, placing another patch (bearing a monocusp in 8) anteriorly. Results All the patients survived the procedure. Reoperation has been needed thus far in one for infection of the prosthetic patch placed at the RV outflow tract, and catheter intervention in 2 for mild obstruction across the RV outflow tract. Postoperative catheterization demonstrated: RA pressure 7 ± 4 (6 ~ 20) mmHg (higher than 10mmHg in 5), RVEDV 124 ± 33 (84 ~ 183) % of the anticipated normal value (greater than 150% in 5), RVEF 55 ± 6 (40 ~ 66) %, and Cardiac Index 3.2 ± 0.5 (2.4 ~ 3.7) l/min/m2. The Konno incision, residual pulmonary hypertension, and coronary arterial obstruction preoperatively present, were unfavourable factors affecting these parameters. The presence of a competent trifoliate valve at the RV outflow tract provided higher diastolic PA pressure (11 ± 7 mmHg) than others (3 ± 2 mmHg; p=0.021). In the 5 patients with postoperative RVEDV greater than 150%, use of a pulmonary homograft should have been recommended for better postoperative cardiac performance. Conclusion. RV performance was unlikely impaired even without a competent pulmonary valve in the majority of our patients. Use of a homograft, however, could be preferred in a selected group of patients with deleterious circumstances on postoperative RV function.

184

Long-term follow-up of mechanical versus biological mitral valve replacement in patients aged 1 – 25 years

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Objective To determine the difference in long-term outcome between mechanical and bioprosthesis after mitral valve replacement (MVR) in a very young population with predominant rheumatic heart disease. Method: Between 1978 and 1993, 178 patients(pts) age 1 to 25 years underwent MVR. Preoperative demographics are shown on the table below. Results: There was 10/99(10%) mortality in group M versus 1% in group B (pvalue= 0.0151). Six(6%) of the deaths in group M occurred in patients < 3 years of age. Patients were followed for up to 20 years, mean 11 +/- 6.2 range 7–20 years. 16% were lost to follow up in group M and 10% in group B. Late mortality was 13% in group M versus 3% in group B (pvalue=0.0262). Actuarial survival at 18 years (excluding hospital mortality) in group M was 48.2% +/- 14 versus 89% +/- 7.6 in group B(pvalue<0.005). Freedom from reoperation at 15 years was 77% +/-6 for group M versus 37% +/- 8.5 for group B(pvalue<0.006). Conclusions. In our population, despite the higher need for reoperation, the overall survival of patients with MVR, using a bioprosthesis is significantly better than in patients with mechanical prosthesis. Biological prosthesis should still be considered in young patients particularly young females who wish to have children or those who cannot be anticoagulated.

185

Surgical management of aortic insufficiency in the pediatric age group: re-emphasis of aortic valve cusp-plasty

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Background: To evaluate if aortic cusp plasty can prevent premature aortic valve replacement in severe aortic insufficiency, aortic valve cusp-plasty was used in a selected group of children and the outcome was evaluated. Population: Patients(n=16) with severe aortic valvular incompetence (grade 3–4/4) due to bilobar valvuloplasty (3), tricuspid aortic valve fusion(1) and prolapsing cusps (2) were included. Age ranged from 2.5 to 15.5 yr and weight from 13 to 74 kg. Preoperative NYHA functional class was II in 2 patients and IV in 4 patients. Technique: In all patients the prolapsing leaflet was brought to the level of coaptation by resuspension of the leaflet at adjacent commissures with mattress sutures and re-enforced with 'pledgets' of patient's own pericardium. Two bicuspid valves required post-dilatation tear-repair, one post-dilation tricuspid valve with a prolapsing cusp needed resuspension. Methods: Pre- and post-operatively aortic insufficiency was graded. left ventricular end-diastolic diameter (LVEDD) and shortening fraction (SF) were measured by a blinded observer by Doppler echocardiography. Results: five of six patients improved on a scale from 1 to 4 by at least 2 grades. One still has a grade 3–4 insufficiency. This improvement was statistically significant (p=0.02). The mean preoperative LVEDD was 56.5mm (SEM 2.8) and the mean post operative LVEDD was 44.0 mm (SEM 2.6) (p=0.03). SF was preoperative 35.5%(SEM 1.8) and postoperative 37.7%(SEM 2.5) (NS). No deaths occurred and no valve replacement was required. Exercise tolerance improved in all patients. Conclusion: Native aortic valve cusp repair seems to be a valuable alternative in the management of aortic insufficiency in children and may prevent or postpone aortic valve replacement. This option is important since it allows a more cavalier approach to neonatal balloon-dilatation of valvular aortic stenosis.

186

Surgery for rheumatic mitral valve disease in patients aged under twenty-one years in the south pacific region

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OBJECTIVE The institute provides a pediatric surgical service for New Zealand and the South Pacific region. The patients encountered are unique in that they often present late and postoperative anticoagulation cannot be monitored in some patients due to geographical or socio-economical reasons. Because of this we perform mitral valve repair on severely affected valves that would otherwise have been replaced. We review our results over the last decade. METHODS: Between 1990 and 1999, 71 patients (41 males) aged 13.5+/-4.0 (5.7–20.5) years underwent rheumatic mitral valve surgery. Neighboring Pacific Islanders (PI), Tahiti, Cook Islands made up 53% of patients, New Zealand (NZ) indigenous Maori 30%, NZ Polynesians 15% and Caucasians 1%. Mitral regurgitation was present in 81%, stenosis 5% and mixed disease 14%. Concomitant aortic or tricuspid disease in 72%. Acute rheumatic carditis was present in 45%. Preoperatively 70% of the patients were in NYHA class III or IV with 8 patients requiring preoperative ICU support. RESULTS: Mitral valve repair was undertaken in 42% and replacement in 58%. Concomitant aortic or tricuspid surgery in 69%. The operative mortality was 4%. No death occurred in the acute cardiac group of the 8 patients who required preoperative ICU care. Patients with acute rheumatic fever were not more likely to require mitral valve replacement than those without active carditis (17/33 vs 26/40, p=ns). Follow-up was 86% complete, mean duration 24 (0.9–76.2) months. There were 3 (4%) reoperations (mean duration 3 months) and 1 late death (cause unknown). At follow up, 46 patients were in NYHA class I and 19 patients in class II. CONCLUSIONS: Despite significant preoperative morbidity and severely affected valves, rheumatic mitral valve surgery in children and adolescents can be associated with low operative mortality and good clinical outcome. Acute rheumatic carditis is not associated with an increased need for mitral valve replacement.

187

Repair of congenital mitral valve dysplasia in infants and children

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Objective: surgical management of congenital mitral valve (MV) dysplasia in our pediatric age group remains a therapeutic challenge for the wide spectrum of morphological abnormalities and the high incidence of associated cardiac anomalies. Methods: thirty-eight consecutive children (M/F=20/18) with mean age of 5.2 years (range 45 days–18 years) were treated surgically for congenital MV disease between January 1987 and July 2000. Six patients (16%) were under 12 months of age, while 22 (58%) were younger than 5 years. Twenty-two patients presented with MV incompetence (or prevalent incompetence), while 16 presented with stenosis (or prevalent stenosis). Associated cardiac anomalies were present in 24 patients (63%). Regular mitral valve repair was possible in all. There was one hospital death (2.6%) in a patient with associated aortic and subaortic stenosis, who died for severe left ventricular fibroelastosis after MV repair and Ross procedure. Four patients required reoperation for MV restenosis (re-repair in one, MV replacement with mechanical prosthesis in 3); 1.3 months, 4 months, 27 months and 5.6 years after repair, with no operative death. There was only one late death for prosthetic valve thrombosis 10 months after surgery. At a mean follow up of 78 months (range 14 months–13.1 years) all survivors are

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

asymptomatic and well. Actuarial survival at 13.1 years is 91%. Echocardiography performed in all of them shows no or mild incompetence or stenosis in 71%, while residual moderate incompetence persists in six. Conclusions: mitral valve reconstructive procedures in infants and children with congenital MV dysplasia may be effective and reliable with low mortality and low reoperation rate. Mitral valve repair should always be attempted, especially in infants, despite the frequent severity of MV disease, to avoid the drawbacks of the currently available prostheses.

188
Use of Medtronic Freestyle bioprosthesis in congenital heart disease
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INTRODUCTION: We have been using the Medtronic freestyle bioprosthesis recently in the hope of overcoming some of the limitations associated with homografts and stented valves. The aim of this study was to review the performance of this prosthesis in congenital heart lesions and examine the rate of calcification in these prostheses. METHODS: Prospectively collected data for all patients having freestyle bioprostheses implanted for congenital heart disease were reviewed. Clinical and echocardiographic data were analysed. Limited CT scans are being performed to quantify the degree of bioprosthetic calcification. RESULTS: Twenty-three bioprostheses were implanted. Fourteen were used as RV to PA conduits. There were 5 subcoronary aortic valve replacements, 3 aortic root replacements and 1 pulmonary valve replacement. Median age at implant was 8.0 years (range 13 days to 22 years). Median implant size was 23mm (range 12–27mm). One neonate with truncus arteriosus died perioperatively due to pulmonary hypertension. One conduit was explanted 27 months after repair of truncal truncus because of obstruction proximal to the conduit anastomosis. Radiographic examination of the explanted conduit demonstrated no significant calcification and the leaflets remained pliable and mobile (see figure 1). There has been no incidence of significant prosthetic regurgitation, thromboembolism or endocarditis at mean follow-up of 19.1 months (range 7 to 35 months) and all gradients are less than 25mmHg at present. CONCLUSIONS: The Freestyle valve has proved to be a versatile bioprosthesis in children for a variety of indications. Post-operative haemodynamic performance and freedom from valve-related complications has been excellent. The results of CT scan quantified bioprosthetic calcification are awaited.

Session 18: General Pediatric Cardiology, Prognosis/Natural History

189
Influence of fetal and postnatal growth on heart rate variability in young infants
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The Barker hypothesis states that fetal undernutrition programs later cardiovascular disease. Heart rate variability (HRV), a measure of cardiac autonomic control, was analyzed in infants to assess the hypothesis that early undernutrition may induce autonomic dysfunction that could play a role in that programming. ECG data were collected in 505 healthy infants aged 5 to 12 weeks (birth weight 2400 to 4830g, mean 3206g). HRV measures were calculated over 400min of sleep. Statistical associations between 5 time-domain HRV indices (SDNN, SDNNi, SDANN, rMSSD, pNN50) 5 frequency-domain indices (spectral power in the very low (VLF), low (LF) and high (HF) frequency regions, total spectral index (TSI) and LF/HF ratio), and early growth indices were established by linear regression analysis. A significant positive correlation ($p<0.05$) between birth weight, neonatal weight to head circumference ratio and postnatal weight gain, and the HRV indices mostly influenced by the sympathetic activity (SDNN, SDNNi, SDANN, VLF, LF, TSI) was demonstrated in 11 and 12-week-old infants. A slighter correlation was found in younger infants between the same indices. Our data suggest the influence of fetal and postnatal growth on the programming of the autonomic nervous system beyond the neonatal period. Increased sympathetic tone characterizing infants with impaired growth. It may be one of the mechanisms that link early impaired growth to later cardiovascular disease.

190
Correlation between one year neurodevelopmental tests and eight year achievement tests in cohort of infant heart surgery patients
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The choice of neurodevelopmental endpoints for trials in congenital heart patients must balance the need for relevance to long-term function against the expense entailed in follow-up of children over many years. The purpose of our study was to assess the predictive validity of scores at age one year on the Bayley Scales of Infant Development for later ability and achievement in children who undergo infant heart surgery. Our goals were pursued using the Boston Circulatory Arrest study database of children with D-TGA who underwent the arterial switch operation before age 3 months. In this study, children underwent testing at age one year with the Psychomotor Development Index (PDI) and Mental Development Index (MDI) of the Bayley Scales and at age eight years with a battery including the WISC-III and the WIAT. We examined the correlation between outcomes using Spearman correlation coefficients, testing both at ages 1 and 8 years was performed in 144 children. One-year PDI scores were significantly correlated with 8-year Full Scale IQ ($r=.22$, $P=.018$), Verbal IQ ($r=.21$, $P=.01$), Performance IQ ($r=.19$, $P=.02$), and Composite Math Achievement ($r=.20$, $P=.01$), and with a trend toward Composite Reading Achievement ($r=.15$, $P=.08$). One-year MDI had somewhat greater predictive value than PDI with significant associations for Full Scale IQ ($r=.30$, $P=.000$), Verbal IQ ($r=.31$, $P=.0001$), Performance IQ ($r=.21$, $P=.005$), Composite Reading Achievement ($r=.24$, $P=.005$), and Composite Math Achievement ($r=.24$, $P=.003$). These data suggest that neurodevelopmental outcomes measured at 1 year of age in children with (CHD) may provide information about long-term function. The correlations, although significant, are modest in magnitude, explaining relatively small amounts of variation in outcomes at age 8. High MDI and PDI scores at one year of age do not obviate the need for long-term follow-up.

191
An evaluation of health outcomes and cost-benefit analysis of a pediatric cardiology outreach program
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Introduction: The Pediatric Cardiology Outreach Program (PCOP) was established to provide clinical and cost effective tertiary care in 8 regional centers across British Columbia. The study reports the results of the program over the past 5 years. Methods: We retrospectively reviewed our clinical database from 1994 to 1998. We identified patterns of referral, problems with patient management, follow up plans, and hazardous or undesirable outcomes. We also assessed program growth and developed a series of cost-to-benefit scenarios to determine whether the program was cost effective for both our hospital and the families of children with CHD. Results: There has been a consistent annual trend towards a reduction in problems with patient management (16.1% to 10.2%), hazardous outcomes (0.9% to 0.8%) and undesirable outcomes (5.7% to 1.7%) involving patients seen in PCOP. Between May 1997 and March 1999, 43% of children were recommended for ongoing follow-up at PCOP, 38% were discharged from cardiac follow-up, and 18% were referred to our tertiary care hospital (BCCH). Between 1994 and 1998 there was an increase in the clinic locations from 4 to 7 (175% increase), clinic days from 11 to 33 (300% increase), and patient visits from 115 to 476 (414% increase). Travel costs for staff (cardiologist, nurse, echocardiographer) to attend clinics range from $1,985 to $4,595, while costs for an individual family to travel to BCCH range from $429 to $2,360. While 12–15 patients are seen at each clinic, the program is cost effective if as few as 8 patients are seen (range 1.7–7.9). Conclusions: The PCOP has grown 400% since 1994 providing a clinically useful and cost effective service. The decreasing trend in problematic outcomes likely reflects increased community awareness of patient management issues.

192
Acquired von Willebrand's (VWD) disease in children with patent ductus arteriosus
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There is only one report of acquired vWD in noncyanotic children with cardiac defects. We prospectively examined 12 consecutive children with a large

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

patent ductus arteriosus (PDA) for acquired vWD. 4/12 children (33%) had a deficiency of the largest multimers similar to vWD type 2B. These children were younger (2.0 vs. 4.8 years) than the remaining cohort. vWF antigen (57%) and collagen binding activity (44.5%) were lower than in the remaining 8 pts. One pt had a prolonged aPTT; FVIII activity, BMBL, fibrinogen and platelet counts were normal in all pts. History did not depict a clear difference in bleeding tendency between the two subgroups. Cardiac catheterization revealed a higher shunting across the PDA in children with vWD (Qp/Qs 1.47 ± 1 vs. 1.33 ± 1). Following PDA occlusion with an Amplatzer-duct-occluder, pts. with vWD showed progressive recovery of the large multimers confirming the acquired nature of the disorder. To our knowledge this is the first report of an association between acquired vWD and PDA. The presence of acquired vWD in 33% of children with clinically relevant PDA suggests a considerable prevalence of this disorder.

194
Tetralogy of Fallot in subsaharian Africa: experience of Institut de Cardiologie d'Abidjan
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From January 1980 to December 1999, 114 cases of tetralogy of Fallot have been hospitalized at Institut de Cardiologie d'Abidjan. The mean age was 8.7 years (extremes: 6 months and 29 years), the mean weight was 16.4 Kg (extremes: 6.2 and 55 Kg). Twenty three cases had cyanotic spells while 91 cases had few symptoms. The echographic and cineangiographic studies showed 65 cases(57%)of regular forms and 49 cases (43%)of irregular forms. Thirty one patients (27.2%)had only medical management with 5 deaths (16%). Twenty five patients (21.9%) had palliative with 2 deaths (8%). Sixty six patients (57.9%) had repair with 20 operative deaths (30.3%). The operative mortality, high in the 5 first years 8/14 (57%) dropped the 10 last years 6/42 (14%). The risk factors have been the low age and irregularity of the institution. The mean follow up of 7 years (extremes: 1 and 17 years) allowed to notice the good quality of life in the survivors with 2 cases (4.9%)of reoperations and 3 cases of late deaths (6.8%)including 2 extracardiac deaths. The inclusion mortality in our study was 23.6% with the best quality of life in operated group without late complications.

195
A new approach to continuous blood pressure monitoring during exercise-ECG
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Conventional non-invasive blood pressure measurements utilizing inflatable cuffs around arm, wrist or finger (Riva-Rocci, oscillometric) only allow readings at discrete times with intervals in the range of minutes. However, during stress ECG tests a continuous blood pressure monitoring indicating dynamic blood pressure changes would enable much better insight in the cardiovascular regulating system. Our new approach is based on the dependence of the instantaneous heart rate, the pulse wave velocity and derived parameters on the blood pressure. Systolic and diastolic pressures are computed on a beat to beat basis using an artificial neural network (ANN). As input for the ANN we calculated generalized input transfer functions, which are normalized with regard to the individual abilities. Sensor requirements are an ECG and the simultaneously measured photoplethysmogram (PPG). Stimulation of blood pressure reactions using standard exercise tests on bicycle ergometers and the orthostatic stress test (Schelling) on more than 30 patients in the range of 8 to 65 years yielded learning results for our system. Placing the PPG-sensor at the ear lobe together with signal enhancement including amplitude regulation and short time correlation of adjacent pulse waves gives a satisfying rejection of motion artefacts even during exercise. Our new method has been verified using a 24-h blood measuring device, several oscillometric and Riva-Rocci instruments and a Portapres system. The investigation reveals during exercise ECG immediate systolic pressure drops during relaxation intervals not visible in the conventionally measured data. The new method is not disturbing the patient at all and yields more accurate information on blood pressure behaviour than previously known.

195
Doubly committed VSD with absent subarterial conus – the "more-spelling" form of tetralogy of Fallot
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A rare form of double outlet right ventricle (DORV) has been described with absent subarterial conus and doubly committed VSD. To assess the incidence and anatomic/clinical features of this anomaly, we reviewed the medical records of all pts diagnosed with tetralogy of Fallot or DORV at Children's Hospital of Wisconsin from 1991 through 1997 (n=190). Nine patients (4.7%) had absent subarterial conus, all associated with mesially related great arteries, dextroposition of the aorta (Ao not abut with at least 50% override of the muscular septum), and concordant AV connections. The roof of the VSD was formed by the contiguous Ao and pulmonary valves in all cases, and mitral-Ao continuity was always present. All had a dilated Ao root (12.2 +/- 2.2mm) and mild-moderate valvar pulmonary stenosis (gradient 42.6 +/- 12.1mmHg) with significant annular hypoplasia (diameter 6.2 +/- 2.1mm) at the mitral necessary only. The PDA was a good size in all pts, the LPA was discontinuous in 1 pt and stenotic at its origin in 3 others. A right aortic arch was commonly present (4/9 pts). The pt with discontinuous PA's required Ao-PA shunt placement, no other pt required palliative surgery, none had hypercyanotic spells, and all had a definite repair at a relatively late age (21.4 +/- 14.5mo) with no mortality. Two patients had balloon pulmonary valvuloplasty at 2 and 4 mo because of increasing cyanosis which delayed surgical repair until > 12 mo of age. In summary, these patients are remarkably homogeneous group that likely represents the extreme form of conal hypoplasia in the spectrum of tetralogy defects. The absence of subarterial conus appears to alleviate the risk of hypercyanotic spells, predicts excellent response to balloon valvuloplasty if necessary, and allows for successful late definitive repair.

196
Transcatheter versus surgical closure of patent ductus arteriosus: Changing trend of treatment modality?
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To evaluate the effects and problems of the application of transcatheter occlusion (TCO), and the changing trend of treatment modality of patent ductus arteriosus(PDA), we made a retrospective analysis of our experience with 228 PDA pts from Nov. 1994 to Oct. 1997 at Samsung Medical Center. Observations made were as follows. 1) 134 pts were managed with TCO, while 94 pts (including 51 neonates) were operated(OP). 2) All 51 neonates resistant to medical management were operated, and most of the pts over 12 months of age were managed with TCO since 1996 (89 among 92 pts). Annual number of TCO/OP over 1 month of age was '94 – 0/1, '95 – 11/15, '96 – 16/4, '97 – 19/6, '98 – 36/7, '99 – 34/6, 2000 – 19/4. 3) Devices used for TCO were single coil in 67 pts (Gianturco detachable(GD) in 37, Duct-Occluder (DOD) in 29, Gianturco embolization coil(GE) in 1), multiple coils in 35 pts, Amplatzer ductal device in 24 pts, Sideris button device(BD) in 7 pts and Rashkind device in 1 pt. All BD and Rashkind cases were pre-1998 pts, and our new multiple coil technique using DO & GD was applied since 1997. Amplatzer device has been introduced in 1999. 4) Among the 94 OP pts, 6 showed residual leak on echo. Dysplasia and 2 of them needed TCO. Among 134 TCO pts, there were 8 technical failure and 1 embolization of the coil and 3 significant residual leak, who required 2nd TCOs, at the early stage of TCO application. Another 15 cases showed clinically insignificant residual shunt. 5) Our current treatment guide line for PDA is ; Neonate resistant to medical management – OP, small ductus – single coil (mostly GD), medium to large ductus & adult ductus – multiple coil or Amplatzer device.

197
Interrupted inferior vena cava: associated anomalies in 179 cases
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To delineate anomalies associated with an interrupted inferior vena cava (IVC), in cases with left atrial isomerism (LAI) as well as in those without, we reviewed all 179 cases occurring at The Hospital For Sick Children, Toronto, from 1950 to 1997. In 151 cases (84%), there were other features of LAI such as polysplenia, bilateral bilobed lungs, left atrial rhythm, congenital AV block. The spektrum of heart defects were more complex in the LAI group ($p<0.001$, chi-square test). The LAI patients had an otherwise normal

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

heart in 21 (14%) versus 9 (32%) in the other group. There were balanced heart defects in 67 (44%) of the LAI group (mainly double outlet ventricle) versus 6(3% in the other (mainly VSDs). In 62 (42%) of the LAI group there were unbalanced defects unsuitable for biventricular repair, versus 7% in the remainder. AV block was present in 10 LAI patients (7%) only. Extracardiac anomalies were found in 45 LAI patients (30%) and 10 (36%) in the other patients (p=0.2, NS). The striking feature in the LAI group was biliary atresia in 14 pats, whereas no typical extracardiac malformation could be noticed in the other group. We conclude that interrupted IVC is rarely encountered in conjunction with LAI but cases without LAI is also found. In those pats with LAI the heart defects are generally more complex whereas the extracardiac malformation frequency is comparable.

198

Rheumatic heart disease in children and adolescents: evolution and predictive factors of significant chronic valvar lesion

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Purpose: To trace the evolution of valvar lesions based on clinic and Doppler echocardiographic evaluations and to identify the predictive factors of significant chronic rheumatic valvar disease. Method: The research was a cohort study on 298 children and adolescents aged between 3.7 and 15.2 years (mean age 10.2 ± 2.6 years). The total follow up period varied between two and 15 years, resulting in 1163 patients-year (mean of 5.4 ± 2.7 years). The diagnosis of rheumatic fever was based on the revised Jones criteria. In the acute phase, 139 patients were submitted to Doppler echocardiography study, while in the chronic phase, all the 298 patients were submitted to at least one Doppler echocardiography exam. The time zero was defined as the date of the beginning of the acute phase. A final event was defined as any type of severe mitral and/or aortic valvar lesions according to the Doppler echocardiographic classification occurring at least two years after the onset of first episode of acute rheumatic fever. The variables associated to the significant chronic rheumatic valvar disease were initially identified through the Kaplan-Meier estimation survival. The differences between the variable categories were evaluated by using the log-rank test. By using the Cox regression model, was estimated the relative risk of significant chronic rheumatic valvar disease. Results: According to the Doppler echocardiographic evaluation, of the 298 patients studied, 91 (15.1%) showed significant lesion of mitral and/or aortic valve. None of the patients that presented subclinical valvulitis developed significant chronic rheumatic valvar disease during the follow-up period. According to the clinical evaluation in the chronic phase, there was larger regression of the valvar lesion (29.4%) when the carditis was of mild degree, smaller regression was observed when the carditis was of moderate degree (15.2%). There was no involution in the cases of severe carditis. Of 112 patients with the clinical evidence of carditis, eight (7.1%) showed heart alteration in the chronic phase, according to clinical evaluation. The variables: color, family income, level of mother's education, weight at the admission, carditis degrees and recurrences were associated to the final event, according to the univariate analysis. After the adjustment for the multivariate model, only three variables were viewed to be independently associated to the significant chronic rheumatic valvar disease: the carditis degree, the occurrence of recurrent attacks and the level of education of the patient's mother. The study also showed the existence of significant interaction between the level of education of the patient's mother and occurrence of rheumatic fever. Conclusion: The evolution for significant chronic rheumatic valvar disease was more frequent among the patients that presented moderate or severe degrees of carditis, recurrences of acute rheumatic fever and those whose mothers had low level of education.

199

Does prematurity predispose to pulmonary vein stenosis?

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Pulmonary vein stenosis (PVS) in normally connected veins is rare, and leads to progressive pulmonary hypertension and cardiac failure in infancy. 8 consecutive patients with this condition presented between June 1982 and July 2000. Age at diagnosis ranged from 1 day to 2.5 years. Initial diagnosis was made by echocardiography (and confirmed by catheterisation) in 4 (50%)patients. The diagnosis was made at the initial cardiac assessment in 2 patients but was identified subsequently in 6 at a median of 3 months after referral. 5 patients were born prematurely, age at referral from 5–6 months, at a corrected age of 3 to 3.2 months, and weight from 2 to 4.3 kg. 2 of the 3 premature infants died 1.5 to 3 months after diagnosis and operation. 5 were full-term infants (FTIs); age of referral 4 hours-2.5 years. Associated cardiac anomalies in 3 FTIs were more complex than those associated with prematurity. Of the 5 FTIs 2 had surgical procedures aged 3 to 7 months. 3 died less than 1 month after diagnosis or operation. 3 of 8 had stenosis of all 4 PVs (1 patient was premature). They tended to be referred earlier than those with 1 or 2 PVS but not significantly. In our study 5 of 8 patients died at a range of 0.75 to 1.5 months after diagnosis or operation. 3 survived but are severely symptomatic. The diagnosis depends on a high index of suspicion. Crosssectional echocardiographic examination of all PVs should be mandatory in patients with pulmonary hypertension, particularly those with a structurally normal heart. Both the number of PVs involved and severity of stenosis influence outcome, which is poor. Our study indicates an association with prematurity.

200

Decreased aortic elasticity in operated versus non-operated Marfan patients

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Background: Following aortic root replacement Marfan patients may develop complications in the aortic tract beyond the aortic root, even without severe aortic dilation. Aortic stiffness parameters are related to aortic wall behaviour and may serve as additional risk factors for aortic complications before the aorta is dilated. Purpose: To compare aortic elasticity between electively operated and non-operated Marfan patients. Methods: 20 Marfan patients with elective aortic root replacement (mean age 36±13 years, 21 female, 9 David) and 20 non-operated Marfan patients (mean age 32±8 years) underwent magnetic resonance imaging of the entire aorta. Aortic diameters and distensibility (D) at 3 levels of the descending aorta were assessed (level 1: descending thoracic, level 2: diaphragm, level 3: above the aortic bifurcation). Furthermore flow wave velocity (FWV) between level 1 and 3 was assessed. Results: Aortic diameters were normal at all levels in the operated and non-operated group. However, the operated patients had a significantly decreased local distensibility (D) at the level of the descending thoracic aorta compared to the non-operated patients (2.5 ± 1.5 vs. 5.6 ± 2.6 x 10−3mmHg-1 respectively P=0.01). No significant difference was found in aortic flow wave velocity (FWV) between the operated and non-operated group (5.6 ± 1.5 vs. 5.7 ±1.3 m/s). Conclusion: Following elective aortic root replacement Marfan patients show decreased local elasticity in the descending thoracic aorta compared to non-operated Marfan patients. This might be of clinical importance in the follow up of operated Marfan patients.

Session 19: Catheter Interventions

201

Results of balloon valvuloplasty of critical neonatal aortic stenosis: up to 12-years follow-up

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The retrospective study was carried out to assess results of valvuloplasty in critically ill neonates with AS. Inclusion criteria were age up to 28 days and critical AS associated with severe LV dysfunction, low cardiac output syndrome or duct-dependent systemic circulation. In consecutive sample of 62 newborns, valvuloplasty was performed at age 0 to 28 (median 2) days. Body weight at the procedure was 3.19±0.63 kg. Balloon-to-annulus diameter ratio was 1.00±0.08. Fluoroscopy time was 16.7±8.5 minutes. In 45 survivors the follow-up period was 2 month to 12.8 years (median 3.6 years). Total mortality was 28.6% (early 9.5% and late 19.1%). Re-intervention rate was 30.2% (19.0% of surgical et interventions). Actuarial probability of survival 12.8 years after the procedure was 70±6% and of survival without a re-intervention 31±14%. Respective values before and after the procedure were as follows: peak gradient (mm Hg) 64±29 and 37±14 (p<0.001), mean gradient (mm Hg) 39±20 and 22±9 (p<0.001), left ventricular shortening fraction (%) 29±12 and 39±9 (p<0.001). The values did not change significantly over the follow up period. Median ECHO-grade of aortic regurgitation increased from 0 to 1 with the valvuloplasty and to 2 at the last follow up (both p<0.001). Aortic annulus grew from 6.6±1.0% of

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

normal value at time of procedure to 85±16% at the latest follow-up ($p<0.001$). We conclude that the valvuloplasty is capable of saving majority of the newborns with the critical AS. It provides lasting reduction of the gradient, improvement of LV function, and potential for aortic annulus growth. However, the aortic incompetence caused by the procedure is progressive and re-intervention rate is high. Accordingly, the valvuloplasty must only be considered a life-saving palliation. Acknowledgement: supported by grant NA5263-3, Ministry of Health, CR.

202

Balloon angioplasty in peripheral pulmonary stenosis

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Efficacy of balloon angioplasty (BA) for branch pulmonary artery (PA) stenosis is still unknown, and it was evaluated at eight institutions in Japan. During a 5-years-study period, 349 patients underwent BA for 420 vessels, including 74 main PA and 356 branch PA, at a mean age of 6.1±15.5 years. Most patients had associated congenital heart diseases, tetralogy of Fallot being the most common. The initial success rate of BA varied depending on the success criteria, 48% (an increase in diameter >50% of pre-dilation value), 60% (post-dilation diameter> 70% of non-stenosed vessel), and 63% (post BA pressure gradient <50% of pre-dilation value). Before BA, the right ventricular / left ventricular pressure ratio was 0.65±0.24 and after BA it decreased to 0.50±0.17. In 61% of patients, it decreased to <0.50. Before BA, lung perfusion scintigraphy revealed only 23±13% of total blood flow went to the affected side, and the value increased by 9±36% after BA. Complications occurred in 20 patients (6%), pseudoaneurysm of the PA being the most frequent complication (5 patients). No death related with BA. Follow-up studies in 49 patients showed a restenosis rate of 48%. Conclusion: BA for branch PA stenosis can be performed safely with a success rate of 50-60%. Further technical improvements is necessary to reduce the restenosis rate.

203

Percutaneous valvuloplasty in juvenile mitral stenosis with metallic commissurotome and Inoue balloon – a retrospective comparison of immediate results

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Valvotomy with Inoue Balloon provides significant symptomatic benefit in patients with juvenile mitral stenosis. Metallic commissurotome (Cribier device) is now being evaluated for mitral valvotomy in adults with mitral stenosis. There are no reports of use of commissurotome in juvenile mitral stenosis. We hereby present data of immediate results of commissurotomy using metallic commissurotome and Inoue balloon in 44 cases (M - 18, F - 26). The age range was 8–18yrs. Mean height was 144 (± 7.9) cm. All the patients were symptomatic (NYHA class II – 22, class III – 20). Two patients were in pulmonary oedema when the procedure was done. Four patients had previous valvotomy (BMV – 2, CMV – 2). Mean mitral valve score was 7 (± 2.7). Procedure was successful in all patients. A metallic commissurotome was used in 20 patients while the remaining 24 underwent valvotomy with Inoue Balloon. In the Inoue balloon group, the MVA increased from 0.92 (± 0.19) sq cm to 1.79 (± 0.23) sq cm, while in the metal commissurotome group MVA increased from 0.94 (± 0.21) sq cm to 2.22 (±0.26) sq cm. Mean gradient across mitral valve decreased to 8.6 (± 3.26) mm Hg from 25.74 (± 7.4) mm Hg in the balloon group. In the metal commissurotome group gradient decreased from 24.94 (± 8.2) mm Hg to 5.2 (± 2.06) mm Hg. Fall in PA pressure was significant in both the groups following valvotomy. One patient developed grade III MR following balloon valvotomy, which was conservatively managed. A retrospective comparison of the immediate results obtained with the two modalities, revealed that the valve areas and gradient reductions achieved with the metallic device were better than those with the balloon ($P < 0.05$).

204

Evolution of a new device for interventional closure of the atrial septal defect (asd): intermediate results in the porcine model

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Over the past years numerous different devices for interventional ASD closure have been suggested and tested. All inherit specific drawbacks and limitations ranging from lack of retrievability, bulkyness, to limited manoeuvrability, risk of embolisation or limitations in the present or future use of NMRI. The new device we thought to develop to include the strongholds of previous devices consists in principle solely of a nitinol tube which is directed by a laser knife and thermally treated to form two interconnected umbrellas. The interconnecting portion caps itself inside the defect. In order to achieve complete closure of the defect then fibre meshes are fixed in each of the umbrellas and the interconnecting portion. Placement of the device is possible through 8 Fr sheath and can be done over a wire. The delivery system allows the device to reach its definitive position before final release and at that stage does not apply any forces to it. This approach allows a mono block design of the device without the need to connect wires. It is available in different sizes ranging from: 18 mm to 28 mm diameter of the umbrellas. The device was tested in a porcine model of ASD (n=10, weight: 20 – 25 kg, age: 8 to 10 weeks) where the the foramen ovale was dilated by angioplasty leading to a ASD of 9 to 12 mm in diameter. Placement of the device was possible in all cases, retrieval was possible when stenosed. Selective angiography into the left atrium demonstrated complete closure and the post-mortem examination revealed correct placement without entanglement of any neighbouring structures. The results confirms the feasibility of the new concept. Chronic studies are necessary and under way before approaching clinical studies.

205

Multiple atrial septal defects : transcatheter closure with single Amplatzer septal occluder

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Purpose: Retrospective analysis of feasibility of single Amplatzer atrial septal occluder (ASO) to close multiple secondum atrial septal defects (ASD). Methods: From 278 of our ASD patients (pts) treated with ASO, 36 at a mean age of 14 (range 1 – 55)y had multiple ASDs. During TEE mean size of biggest defect was 10.9 (range 5–19) mm, smaller defect – 3.6 (range 2–9) mm, and mean distance between both defects – 5.5 (range 2–12)mm. Special attention was paid to keep still guide wire position in the biggest defect and inflation of Amplatzer strong balloon in an attempt to achieve stop flow through additional ASD. Results: In 30 pts the procedure was performed with single ASO. Mean diameter of ASO was 16 (range 5- 28) mm and was equa. to the stretch diameter in 19 pts, while 1–4 mm oversizing was applied in the rest 11 pts. Mean fluoroscopy time was 15 (range 2.2 – 38) min. Complete closure in colour Doppler was achieved in 23/26 (88%) after 24 h, 28/30 (93%) after 1 month, 34/30 (80%) after 3 month, 18/28 (66%) after 1 year and all 16 after 2 years. Residual leaks resolved or diminish with time (were initially observed in 9/ 8 pts in whom the distance between two defects exceeded 7 mm). In 2 pts – one with 18/9 mm ASDs at distance of 7 mm and second with 15/7 mm ASDs at distance of 11 mm – 4 devices were implanted with complete closure after 24 h. No complications occurred. Conclusion: Multiple ASDs can be effectively closed with single Amplatzer atrial septal occluder. Strategy of transcatheter closure depends on the size and distance between both defects and dilatability (performed with Amplatzer sizing balloon) of septum.

206

Transcatheter closure of high pulmonary pressure patent ductus arteriosus with the Amplatzer muscular ventricular septal defect occluder

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Objectives: In the report we described the successful use of Amplatzer muscular ventricular septal defect occluder (AMVSDO) for the treatment of high pulmonary artery pressure PDAs (hypertensive PDAs). Methods: From May 1997 through August 2000, 7 patients (4 girls), aged 5 to 12 years, with

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

large PDA and systemic or near systemic pulmonary artery pressure underwent attempted TC closure using the AMVSDO. The device consists of two low profile disks made of Nitinol wire mesh with a 7-mm connecting waist. Balloon PDA occlusion was performed in all pts before TC from the venous side. The prosthesis size was chosen according to the measured balloon 'occluding' diameter. A 6F to 7F sheath was used for the delivery of the device. All pts underwent a complete hemodynamic and angiographic study one year after occlusion. Results: The mean PDA angiographic size was 9.8 ±2 mm (range 6.5 to 12 mm), and the mean AMVSDO diameter was 11.4±2 mm (range 8 to 14 mm). The Qp/ Qs ranged from 1.9 to 2.2 (mean 2.1±0.1). The mean systolic pulmonary artery pressure before, during balloon occlusion, immediately after the procedure, and at 1-year follow-up was 103±12 mm Hg, 64±6 mmHg, 58±5 mm Hg, and 37±10 mm Hg, respectively. Complete angiographic closure was seen in all pts. Fluoroscopy time was 8.2±3 min (range 6 to 14 min). No complications occurred. Conclusions: AMVSDO is an efficient and safe device for the treatment of hypertensive PDAs. The right disk of the device reduces stability of the occluder across the PDA in the presence of high pulmonary artery pressure.

207

Stenting of branch pulmonary arteries following the arterial switch operation

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12 patients aged 6–12 years (median 8.2) with haemodynamic stenosis of the left and/or right pulmonary arteries following the arterial switch operation underwent transcatheter stent implantation using a Palmaz-Wallstent expandable stent. In 9 patients with bilateral branch stenosis, the right ventricular to aortic systolic pressure ratio fell significantly (mean 0.75 to 0.34) following stenting and balloon dilatation to 12–15mm. In 3 patients with moderate or severe left pulmonary artery branch stenosis, the pressure ratio fell from a mean of 0.42 to 0.28 following dilatation of the stent to 12mm. At follow-up, 0.5–4 years later by cardiac catheterisation and angiography, 2 patients required further balloon dilatation of bilateral stents. There was no angiographic evidence of stenosis in 6 and mild stenosis in 2. There was a mean peak to peak gradient of 12mmHG and 10mmHg across the proximal left and right pulmonary arteries respectively. No patients developed aneurysms, intimal proliferation, occlusion of pulmonary artery branches or myocardial ischaemia. Stent implantation is the treatment of choice for pulmonary artery branch stenosis in older children following the arterial switch operation.

208

Interventional catheterization management of perioperative peripheral pulmonary stenosis- balloon angioplasty or endovascular stenting

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Background: Balloon dilation (BD) of peripheral pulmonary stenosis (PPS) at a surgical site in the early postoperative period is a risk factor for vessel rupture. Methods: We reviewed operative reports and catheterizations in patients undergoing interventional therapy for PPS at a surgical site less than 7 weeks after operation. Successful dilation (SD) was defined as ≥50% increase in proximal to distal diameter. Outcome variables included survival, change in vessel diameter, and complications. Results: From 1984–2000, 17 patients had 19 proximal pulmonary arteries dilated 1 to 46 (median 8) days postoperatively. Median age and weight were 3.1 yr and 12.7 kg. Three arteries were initially totally occluded. Seventeen arteries had initial BD with post intervention imaging available in 15. 8 arteries had SD. The arterial diameter increased from 3.9±2.6 to 5.5±2.8 mm ($p<0.001$). None of these arteries had stents placed without prior BD with diameter increasing from 1.3 to 9 mm and 8.2 to 14 mm. A stent was placed in 1 of 7 arteries prior to 1993 and in 10 of 12 arteries thereafter ($p<0.004$). Two early patients (<1%) had catheterization related deaths due to vessel rupture after BD. Another patient had an luminal tear produced by BD treated with stent placement. Conclusion: BD produces SD in approximately one-half of the procedures but is associated with morbidity. Stent placement produces greater increase in vessel diameter than BD alone. Stents by providing vessel support can prevent vessel tear, reduce the acute complication rate, and avoid early reoperation in this patient group.

209

Predictors of residual defect following placement of Amplatzer ASD occluder device

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Background: Residual defect (RES) is the most common complication following closure of atrial defects using transcatheter techniques. Factors that predict the occurrence of RES have not been identified. Method: Eighty-five consecutive patients who underwent Amplatzer device closure of atrial septal communications between February 1997 – February 2000 were studied. The associations between RES and clinical, echocardiographic and catheterization parameters were explored using contingency table and logistic regression (simple and multiple) analyses. Results: Ninety-six devices were placed in 85 patients. Eighteen patients (20%) had RES by echocardiogram at least once during the follow up period. All but one patient had RES <3mm. Seven patients (8%) had RES at latest follow up. SVC rim less than 8mm ($p=0.008$), smaller LV ($p=0.03$) or MV ($p=0.06$) same chevere atrial septal length at 30° TEE plane ($p=0.04$), use of multiple devices ($p=0.06$), and greater Qp:Qs ($p=0.07$) were associated with greater chance of RES. After indexing to body surface area (BSA), larger device diameter ($p=0.05$), larger diameter of ASD ($p=0.08$), and longer atrial septal length at 0° TEE plane ($p=0.09$) also were associated with increased chance of RES. Multivariate analysis showed that SVC rim less than 8mm (adjusted odds ratio = 10.1, $p=0.001$) and smaller length of atrial septum at 30° TEE plane ($p=0.04$) were independent predictors of RES. Age, gender, weight, height, BSA, type, circularity or stretchability of atrial defect, and presence of atrial septal aneurysm were not associated with the occurrence of RES. Conclusion: Smaller defect rim size and shorter atrial septal length at 30° TEE plane are statistically significant predictors of RES following placement of Amplatzer occluder device. But the significance of the latter is unclear. Better understanding of the atrial septal morphology is necessary.

210

Comparative use of cook detachable coils and Amplatzer Duct Occluders for percutaneous closure of medium to large sized patent ductus arteriosus

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The transcatheter closure of a medium to large sized patent ductus arteriosus (PDA) is challenging especially in infants. The purpose of this study was to analyse further experience with two retrievable closure devices: the Amplatzer Duct Occluder (ADO, approx. 1400 $) and Cook (Jackson) Detachable Coils (CDC, approx. 700 $). In 4 centers we analysed 106 patients (pts); age: 2 months–16 years; 3.2–66 kg); respectively 90 pts had a medium (group A; 2–4mm) and 16 a large (group B; 4–9mm) PDA. Of group A, 40 pts (PDA=2.4 +0.5mm) received an ADO and 50 pts (PDA=2.8 +0.4, $p<n$) received a CDC. A complete closure was possible in 36 pts by using an ADO, 47 pts by using CDC after follow up of one year ($p=n$). Fluoroscopy time was 12.7 min for ADO and 8.9 min for CDC ($p<0.05$). No major complications occurred. Of group B, 14 pts (PDA=4.8) received a ADO and 2 pts (PDA=4 and 5 mm) received two CDC's each. Fluoroscopy time in group B was 11–21 min. A complete closure after one year was possible in 12 pts by using ADO and in 1 pt by using CDC. And obstruction of the main pulmonary artery occurred in one case with an ADO. Conclusion: Both PDA closure devices can be equally used for medium PDA's with a similar high closure rate and no major complication but CDC have a shorter fluoroscopy time. For large PDA's physicians mostly choosed ADO although the costs are higher.

211

Does nickel toxicity occur after transcatheter placement of nitinol (nickel-titanium) devices?

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As a result of the unique shape memory property of nitinol, the alloy has become popular in surgical and transcatheter interventional devices. A family of such devices are the Amplatzer(r) Occluders well advanced in the multicentre evaluation of intracardiac septal defects and patent ductal arterial occluders, with very favourable result. Nickel toxicity has been of particular

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

concern with these devices because of the high metal mass. The purpose of this study is to measure the Nickel level six months after implantation and compared to that before device placement. Since April 1998, 101 patients, age 0.4 – 78.2 years had 118 Amplatzer(r) Occluder placed to occlude atrial septal defects, patent ductus arteriosis, fenestrated Fontan, extracardiac vascular anomalies and ventricular septal defect. In 19 patients, age 2.3 – 34.5 years (mean 11.3, SEM ±1.9, median 9.5 years) weighing 11.7 – 134.0 kg (mean 39.4, SEM ±5.8, median 32.7 kg), blood was collected prior to device placement and repeated 6 months later for trace element analysis. Nickel was analyzed by inductive coupled plasma mass spectroscopy. There Amplatzer(r) PDA Occluders (patent ductus arteriosus n=1, axillary artery fistula n=1 with 2 devices), and 16 Amplatzer(r) Septal Occluders (atrial septal defects n=14, fenestrated Fontan n=2), were 4 – 24 mm were used. The mean nickel level before device placement was 65.8 mol/L (SEM ±6.5, median 71.0 mol/L) and 68.3 mol/L (SEM ±4.6, median 71.5 mol/L) six months later. There was no significant change in the blood nickel level before and after placement of Amplatzer(r) Occluders. There was no clinical presentation of nickel toxicity in any of the patients. In conclusion, the Amplatzer(r) Occluders are biocompatible, and do not cause significant release of nickel six months later. To date clinical nickel toxicity has not been observed.

Session 20: Cardiac Imaging: Echo-2-D, 3-D, TEE

212

Two-dimensional echocardiographic studies on therapeutic effect of catheter intervention of congenital heart disease

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Seventy-six cases of congenital heart disease (CHD), aged from 1 months to 16 yrs (mean 5.6 yrs), including PDA in 57 (group 1), PS in 15 (group 2), in 2 (group 3), after catheter intervention were studied by two-dimensional echocardiography (2-DE) with Doppler technique and Color Doppler Flow Imaging (CDFI). Follow-ups were done after 1 day, 1,2, 6 months, 1, 2 and 6 years after intervention. In group 1, Rashkind devices were used in 44 (aged from 8 to 36 years, mean 10.5 yrs). Cook coil devices 8 (aged from 5 to 12 yrs mean 8.3 yrs). Amplatzer devices in 6 (mean 6.5 yrs). In group 2, single balloon PBPV was used in 12, double balloon in 1, balloon plus modified Park blade catheter in 2. In group 3, BAS was used in 2, balloon plus modified Park blade catheter in 1. In group 1, 3 patients had residual shunts (5.2%), two (one with Rashkind and one with coil device) resolved two months later. The third patient using coil device has a relatively larger residual shunt, which has successfully occluded with Cook coil the second time two months later. Six cases with Amplatzer devices had no residual shunt. In group 2, all 15 patients presented with mild residual stenosis, while the mean pressure gradient between RV and PA decreased from 87.8 mmHg to 28.34 mmHg ($p<0.01$). One case with thick pulmonary valve was treated successfully by PBPV plus modified Park blade catheter. In all the 2 cases of TGA after intervention, mean SaO2 increased from 47.24% to 75%. Clinically significant improvement occurred in a 5 months' old baby after BAS with modified Park blade catheter. Because of the thick intra-atrial septum, the ASD was dilated from 5mm pre-procedure to 11mm after-procedure. In conclusion, 2DE is a safe, accurate method for evaluation of therapeutic effect of catheter intervention in CHD.

213

Myocardial integrated ultrasound backscatter in patients with Duchenne's progressive muscular dystrophy

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To evaluate whether myocardial ultrasound integrated backscatter is useful for the early detection of myocardial involvement in patients with Duchenne's muscular dystrophy (DMD), the magnitude of cyclic variations (CV) and integrated backscatter (IBS) intensity were measured separately in the inner and outer halves of the left ventricular posterior wall in patients with DMD (mean age 17.6±2.7 years). Fourteen healthy individuals were used as an age-matched control. Both the CVs in the inner and outer halves of the left ventricular posterior wall were lower in patients with DMD than in the control group. Both the corrected mean IBS intensities in the inner and outer halves of the ventricular wall (cIBSin and cIBSout, respectively) were greater in patients with DMD than in the control group. Among the patients with DMD, the shortening fraction of the left ventricle was lower in those patients with an increase in both the cIBSin and cIBSout, compared with those patients with normal cIBSin and cIBSout ($p<0.005$), and those patients with an increase in only cIBSout ($p<0.05$). Among the 9 patients with DMD and a normal left ventricular shortening fraction, 6 patients had an increase in the difference between cIBSin and cIBSout (>5 dB). These data indicate that myocardial changes, such as fibrosis, begin in the outer half of the left ventricular wall in patients with DMD, even if global left ventricular function is normal. In conclusion, increased IBS intensity in the outer half of the left ventricular posterior wall is an early echocardiographic sign of myocardial involvement in patients with DMD.

214

Transthoracic versus transesophageal echocardiograhy before interventional occlusion of atrial septum defects – a single center study on 143 consecutive patients with three different devices

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Objective: Standard therapy of ASD II is open heart surgery with low mortality/morbidity. Nowadays interventional closure (IAC) with umbrellas is becoming more popular. Not all kinds of ASD are suitable for IAC. Transthoracic echocardiography (TTE) is used as a screening method. There are no data comparing conformity of TTE versus transesophageal echocardiography (TEE) in such patients. Method: We compared echo findings in 142 consecutive patients (mean age 12 y, mean weight 37.6 kg, mean height 145.6 cm) who had TTE and TEE before IAC between 7/96 and 6/98. Following systems were used: Angel Wings 3/96 – 10/97, CardioSeal/Starflex 11/97 – 6/98, Amplatzer 6/98 – 6/98. Following parameters were analysed: number, size and location of defects. Ballon occlusion diameter (BOD) and device size were analysed according to the different systems as well as reasons for unsuccessful implantation. Results: In 77 of 143 (55.2%) patients a device was implanted. The implantation rate was highest for Amplatzer (75 %) and lower for Angel wings (41 %) and CardioSeal/Starflex (31 %). Patients with Amplatzer devices had higher native size and BOD ($p<0.05$). TTE and TEE findings corresponded well according to defect size/number and poor screening in localisation and rim size. Main reasons for not implanting a device were large defect size in all three systems, followed by rim size in the Starflex and multiple defects in the Amplatzer group. Summary: As a consequence we prefer Amplatzer devices for normal ASD, CardioSeal/Starflex for PFO and multiple ASD.

215

Abnormal diastolic myocardial velocities: a new sign of early anthracycline-induced cardiotoxicity?

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Previous studies with tissue Doppler imaging (TDI) have shown significant abnormal myocardial velocities in late survivors of childhood malignancies. The purpose of this study was to assess myocardial velocities during and shortly after treatment. A prospective study was performed in 17 children (age range 5–16 yr) during treatment with anthracyclines (cumulative dose range 150–360 mg/m2). TDI from the apical 4- chamber view (4-CV) was carried out at baseline (before start chemotherapy), after each intermediate dose, and 6 months after end of therapy. All patients were evaluated during therapy. The 6 months follow-up was completed in 14 patients. Paradoxical mid-diastolic myocardial velocities of LV and/or IVS walls were detected, using both single gated- and 2D color TDI. The frequency of appearance of this phenomenon (see figure) increased significantly with increasing cumulative dose of anthracyclines ($p<0.001$), and decreased within 6 months after end of therapy ($p<0.05$). Abnormal myocardial velocities were more often detected in the LV free wall (82%) than in the IVS (35%). The authors conclude that TDI has the potential to detect subclinical myocardial abnormalities in children, both during and shortly after receiving moderate doses of anthracyclines. The abnormal mid-diastolic myocardial velocity might be the first sign of acute anthracycline-induced cardiotoxicity. Its clinical implication should be further studied.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

216

What is the outcome of infants and children who have been returned to bypass for further surgery? A decade of intraoperative transesophageal echocardiographic experience

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The purpose of this study was to determine the longterm outcome of infants and children who were returned to bypass (RTB) for further surgery or myocardial cooling, based upon the detection of residual problems during intraoperative transesophageal echocardiography (TEE). From 1990 through 1999, 1781 infants and children had TEE examinations during repair of a primary congenital cardiac defect; 123 (6.9%) were returned to bypass utilizing TEE information. In 122/123 the problem evident on TEE was confirmed by pressure or oximetry measurements, and/or direct surgical inspection. In the TEE database, cases are coded as 1 of 4 outcome categories following RTB: (1) problem relieved, no sequelae; (2) problem improved, some hemodynamic residual, good outcome; (3) problem relieved, patient died; (4) no effect or unable to relieve problem, patient died. Group 1 contained 45.1% of RTB patients, with the identified problem completely relieved. No patient in this group has required re-intervention for the original problem. Group 2 contained 12.3% of RTB patients, and one has required re-intervention for incomplete relief of the original problem. Thus 57.4% of patients who had RTB are alive and doing well with a median follow-up of 5 ½ years. In contrast, 42.6% of patients undergoing RTB did not survive. Group 3 contained 14.7% of patients while Group 4 contained 27.9%. Problems encountered in Group 4 patients that could not be overcome included ventricular dysfunction and atrioventricular valve regurgitation in 47%, and hypoxia in 16%. Stage 1 Norwood patients comprised 26% of Group 4. In conclusion, TEE has a high accuracy in the definition of residual problems during repair of congenital cardiac defects. With RTB and immediate revision, the majority of patients have had a highly beneficial outcome, avoiding late re-operation. Problems identified by TEE that are less often associated with good outcome following RTB include ventricular dysfunction and atrioventricular valve regurgitation.

217

The product of aortic velocity time integral and heart rate reflects cardiac output and changes in cardiac output, and differentiates neonatal children from those with dilated cardiomyopathy

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BACKGROUND: Cardiac output (CO) can be estimated from the aortic velocity time integral (VTI), cross sectional area (CSA) and heart rate (HR). We sought a method to reflect cardiac output while not incorporating CSA, because of errors induced in its calculation. OBJECTIVE: To determine the range of VTIxHR in normal children, to assess serial change in VTIxHR and thermodilution CO with and without pressors, and to compare VTIxHR in patients with dilated cardiomyopathy (DCM). METHODS: 116 children without heart disease were studied along with 23 with DCM, and 6 patients having thermodilution CO measurements. We measured aortic VTI from 4 chamber views, HR, shortening fraction (SF), ejection fraction (EF), and the Tei myocardial performance index. The thermodilution patients had VTIxHR and thermodilution CO measured before and after pressor augmentation. RESULTS: In normal children, all conventional indices of function were normal. The VTIxHR was 4013 +/- 501(SD), having a slightly negative slope when plotted against BSA. In DCM patients, EF was 34.3%, SF 13.9%, Tei index 0.77, all abnormal mean values. The VTIxHR in DCM was 1271 +/- 259, p=0.001 vs normal children. In the thermodilution patients, resting CO was 2.4, increasing by 46.3% with pressors, while resting VTIxHR was 1410 +/-233 increasing by 50.1% with pressors (p=0.93). CONCLUSION: Serial assessment of VTIxHR in individual patients given pressors indicates that changes in CO are reflected in similar magnitude changes in VTIxHR. The lower values of VTIxHR in DCM patients suggest that the reduced function reflected in conventional indices is associated with reduced CO. The VTIxHR, easy to obtain, appears useful in serial assessment of changes in CO in individual patients and in assessment of differences in output states between groups of patients.

218

Three-dimensional echocardiographic reconstructions of trabecular ventricular septal defects

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The pathophysiological mechanisms responsible for late hypertension and cardiovascular morbidity after coarctation (Coa) repair have not been clearly assessed. We studied 70 normotensive subjects at rest (age: 14±5 y, pressure: 116±13/56±9 mmHg), who had good repair of Coa defined by the absence of gradient between upper and lower right limb (0: -2 mmHg). After exercise testing, we defined 2 groups: Coa HT: systolic hypertension at exercise (> 200 mmHg, n=20: 228±23 mmHg) and Coa NT: normal systolic pressure at exercise (n=10, 163±24 mmHg). These subjects were sex-, age- and blood pressure-matched to 20 controls (age, 15±3 y, pressure 115±10/56±6 mmHg). Using echo-tracking technique, we measured common carotid artery (CCA) diameters and the intima media thickness (IMT). Compliance (CSC), distensibility (CSD) and elastic modulus (Ems) were calculated. CCA pressure waveform and the local pulse pressure were determined in 20 subjects to define augmentation index (AI). Vasodilatation of the brachial artery in response to reactive hyperaemia and to glyceryltrinitrate (GTN) were measured. The IMT was increased in the whole Coa group (p<0.001) (0.57±0.01 mm in Coa HT vs 0.54±0.05 mm in Coa NT). The CSD was decreased and the Ems was significantly higher all patients. The carotid pulse pressure was higher in the Coa HT (41±14 vs 33±9 mmHg, p<0.05). The AI was increased in both Coa groups. Flow mediated dilation and GTN-mediated dilation of the brachial artery were impaired in the Coa group (p<0.01). GTN mediated dilation was inversely correlated with maximum systolic blood pressure at exercise (r=-0.38, P=0.03). The combination of distensibility decrease in the proximal arterial bed with an impairment of distal artery reactivity can account for the elevation of exercise blood pressure after Coa repair.

219

Noninvasive diagnosis of neonatal coarctation of the aorta in association with a patent ductus arteriosus

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Identification of coarctation in the neonatal period can be very difficult when a widely open ductus arteriosus is present. From 1994 to 1999, 45 neonates (four of them were premature babies) with coarctation of the aorta confirmed after arteriogram or surgery were enrolled in this study. Six of them were excluded because the ductus arteriosus closed before our diagnosis. We performed 2-D and Doppler echocardiography on all patients. The inner diameters along the aortic arch were measured and Doppler flow mapping was undertaken over isthmus, ductus arteriosus and descending aorta. In addition, 18 neonates (including three premature babies) with isolated patent ductus arteriosus (PDA) were selected for control. The most significant diagnostic index was the ratio of isthmus/descending aorta diameters (I/D ratio). The I/D ratio in coarctation group ranged from 0.30 to 0.68, (mean ± SD: 0.49 ± 0.15, 95% CI = 0.45, 0.54) whereas the I/D ratio in control group ranged from 0.65 to 1.0, (mean ± SD: 0.84 ± 0.14, 95% CI = 0.77, 0.91). Based on the findings of this study, the diagnostic dilemma for neonatal coarctation associated with PDA can be solved by our new diagnostic criteria. If any neonate who had one of the three conditions below, we can establish the diagnosis. First, there is significant blood pressure discrepancy between arm and leg without interrupted aorta, as indicated by echocardiogram. Second, a posterior infolding at the aorta is demonstrated by echocardiogram. Third, the I/D ratio is lower than 0.68. Using above criteria, we found the sensitivity of diagnosis achieved 95% in our patients without false positive in our control group. These criteria cannot only be applied to patients with or without other intracardiac lesions but also be applied to premature neonates.

220

Diagnostic value of contrast echocardiography in the examination of congenital or acquired heart disease

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Echocardiographic contrast agent SHU 454 provides microbubbles of defined size (median 3µm), a solution of galactose and SHU 508A is

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

specially manufactured (9899.9) galactose microparticles and (900 1) palmitic acid (bubble size less than 2.5μ). Myocardial gas bubbles are known to have only a limited stability in blood. SHU 454 are absorbed in the capillaries of the lung after intravenous injection and do not reach the left side of the heart, but SHU 508A are not absorbed in the capillaries of the lung and reaches the left side heart. Thus the agents can be used by peripheral venous injection for detection of right-sided pulmonary, mitral and aortic valve insufficiency as well as detection of intracardiac shunt or for anatomical identifications in complex cardiac defects. In this study we aimed to demonstrate the role of peripheral venous injection SHU 454 and SHU 508A in the diagnosis of congenital or acquired heart disease. MATERIAL and METHODS: Four hundred thirty patients (210 girls, 220 boys, age range 1 month-17 y) were involved in the study (January 1994-August 2000). The subject had right heart lesions (370 preserial septal defect/ASD, VSD, Pulmonary venous, pulmonary arteriovenous fistula, urology of Fallot, atrioventricular septal defect) and left heart lesion (60 pts: Aortic and mural valve insufficiency, coronary arteriovenous fistula and coronary ectasia). The echocardiographic examination was performed to examine apical four chambers, parasternal long and short axis views. The dosage SHU 454 was 0.5 ml/kg/per inj max. 10ml/per inj in five times and SHU 508A was 0.5–2ml/per inj in five times. RESULT: Each patient received single injection SHU 454 and SHU 508A gave great information about of right and left ventricle, pulmocary vascular structure and anatomy in urology of Fallot, aortic stenosis, about surgical indication of ASD VSD silent patent ductus arteriosus and valve morphology in other anomalies. IN CONCLUSION: SHU 454 and SHU 508A are significant enough to advance the technique not only to replace the other conventional methods for diagnosis but also to create new diagnostic capabilities.

221
Age-related changes in coronary flow reserve and contractile state response to dobutamine from infancy to childhood
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Left ventricular (LV) functional reserve response to inotropic stimulation is known to be age-dependent, however, there is limited information about age-related changes in the effects of dobutamine on coronary blood flow reserve (CFVR). To assess the effects of age on CFVR, dobutamine stress (5 μg/kg per minute) transthoracic echocardiography was performed in children. The study group consisted of 29 children aged from 1 month to 16 years (mean 8±7 years). Peak diastolic velocity in the left descending coronary artery (CFV) was recorded by pulsed-Doppler under the guidance of color Doppler flow mapping. Coronary flow velocity reserve (CFVR) was calculated as the ratio of maximal CFV at dobutamine infusion to basal CFV. LV contractility was calculated by two dimensionally directed M-mode echocardiography. The rate-corrected mean velocity of circumferential fiber shortening (mVcf/c) and LV end-systolic wall stress (LSS) were used as indices of contractility. CFV at dobutamine infusion increased significantly compared with the control values (+32 %, p < 0.01). CFVR in the younger children was low and increased significantly with age (r = 0.68, p < 0.01). Dobutamine induced an increase in mVcf/c (+32 %, p < 0.01) and a decrease in LSS (-16 %, p < 0.01). The percentage of increase in mVcf/c (Δmvcf/c) during dobutamine infusion was low in infants and increased significantly with age (r = 0.62, p < 0.01). CFVR correlated significantly with ΔmVcf/c (r = 0.65, p < 0.01) during dobutamine infusion. Responses of CFV to dobutamine are less sensitive in younger children. Age-related increase in CFVR is associated with age-related changes in LV functional reserve.

222
Left and right ventricular volume determination in children with congenital heart defects (CHD): 3D-echo versus angiography
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Angiography has commonly been used to determine ventricular volumes in children with CHD. Three-dimensional echocardiography (3D-echo) may allow more accurate volume assessment since it is independent of geometrical assumptions. We compared LV and RV volumes determined by 3D-echo and angiography in children with different types of CHD. We studied 102 patients aged 3 days to 27 years (median 2.25) with body surface area 0.21–1.79 m2 (median 0.53). Biplane angiography was obtained during diagnostic cardiac catheterisation. 3D-echo was performed immediately after catheterisation using a rotating transducer (Acuson) usually from the subcostal window. For 3D-reconstruction a Tomtec-System was used. Angiographic volumes were calculated during end-diastole and end-systole using Simpsons rule. 3D-echo calculations summarized the volumes from multiple short axis slices (distance 2mm) after manual tracing of ventricular borders. Volume calculations by 3D-echo was possible in 84 pts. (82%) for LV, but only in 15 pts. (32%) for RV. LV volumes by 3D-echo correlated well with angiographic volumes in systole and diastole (sys. r2=0.96, dia. r2=0.93). RV volumes by 3D-echo correlated poorly with angiographic volumes (sys. r2=0.7; dia. r2=0.79). Comparison of both methods showed larger volumes determined by angiography, particularly for the RV. (EDVsys. 0.6±3.3 ml, 0.9±25.8 %, LV dia. 7.1±28.4 ml, 7.4±12.1%, RV sys. 1.8±6.9 ml, 19.9±24.1%; RV dia. 6.4±9.4, 42.5 ± 33.6%). Different types of CHD did not influence the differences between the two methods (p>0.005). We conclude that 3D-echo allows more accurate ventricular volume determination in CHD-patients. Its practical use, however, may be assured by appropriate image acquisition.

223
Left ventricular outflow tract pseudoaneurysms in congenital heart disease
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Left ventricular outflow tract (LVOT) pseudoaneurysm occur rarely after LVOT surgery in adults, but little is known about them in children. We retrospectively reviewed the Michigan Congenital Heart Center database to define size and location of pseudoaneurysm, clinical characteristics, and results of surgical repair. Eight patients with LVOT pseudoaneurysms were identified. Aneurysms occurred at 8.4 years of age (range: 0–37 years) and measured 6 to 50mm (mean 26.3±16 m) at diagnosis. Pseudoaneurysms occurred in two locations, originating from the aorto-mitral intervalvular fibrosa (6 patients) and in the native RV free wall after interventricular tunnel of the LV outflow across a VSD to the pulmonic valve (2 patients). Of those in the intervalvular fibrosa, 1 was seen at birth, 3 after Ross procedure, 1 after 2 subaortic resections, and 1 after endocarditis and cardiac catheterization. Post-operative aneurysms were detected 10 days to 12 months (mean 5.5 months) after surgery. Five pseudoaneurysms impinged on neighboring structures. In 4 patients, the pseudoaneurysm was repaired with Gore-tex patch exclusion and partial excision. One of these required 3 months post-operatively, necessitating a second operation. One pseudoaneurysm was resected and suture closed. One pseudoaneurysm remains stable without intervention. Six of 8 patients remain well, 1 died intra-operatively and 1 died after Cooley-Yacoub procedure without pseudoaneurysm recurrence. LVOT pseudoaneurysms associated with congenital heart disease occur after LVOT surgery and congenitally. Surgical repair by orifice exclusion and complete or partial resection is successful in most cases. The pseudoaneurysms were located in the aorto-mitral fibrous continuity or in the right ventricular free wall, suggesting that these locations are inherent anatomic weak points at risk during surgical procedures.

MAY 30 Time: 11:00 – 12:30

Session 21: Cardiac Nursing

224
Congenital heart disease and parenting stress
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Parents of infants with congenital heart defects (CHD) experience increased stress, often recognized by health professionals at the time of diagnosis and first hospitalization. Since stressful parent-child systems may have a negative impact on child psychosocial outcomes, we assessed parenting stress in 42 parents of children > 2 years of age (mean 5.6 yrs, range 2 – 12.4 yrs) with CHD. Parents of 19 female and 23 male children with simple (n=14) and complex (n=28) CHD participated. Parents completed Abidin's Parenting Stress Index (PSI) in the outpatient clinic. The median PSI total stress score was 78, compared to 69 in a normative sample. On the child domain PSI subscale, 30% (2 × expected) had scores at or above the critical cutoff score for high stress. (High scores in the domain reflect parental perceptions that the child has qualities or behavioral characteristics that make them difficult to parent.) There were no significant correlations between parenting stress and the child's age (r=.249). Interventional most recent surgery (n=38, t=.068), or severity of CHD (mean 76.1 simple CHD vs 69.7 complex CHD, t=2.0,

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

p = 0.3). Parenting stress was significantly higher in single parent homes (mean 88.5) than in 2-parent homes (mean 67.8, p = 0.2 We conclude that parents of children with CHD are at increased risk for chronic high stress related to parenting the child with CHD. High stress is unrelated to the severity of CHD. Ongoing counseling of all families regarding the meaning of the cardiac diagnosis, including changes along the developmental continuum, is needed

225
Creating a heart healthy environment for our children
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Prevention of heart disease begins in childhood. The promotion of smoke-free living, healthy eating and regular physical activity in children is the mission of Heart Health Hamilton-Wentworth. In 1998 the Health Promotion Branch of the Ontario Ministry of Health provided funding for five years to Heart Health Hamilton-Wentworth (HHH-W). The Child Youth Heart Health Work Group is one subcommittee of HHH-W. It is comprised of representatives from health, education recreation and volunteer sectors. Initially 441 students from Grades 6–8 in Hamilton-Wentworth Catholic School Board were surveyed. The objectives were to determine student's knowledge, habits and attitude about physical activity, nutrition and tobacco. One important finding was that youth want to take action towards a healthier lifestyle but there are barriers towards achieving this. From the survey, schools were chosen as the avenue to reach the youth. Presently two pilot schools have volunteered to participate as Heart Healthy Schools with other schools planning to join. Enthusiastic students, parents and staff from these pilot schools have developed heart healthy activities either via the curriculum, or as after school activities. Examples of these include cooking clubs, walk to school day and an anti-smoking game in the classroom. Other exciting initiatives will be presented. Prevention of heart disease in children by reducing risk factors such as obesity, hypertipidemia, smoking and sedentary lifestyle needs to be a goal of the health profession and the community. This will reduce heart disease in our future adult population.

226
Neonatal transplantation. Where to wait
Erin Messinger, Catherine Mery
Hospital For Sick Children, Toronto, Canada

The neonate awaiting heart transplantation requires specialized care from all members of the interdisciplinary team. Infants with single ventricle physiology present a unique set of challenges. Often these neonates endure lengthy waiting periods in critical care environments. Limited and costly critical care resources have forced The Hospital for Sick Children (HSC), Toronto, Canada, to address the feasibility and appropriateness of caring for these neonates and their families beyond the critical care setting. At HSC, the alternative setting was an observation room on the inpatient cardiac unit. Initiating this change in the care environment revealed several issues and gaps. They included knowledge and resource deficits around the care of a patient in a hypoxic gas environment, ensuring appropriate monitoring and timely intervention in the event of any deterioration, and the ability to adequately support the family and the ongoing developmental needs of the infant. In response, an interdisciplinary 'Pre-transplant Working Group' was formed to identify and secure the appropriate resources for the care of this fragile patient population. This poster presentation will identify the challenges faced, solutions and recommendations for the future. Current literature on outcomes and treatment of single ventricle and neonatal heart transplant recipients from a physiological, psychosocial and developmental perspective will be included. Recent research related to the needs of the family and the team will be incorporated. Experience to date at HSC will highlight the benefits of this initiative while addressing fiscal and resource implications for both the critical care and inpatient units.

227
Predictors of Oral Feeding at Discharge in Post Cardiac Surgery Neonates
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Background: Changes in technology and cardiac surgical procedures over the past decade have led to an increase in the number of neonates undergoing cardiac surgery. Feeding difficulties after cardiac surgery in the neonate can prolong hospital stay and complicate post-operative care. The purpose of this study was to identify independent predictors of post operative feeding difficulties. Methods: A retrospective chart audit of 101 consecutive neonates who underwent cardiac surgery was conducted. Neonates with significant structural or functional defects that would affect oral feeding were excluded. Ten variables were analyzed as possible predictors of post-operative feeding difficulties including: diagnosis, demographics, details of surgery, and post-operative course. Results: At hospital discharge 71.3% (N=72) of neonates were orally feeding and 28.7% (N=29) were not. Overall mean hospital length of stay was 17.7 +/- 16.4 days. Neonates with feeding difficulties were more likely to undergo operations involving the aortic arch (31% vs 15%, Chi-Square=3.23, p=0.07). Neonates not feeding orally at discharge had longer ICU lengths of stay than those who fed (16.5 +/- 18.4 vs 7.3 +/- 5.8 days, p=0.01). They also had longer post-operative stays (30.3 +/- 24.2 vs 12.7 +/- 7.5 days, p<0.001). Neonates with feeding difficulties had a higher incidence of vocal cord injury (24.1% vs 1.4%, Chi-Square=14.67, p<0.001). Multivariable logistic regression analysis revealed vocal chord injury (odds ratio 17.5) and post-operative ICU stay (odds ratio 1.1 per day) as independent predictors of failure to feed orally at discharge from hospital. Conclusion: Risk factors for feeding difficulties in the post-cardiac surgery neonate are vocal chord injury and prolonged ICU length of stay. Early identification of neonates at risk followed by definitive intervention strategies will lead to improved patient care and resource utilization.

228
Risky business: assessing risk factors for coronary artery disease after Kawasaki disease
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Kawasaki Disease (KD) is the leading cause of acquired heart disease in children. KD causes a systemic vasculitis, potentially resulting in the formation of coronary aneurysms. This general vasculitis causes an increase in endothelial dysfunction resulting in increasing the risk for premature myocardial infarction. Our purpose was to evaluate patients who had Kawasaki Disease and determine if they were at higher risk for CAD than subjects in the normal population. We evaluated KD patients >5 years post initial diagnosis with known risk factors for CAD. Patients were grouped into persistent aneurysms, regressed aneurysms and no aneurysms. Subjects were matched by age and gender with a control group of subjects who never had KD. All subjects were assessed for cardiovascular risk factors including diet, BMI, family history and activity level. Ancillary tests included renal and endocrine function, lipoproteins, C-reactive protein, homocysteine and fibrinogen. All patients had 24 hr BP monitoring, 2D ECHO, Carotid and Brachial Artery ultrasound. The results of this study will determine if patients who have had KD, regardless of their subgroup, are at higher risk for developing premature CAD than those in the control group. The results of this study will also support the need for evidence based practice. Health Care Professionals will be able to apply the knowledge when counseling newly diagnosed families whose children are being treated for KD.

229
The impact of prenatal versus postnatal diagnosis on psychological distress in parents of children with severe congenital heart disease
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Objective: To evaluate symptoms of psychological distress at the time of birth and six months later in parents of children prenatally diagnosed with severe congenital heart disease (G1) and parents of infants with similar forms of heart disease who are diagnosed postnatally (G2). Methods: From 2/99 to 10/00, parents of ten infants prenatally diagnosed by fetal echocardiogram and seven infants postnatally diagnosed with severe congenital heart disease were evaluated for evidence of psychological distress. Qualitative data was obtained by taped, transcribed semi-structured interviews and examined for recurrent themes of psychological distress. Quantitative data was obtained using the Brief Symptom Inventory (BSI), a self-report inventory that measures global symptoms of psychological distress. Data was collected at the time of diagnosis, at the time of birth (if different from the time of diagnosis) and six months after the birth and compared between the two groups and to a normative sample. Results: Guilt, disbelief, fear and anger were common themes across both groups regardless of timing of diagnosis. There was no statistical difference in global symptoms of psychological distress scores in G1 from the time of diagnosis to the time of birth. There was no statistical difference in global scores between groups at the time of the child's birth.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

In both groups, scores for depression, anxiety, and global symptoms were significantly higher than those of the normative sample at the time of the child's birth ($p = 0.001$) but normalized six months later. Conclusions. Parents of children with severe congenital heart disease experience similar symptoms of distress regardless of when the diagnosis is made. Although the information does not appear to lessen parental distress at the time of birth, most families expressed being grateful for the prenatal diagnosis.

230

Care of adult congenital heart surgery patients in pediatric intensive care units

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Background: With advances in treatment of congenital heart disease (CHD), the population of adults with CHD is growing. In our institution, adults with CHD who are referred to the pediatric cardiac surgeon are cared for in the pediatric intensive care unit (PICU) postoperatively. A study was done to assess standard practice for postoperative ICU care of adults with CHD in the USA. Methods: A questionnaire was developed to assess this practice. It was sent to advanced practice nurses (identified in the directory of the Society of Pediatric Cardiovascular Nurses) at 35 institutions. Results were evaluated in the 22 responses received. Results: Twenty one of 22 (95%) of hospitals studied provide care for adults with CHD in their PICU. Nine of the institutions are free standing children's hospitals. The patient population per institution ranged from 1 to 75 adult CHD patients per year (mean of 5% of the annual total CHD surgery population). The patient age range was 18 to 72 years. Surgery was performed by pediatric cardiac surgeons in 15 (75%) and by surgeons who do both pediatric and adult cardiac surgery in 5 (25%) of the institutions (1 hospital did not respond). None of the PICUs had policies restricting which of their nurses may care for these patients. Postoperative medical care was provided by pediatric professionals including intensivists in 15, surgeons in 13, fellows/residents in 12, cardiologists in 12, anesthesiologists in 3, nurse practitioners in 7, and physician's assistants in 4. Conclusions: It is standard practice to care for postoperative adult CHD patients in a PICU in many institutions. Preoperative and postoperative care for these patients may be best provided by the experienced and specialized pediatric team regardless of patients' age.

231

The development of a transition process from a paediatric to an adult care centre

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With the tremendous advances seen in congenital heart disease treatment and surgery in the past 20 years there is a growing population of youth with complex cardiac health needs. At British Columbia's Children's Hospital in Vancouver approximately 300 youth per year transition to St. Paul's Pacific Adult Congenital Heart Clinic (PACH). As they moved into the adult health system some method of enabling a successful transition became necessary. In consultation with the Pacific Adult Congenital Heart Team (PACH clinic) a transition process was developed. With support from the Youth Health Program, a multi-disciplinary cardiac transition team was formed. The team members come from both the paediatric and adult clinics and worked closely together. A variety of educational tools, information exchanges and eventually a transition clinic evolved from these meetings. Several problems were identified including the need for more youth education, autonomy and a general reluctance towards the transfer of care. Strategies enhancing the youths understanding of their health condition and promote independent behaviours and self-advocacy have been incorporated in a regular components of outpatient visits. Transition issues are now discussed starting at an early age and reinforced on subsequent visits. A half-day Transition clinic, held at the adult centre, further facilitates the transfer of care. At this clinic the focus for the youth is to promote self-advocacy within the adult health care system. For the families the focus is on fostering the youth's independence and relinquishing their role as primary care provider. Evaluation of this process including feedback from graduates

232

School performance following pediatric heart transplantation

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Following heart transplantation (TX), children are at risk for learning problems and falling behind grade level. Multiple factors, including prior congenital heart disease and end stage CHF, chronic illness, neurologic complications, and lengthy school absences likely contribute to school problems. This study describes the school experiences of patients following TX. By retrospective chart review, pre and post TX data were retrieved for all children followed in our heart TX program who were at least 1y post TX. 35 children (23m, 12f) were in grades 1–12. School performance was defined as presence of school- based special services (tutoring, special education services), age appropriate grade level and presence of attention deficit disorder/learning disabilities. Age at TX was 7m-16y, 19 children were enrolled in school at time of TX. Pre TX diagnosis was cardiomyopathy (CMPY) in 17 and congenital heart disease (CHD) in 18. RESULTS: All children were attending school full-time though 20/35 (60%) require special services for some period of time. 17/35 (48%) children were behind expected grade level. Of these, a third were behind grade level before TX. 12/17 (70%) with primary diagnosis of CMPY were at grade level while 9/18 (50%) with CHD were at grade level. 11/35 (31%) were identified with attention deficit disorder/learning disabilities, half prior to TX. CONCLUSION: Following heart TX, all students returned to school full-time though most require special services. Those with CHD have significantly more school problems than those with pre TX diagnosis of CMPY. Many learning problems predate TX. Children post TX require assessment of learning problems and intervention to enhance academic achievement.

233

Blake drain study

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PURPOSE: To determine if the implementation of the usage of Blake drains, rather than the traditional chest tube system will affect hospital charges and length of stay (LOS) following the bi directional Glenn (BDG) and Fontan Procedures. METHODS: A retrospective chart review was done on all BDG (n = 51) and Fontan (n = 34) completion patients from April 1997 through October 2000. RESULTS: Patients undergoing BDG operation with conventional chest drains (n = 24) had an average LOS of 7.46 days +/- 5.0. Use of Blake drains in similar patients undergoing the BDG operation (n = 27) resulted in an average LOS of 5.3 days +/- 1.2 ($P < 0.05$). Hospital charges for the BDG conventional chest tube group averaged $67806 +/- $29581. Charges for the BDG Blake group averaged $48557 +/- $13841 ($P < 0.05$). Patients who had Fontan Completions with conventional chest tubes (n = 15) had an average LOS of 9.47 +/- 5.7 days vs patients utilizing Blake drains post operatively (n = 17) who had an average LOS of 6.12 +/- 1.9 days ($P < 0.05$). Hospital charges for the Fontan conventional chest tube group averaged $58264 +/- $18403. Charges for the Fontan Blake group averaged $48116 +/- $7519 ($P < 0.05$). CONCLUSIONS: Blake drain use in the Fontan and BDG populations significantly decreases hospital charges and LOS.

234

Quality of life perceptions in adolescents with congenital heart disease: a comparison between patients and parents

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Purpose: This study was conducted to explore the perception of health-related quality of life (HRQL) in adolescents with congenital heart disease (CHD) compared with those of parents. Methods: HRQL was measured using the General Health Assessment for Children, modified for adolescents with heart disease. This is a developmentally-sensitive, disease specific outcomes chart measures HRQL in the following domains: overall health ratings, physical function, psychological well-being, social and role function, health care utilization, and symptom related distress. Initial pilot testing suggested the reliability and validity of this instrument in this population. Eleven adolescent patients (age 12-17 years) and their parents completed parallel questionnaires. Pearson correlations were used to examine relationships between the groups. Results: Adolescent and parent HRQL scores were significantly correlated for overall health perceptions ($r=.73$, $p=.01$) and

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

health care utilization (r= 63, p= 03). No significant association was found between their perceptions of physical function (p=.814), psychological well-being (p=.167), social and role function (p=.420), or symptom related distress (p= 252). Conclusions: Adolescent and parent perceptions of overall health status and number of contacts with the health care system were in agreement. However, their perceptions differed regarding the impact of CHD on physical function, psychological well-being, social and role function, and symptoms-related distress. These differences in perceptions should be considered when counseling families about the impact of CHD, so as to facilitate communication and optimize health promotion strategies.

235

Beta blockade therapy in children: a protocol for the ambulatory setting Toronto

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We present an interventional protocol for the administration of beta blockade therapy (Carvedilol) to children with chronic congestive heart failure (CHF). Our approach is extrapolated from adult studies of initiation of beta blockade therapy and details the frequency of patient assessment, duration of Carvedilol, communication between team members, and documentation. Patients are seen by a clinic nurse weekly during the initiation of Carvedilol therapy, and by their cardiologist monthly, or more frequently as the condition demands. The cardiologist establishes dosing, usually beginning at 0.2 mg/kg/day, to a maximum of 1 mg/kg/day, with the flexibility to adjust dosing according to the patients' condition. The patient population consists of children between the ages of 1 month and 16 years, with dilated cardiomyopathy or end-stage congenital heart defects, with a left ventricular ejection fraction of < 45%. These patients are on triple therapy of digoxin, diuretics, ACE inhibition, and aldactone, and are volume stable. Over an implementation period of 18 months, a low hospital admission rate for CHF (1 patient of 12) indicates effective outpatient management of symptoms. Additionally, positive feedback from both staff and families demonstrates the validity of our methods. Our experience indicates that this protocol is both a safe and workable approach to administering beta blockade therapy in children with chronic CHF in the outpatient setting.

Session 22: Arrhythmias, Electrophysiology, Sudden Cardiac Death

236

Sino-atrial node reentrant tachycardia; an under-recognized clinical entity in infants with congenital heart disease

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Background: Whereas sino-atrial node reentrant tachycardia(SANRT) is well described in adults, it is not well described in children. Methods: The MUSIC pediatric electrophysiology study(EPS) database was reviewed for cases of SANRT, a reentrant SVT with a p wave morphology nearly identical to sinus, diagnosed between 1/99 and 11/00. Demographic, clinical, and electrophysiologic variables are described. Results: Of the 106 pts, 0–18 yrs, who underwent EPS for documented SVT, 6(6.6%) met criteria for SANRT. Presentation age was 1 day-14 mos, median 0.4mos. One pt had cardiomyopathy while 6 had congenital heart disease(CHD), at which 5 had recently undergone CHD repair(0–11 days, median 5 days). At presentation, SANRT was sustained in 6 pts and caused variable hemodynamic compromise (none 5 pts, mild 1 pt, moderate 1 pt). One pt, not compromised at presentation, had moderate compromise with SANRT during EPS. The pt with cardiomyopathy had an improvement in shortening fraction from 14% at presentation to 30% 5 days after termination. Electrophysiologic variables were: cycle length(280–383 ms, median 315 ms), VA(160–280 ms, median 180 ms), AV(100–160 ms, median 120 ms) and VA:AV(1–2.8, median 1.4). In 2 pts, cycle length varied by 20–40 ms. Adenosine was given to 6 pts and terminated the tachycardia in all. All pts were initially treated with digoxin. SANRT was inducible in 2 pts undergoing repeat EPS. At follow up (1wk–9 mo, median 4.5 mos), 4 pts remain on digoxin, 1 was changed to sotalol, 1 was taken off medication and 1 died following CHD repair, but none had a clinical recurrence. Conclusion: SANRT occurs in infants, particularly those with CHD, and can cause hemodynamic compromise despite its slow rate. It may be recognized acutely with adenosine and is suppressed by chronic drug therapy.

237

Experienced and calculated battery service life data of autocapture devices based on long-term follow-up

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Background: Substantial stimulation energy savings can be achieved with AutoCapture (AC) devices. A question of major clinical interest is now to what extent these energy savings may have the potential to prolong battery service life. Methods: In 67 children, aged 6.8±6.4 months, AC devices (Microny n=17, Regency n=26, Affinity n=24, Integrity n=2, St. Jude Medical) were implanted together with various bipolar endocardial lead models in 19 and epicardial leads (Medtronic CapSure Epi 10366 or 1968) in 48 children. During a median follow-up of 12 months (range 1–39 months) AC controlled ventricular pacing was applied in 56/59 children. In 40 children with at least 80% ventricular pacing, battery impedance data and battery service life prediction were obtained. The calculations are based on the actual percentage pacing, the pacing mode (VVIR 24, DDD 16 patients), Holter acquired mean heart rate, lead impedance and stimulation output (0.25–0.1 Volt above actual threshold). Results: See table. The ventricular stimulation output measured was 1.14±0.34 V and the battery impedance remained < 1 kOhm in all AC activated devices. Conclusion: Long-term follow-up data indicate the consistency of low energy pacing in AutoCapture devices, which is confirmed by low battery impedance values. Thus a remarkable extension of battery service life can also be expected in smaller devices. The resulting lower incidence of pulse generator replacements constitutes a substantial clinical advantage in pediatric pacing.

238

Medium-term follow up comparison of steroid-eluting and non steroid-eluting epicardial pacing leads

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Purpose: To compare the medium-term failure rate, pacing and sensing characteristics of steroid eluting (SE) and non-steroid-eluting (NSE) epicardial pacing leads. Methods: Retrospective review of pacing and clinical data recorded at implant and during follow-up of 127 leads (104 SE, 23 NSE) placed in 65 patients (39 male) aged 3 days to 21 years (yrs) (median 3.5), 55 with congenital heart disease, from October 1988 to Nov 2000 at a single institution. Results: Follow up was 2 weeks – 7yrs (median 1.5yrs) for SE leads, and 2 weeks – 12yrs (median 2.5yrs) for NSE leads. Exit block necessitating lead replacement occurred in 5.7% of SE leads and 47% of NSE leads ($p=0.0001$); tending to occur earlier in NSE leads. Pacing threshold increased in NSE leads peaking at 6 weeks: 3.9V+2.8, and remained higher throughout follow-up for surviving leads: 2.3V+1.4 at 2.5 yrs, 2.7V+2.5 at 4 yrs. Pacing thresholds at SE leads remained stable, 1V+0.4 at 6 weeks, 1.4V+1.2 at 2.5 yrs, 1.4V+0.6 at 4 yrs. Ventricular sensing thresholds showed no significant difference between SE and NSE leads at any stage. Conclusion: steroid eluting epicardial leads have a reduced incidence of lead failure, with lower pacing thresholds being maintained.

239

Influence of D-net (European GSM-standard) cellular phones on implanted pacemakers in children: Ankara, Turkey

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This study was designed to evaluate possible interactions between digital cellular phones (CP) and implanted pacemakers (PM) in children. To our knowledge, there is no published study about electromagnetic interference in implanted pacemakers in children. The study comprised 95 patients (pts) (53 boys and 42 girls) with a mean age of 11.5 ± 4.6 years (range 1 – 22), the average time from PM implantation was 3.5 years (range 1 month – 12 years). Fourteen devices (15%) were dual-chamber and the remaining were single-chamber PM. The following companies manufactured the PM tested: Medtronic (42), Biotronik (9), Vitatron (16), Pacesetter (19), CPI (8), and Biotronik (1). Transvenous PM were located in the right pectoral region, subcutaneously in 45, subpectorally in 40 and 10 PM were implanted epicardially. All the pts were tested in the supine position during continous ECG monitoring. After completion of the routine PM check, the effects of

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

European Global system for mobile communication (GSM) was tested using two CP models (Ericsson GA 628 and Siemens S 25.2 W Power). The GSM works with pulse-shaped amplitude-modulated (AM) signals at 900 MHz. For the purpose, atrial and ventricular sensitivity settings were programmed to their most sensitive values and the tests were carried out in the unipolar and bipolar sensing modes. The evaluation was performed during ringing, switching on/off and conversation phase with the CP positioned over the pulse generator and around the PM pocket. A malfunction of the PM was not observed in any pt. Only 1 (1%) of 95 pts showed brief undersensing problem during calls with the CP. In this case, a Medtronic (Prodigy SR, 8162) AAI-R PM was implanted transvenously in a subcutaneous pocket and the sensing defect occurred only with the unipolar sensing mode. Once the source of interference was removed, no sensing defect was detected and the pt remained asymptomatic. No PM inhibition was observed and at no time were symptoms experienced in the study. In conclusion: Although, we didn't observe any PM inhibition, we believe that PM-dependent pts should be tested for possible interferences before they use digital CP. The CP should be more than 20 cm from the PM to the ear contralateral to the PM should be used.

240

18 years experience of cardiac pacing in children with complete heart block

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Aim: To assure the experience of permanent cardiac pacing in children in BCCVS from 1982 to 2000 and to find the most save and effective method for pacemaker implantation. Material: 292 pacemaker implantations in 274 children were reviewed. Congenital heart block was in 44 patients (pts) (21.4%), heart block after open heart surgery was in 153 pts (74.2%). Surgery performed: IVSD repair 56 (36.6%), IASD repair 30 (19.6%), correction of great vessels transposition 22 (14.4%), tetratology of Fallot 24 (15.7%), atrioventricular communications 17 (11.1%). In 55 pts (27.6%) the transventricular approach was used. 127 (61.6%) recieved myocardial and 22 (10.7%) epicardial unipolar leads. 164 pts (79.6%) paced in VVI(R) mode, 42 pts (21.4%) – in DDD(R) mode. Results: Acute stimulating threshold was low (mean 0,4 + V) in endocardial leads and grew on the same position in long term follow up. In small children it is possible to make the loop in the atrium which allows child to grow without damage of the lead. In myocardial lead pacing threshold may be low in acute phase but in chronic one it increased to high level (5 + 2V). Steroid-eluted epicardial leads showed good pacing parameters. Conclusion: Active fixation new epicardial leads and DDD or VDD pacing modes are preferable in low weight children with bradicardia.

241

Long-term follow-up of a steroid-eluting bipolar epicardial pacing lead in a paediatric population

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Background. In an effort to assure reliable pacing in children and patients with congenital heart defects that preclude the insertion of transvenous pacing leads, an epicardial bipolar steroid-eluting pacing lead was developed and its long-term performance evaluated. Methods. 42 children aged 5.1±4.6 years, were implanted with 76 (31 atrial; 45 ventricular) epicardial leads (Medtronic CapSure Epi, models 10366 and 4968) and connected to various pulse generators. The lead features two dexamethasone eluting, platinized, porous electrodes (6 mm2 cathode and 14 mm2 anode) with 2 suture holes. Atrial (A) and ventricular (V) lead performance was obtained at implant, pre-discharge, 1, 3 and 6 months, and every 6 months thereafter. Pacing thresholds were screened for differences in pulse width. The data were analyzed using longitudinal methods. Results. The mean follow-up was 2.8±1.8 years. Kaplan Meier estimates of lead survival at 1, 3, and 5 years are 93%, 93%, and 83% for A leads; and 100%, 93%, and 86% for V leads, respectively. Failures were due to fractures (1A, 2V), undersensing (2A, 1V) and dislodgement (1V). Two patients received transvenous replacement; systemic A impedance significantly increased from 556±96 ohms at 1 month, to 695±84 ohms at 5 years ($p<0.05$). Mean values of A sensing (2.52±1.38 mV at 1 month and 2.78±1.18 mV at 5 years), A pacing threshold (1.39±1.86 and 0.73±0.24 V), V sensing (8.75±4.1# and 5.06±3.37 mV), V impedance (643±164 and 651±114 W), and V pacing threshold (1.04±0.93 and 1.86±1.26 V) did not change significantly over time. Conclusion: Long-term follow-up data prove a high probability of survival for the Medtronic bipolar epicardial lead with consistently low pacing thresholds and stable sensing and impedance values. These results suggest that this lead can be reliably used in the paediatric population.

242

A novel rabbit model of variably compensated complete heart block

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Complete heart block (CHB) provides a useful substrate for study of bradycardia-dependent ventricular arrhythmias such as torsades de pointes. Existing animal CHB models are limited by surgical recovery issues and by reliance on intrinsic escape rhythms. We describe a novel model of CHB involving radiofrequency AV node ablation and ventricular rate control with programmable pacing in the rabbit. New Zealand White rabbits (2.5–4.0kg) were inhalationally anesthetized. The right internal jugular vein was surgically exposed and cannulated with a 3F passive fixation pacemaker lead and a 5F introducer sheath. A 5F quadripolar catheter was guided to the AV junctional area fluoroscopically and by appearance of typical atrial, ventricular, and His bundle electrograms. Radiofrequency energy was applied for 30–60 s. Unipolar VVI or VOO pacing at the RV apex was initiated after onset of AV dissociation, with subcutaneous interscapular pacemaker implantation. Permanent CHB was achieved in 32/37 attempts overall, with application of ≤5 RF lesions and ≤5 minutes of fluoroscopic exposure in 13/14 of the most recent attempts. Accuracy of RF lesion placement was confirmed with gross and histopathologic postmortem examination. Peak cardiac output decreased significantly from 2.2±0.3 (mean±SEM) L/min in pre-ablation sinus rhythm at 206±13 beats/min (n=7) to 1.1±0.1 L/min with post-ablation pacing at 140 beats/min (n=6), recovering to 1.7±0.2 L/min with pacing at 280 beats/min (n=5). Acute periprocedural mortality due to cardiac perforation and tamponade (n=2), airway complications (n=2), and unknown causes (n=5) mainly occurred early in the series. Survivors were maintained for up to 15 days post-procedure with no sign of hemodynamic impairment while being chronically paced at either 140 (n=25) or 280 (n=5) beats/min. Our approach provides a novel, reproducible, and minimally invasive CHB model with adjustable ventricular rate control.

243

QT dispersion in children before and after repair of tetralogy of Fallot

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The objective of this study was to investigate the changes in durations of QT, QRS and QT dispersion (QTd) of children before and after surgical repair of tetralogy of Fallot (TOF). Thirty children (16 boys, 14 girls) and 30 healthy age-matched controls were studied. The mean age of TOF children at corrective surgery was 26.9 months. Mean duration of post-operative follow-up was 2.6 years (range 1 to 5 years). All TOF children had cross sectional colour Doppler echocardiography done before surgery and during post-operative follow-up. QT, QRS and RR intervals were manually measured from each of the 12 leads of surface ECG. QT was corrected (QTc) using Bazett's formula. QTd was the difference between maximum and minimum QT of the measurable leads. Within the first 3 post-operative years, mean(SD) QTc, QRS and QTd were significantly higher than that before operation: QTc: 471(38) vs 417(29) msec., $p<0.0001$; QRS: 117(19) vs 79(14) msec. $p<0.0001$. QTd: 54(20) vs 42(12) msec., $p<0.0001$. QTc and QTd improved somewhat on 3 to 5 years after surgery. After surgery, children who had right ventriculotomy, right ventricular enlargement or right bundle branch block had more prolonged QTc, QRS and QTd compared to those without. Increased ventricular conduction time and inhomogeneity of ventricular repolarization may occur in children after surgical repair of TOF and may contribute towards susceptibility to ventricular dysrhythmia. These electrocardiographic markers showed some improvement by 5 years post-operation.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

244
Is there any circadian variation of QTc dispersion in children with vasovagal syncope?
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QTc interval dispersion is an indirect measure of the heterogeneity of ventricular repolarization. Cardiac and systemic autonomic nervous function may be impaired in patients with vasovagal syncope (VS). OBJECTIVE: To determine the sympathetic nervous function in patients with VS using QTc interval dispersion. DESIGN: Prospective comparison of QTc dispersion measurements in 69 VS patients (31 boys, 38 girls; mean age 13.4). Results of the head up tilt test (HUT) positive and negative groups were compared. RESULTS: HUT was positive in 38 patients and negative in 31. QTc dispersion was significantly higher in the HUT positive group both in the early morning and at night compared to the HUT negative group ($p<0.001$). Although there was a circadian rythm in HUT positive group, there was no change on the QTc within a day in the HUT negative group. CONCLUSION: QTc dispersion in the HUT positive group was high in concordance with their high sympathetic nervous stimulation. Also, QTc dispersion is higher at night and early morning than the rest of the day. This may explain why the tilt positive patients have their syncope attacks early in the morning usually.

245
Catheter ablation of atrioventricular nodal reentrant tachycardia in children: increased efficacy and safety by continuous monitoring of anterograde conduction and Localisa
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The aim of the study was to assess the results and safety of slow pathway ablation in atrioventricular nodal reentrant tachycardia (AVNRT) in children. While RF ablation is highly successful the risk of AV block may be higher in children. 20 consecutive children (median age 8 years, range 2–14) underwent catheter ablation for AVNRT. The arrhythmia was induced, and the mechanism confirmed by standard pacing and mapping techniques. Localisa, a unique mapping system which allows precise 3-dimensional localisation of standard intracardiac electrodes, was used in combination with local electrograms showing slow potentials, to accurately target the slow pathway component of the AVNRT circuit. Continuous monitoring of catheter tip location by Localisa and of anterograde conduction using a high speed (1000 cm/sec) triggered monitor was performed during RF delivery. Accelerated junctional rhythm occurring during RF lesion application was seen in all patients, suggesting slow proximity to the compact AV node. Between 2 and 5 lesions were applied in the area. Successful slow pathway ablation defined as loss of dual AV nodal physiology, absence of atrial echo beats and non inducibility of AVNRT, was achieved in 19/20 patients in a single session. One patient who had persistent atrial echo beats at the end of the first procedure developed recurrence of AVNRT 2 weeks later and was successfully treated during a second procedure. None of the patients developed AV block, and during a median follow-up of 12 months (3–24), none has had recurrence of AVNRT. Continuous and accurate localization of intracardiac electrodes by Localisa in combination with continuous monitoring of anterograde AV conduction enables successful and safe slow pathway ablation in children with AVNRT.

246
QTc and JTc prolongation after radiofrequency ablation (RFA) in children
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Background: Transient T wave abnormalities and QT changes have been reported in patients following RFA. Our observation of prolongation of the QTc in children during the first 24 hours following RFA, along with a case report of torsades de pointes in a patient 18 hours following RFA, prompted us to investigate the evolution of the QTc and JTc interval after RFA. **Methods:** We compared calculated QTc and JTc intervals in sinus rhythm in 137 patients using manually measured QT, JT and RR intervals as per-RFA, at 24 hours and at 2 months after the procedure. Only JTc interval was measured when pre-excitation was present, and other forms of aberrancy were excluded. **Results:** We observed an increase in the QTc and JTc interval at 24 hours after the procedure ($p=0.02$), although JTc shortening at 2 months did not reach significance ($p=0.8$). No ventricular arrhythmias occurred. Population means and standard errors are shown. **Conclusion:** There is significant prolongation of the repolarization in children 24 hours after RFA. The causes and evolution of QT prolongation after RFA should be further investigated.

247
Effect of respiration on intracardiac electrograms and radiofrequency ablation parameters
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Introduction: stable intracardiac electrograms with the desired morphology are critical for determining the site of successful radiofrequency(RF) ablation. Respiratory activity changes the heart-catheter relation and may cause dramatic electrogram and RF ablation parameter variability. Methods: Target intracardiac electrogram amplitude(mV) and 3 RF ablation parameters, power (P, watts), temperature (T,°C) and impedance (R, ohms) were examined for the effects of respiratory activity in 5 patients (28 mos- 17 yrs) undergoing catheter ablation therapy for SVT under general anesthesia. Four patients had an accessory mediated tachycardia and 1 had AV node reentry. The following respiratory sequence was used prior to and during RF application at the site of successful ablation: 5 inspiratory/expiratory (I/E) cycles (20 sec/cycle) at a tidal volume of 10 ml/kg and a brief E during the first 45 sec of RF application followed by a 10 ml/kg I held for the final 15 sec of RF. Data were analyzed for intra-patient and group I-E differences where appropriate. Results: Electrogram amplitude varied from I in E for 4 of 5 individual patients ($p<0.05$), and for the group. Three of the patients had a larger amplitude during inspiration, while 1 had a larger amplitude during expiration. During RF application, all 3 RF parameters (T,P,R) varied significantly between respiratory states with differences of 6.8 °C (range 1–32), 2 watts (range 1–5), and 4 ohms (range 1–7), respectively. A higher T was achieved during I in 3 patients and during E in 2 patients. In 2 patients, a shift of respiratory state caused T from above to below 50 °C. Conclusion: Respiratory activity affects intracardiac electrogram amplitude at the AV groove, indicating a changing relation between the catheter and the heart during ventilation. Further, ventilation related movement and possibly blood flow changes significantly affect the parameters which determine tissue heating. Tip temperature can vary above and below a target limitation of 50 °C, depending on respiratory phase. These observations suggest respiration may affect acute and long term procedure outcome.

Session 23: Surgical Management and Results: Univentricular Heart/Hypoplastic Ventriculoarterial Discordance

248
Should persistent fenestrations be closed late after a Fontan repair?
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Our unit has not had a policy of early routine fenestration closure after the Fontan IVJ operation as some fenestrations undergo spontaneous closure. Nevertheless continued fenestration may contribute to ongoing ventricular dysfunction and symptomatology. We recently undertook to close fenestrations in patients whose saturations were >90%. The purpose of the study was to review these patients. Six patients, aged 6–17 years, underwent transcatheter closure of surgically created fenestrations 4.7 + 1.7 years after the F. All patients had undergone a lateral channel F using Gortex with a single 4mm fenestration between 1994–8. Symptoms included exercise impairment (4) and headaches (1). 2 were asymptomatic. Ventricular function was normal/mildly impaired in 4, moderately impaired (1), severely impaired (1). Saturations were 81+ 3% All patients underwent cardiac catheterization including balloon test occlusion of the fenestration. The mean transpulmonary pressure rose from 9 + 2 to 12.5 + 3 ($p < 0.001$) but there was no significant decrease in cardiac output with the arteriovenous difference increasing from 22 + 7 to 26 + 6 (p = NS). Fenestration closure was by CardioSeal umbrella (2), Gianturco coils (3), Amplatzer Septal Occluder (1). Two patients had small concomitant baffle leaks with no attempt to close later. At most recent follow up saturations are 92 + 2% and 5 of 6 patients report improvement, 4 improved exercise tolerance and 2 improved mental performance. Conclusion: Late fenestration closure is associated with

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

spontaneous improvement and can be considered at any time interval after the Fontan repair. Ventricular dysfunction is not necessarily a contraindication to fenestration closure.

249 ✓

Understanding ventricular remodeling in hearts with tricuspid atresia: eccentric ventricular hypertrophy and decreased capillary-to-myocyte ratio

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The use of the Fontan principle has improved the survival and functional capacity of patients with univentricular atrioventricular connection. However, in some patients, myocardial hypertrophy expressed by a greater wall thickness jeopardizes the early and long-term outcome of the procedure. Little is known about the myocardial structure of these volume overloaded hearts. Methods: We examined 32 hearts with tricuspid atresia (10 with discordant ventriculoarterial connections) and 27 normal hearts (mean ages 9,6 months for both). For analysis, we considered two age groups: 0–2 months (15 hearts) and older than 2 months (17 hearts). Wall thickness was measured at the left ventricular (LV) inlet. Inlet and outlet lengths were also measured, as an indirect estimation of the cavity size. Myocyte diameter and endocardial thickness were measured by computer-assisted morphometry (Quantimet Leyra) at the LV inlet, apex and outlet. Immunohistochemistry for von Willebrand factor was used to label myocardial capillaries. Using a grid of known area, myocyte, nucleus and capillary profiles were counted and the myocyte-to-capillary and myocyte-to-nucleus ratios were calculated. Results: The wall thickness in the malformed hearts did not differ from the controls. Also, we did not find significant differences in the myocyte diameter in any of the regions and age groups analyzed. Myocyte diameter increased with age ($r = 0,45$ $p=0,02$). Endocardial thickness was greater in the malformed hearts. Myocyte to nucleus ratio was significantly greater in the malformed hearts ($p=0,008$). Myocyte-to-capillary ratio also tended to be greater, although without statistical significance. Conclusions: The findings are compatible with eccentric hypertrophy. Decreased number of capillary profiles in relation to myocytes could indicate an inadequate expansion of the capillary network in these malformed hearts, and possibly a greater vulnerability of the myocardium to ischemia.

250

Efficacy of total cavopulmonary connection without use of cardiopulmonary bypass

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Background: To determine the efficacy of total cavopulmonary connection (TCPC) without use of cardiopulmonary bypass(CPB). Methods: Since April 1996, 87 patients have undergone TCPC. Of these, the procedure was established without use of CPB in 41 (off-CPB group), while CPB was used because of intracardiac maneuvers concomitantly needed in the other 46 (on-CPB group). No significant difference was seen, between these groups, in age at operation, in preoperative mean PA pressure, and in preoperative pulmonary resistance. Results: Amounts of blood loss during operation and overall blood transfusion were smaller in the off-CPB group than in the on-CPB group ($p=0.02$). Duration of postoperative significant fluid sequestration was shorter, and the chest drainage tubes could be removed earlier in the off-CPB group than in the on-CPB group ($p=0.001$). In the off-CPB group, postoperative Respiratory Index ($PaO2/FiO2$ ratio) was higher ($p=0.01$), and duration of tracheal incubation was shorter ($p=0.03$). Particularly in the subset of patients under 2 years of age, pressure gradient between the pulmonary arteries and the atrium was smaller in the off-CPB group, and duration for inhalation of nitric oxide was shorter ($p=0.04$). Furthermore postoperative maximal concentrations of GOT, GPT, LDH and CK in serum were lower in younger patients of the off-CPB group. Measurements of complements and cytokines in serum demonstrated less activated inflammatory reactions in the off-CPB group; changes in levels of C3a, IL-6, IL-8, TNF-alpha, PMN elastase, and thrombomodulin being statistically milder ($p<0.04$) immediately after and 2 hours after the procedure. Conclusion: TCPC without use of CPB is an attractive surgical alternative, being less invasive, particularly in younger patients, if no intracardiac maneuvers are needed concomitant with establishment of the Fontan circulation.

251 ✓

Extra and intracardiac Fontan procedure after bidirectional cavopulmonary shunt

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The Fontan operation has been used to palliate patients with a functional single ventricle. In many such patients the operative risk for a Fontan procedure is high and bidirectional cavopulmonary shunt (BCPS) is a useful interim palliation. Outcome of the Fontan operation was analyzed to assess the effect of a prior BCPS and to compare the extracardiac conduit with intracardiac lateral tunnel, utilized as connection between the inferior vena cava (IVC) and the pulmonary arteries. From January 1992 to August 2000, 55 consecutive patients were submitted to Fontan procedure after a prior BCPS. In 24 patients (Group I) the connection between IVC and pulmonary arteries was accomplished by means of an intracardiac lateral tunnel; in 31 patients (Group II) an extracardiac conduit was utilized. Mean interval between BCPS and Fontan operation was 28 months. In 48 patients pulmonary arteries were judged normal sized; in 7 patients a significant kinking/stenosis were presented. There was no statistically significant difference between the two groups in terms of age, diagnosis and preoperative risk factors. In all patients the Fontan procedure was performed with the aid of cardiopulmonary bypass, in 54 patients through a median sternotomy and in one through a right lateral thoracotomy. 3 patients died in the hospital (5%) and one patient died after four month for neurological complications. After Fontan procedure there was no significant difference between the two groups in terms of hospital mortality, pleural effusion and protein losing enteropathy. Incidence of rhythm disturbances and ICU stay were lower in Group II. Fontan procedure after BCPS can be performed with a low mortality and morbidity. The use of an extracardiac conduit is a safe and reproducible technique, which may decrease the incidence of postoperative arrhythmias.

252 ✓

Subaortic outflow tract obstruction in univentricular atrioventricular connection

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Object: This study analyses risk factors for SAO and the results of surgical strategies for SAO treatment. Methods: All Pts with UAVC first treated at our institution from 12/2/1981 to 29/3/2000, are included. Pts' record, outpatient's record and the operative registry were analysed. Analysis of variables considered was performed by BMDP for Windows. Results: 24 pts were neonates or infants with UAVC not previously treated. Five Pts (20.8%) had coexisting coarctation of the aorta (CoA). Eight Pts (33.33%) had pulmonary stenosis. Median age was 7.5 days (range 1–370). Median weight was 3.5 kg (range 1.9–6.3). 17 pts underwent PAB (2 operative deaths in pts with associated arch hypoplasia). One pts with aortic apoplasia and SAO underwent a Damus-Kaye (DKS) procedure and died. Five pts required a shunt. One pts underwent a Glenn anastomosis (BDG). Operative mortality was 12.5%. The only factor affecting mortality was CoA ($p=0.0049$ by Fisher exact test). SAO developed in 10 pts (41,66%): 3 (12.5%) at birth; in 3 after PAB, in 4 (16.66%) after a BDG. The anatomic type of UAVC was predictive of SAO (TA/TGA vs DILV/TGA: $p=0.0022$). SAO was due to a restrictive VSD in 8 pts. Of these, 5 were treated by VSD enlargement. All these pts are currently well. SAO recurred in 2 pts: one underwent again VSD enlargement. None of the procedures performed was complicated by complete heart block. Follow-up is 100% complete. One patient died after a BDG (overall mortality: 5%,67%). A Fontan repair was achieved in 11 pts (45,83%). One patient underwent Fontan takedown. We conclude that pts with TA/TGA have a greater risk of developing SAO. Treatment of SAO by VSD enlargement is safe and doesn't vitiate the results of the Fontan operation.

253 ✓

Left ventricle is better suited as the pulmonary ventricle in simple transposition with pulmonary hypertension

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Ten infants with D-Transposition of the great arteries with essentially intact interventricular septum (DTGA IVS) and severe pulmonary arterial hypertension (PAH) underwent surgical treatment. Age ranged from 3 to 6 months (mean 4.2 months). One of these patients had a large shunt with left to right shunting but the others had no intra or extracardiac shunt to account for their pulmonary hypertension. All 10 had 'prepared' left ventricles.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

The first 4 children underwent an arterial switch operation (ASO). Uneventful surgery was followed by prolonged ventilator dependence in all 4 with recurrence of severe PAH every time weaning from ventilation was attempted. This was accompanied by metabolic acidosis and features of right heart failure. Only 1 patient (DORV IVS PDA) could be extubated and discharged from hospital. Subsequently, the other 3 infants underwent a Senning repair. There was no early mortality. All patients were separated from mechanical ventilation within 48 hours of surgery without blood gas derangement or heart failure despite elevated pulmonary artery pressures (PAP) in all. The child with the ASO has PAP at 50% systemic 3 years following repair while among the Senning group 2 patients continue to have PAP > 60% of systemic while 4 have normal PAP at a mean follow up of 8 months. Atrial level repairs seem to perform better than arterial level repairs in children with TGA with persistent PAH. Better tolerance of PAH in this group is probably consequent to the superior ability of the left ventricle to tolerate a pressure load in the early postoperative period.

254 J

One and a half ventricle repair: common and uncommon indications

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Patients with hypoplastic subpulmonary ventricle may benefit from the inclusion of a small ventricular chamber in the circulatory pathway. Method: Twenty-one pts were treated since November 1993. Mean age and weight at surgery were 51 mos (range 1–222) and 17 kg (range 2.6–53). All pts had a hypoplastic subpulmonary ventricle, this was morphologically right in 17 cases, morphologically left in 2 and morphologically outlet chamber (OC) in 2. Nineteen pts had been operated previously. Complete repair (absence of intra-atrial shunting) was achieved in 12 pts; a PFO or ASD were left intact in 9 pts. The repair included a BCPA in all pts, associated with procedures aps to regulate the pulmonary flow: S-P shunt division (10 pts), VSD closure (6), RVOTO relief (4). Two pts had a homograft conduit interposed between RA and OC. Results: three pts died (hospital mortality 14%). The youngest patient in the series (PA, IVS) and one patient with CAVSD, Down syndrome, did not survive; the third patient, with tricuspid stenosis, died of unexplained brain damage despite low SVC pressure. Tall systolic waves common in the SVC tracings postoperatively usually subsided within 48 hrs. In a mean follow-up interval of 35 mos (range 4–86), no late deaths occurred; 9 pts are asymptomatic, 10 have mild cyanosis or effort intolerance, 1 (the only pt with postoperative av block) had a poor result and required heart transplantation 6 years after 1,5 ventricle repair. Conclusion: the 1.5 ventricle repair can be applied to complex malformations with concordant, discordant or univentricular connection, provided an unobstructed BVCT and $Qp/Qs \leq 1$ or lower. The hazards/drawbacks inherent in biventricular or Fontan repair are thereby avoided.

255 J

Surgical strategies for isomerism heart with common atrioventricular canal

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Background: As isomerism heart is usually associated with common atrioventricular canal (CAVC) and abnormalities of systemic venous return, biventricular or univentricular repair (BVR or UVR) should be chosen by assessing the type of systemic venous return and CAVC, and systemic ventricular volume. We have performed BVR for isomerism heart with bilateral atrioventricular connection, over 70% of systemic ventricular volume and over 50% of ejection fraction. In the present study, early and late outcomes of BVR or UVR for isomerism heart were evaluated. Patients and Methods: 16 patients (pts) with isomerism heart (8 left) underwent definitive surgery. 7 of 8 pts with bilateral connection underwent BVR (G-BVR) and the remaining 1 and 8 pts with double inlet right ventricle underwent UVR (G-UVR). In G-BVR, 5 underwent RV outflow reconstruction and 5 underwent atrioseptal rerouting using PTFE patch. In G-UVR, staged Fontan operation was done in 8 pts with high risks such as TAPVR, severe CAVV regurgitation and hypoplastic pulmonary arteries. Two pts underwent fenestrated Fontan operation and fenestration was finally closed in both pts. Results: All pts in G-BVR are surviving with 1 to 15 years of follow-up and current NYHA status is 1 in 6 and 2 in 1, while two underwent mitral repair after primary repair. In G-UVR, one died of cerebral infarction early after surgery and one died of pulmonary arteriovenous fistula 2 years after surgery. The remaining 7 are surviving with 1 of NYHA status. PeakVO2 in G-BVR was significantly higher than that in G-UVR (28.9 vs 15.8 l/min/kg). Conclusion: Regards to survival and QOL after surgery, BVR should be selected if surgical criteria is matched. Even Fontan candidates with high risks can safely undergo UVR if staged operation is selected.

256 J

Coagulation factor abnormalities in children with single ventricle precede the Fontan procedure

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Objective: Thromboembolic events in post Fontan patients have been reported as high as 20%. A hypercoagulable state with deficiencies in proteins C and S has been implicated in these patients. Using age matched controls, this study evaluated whether an altered coagulation state is present earlier in the course of staged single ventricle (SV) repair. Methods: With informed consent, coagulation factors were assayed in 36 infants (mean age 8±4mo) with single ventricle (SV) cardiac defects, immediately prior to the bidirectional Glenn procedure (BDG). 32 infants (mean age 8±2mo) without cardiac disease were assayed as controls. Factors II, V, VII, VIII, IX, X, AT III, Plasminogen, Protein C and S were measured. Regression analysis was used to establish 95% normal reference intervals based on the control patients. Results: Reduced levels of multiple pro-and anticoagulant factors were detected prior to the BDG in all ($p<0.01$, Student t test). Eleven patients had two or fewer abnormalities whereas 25 patients had 3 or more. Most common abnormalities were low levels of Factors: II (25/36), V (26/36), X (25/26) and Protein C (23/36). Based on multiple logistic regression, patient demographics, SVC saturation, ventricular function, right atrial pressure and Qp/Qs ratio were not predictors of abnormally low factor levels. Conclusion: The study demonstrates that pro- and anti-coagulant factor abnormalities occur early in the course of SV and precede the cavopulmonary connection. It remains to be determined whether additional mechanisms, which might contribute to a potential hypercoagulable state exist, or whether these defects are congenital.

257 J

Risk factors for mortality following stage I reconstruction for hypoplastic left heart syndrome

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We reviewed a single-center, 4 surgeon experience with Stage I reconstruction for hypoplastic left heart syndrome (and variants) to determine risk factors for early survival in the current era. All 175 patients who were eligible for Stage I between 1/1/97 and 10/31/00 were included. Only 9 did not undergo surgery, secondary to multiple congenital anomalies (5), multiorgan system failure secondary to shock at presentation (3); and preoperative sepsis (1). These patients are excluded from further analysis. Of the remaining 166, 75 had acute atresia; 19 had additional congenital anomalies and/or genetic syndromes; 22 weighed < 2.5 kg; 10 were premature (<37 weeks); 7 had heterotaxy; and 5 had preoperative obstruction of pulmonary venous return. Measures of intraoperative support (cardiopulmonary bypass [CPB], circulatory arrest [CA] and total support [CPB+CA] times) were also evaluated as possible predictors of mortality. There were 152 patients (80.5%) who survived to hospital discharge (or 30 days, whichever came later). In multivariable analysis, preoperative obstruction to pulmonary venous outflow (OR=8.4, p=0.026), and an increase in CPB, CA or CPB+CA times (OR=1.02/min, p<0.001) were significantly associated with death. There was a trend towards increased mortality with additional congenital anomalies (OR=2.9, p=0.07). Aortic atresia, heterotaxy, weight, prematurity and surgical date were not associated with death. Median (range) times (min) were: CA = 42 (11–77); CPB = 41 (35–196) and CPB+CA = 84 (68–240). All operative times were highly linked with surgeon, but not with patient characteristics, such as low weight or the presence of aortic atresia. We conclude that preoperative patient characteristics (other than obstructed pulmonary veins) did not identify a subset of patients at increased risk for operative mortality. Efforts to improve results, therefore, should focus on intraoperative management, rather than patient selection.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

258
Survival in patients undergoing delayed stage I Norwood palliation: The Children's Hospital experience 1980–2000
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OBJECTIVE: To evaluate survival and determine risk factors in patients who underwent stage I Norwood palliation > 30 days of age. METHODS: 58 consecutive patients > 30 days of age who underwent stage I Norwood palliation between January 1980 and December 2000 were evaluated. Anatomic malformations in 10 patients were classified as hypoplastic left heart syndrome (HLHS) defined as aortic and mitral atresia or stenosis, normal segmental anatomy, intact ventricular septum and hypoplastic left ventricle. Other cardiac malformations included double-outlet right ventricle (n=3), double-inlet left ventricle (n=2), unbalanced complete AV canal (n=2) and L-TGA with tricuspid atresia (n=1). Median age at diagnosis was 10 days (range 0–221 days) and at operation was 54 days (range 32–696 days). RESULTS: All 10 patients underwent stage I Norwood palliation. There were 2 intraoperative deaths (10.3%). Actuarial survival derived by the Kaplan-Meier method was 82% at 1 month (95% confidence interval = 70–90%) and 66% at 3–12 months and thereafter (95% confidence interval = 50–82%). Median follow-up was 37 months (range 1.1–216 months). Eight patients have since undergone fenestrated Fontan procedures, 2 have undergone bidirectional cavopulmonary anastomoses (BDCPA), and 2 await further palliation. Preoperative PRISM III physiology score was the only significant risk factor for mortality according to the Cox regression model: each additional point was associated with a 30% increased mortality risk of death (risk ratio = 1.3, 95% confidence interval = 1.1–1.5, p = 0.002). Anatomic diagnosis, reason for delay, age at diagnosis, presence of restrictive ASD, age or weight at operation, size of Blalock-Taussig shunt, and year of operation were not predictive of outcome in univariate or multivariate analyses. CONCLUSION: Stage I Norwood palliation is appropriate for patients > 30 days of age with HLHS or other unstable malformations.

259
Differences in anaerobic threshold predict mid-term survival after the Norwood operation for hypoplastic left heart syndrome
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We previously reported the relationship between SvO2 and anaerobic metabolism in neonates following the Norwood procedure (NP) for HLHS, finding a high early survival rate (93%) utilizing a management strategy to maintain SvO2 above an apparent anaerobic threshold of 30%. In this report, we compare the early anaerobic threshold between survivors vs. non-survivors to bidirectional Glenn operation. Neonates undergoing the NP for HLHS underwent standardized perioperative management with physiologic parameters recorded prospectively for the first 48 postoperative hours. SvO2 was monitored continuously using a fiberoptic catheter in the SVC. Anaerobic conditions were defined as a standard base excess (BE) less than -4 mEq/L or a change in BE of -2 mEq/L/hour. The risk of anaerobic conditions at strata of SvO2 between survivors and non-survivors was tested by the likelihood ratio test, and by ANOVA for repeated measures with Tukey's correction for multiple pairwise comparisons with p<0.05 considered significant. Of 51 consecutive patients, 39 (76%) survived 6 months or more to stage II. There were no differences in age, weight, normalized shunt size, or CPB time. The mean SaO2 (74.2% vs 77.5%, p=0.024) was lower in non-survivors. Although the mean blood pressure or SvO2 did not differ between groups, the risk of anaerobic metabolism was significantly higher at SvO2 less than 50% in the non-survivors (18.5% vs 8.1%, p=0.0038, see table). The risk of anaerobic metabolism was low in survivors until SvO2 <30%, while the risk rose significantly at SvO2 <50% in non survivors (p<0.0001). The anaerobic threshold after Norwood palliation of HLHS is higher in patients who suffer mid-term mortality compared to survivors, possibly indicating intrinsic biochemical vulnerability in non-survivors.

Session 24: General Pediatric Cardiology, Prognosis/Natural History

260
Management and outcomes of 116 infants and children with right atrial isomerism
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We report on the management and outcomes of 116 infants and children (71 males) with right atrial isomerism, the largest clinical series to date from a single institution. The data of patients diagnosed with right atrial isomerism between 1/1980 and 11/2000 were reviewed. Actuarial survival was assessed by Kaplan-Meier estimates, while effects of covariates on survival were analyzed by Cox regression model. The median age at presentation was 1 day (range 1 day to 3.7 years). Cardiac morphologic anomalies included dextrocardia (25%), common atrium (5%), common atrioventricular valve (92%), single main ventricle (83%), abnormal ventriculoarterial connection (95%), pulmonary outflow obstruction (85%), anomalous pulmonary venous (PV) drainage (51%) and PV obstruction (15%). Surgery was performed in 36% (42/116), 76% (32/42) of which died. The surgical mortality for PV repair was 43% (3/7), Fontan procedure 32% (6/19), and cavopulmonary shunt 15% (2/13). Late mortality was related to infection (n=9), sudden death of unknown aetiology (n=6) and arrhythmia (n=1). The proportions of patients surviving at 1 month, 1 year, 5 years were 80%, 65% and 51%, respectively. Independent risk factors for mortality included PV obstruction (relative risk [RR] 3.6, p=0.001), PV drainage (RR 0.33 for normal drainage, p<0.0001), single ventricle (RR 2.9, p=0.016) and oxygen saturation (RR 0.96, p=0.011). Survival estimates for children with normal PV drainage were 93% at 1 month, 90% at 1 year and 86% at 5 years. No risk factors were identifiable. The outcomes of children having right atrial isomerism with or without PV obstruction remain discouraging despite surgical intervention.

261
Natural history of aortic root dilatation following correction of tetralogy of Fallot (TET)
Hawker R, Smolc C, Goldsmith R, Celermajer D
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Natural history of aortic root dilatation following correction of tetralogy of Fallot (TET). Hawker, R., Smolc, C., Goldsmith, R., Celermajer, D. Sydney, Australia. This longitudinal review of the aortic root dilatation (root dil.) in patients (pt.) with corrected TET studies: 1. Whether pre-op root dil. persists or progresses. 2. Whether early correction will lessen late root dil. > From echo databases of our two units, M-mode aortic root size in mm was obtained from 533 studies in 265 pt. followed 1–23y. after correction of TET and plotted using a polynomial fit against the normal data of Henry et al. Root dil. (y axis) persisted in 456 studies of 242 pt. Repaired beyond 1y. of age (Fig 1). In 69 studies of 43 pt. corrected before 1y., root size approached the normal range by about 6y. (Fig 2). Pat. Fig1 here. Pat. Fig2 here. Conclusion: 1. Root dil. persists in post op TETs. It will be aggravated by hypertension & aortic sclerosis. 2. Early correction may allow TETS to grow into

262
Aortic regurgitation post-balloon aortic valvuloplasty: predisposing morphology
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Objective: To identify by echocardiography the morphology of the aortic valve (AV) and aortic annulus (AA) associated to the development of aortic regurgitation (AR) after balloon valvuloplasty. Methods: between 1990–99, 51 patients (pts) with aortic valve stenosis (PG > 50mmHg) underwent a balloon valvuloplasty at age x: 8m (1day–16 years). An echo-doppler study was performed prior, immediately after and every 6 months during a follow-up of 1 to 9 years, x: 4 ys. Results: the degree of AR immediate after the procedure was absent in 4, trivial or mild in 45 and moderate in only 2 pts associated to avulsion of the aortic cusp. During follow-up 3 groups were clearly identified: AR did not progress and remained mild, Group I: 30 pts (61.2%); AR progressed and became moderate, Group II: 16 pts (32.6%) and turned severe, Group III: 3 pts (6.1%). In Group I a bicuspid valve was found in 90% of pts and a circular shape

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

263

Natural history of congenital heart block in pediatric patients

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Background The natural history of isolated congenital complete atrioventricular block (CCHB) is considered favorable in adults. The prognosis is essentially related to the severity of underlying condition such as ischemic, hypertensive, or valvular heart disease. However, this belief is based on studies comprising patients without symptoms during their childhood. Objective To elucidate the natural history of CCHB in pediatric patients. Patients and Methods: 10 patients (4 female and 6 male) with isolated CCHB admitted at our institute from 1979 to 2000 were reviewed. The mean age at follow up or at death was 10 years, ranging 0 to 18 years. The hospital records were studied retrospectively, including the age at pacemaker implantation (PMI), LVFS, titers of anti-SSA/Ro antibodies in maternal sera (SSA Ab), and autopsy findings. Results: 9 patients received PMI at the age of 0 in 5 and 1, 2, 4, 6 years in 1 each. The only one patient is doing well during his 15 years of life without PMI. There were 5 deaths. The causes of death were endocardial fibroelastosis in two, and the first attack of ventricular arrhythmia in one patient. The ages of death were 1 day, 2years, and 15 years respectively. Of 5 living patients without symptoms, 4 showed reduced LVFS below 30%. 18 years old female having mitral regurgitation, LVFS of 24%, and frequent VPC experienced syncope. The titers of SSA-Ab were extremely high (>1:800) in 4 patients, 3 died and 1 experienced syncope. Conclusions: Natural history of CCHB in pediatric patients is not so favorable as in adults. Pericarditis caused by autoantibody during fetal period might be responsible for myocardial disorders as well as conduction system. Careful follow up should be continued even after PMI.

264

Increased brain and atrial natriuretic peptides in patients with chronic right ventricular pressure overload

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Objective: To evaluate the role of plasma neurohormones in diagnosis of asymptomatic or minimally symptomatic RV dysfunction. Setting: Tertiary cardiovascular referral center. Methods: Plasma brain natriuretic peptide (BNP) and atrial natriuretic peptide (ANP) levels were measured in 21 asymptomatic or minimally symptomatic patients with chronic RV pressure overload due to a congenital heart disease and in seven healthy volunteers. RV ejection fraction (EF) was determined using magnetic resonance (MR) imaging. Results: RVEF of the volunteers was significantly higher than RVEF of the patients (59.0(8.2)% v 38.0(12.0)%, respectively, p<0.0001). Left ventricular (LV) EF in volunteers and patients was 72.3(7.8)% v 69.1(11.0)%, respectively, p=NS. Between patients and volunteers there was a significant difference in the plasma concentrations of BNP (5.3(3.5)pmol/L v 2.3(1.7)pmol/L, respectively, p=0.009) and ANP (7.3(4.3)pmol/L v 3.6(1.4)pmol/L, respectively, p<0.05). Both in patients and volunteers mean ANP plasma concentrations were higher than mean BNP plasma concentrations. RVEF was inversely correlated with BNP and ANP (r=0.65, p<0.0002 and r=0.61, p=0.002, respectively). No correlations were found between LVEF and BNP (r=0.2, p=NS), and LVEF and ANP (r=0.32, p=NS). Similarly, no correlation was shown between the level of RV systolic pressure and plasma neurohormones BNP (r=0.20) and ANP (r=0.07) respectively. Conclusions: Our study shows a significant inverse correlation between RVEF and the plasma neurohormones BNP and ANP in asymptomatic or minimally symptomatic patients with RV pressure overload. Monitoring of changes in BNP and ANP levels may provide quantitative follow up of RV dysfunction in these patients.

265

Left coronary Doppler flow dynamics in neonates

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Background: Evaluation of coronary flow with transthoracic echocardiography and Doppler has recently become possible in children. Normal values for left coronary artery Doppler flow velocities in neonates are not yet known. The aim of this study was to assess the relation between left coronary flow dynamics and age, left ventricular size and function. Methods: Fifty four healthy neonates were examined with transthoracic echocardiography including pulsed wave Doppler registration in the proximal LAD (90 systoles). Their median age at examination was 3 days (range 1 to 30 days). The LAD Doppler curve was analysed regarding peak flow velocity in diastole (PFVd) and systole (PFVs) and velocity time integral (VTI). VTI per minute (VTI/min) was calculated by multiplying the sum of diastolic and systolic VTI with heart rate. Results are presented as mean (SD). Results: PFVd was 25.7 (8.2) cm/s. It increased linearly with age (r=0.45, p=.0003), and was linearly related to aortic peak flow velocity and the aortic VTI (r=0.51, p<.0001). PFVs was 13.7 (5.0) cm/s. It increased linearly with age (r=0.60, p<.0001), and had a weak linear relation to aortic VTI, and left ventricular mass. The sum of diastolic and systolic VTI was 6.0 (1.9) cm. VTI increased linearly with age and left ventricular mass (r=0.59 and 0.55 respectively, p<.0001). VTI/min was 7.32 (2.96) m. It increased linearly with age, aortic VTI and left ventricular mass (r=0.70, 0.46, and 0.43, p<.0001, = .0010 and .0029, respectively). Conclusions: Normal values for LAD Doppler flow velocities, velocity time integrals and flow are reported for neonates. In healthy neonates, coronary flow parameters are linearly related to age, left ventricular mass, and left ventricular function.

266

Williams syndrome outcome multi-center pediatric cardiac consortium

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Introduction: The data for outcome management of cardiac catheterization and surgery in Williams syndrome (WS) patients is limited due to its rare occurrence. The Pediatric Cardiac Care Consortium (PCCC) data collection offers an understanding of cardiac anomalies distribution and surgical therapy. Using the PCCC database we reviewed 244 children with Williams syndrome and cardiac anomalies. Methods: The PCCC is a collaborative effort of 45 pediatric cardiac centers in 20 states. Data regarding cardiac catheterization, cardiac operation and autopsies are collected from participating centers for analysis with the goal of improving cardiac care in children. The records of 244 patients with WS were reviewed from over 62,000 records in the PCCC database from 1984- 1999. There were 149 (61%) males with WS. Ages at the first catheterization or operation ranged from 1 day to 21 years, the week the average age of 57.3 months. The average weight was 17.1 kg. Results: Among the 244 patients, 294 cardiac catheterizations and 144 operations were performed. Eighteen deaths occurred, 11 within 30 days of operation, 6 within 48 hours of a catheterization, and the remaining patient died unrelated to a procedure. Twelve deaths occurred in 80 patients with both supravalvular aortic stenosis (SVAS) and pulmonary artery stenosis (PAS): 6 following operation and 6 following catheterization. In contrast, only 1 death occurred in 70 patients with SVAS alone and one in 29 with PAS alone. Three remaining deaths occurred in 58 patients with SVAS or PAS with another cardiac lesion. Conclusions: Supravalvular aortic stenosis is the most common cardiac lesion associated with Williams syndrome. The patients with SVAS or PAS have a low rate of mortality in comparison with other lesions. However, when the two lesions occur in combination, the mortality rate is significantly increased for both cardiac catheterization and cardiac operation.

267

Percutaneous versus surgical closure of secundum atrial septal defects

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Background: Surgical ASD closure provides excellent results. With the current risk of percutaneous techniques, a comparison is needed. Aim: To compare percutaneous and surgical closure of secundum ASD. Patients: From 4/96 to 10/00, 493 consecutive pts underwent percutaneous ASD closure. ASD (Group A); From 4/92 to 10/00, 400 consecutive pts underwent ostium secundum ASD surgical repair (Group B). There was a slight difference in mean age between the two groups (Group A: 26.6(19) yrs vs 24.5(17.5)yrs, p=0.05). Results: Hospital stay was lower in group A (3.1(0.6)days vs 9.7(2.6)days, p<0.0001). Total number of complications was higher in group B (39 % vs 1.5%,p<0.0001). In Group A complications included: (a) device embolization needing surgical retrival (1.4 %); (b) moderate pericardial effusion (0.2 %), (c) Haemopericardium needing pericardial drainage (0.2 %); (d) iliac vein dissection needing percutaneous stent implantation (0.2 %);

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

(n) transient atrial fibrillation (0.8 %); (f) severe haematoma of the groin (0.2 %), thrombus formation on the device needing anticoagulant therapy for 6 months (0.2 %). In Group B complications included minor transient complications in 86 % of the pts (respiratory, arrhythmias, pericardial effusion, anemia, pneumothorax), severe transient in 8 % (arrhythmias, severe bleeding, heart failure), minor with sequelae in 4 % (arrhythmias) severe with sequelae in 2 % (neurologic, complete AV block needing PM implantation). Transfusion rate was 2 %. Chest re-opening rate for severe bleeding was 1 %. No death occurred. Residual shunt at discharge was trivial in 6 % in Group A vs 3 % in Group B. Five pts of Group A were treated for a significant residual shunt after surgical ASD closure. Conclusions: Percutaneous ASD closure provides excellent results with lower complications, no sequelae and shorter hospital stay.

268

Acceptability Of finger-prick anticoagulation control in children

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Background: Oral anticoagulation is indicated for children at risk of thrombosis. Maintenance of the international normalised ratio (INR) in the desired range can be difficult, necessitating frequent blood sampling. We present data on children whose anticoagulation was monitored with finger-prick samples using CoaguChek(r) machines. Methods: Patients on warfarin (coumadin) were identified using computerised patient records and INR cards. Parents provided their views on the CoaguChek system using a telephone questionnaire. Haematology laboratories tested and quality-controlled hospital CoaguChek machines, concurrent quality-control samples were used for home monitors. Results: Finger-prick sampling of INR commenced in 1994 in Cardiff and 1998 in Bristol. Prior to this, blood samples were obtained by venepuncture. 34 patients (aged 3 months-16.5 years) having finger-prick samples were identified. 24 had undergone venepuncture previously and 10 had only used the CoaguChek system. Indications for anticoagulation included prosthetic valve or conduit (19), Fontan operation (13) or dilated cardiomyopathy (2). Finger-prick sampling led to increased frequency of INR testing in 2 patients. Accuracy of the machine was tested against venepuncture only if the INR was out-of-range. There was close agreement (to within 0.4) between the two methods of INR analysis, unless the INR was >8. There were no thrombotic or haemorrhagic complications in the follow up period. 32/34 parents expressed a preference for finger-prick analysis over venepuncture because of rapid availability of results, ease of use and reduced patient discomfort. 2 patients reverted to venepuncture due to erratic

269

Late systolic hypertension at exercise after coarctation repair is associated with reduced distensibility of elastic arteries and impaired endothelial function

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The pathophysiological mechanisms responsible for late hypertension and cardiovascular morbidity after coarctation (Coa) repair have not been clearly assessed. We studied 20 normotensive subjects at rest (age:14±5 y, pressure:116±13/56±9 mmHg) who had a good repair of Coa (defined by the absence of gradient between upper and lower right limb (0. -20 mmHg). After exercise testing, we defined 2 groups: Coa HT : systolic hypertension at exercise ≥ 200 mmHg (n=20, 228±23 mmHg) and Coa NT : normal systolic pressure at exercise (n=10: 163±24 mmHg). These subjects were sex-, age- and blood pressure-matched to 20 controls (age, 13±3 y, pressure 115±10/56±6 mmHg). Using echo-tracking technique, we measured common carotid artery (CCA) diameters and the intima-media thickness (IMT). Compliance (CSC), distensibility (CSD) and elastic modulus (Einc) were calculated. CCA pressure waveform and the local pulse pressure were determined in 30 subjects to define augmentation index (AI). Vasodilation of the brachial artery in response to reactive hyperaemia and to glyceryltrinitrate (GTN) were measured. The IMT was increased in the whole Coa group (p<0.001) (0.57±0.04 mm in Coa HT vs 0.54±0.05 mm in Coa NT). The CSD was decreased and the Einc was significantly higher all patients. The carotid pulse pressure was higher in the Coa HT (48±14 vs 33±7 mmHg, p<0.05). The AI was increased in both Coa groups. Flow-mediated dilation and GTN-mediated dilation of the brachial artery were reduced in the Coa group (p<0.01). GTN-mediated dilation was inversely correlated with maximum systolic blood pressure at exercise (r=-0.31, P=0.03). The combination of distensibility decrease in the proximal arterial bed with an impairment of distal artery reactivity can account for the elevation of exercise blood pressure after Coa repair.

270

Recombinant human alpha-glucosidase from rabbit milk has effect on cardiac muscle in patients with Pompe's disease

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Infantile Pompe's disease is a fatal muscular disorder caused by lysosomal alpha-glucosidase deficiency. Hypertrophic cardiomyopathy is characteristic for the disease, which is commonly fatal in the first year of life. In an open label study 4 babies with typical symptoms of infantile Pompe's disease and virtual absence of alpha-glucosidase were treated with intravenously administered recombinant human alpha-glucosidase (rhGAA) from rabbit milk in dose starting with 15 or 20 mg/kg and later 40 mg/kg. No major side effects were seen. At inclusion 2 patients had end stage disease, ages 7,8 months; 2 were included before age 3 months. Clinical improvement was seen in all patients, but the most prominent effect was seen on the heart. Based on 2D echo, left ventricular mass index decreased significantly over 64 weeks of treatment from 171,203,318 and 599 g/m2 at the start to 77,155,104 and 115 g/m2, each p <0.05. In the 2 youngest included the mass index was low etc. their clinical condition is best. Symptoms of cardiac instability disappeared, 3 patients are on mild diuretic medication now at the age of 26, 25, 24 and 29 months resp. Indexed mass is still increased but systolic function is normal. Diastolic function is mildly restricted in 1 patient. In conclusion rhGAA from rabbit milk has therapeutic effects in infantile patients with Pompe's disease. It is recommended to start treatment early.

271

Prognostic value of aortic elasticity on aortic complications in patients with Marfan syndrome

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Background: In Marfan patients survival is mainly determined by aortic complications at a relatively young age. The occurrence of acute dissection and rupture in Marfan patients is difficult to predict by mere aortic dimensions. Assessment of aortic elasticity may be of additional value for risk stratification. Methods: To assess the prognostic value of aortic elasticity on aortic complications (defined as: 1 aortic root diameter increase > 2 mm/year, 2 aortic dissection or 3 death) 73 Marfan patients (aged 31 ± 8 years, 41 men and 32 women) underwent magnetic resonance imaging of the entire aorta in 1997 and were followed up for 3 years. Aortic diameter and ascending aortic distensibility were assessed. MR velocity mapping was used to assess flow wave velocity along the descending aorta as an additional index of elasticity. Results: 10 patients (13.6%) of the 73 patients examined in 1997 reached one of the endpoints (7 patients with an increase in aortic root diameter > 2 mm/year, 3 acute dissections) after 3 years (3.1 ± 0.2 years). The patients were divided in a complicated and a non-complicated group. There was no significant difference in baseline characteristics and in aortic root diameter (47 ± 4.4 vs 43 ± 7.3 mm, respectively) between the two groups. However the 10 complicated patients had a significantly decreased local ascending aortic distensibility (2 ± 1 vs 3 ± 1 x 10-3 mmHg-1, respectively) and significantly higher descending aorta flow wave velocity (6.6 ± 1.6 vs 5.8 ± 1.3 m·s-1, respectively) compared to the non-complicated Marfan patients. Conclusion: Descending aorta flow wave velocity and ascending aorta distensibility are related to the occurrence of aortic complications in Marfan patients and could be of additional prognostic value for risk stratification.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Session 25: Catheter Interventions

272
Amplatzer and Cardioseal devices for ASD closure: results of a 'geographically' randomised study
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Background: Comparison of results of ASD closure with Amplatzer and Cardioseal devices has been hampered by non-randomisation for device type. Methods: We compared ASD closures (intention to treat) with both devices in 2 university hospitals within the same region. Each centre used 1 type of device. Device type did not influence referral patterns. Patient selection followed criteria of manufacturers for clinical trials in use. Amplatzer: central ASD, 5 mm rims. Cardioseal: stretched diameter < 21 mm, device < 80% of septal length. Results: In a 3-year period, the Amplatzer centre (Amp) performed 28 procedures, the Cardioseal centre (Car) 25, in children. Age at catheterisation (Amp vs Car) (6 ± 4 vs 7 ± 5 years), weight, Qp/Qs (2.2 ± 0.8 vs 2 ± 0.5) and stretched ASD diameter (16 ± 5 vs 21 ± 5.5 mm) did not differ between the groups. In the Amp group 2/28 of the procedures were unsuccessful, in the Car group 4/25 (p=n.s.), with complications in 1 Amp patient (air embolism into coronary arteries) and 2 Car patients (1 inappropriate position, retrieval and subsequent surgery, 1 surgical removal; p=n.s.). Duration of follow-up (Fu) (6 ± 6 vs 13 ± 6 months) was significantly longer for the Car group. Residual atrial shunts at 6 months Fu: 0/16 for Amp, 5/16 for Car (p=0.015), at 12 months Fu: 0/7 vs 4/13 (p=n.s.). Complications did not occur during Fu. Conclusions: In similar populations, results of Amplatzer and Cardioseal ASD closure are highly comparable. For Cardioseal devices, % residual shunt is higher as short term Fu. Restrictions in ASD size are the most important limitation of the Cardioseal device.

273
Bridge stents in the management of obstructive vascular lesions in children
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Balloon expandable Palmaz stents have almost exclusively been used in the treatment of vascular stenoses in children. Whereas these stents have generally been useful, problems of balloon rupture, stent migration, requirement of a large delivery sheath and longitudinal rigidity continue to exist. To circumvent these problems we have utilized bridge stents (balloon expandable flexible biliary/peripheral stents manufactured by Medinvent AVE). During a 9-month period preceding July 2000, 13 children, ages 1–16 years, underwent stent implantation to treat long segment stenosis of branch pulmonary arteries (N=8), right ventricular outflow conduit (N=2), aortic coarctation (N=2) and post-surgical superior vena caval obstruction (N=1). The stents were delivered to the implantation site without a long sheath in six patients and via a 8-F long blue Cook sheath in the remaining seven. No difficulties were encountered in traversing tortuous course nor were there any instances of balloon rupture. Increase of stenotic segment diameter (3.7±1.2 vs 10.2±2.7 mm, p<0.001) and reduction in pressure gradient (28±9 vs 6±2 mmHg; p<0.01) occurred. Quantitative pulmonary perfusion scans (23±9 vs 41±8%, p<0.05) showed improvement in ipsilateral lung perfusion. Follow-up echo-Doppler studies were available in all patients (1–12 months) after stent deployment and continue to show improvement in Doppler derived gradients. The data suggest that it is feasible to implant bridge stents via a small caliber or without a long sheath and without dislodgement or balloon rupture and that appears to be related to creation of

274
Various reasons for re-dilation of stented pulmonary arteries in paediatric patients
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Between 1994 and 2000 46 pulmonary stenoses in 38 patients were treated by implantation of 56 balloon expandable stents. 14 native stenoses and 32 postoperative lesions were stented. The age of patients ranged between 6 days and 34 years, mean 6.9 years, the bodyweight from 2.5 Kg to 75 Kg, mean 20.5 Kg. 52 Palmaz stents 3 Corinthian and one Jupiter stent were implanted in 16 main pulmonary artery stenoses, 19 right and 27 left pulmonary artery stenoses. At time of implantation the quotient between stent diameter and stenosis was measured 1.25 to 12, mean 3.1; the quotient between stent and surrounding vessels 0.8 to 2.4, mean 1.1. During follow up time of 3 months to 6 years, mean 2.2 years 42/56 stents were re-catheterized. 39 re-dilations were performed on 28/42 stents. Out of the 28 stents re-dilation was performed twice on 8 stents, 3 times on four stents. The mean time period between implantation, re-dilations and re-re-dilations was 15.5 months, the mean gain of weight during these period 3 Kg. 27 re-catheters were accomplished on 21 stents in 18 lesions due to the growths of the patients. Three patients with four stents in left pulmonary artery stenoses were re-dilated 6 times because of intimal proliferation. The patients with two stents showed repair intimal proliferation. A pulmonary homograft stenosis, compressed from outside, was reserted twice and re-dilated three times. Conclusion: Re-dilation of stented pulmonary arteries was mainly performed in order to adapt the stent-diameter to patients growths (69%). However, 15.5 % of the re-dilations, all performed on left pulmonary artery stents, were done because of intimal proliferation and 15.5 % of re-dilations had to be carried out due to compression from outside surrounding structures.

275
Inflammatory response and myocardial trauma during and after amplatzer occlusion and conventional surgery of ASD
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Aim: The aim of this study was to compare the acute immune response and myocardial damage during and immediately after convential surgery as compared to Amplatzer occlusion of ASD. Cardiopulmonary bypass is associated with the sequential release of numerous stimulatory (interleukin-6, -8) and of inhibitive cytokines (IL-10) and complement activation. Additionally, a relevant myocardial damage. Method: This prospective study was performed in children (age 3 to 12yrs) who underwent surgery for ASD (n=20), Amplatzer occlusion (n=10) or diagnostic cardiac catheterisation (Control, n=20). Blood samples were obtained 1 d before, at, and 4h, 1d and up to 1 month after intervention and analyzed serologically and for cellular components. Results: Patients responded to ASD surgery by an acute inflammatory response with leukocytosis, neutrophilia, massive release of IL-6, -8, -10 and complement activation. None of these signs were found in the Amplatzer and control group. ASD surgery induced myocardial damage as detected by elevation of serum Troponin I. Much lower Troponin I was found in the Amplatzer and no elevation in the control group. The effect of Amplatzer occlusion on platelet activation will be detailed. Conclusion: Amplatzer ASD occlusion of ASDII is associated with minimal trauma and no detectable impairment of the immune system. A transient platelet activation is induced, possibly at the initial step in closing the occluder. Furthermore, it induces only minor acute myocardial damage as compared to the conventional approach. The variables do not reflect the existence of the 'foreign body' uncovered by endocardial tissue.

276
Percutaneous pulmonary valve implantation
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Stenosis and insufficiency of the valve in right-ventricle to pulmonary-artery prosthetic conduits is a common problem leading to reoperations for conduit replacement. Conduit stenting has emerged as an alternative technique delaying the time of the surgery but creating or increasing the pulmonary insufficiency. We developed a device for percutaneous stent implantation combined with valve replacement. Here we describe the experience of the first non surgical heart valve replacements in the human. Two 12 years old children with an 18 mm prosthetic conduit between the right ventricle and the pulmonary artery underwent percutaneous pulmonary valve replacement. They were symptomatic because of significant stenosis and insufficiency of the conduit. An 18-mm biological valve was mounted in a valvular stent and then cross-linked with a glutaraldehyde solution. Catheterization through the femoral vein was performed under general anesthesia. Hemodynamic evaluations and angiographies were obtained to identify the position of the stenosis and to confirm the indications of valved stent placement. The valved stent was mounted onto a specially designed 18F with an 18-mm balloon catheter. The system was then connected and advanced in the pulmonary trunk on a previously positioned guide wire. The balloons were thereafter inflated deploying the valved stent at the position of the

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

obstruction. The system was subsequently removed and angiographic and hemodynamic studies ended the procedure. The two valved stents were successfully delivered. In both patients the newly implanted valve was competent in diastole and the systolic obstruction of the conduit was reduced. There were no complications and the valve was perfectly functioning during the early follow up.

277

Heart Function and hemodynamic changes after secundum ASD occlusion with the Amplatzer device

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Objectives Although transcatheter closure with the use of occluder devices has been increasingly used in the management of Atrial Septal Defect (ASD), there are no studies examining the impact of such an intervention to the Left Ventricular (LV) and Left Atrial (LA) mechanical function. This study determined LV and LA Volumes and function in a group of ASD patients 24 hours follow implantation of the Amplatzer Septal Occluder (ASO). Methods: 42 pts with ASD (age 8.5+/-3.8 years) successfully treated with ASO (device size 18 to 30 mm, mean 19.5+/-3.8 mm) were studied with M-mode and 2 Dimensional EchoCG. Results: Before ASD occlusion there is a type D paradoxical motion of the IVS due to RV volume overload. This causes decrease of LV end-diastolic and of LV end-systolic diameter and volume. As a result the Stork Volume and Ejection Fraction are decreased. After occlusion the paradoxical motion of the IVS disappear. In addition there is increase of LV end-diastolic (11.3%) and decreasing of LV end-systolic (87.66%) diameter and volume. This is result increasing of the stork volume and ejection fraction (108% and 104% respectively). All before closure data were considered as equal to 100% for normalization of the age, weight height body surface of the different ages children. After occlusion it was significant changes also in the LA function: systole diameter of the LA decreases and diastole diameter increases, the Volume of the Fast Filling, Volume of Slow Filling and Volume of Atrial Systole increase comparatively with these initial level. Conclusion: Transcatheter device closure of atrial septal defects has a positive impact on left ventricular and left atrial function, which is observed early follow.

278

Stenting of native coarctation of the aorta – technical overkill or acceptable treatment

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The implantation of stents even in native coarctation of the aorta (CoA) offers a possibility to achieve long-lasting increase of the diameter minimizing the pressure gradient immediately. We report our clinical experience of stenting native CoA as the first treatment to judge safety during midterm follow-up. Methods: In 14 patients (P) (mean age 7.9 years (Y) (0.1 – 39.4)) retrograde BD of native coarctation failed with a remaining gradient over 25 mmHg. All these patients underwent subsequent stent implantation via a retrograde approach during the same procedure. The diameter of the balloon catheter inflating the stent was chosen according to the diameter of the transverse arch. In P younger than 10 Y of age a stent with a possible expansion up to 15 mm diameter and in older P stents up to 25 mm were used. Results: The post-stent systolic gradient was median 5.0 mmHg (4.5 – 7.0)) and disappeared completely after implantation (p<0.001). In all P total expansion of the stent could be achieved with a mean proportional increase of the stenosis of 116.7% (61.5 – 200). No vascular complications or thromboembolism have occured. During a median follow-up of 1.75 Y (1.02 – 7.7) only in 1 P a neointima layer caused a Re – CoA, which could be unproblematic treated by ballooning 1.5 Y after implantation. No adverse systemic hypertension has been noticed in any patient after stent implantation. During follow-up only 24% of the P need further antihypertensive treatment. Conclusion: Stent implantation in native CoA seems to be an alternative treatment to surgery if BD failed. The mid term results are very promising. To prove whether adaptation to growth could be managed by redilatation long term follow up is required.

279

A new Amplatzer device to maintain ductus arteriosus patency: preliminary results in lambs

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Background: Systemic to pulmonary artery shunt (SP) is a standard palliation in patients with cyanotic congenital heart disease. The purpose of the study was to evaluate a new device which would maintain short-term patency of the ductus arteriosus and avoid SP shunt until the patient is ready for complete repair. Methods: Amplatzer Duct Occluder was modified to create a 4–6 mm tunnel through its waist. The length of the device ranged from 7–10 mm. The edges of the device were flared to secure retention into the vessel. The loading, delivery and deployment were similar to the Duct Occluder. Six newborn lambs weighing 2.5–4.5 kg underwent cardiac catheterization for device placement. Femoral artery and vein were accessed percutaneously. After measuring the size of the vessel by angiography, the devices were deployed in the systemic artery (n=1), pulmonary artery (n=3), and ductus arteriosus (n=2). Results: Technical success rate was 100%. There was no incidence of device embolization. One lamb was euthanized immediately placement of the device for acute assessment of the device position. Two lambs were euthanized after 6 weeks and three lambs died two weeks after placement of the device because of pneumonia. Pathological examination revealed that the device was patent in all lambs. A thin gray white reaction of film of pseudointima formation was seen in the lambs who died after two weeks. In one lamb, there was near occlusion of the lumen because of minimal hyperplasia. Conclusions: The Amplatzer ductus device appears feasible in maintaining short term vessel patency. Minor modifications and further studies are needed before human trials are conducted.

280

Dilatation of the pulmonary valve under real-time magnetic resonance imaging (MRI) guidance in an animal model

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Purpose: We sought to investigate the feasibility of balloon dilation of the pulmonary artery valve under real-time MRI guidance. Drawbacks of conventional x-ray guided interventions in pediatric cardiology include radiation exposure, a poor soft tissue contrast and the inability to acquire three-dimensional cross-sectional views. Methods: In three healthy animals (one dog, two pig) we placed a balloon-catheter (5 mm) with a diameter according to the valve size in the main pulmonary artery. Under real-time MRI guidance (MR fluoroscopy with steady state free precession true FISP MR pulse sequence; 7 frames/sec) the balloon was partially filled with air or gadolinium solution and tracked towards the valve plane and then fully dilated. Balloon valvuloplasty was monitored by high resolution real-time imaging (flip angle 60°, 128 read-out points x 100 phase encodings, 300 mm field of view) real-time imaging with a temporal resolution of 320 ms. Following the procedure gradient echo cine and spin-echo anatomical imaging was applied to rule out any vascular or cardiac damage. Results: Real-time MRI made it possible to follow the position of an angioplasty balloon within the pulmonary artery and its main branches. The inflation and deflation of the balloon positioned in the valve plane could be monitored. No vascular or myocardial damage was detected. Current limitations are caused by a time delay between image acquisition and image display that will be overcome with faster computer hardware for image reconstruction. For improved catheter tracking and high resolution intravascular imaging dedicated MRI guidewires may be helpful. Conclusions: These preliminary data suggest that balloonvalvuloplasty of the pulmonary valve can be monitored under real-time and high resolution MRI. These results are the basis for ongoing experimental trials and developments for MRI-guided catheter interventions.

281

Nickel release after implantation of Amplatzer Septal Occluders in patients with atrial septal defects

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Nitinol based alloys are widely used for medical devices because of its exceptional physical, chemical and mechanical properties. But, there is still less known about the biocompatibility of these nickel containing devices. One of these nickel based devices is the Amplatzer Septal Occluder (ASO), used for

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

transcatheter occlusion of atrial septal defects. Aim of this prospective investigation was to determine nickel serum concentration in patients before and after implantation of ASD. Methods: In a collective of 35 patients (aged 16 to 75 years) nickel serum concentrations (standard value: 0–2 ng/ml) were measured before and after implantation at certain times: 24 hours before, and 24 hours, 1, 3, 12 and 24 months after implantation of ASD. Results: In 35/35 pts nickel concentration before intervention was normal. 24 hours after implantation in 7/35 pts serum concentration of nickel increases above supranormal values, after 1 month in 18/24 pts, after 3 months in 3/24 pts, after 12 months in 0/18 pts and after 24 months in 0/14 pts. There is a significant increase of serum nickel concentration in the total number of pts after 1 months after implantation of ASO. No pts showed signs of incompatibility. Conclusion: There is a significant release of nickel after the implantation of the ASO with a peak after 1 months after implantation. The following decrease of nickel serum concentration and normalization 3 to 24 months after implantation may indicate the endothelialization of the device. After 12 months after implantation all pts had normal nickel serum levels.

282
Should interventional cardiac catheterization be performed in the early post-operative period?
Dabestani N.G., Nykanen D.G., Chang A.C., Lamb M., Tagnesda R., Hannan R.L., Bunke R.P., Zahn E.M. Mattel Children's Hospital, Miami, FL, USA

Background: Cardiac catheterization and transcatheter intervention in the early post-operative period is typically viewed as high risk, and is often avoided. We hypothesized that with a multidisciplinary team approach, cardiac catheterization including intervention could be safely and effectively performed. Methods: We retrospectively reviewed all cardiac catheterizations performed within 6 weeks from the time of surgery between August 1995 and October 2000. Procedures were performed on any patient meeting clinical indications independent of the time elapsed from surgery. An interventional cardiologist with a cardiac anesthesiologist performed all cases. In addition, a cardiac intensivist, surgeon and perfusionist were present or immediately available on site. Results: Sixty patients, median age 4 months (2 days – 11 yrs), weight 4.7 kg (2.3 – 45 kg), underwent 64 catheterizations at median post-op day 9 (0–42 days). Thirty-five cases involved 58 interventional procedures including angioplasty (26), stent implantation (15), vascular occlusion (14), septal occlusion (1), and pulmonary valvotomy (4). Seven patients were on cardiopulmonary bypass support at the time of catheterization, 5 of whom underwent interventional procedures. Success rates by procedure were: angioplasty 69%, stent implantation 80%, vascular/septal occlusion 100% and valvotomy 100%. Complications included stent migration (2), transient tachycardia (1), and loss of pedal pulse (1). Suture disruption did not occur. There were no complications related to patient transport. No patients died during the procedure. Conclusion: Cardiac catheterizations including intervention can be successfully performed in the early post operative period with a low complication rate. These procedures can improve patient outcome, however they should not be performed without the support of a multidisciplinary team in order to maximize patient safety.

283
Covered stents in congenital heart disease
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Despite many potential applications, covered stents have been used rarely in patients with congenital heart disease. This report describes our experience with covered stents in 12 patients. Balloon expandable Palmaz stents (Johnson & Johnson) were covered with expanded polytetrafluoroethylene (ePTFE) surgical membrane with the stent and covering size based on the lesion. The membrane was cut and sewn into a tube using 7-0 prolene. The tube is then attached to the outside of the stent using a single suture. When expanded, the covered stent forms an occlusive tube. The covered stent is then delivered and implanted using conventional techniques for stent implantation. Since 1992, covered stents have been inserted in 12 patients. In 3 patients they were successfully used to simultaneously dilate a left pulmonary artery and close a Potts' shunt. In single patients they have been used to close an iatrogenic aortopulmonary window (previously reported), to recreate an aortic SVC 1 year after cardiopulmonary bypass, or manage an aneurysm following balloon dilation of a right ventricle to pulmonary artery homograft and to close an extensive Fontan baffle leak. They have been placed in 4 patients with pulmonary vein stenosis and 1 patient with SVC stenosis, due to progressive idiopathic mediastinal and systemic fibrosis, in an attempt to reduce the restenosis rate. The covered stent was successfully implanted in each case. Sheaths 1–2 Fr sizes larger than that for uncovered stents were required, but complications were otherwise no different than for conventional stents. Thrombosis has not occurred using the same anticoagulation protocols as for uncovered stents. Covered stents expand the options for transcatheter therapy of congenital heart disease.

MAY 30 Time: 14:00—15:30

Session 26: The Adult with Congenital Heart Disease, Pregnancy/Delivery for the Women with Congenital Heart Disease

284
Breathing oxygen-rich air does not improve exercise capacity of patients with eisenmenger syndrome
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To assess the effect of breathing oxygen rich air before exercise on physical work capacity of patients with Eisenmenger syndrome. Fourteen Eisenmenger syndrome patients (4 M/ 10 F), mean age 38±12 years, had two consecutive exercise tests each on a random order, as follows: 30 mins of rest, breathing either oxygen rich air by face mask or room air, followed by a modified 6 minutes walk test. Total walking distance, O2 saturation, heart rate, blood pressure and ECG were monitored throughout the test. O2 saturation was significantly higher after 30 minutes of breathing oxygen-rich air compared to room air (93.1±6% vs 87.1±6%, respectively, p=0.02) but the lowest O2 saturation during exercise was not different (86±15% vs 89±13%, p=ns). Walking distance was unaffected by breathing oxygen-rich air (161±74 m for room air and 161±66 m for oxygen-rich air, p=ns). There was no difference between the tests in any other parameter. Breathing oxygen-rich air for 30 minutes before exercise does not improve exercise capacity of patients with Eisenmenger syndrome despite the increase in pre exercise oxygen saturation.

285
Buttoned device in the management of platypnea-orthodeoxia
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Dyspnea and arterial desaturation on upright position in elderly subjects is described as platypnea-orthodeoxia syndrome(POS) and in some patients, it is due to right-to-left shunt across an atrial septal defect(ASD)or patent foramen ovale (PFO). The objective of the presentation is to describe the use of buttoned device in effectively occluding the ASD/PFO to relieve hypoxemia of POS. During a four year period ending January 2000, ten patients age 71 ± 9 (range 56–82)years with POS underwent buttoned device closure of their ASDs/PFOs. In her angiographic and balloon stretched atrial defect sizes were 8 ± 3 mm and 12 ± 3 mm respectively. The defects were occluded with devices ranging in size from 25 to 40 mm delivered via 9-French, long blue, Cook sheaths; eight had an additional 25 or 35 mm covered occluders placed on the right atrial side as well. The oxygen saturation increased (p<0.001) from 76 ± 7% (range 69–85%) to 95 ± 2.6 (range 92–98%). No complications were encountered. Relief of symptoms was seen in all patients. Follow-up 1 to 36 months (median 12 months) revealed persistent improvement of symptoms with a pulse oximetry oxygen saturations >92%. Based on these data it is concluded that buttoned device occlusion of ASDs/PFOs to relieve hypoxemia of POS is feasible, safe and effective and is an excellent alternative to surgery.

286
Correlates of educational, occupational, and psychosocial outcome in adolescents and adults with congenital heart disease
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While advances in the treatment of congenital heart disease have dramatically increased survival rates, little is known about the factors that mediate good

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

versus poor psychosocial outcome in adolescents and adults with congenital heart disease. The goals of this correlational study were to 1) identify factors which underlie good versus poor academic or occupational achievement in adolescents and adults with congenital heart disease and 2) identify factors that underlie good versus poor psychosocial adjustment in adolescents and adults with congenital heart disease. Approximately 200 patients were recruited from the Toronto Congenital Cardiac Centre for Adults and the Hospital for Sick Children over a period of 3 years. In order to maximize homogeneity of the sample with regard to the physiological and psychological impact of disease, subject selection was limited to patients with either Tetralogy of Fallot or Transposition of the Great Arteries. Outcome variables included educational achievement, occupational status, social relations, self-esteem, anxiety, and depression. Predictor variables included attributional style, beliefs about personal cardiac health and lifestyle restrictions, knowledge of cardiac condition, expectations for academic/vocational success and achievement motivation. Control variables included physical functional capacity, IQ, socio-economic status and actual disease severity. Results confirmed hypotheses that a more pessimistic attributional style, poor knowledge of one's heart condition, beliefs about disease severity and required restrictions were related to poorer quality of life outcomes independent of actual disease severity and actual disease restrictions. These findings have important implications for interventions with this population in that their quality of life appears to be significantly affected by factors that are very amenable to modification.

287

Survey of the special care facilities for adults with congenital heart disease

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OBJECTIVES: This study surveys the differences and similarities of special care facilities for adults with congenital heart disease (CHD), and defines the evolving role of such centers for the ongoing care of these patients. BACKGROUND: There has been a large increase in adults with CHD who require specialized tertiary center care. Characteristics of care of these patients varies. METHODS: Questionnaire on analysis of the characteristics of five specialized care facilities for adults with CHD in the North America and Great Britain were assessed. RESULTS: All facilities were established over 20 years ago, with the number of registered patients 1,200 to 6,000. Cyanotic CHD ranged between 5 and 20 percent of total registered patients in each center, and post surgical patients ranged between 55 and 81 percent. Close collaboration among medical and pediatric cardiologists, cardiac surgeons, cardiac and non-cardiac specialists were operative in all 5 centers. Training and education for adult CHD extended to medical and pediatric cardiology fellows, residents, international visitors and referring physicians. Out-patient and in-patient care was mainly in an adult setting. The number of admissions/unit/year ranged between 100 and 500. Cardiac surgeries were performed in 50–175/unit/year. Twenty-five to 80 percent of surgeries were reoperations. Low overall surgical mortality was around 3%/unit/year. CONCLUSIONS: Characteristics necessarily differ among these specialized facilities, but the differences were more matters of detail than of principle. Patients are managed by close interdisciplinary collaboration among physicians, nurses and non-physician staffs. Developments of these facilities are considered.

288

Quantitative assessment of dural ectasia as a marker for Marfan syndrome

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Purpose: To establish normal values for lumbosacral dural sac dimensions by magnetic resonance imaging (MRI), and to use these normal values to assess sensitivity and specificity of dural ectasia as a marker for Marfan syndrome. Materials and methods: MRI was used to measure dural sac diameters from L1 to S1 in 44 adult Marfan patients and in 44 matched controls. Dural sac diameters were corrected for vertebral body size, yielding dural sac ratios. The controls served to establish the upper limit of normal values for dural sac ratio at the levels L1–S1. Sensitivity and specificity of dural ectasia as a marker for Marfan syndrome were determined. The presence of other major manifestations of Marfan syndrome was evaluated in the Marfan patients. Results: Cut-off values for normal dural sac ratio for levels L1 through S1 were: 0.64, 0.55, 0.47, 0.48, 0.46 and 0.52, respectively. Significant differences in dural sac ratios at all levels between Marfan patients and controls were shown ($p<0.0001$ at all levels). At the levels L1–S1, sensitivity of dural ectasia as a marker for Marfan syndrome ranged from 45 to 77% and specificity was > 95%. By combining levels L3 and S1, dural ectasia as a marker for Marfan syndrome yielded a sensitivity of 95% and a specificity of 98%. Aortic dilatation was present in 89% of the Marfan patients, ectopia lentis in 45%, familial Marfan syndrome in 70% and major skeletal manifestations in 18%. Conclusion: Abnormal Dural Sac Ratio's at either L3 or S1 identifies Marfan syndrome with 95% sensitivity and 98% specificity.

289

Coronary ostial aneurysms after aortic root surgery in patients with Marfan syndrome

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Background: After aortic root replacement in Marfan patients coronary ostial aneurysms have been described. However the clinical importance is not yet known. Purpose: 1) to analyze the prevalence of, and 2) to assess risk factors for development of ostial coronary aneurysms in Marfan patients after elective aortic root replacement. Methods: 40 Marfan patients (mean age: 36±12 years) underwent MR imaging 3 months to 19 years after elective aortic root surgery. Diameters of the proximal coronary arteries were measured on post gadolinium sagittal 3D MR images. Patients were divided in 2 groups: coronary arteries > 10 mm (group A) and ≤ 10mm (group B). We investigated: 1) surgical techniques (button technique or direct suture, David or Bentall), 2) age at time of surgery, 3) time after surgery, 4) family history, and 5) descending aortic elastic properties in 30 of the 40 patients. Aortic distensibility (D) at 3 levels and aortic flow wave velocity (FWV) from level 1–3 were assessed (see figure). Results: 17 of the 40 (43%) patients had coronary ostial aneurysm (> 10 mm, group A). Patients in group A were significantly younger at time of operation (mean age 27±8 vs. 35±12, $P=0.03$). No significant difference between the two groups were found for surgical techniques, time after surgery, aortic distensibility, aortic flow wave velocity, family history for Marfan syndrome and family history for dissection (see table). Conclusion: Coronary ostial aneurysm is a common finding after aortic root surgery in Marfan patients. In our study only younger age at operation was associated with coronary ostial aneurysms.

290

Coarctation of the aorta in adults. Surgical aspects and mid term results

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We've displayed the experience in surgical treatment of the coarctation of the aorta (CoAo), in adult patients, evaluate the immediate and mid term results. A series of 50 consecutive adult patients with age ranging from 18 to 59 years old (25.4) underwent surgical treatment of CoAo between jan/87 and march/00. Systemic hypertension with mean systolic pressure of 147 mmHg (125–220 mmHg) was present in 43 (90%) patients the mean gradient at coarctation area of 51.54 mmHg (18–125). Other associated lesions not corrected at the same time included: mitral regurgitation (4), aortic regurgitation (9) ventricular septal defects (3) and aortic stenosis (4). In 33 patients (66%) left ventricle hypertrofy was detected by Echo and 10 (20%) patients had left ventricle disfunction. Resection of the CoAo with direct end-to end anastomosis was performed in 20 (40%), aortoplasty of CoAo with a bovine pericardium patch in 22 (44%) and insertion of a synthetic tube graft in 8 (16%). No deaths were observed. One patients required reoperation from surgical bleeding. The main complication was hypertension observed in 48 (96%) patients controled with antihypertensive drugs. The mean time of the follow-up was 32.15 months (1–145) in a group of 45 patients (90%). The mean residual gradient was of 18.71 mmHg. Normal arterial pressure has founded in 40 patients (80%), being 47% no antihypertensive drugs. 53% were receiving with ACE and beta-blockers. In the study 93% of them had no symptoms, keeping in CLINITEA. Three patients were submitted later to other cardiac surgeries: 1 for a permanent pacemaker, two for a valvar replacement, and one had a native valvar endocarditis. The last patient died later with a sepse due to endocarditis. Conclusion: The surgical treatment of CoAo even in adult's patients are strongly recommended as an effective therapeutic method, with low morbidity and good midterm evolution.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

291
Posterior pericardial ascending to descending aortic bypass: an alternative surgical approach for complex coarctation of the aorta
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Background: Aortic coarctation (CoA) is commonly associated with CV disorders that require intervention. The best approach is uncertain. Ascending to descending aortic bypass via posterior pericardium (CoA bypass) allows simultaneous intracardiac repair and an alternative approach for pt with complex CoA or reCoA. Methods: The outcome of 13 males and 5 females, mean age 43 yr, who underwent CoA bypass (1985 - 2000) was reviewed. Mean preop NYHA class was II. 15 pt (81%) had preop hypertension. Mean preop EF was 57%. One or more previous CV operations were performed in 12 pt (67%); 10 had CoA repair (3 pt had 2 CoA repairs, and 1 had CoA repair and subsequent CABG). Two pt had prior non-CoA CV surgery. Results: All pt had CoA bypass via sternotomy. 14 pt (78%) had concomitant procedures: AVR (9 pt), CABG (3 pt), MV repair (2 pt), septal myectomy, and MVR, aortoplasty, sub-AS resection, VSD closure, and ascending aortic replacement (1 pt each). Mean cross-clamp time (n=12) was 52 ± 29 min; mean cardiopulmonary bypass time (n=18) was 118 ± 51 min. Circulatory arrest was used in 4 pt for 20 ± 9 min. All survived operation and were alive with patent CoA bypass at mean follow up of 45 ± 48 months (range 1-177). No late graft complications occurred. No stroke or paraplegia was noted. Morbidity included 1 PPM implantation and 1 reoperation for perivalvular MR after MV replacement. Preop mean systolic BP was 159 mm Hg vs 125 mm Hg postop. EF improved in 2 pt with severe preop LV dysfunction. Conclusions: CoA bypass carries low morbidity and mortality. Although management must be individualized, this is an excellent single stage approach for pt with complex CoA or re-CoA and concomitant CV disorders.

292
Transcatheter coil occlusion of adult patent ductus arteriosus
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Background: The optimal method of closure of the patent ductus arteriosus (PDA) in adults, particularly in the elderly remains undefined. The purpose of this study is to review the outcome of transcatheter coil occlusion for PDA amongst these patients. Methods: A retrospective review of all patients older than 18 years who underwent transcatheter occlusion for PDA between 10/1995 and 09/2000 at The Cleveland Clinic. Results: During (1996) out of 107 patients undergoing transcatheter PDA coil occlusion were adults (7 males, 13 females). Ages ranged from 20 to 77 years (40 21.4 8). Symptoms and complications (dyspnea, chest pain, atrial fibrillation, pulmonary hypertension) were seen at presentation in 9/10 patients older than 55 years compared to 1/10 (p<0.01) patients younger than 55 years despite similar Qp/Qs ratios (1.29 vs 1.46). PDA size ranged from 1 to 7.5 mm (3.1±0.3). Nine patients (45%) required a retrograde approach as was the PDA due to inability to cross the ductus prograde. Nineteen patients had placement of one to four (0.038 Gianturco coils (1.7±0.2). One patient had PDA closure with a 9mm Gianturco coil after an initial unsuccessful attempt using a 0.052

293
Patterns of respiratory pathophysiology in adults with the Fontan circulation
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Introduction: Despite compelling evidence for complex interaction between the respiratory and cardiovascular systems in optimizing cardiac output in patients with the Fontan circulation, there is little data documenting their respiratory abnormalities. Methods: Fontan patients (>18years) were identified from the University of Toronto Congenital Cardiac Centre (UTCCCA) database and prospectively recruited. Patients had assessments of pulmonary function, respiratory muscle strength (RMS) and stage I exercise testing. Patients with a history of smoking, bronchiectasis, asthma or wheezing were excluded. Results: Of the 32 study patients 4 had undergone lateral tunnel, 22 atriopulmonary and 6 atrioventricular Fontan procedures. Fourteen of 27 (52%) patients able to perform the pulmonary function maneuvers had evidence of a restrictive pattern; and 5 (21%) had an obstructive pattern. Mean RMS was reduced, 58%, SD 22% of predicted for maximal inspiratory pressures and 17%, SD 19% of predicted for maximal expiratory pressures. Average peak VO2 was reduced to 41, SD 12 % of predicted. Patients with reduced RMS had higher O2 pulse (12.2, SD 3.7 vs. 8.4, SD 2.8 and 12.6, SD 3.4 vs. 9.2, SD 1.6 ml/beat) and higher respiratory rates (30, SD 9 vs. 27, SD 6/minute and 34, SD 10 vs. 31, SD 9) at anaerobic threshold and peak exercise respectively. Patients with reduced FVC had higher respiratory rates at anaerobic threshold (30, SD 9 vs. 27, SD 6), but this difference did not persist at peak exercise. Conclusion: Adult Fontan patients commonly have respiratory abnormalities including restriction, obstruction and severe RMS weakness. Their exercise capacity is markedly reduced. Patients with reduced RMS and pulmonary restriction may compensate by increasing stroke volume and increasing respiratory rate. Our data supports the notion that higher ventilatory rates at submaximal exercise may in fact augment cardiac output in these patients.

294
Tetralogy of Fallot in adults with hypoplastic pulmonary arteries carries a higher risk for adverse events
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The surgical management of adults with Tetralogy of Fallot is associated with significant morbidity and mortality with special consideration in the perioperative management. Between 1987 and 2000, ninety adult patients with Tetralogy of Fallot with pulmonary stenosis were operated on of a total of 362 patients with Tetralogy of Fallot. The median age of these patients was 27.3 years (range 16 years - 49 years). Twenty two patients out of the 90 underwent a palliative procedure previously. Of which 15 had undergone a BT shunt, 3 patients a Potts shunt and 4 had a Waterston shunt. Echocardiogram: shows adequate PA anatomy in 79 patients while hypoplastic varieties were seen in 13 patients. A cardiac catheterisation was done in 32 patients which showed hypoplastic main pulmonary arteries in 15 patients. Patients with pulmonary atresia were excluded from this study. RVOT reconstruction was done with transannular monocusp patch in 37 patients of the 44 patients (84.2%) who had transannular patch. 28 patients (31.1%) required RV - PA homograft conduit, while one patient required aortic valve replacement as well. A hospital mortality of 21 patients (23.33%) was noted of which 15 have right ventricular failure with low cardiac output, 3 died of septicemia and 3 due to intractable arrhythmias. At follow up of a mean of 6.2 years (range 6 months to 12 years) 94.8% of the survivors were asymptomatic. RV dysfunction was seen in 3.6 % of patients, residual VSDs were seen in 2 patients. Patients with hypoplastic pulmonary arteries requiring a homograft conduit reconstruction of the RVOT had a higher risk for mortality as compared to those who required a transannular patch.

Session 27: Surgical Management and Results: Abnormal Venous Return, Left Ventricular Outflow Obstruction/Aortic Stenosis, Pulmonary Atresia

295
Fate of the aortic root after arterial switch operation
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Study Background: Concerns have been voiced about dilation and insufficiency of the neo-aortic valve after the arterial switch operation (ASO). Aim of the study: Determination of growth of the aortic root after ASO and prevalence of insufficiency or stenosis. Patients and methods: Since 1977, 144 consecutive patients underwent ASO for transposition (TGA). Median follow-up was 8.65 years (0.1-28.5 years). Simple TGA was present in 97 pts and 47 had TGA with VSD. Echocardiography included 608 measurements of the aortic root, which were compared to normal values. Results: The mean aortic valve and aorta annulus follow z-score was 1.9. Under 4 months, mean valve z score was 0.61 ± 2.30, between 5 and 12 months 2.56 ± 2.10 (p<0.0001). Gradual growth occurs thereafter. The aorta at the anastomosis is initially smaller than normal (z-score -0.64). After 4 months the z-score is 0.83, followed by continued growth of 0.1 z-score per year. At the last visit, the aortic valve z-score was above 2 in 31 patients, between -2 and 2 in 72

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

and insufficiency ≥ 2 in 6 patients, none of whom had a flow velocity above 2 m/s. Aortic insufficiency was grade 2/4 in 2 patients, grade 3/4 in 1 and grade 4/4 in 1. Conclusion: After ASO the neo-aortic valve and sinus are larger than normal. In the first year of life, rapid dilatation of the new aorta is observed, followed by growth towards normalization of the valve and sinus. Aortic dilatation is rarely associated with significant insufficiency.

296
Impact of beraprost sodium (oral prostacyclin analogue) on pulmonary vascular resistance of high-risk Fontan candidates
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Background: For high-risk Fontan candidates with the elevated pulmonary vascular resistance (PVR) and mean pulmonary arterial pressure (PAp), the administration of beraprost sodium (oral prostacyclin analogue, prostaglandin I2) patent dilatus of pulmonary artery, may extend the indication for the Fontan procedures. The purpose of this study is to evaluate the mid-term effects of prostaglandin on PVR and PAp as well as surgical outcome in high-risk Fontan candidates. Methods: Twenty-two consecutive Fontan candidates who underwent catheterization for Fontan indication were reviewed. PVR and other hemodynamic data were calculated by Fick's principle or by direct measurement of pulmonary flow with Doppler wire. Of 22, 10 with both PVR > 2.5 Woods units*m2 and PAp > 16mmHg were considered as high-risk Fontan candidates (H) and 12 as standard (N). These 10 H were reclassified after starting oral prostaglandin (4.99 ± 2.1ng/Kg/day) with a interval of 26.3 ± 13 months. Hemodynamic data were compared by Mann-Whitney U test or paired t-test and p<0.05 was considered as statistically significant. Results: PAp and PVR in H were significantly higher than those in N: 19.6 ± 4.2 vs 12.5 ± 3.1mmHg (p<0.001), 3.3 ± 1.2 vs 1.9 ± 0.6 U*m2 (p<0.01), respectively. In H, PAp was increased by prostaglandin from 19.8 ± 4.2 to 12.7 ± 3 mmHg (p<0.001) and so as PVR from 3.3 ± 1.2 to 2.2 ± 0.9 U*m2(p<0.01). No adverse effects of prostaglandin were noted. Seven patients completed successful Fontan operation and 3 was out of indication medically(2) and socially(1) in H, while 11 completed Fontan and 1 had sudden death in N. In conclusion, beraprost sodium is a useful internal medical palliation in high-risk Fontan candidates.

297
Current results with early primary repair of all forms of tetralogy of Fallot
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The approach to Tetralogy of Fallot (TOF) continues to evolve. We describe a single institutional experience with a philosophy of early complete repair for all forms of the lesion. Data was collected prospectively on 110 consecutive patients presenting to our institution with TOF. Preoperative and intraoperative data and outcome variables were analyzed. Ninety-three patients presented to our institution in the first year of life. Eighty-five (91.4%) underwent primary repair (75 in the first six months of life), including three children with associated atrioventricular canal (TOF/AVC) and five with absent pulmonary valve syndrome (TOF/APV). Eight infants (8.6%) underwent palliative procedures, usually because of concomitant serious illness. Seven of the eight infants palliated at our institution have gone on to complete repair. Seventeen patients presented late to our institution (four palliated elsewhere) and were repaired after one year of age. Overall, 99/110 children underwent primary repair (89%). Hospital survival was 99.1% (109/110). Hospital LOS in uncomplicated TOF in the first year of life was 12.6 +/- 4.8 days and did not show a significant correlation to age or weight at repair (r=0.07 for age, r=0.05 for weight). Mid-term follow-up (median 32 months, range 60 months) is complete on all patients. Mid-term survival is 98.2% (108/110) with one late death from non-cardiac causes. Two children (1.8%) have required re-operations for conduit replacements and fifteen (13.6%) have needed interventional catheterization procedures (overall freedom from re-intervention 84.6%). Primary early complete repair of TOF can be accomplished in nearly all infants. Palliation is usually reserved for children with severe concurrent systemic illness. Previously reported long-term benefits of early complete repair can be attained with very low mortality and morbidity, acceptable hospital stays, and a low requirement for re-intervention during mid-term follow-up.

298
Banding of the pulmonary artery to train the morphologic left ventricle in the setting of systemic right ventricle
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Objectives: To assess the efficacy and timing of pulmonary artery banding (PAB) in patients with transposition of the great arteries (TGA) following Mustard/Senning or preparation for arterial switch and in patients with congenitally corrected transposition of the great arteries (CTGA) in preparation for a double switch procedure. Methods: Thirty three patients underwent PAB for training the left ventricle (LV) between 1990 and 2000. Eighteen had CTGA and 15 had TGA. The median age at the time of PAB for CTGA patients was 18 months and for TGA patients was 16.5 years. The LV dimensions, posterior wall thickness, LV/RV pressure ratio, ventricular and tricuspid valve functions were assessed before and after banding. Results: There were no operative deaths following PAB. Four patients required tightening of the band, one of whom was unsuccessful. LV systolic and diastolic dimensions increased significantly (1.4 to 3.6 and 3.1 to 4.5 respectively, p=0.02). The posterior wall thickness during systole and diastole increased (11 to 12, NS, and 6.6 to 9.0, p < 0.001 respectively). The LV/RV pressure ratio increased from 0.5 to 0.8. Tricuspid regurgitation decreased following PAB (2.4 to 1.6, p=0.001). Neoaortic regurgitation developed in 9 (33%) patients requiring valve replacement in two. We found no correlation between the age at banding or the interval of banding and complete correction and neoaortic insufficiency. When age was analysed as a continuous variable it was not found to affect the increase in ventricular mass. Conclusions: PAB is effective in training the LV in patients with failing systemic RV. Where this has not been possible, functional improvement has been achieved indicating that PAB may serve as a therapeutic end-point.

299
Simplified single patch technique for repair of atrioventricular canal. Medium term follow up of 72 consecutive patients
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Objective: To examine the preoperative experience of 72 consecutive patients with complete atrioventricular septal (A-V canal) defect, repaired by a simple one patch technique. Method: All patients, regardless of the size of the interventricular communication, were treated by a single pericardial patch to close the atrial septal defect after direct suturing of the common atrioventricular (AV) valve to the crest of the ventricular septum. There was no division of A-V valve leaflets, all clefts in the left A-V valve were closed and all patients underwent annuloplasty by shortening the length of interventricular septal crest to increase leaflet apposition. Results: Early mortality 2.8%. Cardiac ischaemic time reduced by 20% (mean 70+/-15). VSD size (large 72%, moderate 21%, small 7%). Median follow up 3.2yrs (Interval 0.5 yrs). No late mortality. All patients in sinus rhythm, early and late. No early or late left ventricular outlet obstruction. No significant residual ventricular septal defect (80% no defect, 20% trivial defect). Echocardiographic assessment of Left A-V valve function - normal (65%), mild regurgitation (29%), moderate regurgitation (5%). No deterioration in valve function in the period of follow up. Conclusion: Direct suture of the common AV valve to the crest of the ventricular septum does not interfere with post operative valve function or cause left ventricular outflow obstruction and greatly simplifies and expedites the repair of the defect. The technique is applicable to all types of ventricular septal defect. Post operative A-V valve function is excellent and has not deteriorated in the medium term.

300
Outcome after repair of complete and partial atrioventricular septal defects
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Repair of atrioventricular septal defects (AVSD) remains a challenging procedure due to the complex three-dimensional malformation of the septa and the AV-valves. However, repair of partial AVSD is generally considered easier, but comparative contemporary data on mitral valve performance and

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

reoperations are lacking. We conducted a retrospective study of 365 patients with repair of complete, complex, and partial AVSD between 1990 and 1998. Malformations in 195 patients were complete AVSD (Rastelli A 73%, B 5%, C 22%); 30 patients had complex AVSD (AVSD and TOF n=19, others n=11) and 140 had partial AVSD (transitional AVSD 37%, ASD I 63%). Median age was 4.6, 9.1 and 17.4 months, respectively (p< 0001). Trisomy 21 was present in 54% (complete AVSD 75%, complex 53%, partial 30%, p< 001). Correction was performed with single patch technique in 84% and two-patch technique in 16% in complete and complex AVSD. The mitral cleft was completely closed in 56% (complete AVSD 52%, complex 43%, partial 62%, p=.09). Survival was lower for patients with complex AVSD compared to complete or partial AVSD (operative survival 89%, 97% and 97%, 1-year 77%, 96% and 97%, 5 year 77%, 96% and 95%, p= 002). Age, weight, anatomical subgroups and surgical technique did not influence operative survival. Freedom from reoperation at 1 and 5 years was 87% and 67%. Risk factors for late development of moderate to severe mitral valve regurgitation or mitral valve reoperation in multivariate analysis were absence of trisomy 21 (p= 003) and incomplete mitral cleft closure (p=.01). Survival of AVSD repair is only influenced by complex additional cardiac malformations. The outcome of repair of complete and partial AVSD is comparable. Absence of trisomy 21 and incomplete cleft closure are risk factors for development of postoperative mitral regurgitation.

301

Abrupt aortic root dilation after the Ross procedure: is this a progressive phenomenon?

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Variable patterns of neo-aortic root growth have been reported in patients who have undergone the Ross procedure. This study evaluated serial changes in patient neo-aortic root geometry and determined variance from normal, if any. Methods: Postoperative echocardiographic measurements of the aortic annulus and sinus diameters were obtained from 46 patients (mean age 16yr at time of surgery). Paired analyses were performed between indexed aortic root measurements measured at time of discharge (n=45) and at follow up intervals of 6–12mo (n=31), >12–36mo (n=18) and >36mo(n=12). Aortic root Z-scores were derived from 217 normal healthy controls. Results: Compared to the preoperative pulmonary root, there was immediate stretching of the neo-aortic root annulus (annulus 1.7 vs 1.9 cm/m^2 and sinuses from 2.1 vs 2.8 cm/m^2, p< 0.001*) at mean f/u of 6.2 mo. Further dilation in comparison to time of dc was evident at intervals 6–12mo f/u (annulus 1.9 vs 2.0cm/m^2* and sinuses 2.8 vs 3.1 cm/m^2, p<0.001*). However, additional dilation after the first year was not observed up to the >36mo f/u interval. The aortic root measured at the annulus and sinuses of Valsalva were 1.8 and 2.4 SD, respectively from the normal mean at late f/u (>36mo). This was accompanied by a decrease in left ventricular size from preoperative to >36mo f/u without any change in blood pressure or degree of aortic insufficiency. One patient developed severe aortic root dilation at >36mo f/u and underwent successful aortic valve and root replacement. Conclusions: Aortic root dilation appears to occur up to the first year after the Ross procedure but does not seem to progress beyond this time. Therefore close blood pressure monitoring and prompt management of hypertension are indicated in these patients.

302

Late results of bioprosthetic tricuspid valve replacement in Ebstein's anomaly

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Historically, porcine bioprosthetic valves have poor durability in pediatric patients; nearly half will require replacement within five years. However, our early experience with patients having Ebstein's anomaly suggests that tricuspid bioprosthesis in this anomaly might have better durability. One hundred fifty-eight patients who received a primary tricuspid bioprosthesis because of tricuspid valve anatomy unsuitable for repair between April 1972 and January 1997 were reviewed. Results were analyzed and Kaplan-Meier curves were constructed to estimate patient survival and probability of remaining free of reoperation. Follow-up of 149 patients (94.3%) who survived 30 days ranged up to 17.8 years (mean 4.5 year). Ten-year survival was 92.5%±2.5 SE, 129 late survivors (92.1%) were in Class I or II, and 97.6% were free of anticoagulation. Freedom from bioprosthesis replacement was 97.5%±1.9 SE at 5 years and 88.6%±7.6 SE at 10 and 15 years. Bioprosthesis durability in the tricuspid position in patients with Ebstein's anomaly compares very favorably with bioprosthesis durability in other cardiac valve positions, especially for pediatric patients, and also compares favorably with tricuspid bioprosthesis durability in patients with other diagnoses.

303

Intermediary Results of Truncus Arteriosus repair without extracardiac conduits

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The aim of this study is to report our results on Truncus Arteriosus (TA) repair without extracardiac conduit (Barbero-Marcial technique). Since 1987 this technique has been used in selected patients with TA type I or II aged less than 5 months with pulmonary vascular resistance index less than 5 Wood units and without coronary anomalies. After aortic cross-clamping, a longitudinal incision is made in the LPA and extended towards the common trunk. A pericardial patch is trimmed and sutured dividing the common trunk in two components: aorta and pulmonary artery. The right ventricle is sutured immediately bellow the left Valsalva sinus and the VSD is closed. The lower edge of LPA incision is pulled down and anastomosed directly to the left superior margin of the ventriculotomy with several interrupted stitches, to form an almost horizontal suture line, reconstructing the posterior wall of the RVOT. Finally, the anterior wall is reconstructed using a bovine pericardial patch with a monocuspid valve. Forty seven infants were operated with this technique with a median age 4 months. Median follow-up was 6.6 years. Overall mortality was 27.6% and the most common cause of early death was low cardiac output. There was two late deaths 5 and 19 months after repair, due to pulmonary infections and sepsis. Actuarial survival was 67.5% in 11.4 years. Pulmonary regurgitation was found in 19 pts (40%), being severe in three. Pulmonary stenosis was present in 8 patients (17%), two of them successfully repaired by balloon dilatation. Five patients needed reoperation and actuarial freedom from reoperation was 89%. In conclusion, intermediary results are satisfactory despite the initial high early mortality.

304

Clinical and hemodynamic comparison between Ross and Konno procedure for congenital aortic valve disease

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There are no milder prosthetic valves in the pediatric age group. The Konno procedure was our choice of procedure for aortic valve disease in children. Recently we started applying Ross procedure in patients with small aortic annuli. The purpose of this study is to evaluate and compare acute clinical and hemodynamic status after both Ross procedure and Konno procedure. Method: Ten patients (mean age 7.5 years, range 1–12) with congenital aortic valve disease (CAVD) underwent Ross procedure between 1995 and 1998. Ten CAVD patients without subaortic stenosis underwent Konno procedure (mean age 10.1 years, range 2–34). There were no significant preoperative differences between two groups in terms of age, body weight, left and right ventricular volume, degree of aortic regurgitation or pressure gradient at the aortic valve. Postoperative catheterization was performed one month after operation. Right and left ventricular volumes (RVEDV, LVEDV) and ejection fraction, LVEDV reduction (VR), end diastolic pressure (EDP) and right atrial pressure (RAP) and pulmonary wedge pressure were compared. Cardio-thoracic ratio (CTR) on chest X-ray at discharge, ICU and hospital stays were also compared. Results: There were no hospital deaths in either group. There were no significant differences between groups in terms of CTR, ICU and hospital stay. VR one month after operation was greater in Ross group. RVEDV and RAP were significantly lower in Ross group. There were no any significant differences between groups with respect to the other parameters. Conclusion: In terms of postoperative left ventricular volume, the Ross procedure resulted in a more substantial recovery toward the normal range than the Konno procedure. Right ventricular and atrial parameters were better in Ross group. From the acute hemodynamic point of view, Ross procedure should be the procedure.

305

Ebstein's Anomaly: 1 1/2 ventricle repair

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Background: Ebstein's malformation is a complex anomaly involving the tricuspid valve and the right ventricle. Various operations have been undertaken with varying degrees of results. Innovative surgical approaches must be evalu-

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

sted to improve the outcome of patients with this anomaly. Methods: Between 8/1994–10/2000 49 patients with Ebstein's Anomaly and moderate to severe tricuspid regurgitation (TR) were retrospectively reviewed. All patients were symptomatic in NYHA functional class II (14) or III (2). The surgical procedure, besides aiming to optimize the tricuspid valve function, was also focused on the unloading of the right ventricle (1 1/2 ventricle repair). Our emphasis was on aggressive valve repair to avoid replacement. The patients were treated with bidirectional Glenn, tricuspid valve procedure, and closure of ASD. Median age was 4 years. Two patients who underwent prosthetic tricuspid valve elsewhere, required replacement. Results: There were no early or late deaths. Early operation was necessary in one patient (5%) for right ventricular failure requiring re-opening of the atrial septal defect. One patient with a ring annuloplasty required ring replacement ten months postoperatively for recurrent regurgitation. At the mean follow up of 32 months, all patients are in functional NYHA class I and none have more than mild or mild TR. Conclusions: The surgical approach focused not only on the preservation of the tricuspid valve but also stresses the importance and related benefits of the unloading of the right ventricle. At the midterm follow-up, all patients have excellent clinical outcome. Further follow-up will be necessary to evaluate the long-term outcome of this approach.

306

Staged surgical treatment for patients with late systemic right ventricular failure following Mustard/Senning procedure for d-transposition of the great arteries

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Background: Right ventricular failure occurs as the most severe late complication in 10–15% of patients who underwent Senning or Mustard procedures for transposition of the great arteries (TGA). Surgical options for these patients include staged conversion to the arterial switch operation (ASO) or heart transplantation. Methods: From 1993 – 2000 twelve patients (7 female, 5 male) underwent atrial switch procedures between 1970 and 1993 and were diagnosed with severe right ventricular failure. In these patients, the results of the institutional treatment program consisting of conversion to arterial switch following staged "retraining" of the left ventricle by pulmonary artery banding (PAB) were analyzed. Results: PAB in preparation for an arterial switch for severe right ventricular failure was performed at a mean interval of 9.7 years (SD: 6.2 years) following the initial atrial switch procedure. Readjustments of PAB was necessary in 4 cases in 7 patients reflecting the time course and changes in afterload needed to retrain the left ventricle. Adequacy of PAB was evaluated and monitored by echocardiography. The ASO was completed in 6 patients at 13.5 ± 6.5 months following the initial PAB procedure. In one patient with left ventricular outflow tract obstruction a Rastelli procedure was performed without previous PAB. Two patients died at 172 and 259 days following PAB in unmanageable left ventricular failure. 3 patients until mean ASO. 3 patients died 1, 28 and 30 days following ASO. At 4.5 ± 1.6 years following conversion to arterial switch 4 patients are alive and in NYHA class II-III. Conclusions: Staged conversion of the atrial switch on the arterial switch procedure offers an alternative for patients who are not candidates for heart transplantation. However, the mortality of this staged procedure is substantially high. When available, heart transplantation may be the preferred approach.

Session 28: Fetal Cardiology

307

Prenatal diagnosis of lung hypoplasia in congenital diaphragmatic hernia (CDH) by measurement of pulmonary artery diameter

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Prenatal diagnosis of lung hypoplasia has been difficult for long time. We have the hypothesis that lung volume is correlated with pulmonary artery diameter. To verify our hypothesis, we measured diameters of right and left pulmonary artery in autopsied hearts that died within 24 hours after birth. The diameters in 132 cases without lung and heart anomaly (control group) were strongly correlated with lung weights. Diameters in 15 cases of CDH were significantly smaller than control group whose gestational age and body weight were matched. Secondly, we measured the diameter of pulmonary artery and descending aorta in 10 cases of fetal CDH and compared them with their clinical course after birth. One case died in utero and nine cases have survived. We made two parameters from diameters, the first, RPA+LPA/descending aorta, and the second, LPA/RPA. We compared two parameters with AaDO2 (alveolar arterial oxygen difference) soon after birth and need for NOI (Nitrous oxide inhalation) which may be related with severe lung hypoplasia. The first and second parameters were strongly correlated with AaDO2(r=0.81 and 0.95). We can accurately predict the need for NOI from them, the first parameter less than 1.2 and the second parameter less than 0.85. Sensitivity and specificity became 100%. We conclude that prenatal diagnosis of lung hypoplasia in CDH by measurement of pulmonary artery diameter seems to be possible but we need more cases before definite conclusion.

308

Right coronary flow dynamics during acute asphyxia in the fetal lamb

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Background: Chronic intrauterine asphyxia has been shown to cause increased coronary Doppler flow velocity in the human fetus. The effect of acute severe asphyxia on the fetal coronary circulation have not been clearly investigated. Methods: 11 exteriorized near-term fetal lambs (134 – 138 days) were exposed to acute total umbilical cord occlusion, while 5 fetuses served as controls. Fetal heart rate, blood pressure, blood gases and ECG were recorded. Right coronary artery (RCA) flow velocity was registered by transthoracic Doppler, and myocardial perfusion was measured by radioactive microspheres before and twice during umbilical cord occlusion. Results: Umbilical cord occlusion caused bradycardia and fall in blood pressure. The survival time was 6–20 (median 13) minutes before cardiac arrest occurred. Despite a fall in blood pressure the RCA diastolic peak flow velocity (PFVd) and velocity time integral/minute (VTI/min), showed an immediate increase after umbilical cord occlusion. The mean (SD) maximal value for PFVd was 202%(117%) and VTI/min 280% (237%) of pre-occlusion values. The maximal values were observed within 0.5 to 9 (median 3) minutes of umbilical cord occlusion. PFVd and VTI/min remained above pre-occlusion values 44% of the survival time. There was a linear correlation between PFVd (r=+0.62, p< 0.0001), VTI/min (r=+0.59, p< 0.001) and myocardial perfusion measured by radioactive microspheres. Conclusions: Acute severe asphyxia induced by umbilical cord occlusion in the fetal lamb causes an immediate marked increase in RCA flow velocity and right myocardial perfusion despite a fall in blood pressure. Coronary flow velocity and myocardial perfusion are maintained until a few minutes before hemodynamic collapse and cardiac arrest.

309

Fetal cardiac tumor and tuberous sclerosis : a multicenter study

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Purpose: Fetal cardiac tumors are rare with late-onset appearance. And it is frequently associated with tuberous sclerosis (TS). To investigate the manifestations of fetal cardiac tumors with TS, a multicenter study was performed. Methods: The 36,000 records of fetal echocardiography of 6 cardiac centers during the 12 years were reviewed retrospectively. Medical records and fetal echocardiogram were reviewed for the number, location and type of the tumor, family history of TS, clinical course and gestational age at diagnosis. Results: 21 cases of fetal cardiac tumors were found. Tumors were multiple in 14 cases, single in 7 cases. Among 14 cases with multiple tumor, 12 patients had TS. 1 patient did not have TS and 1 patient with complete heart block was unknown due to termination of pregnancy. Among the cases with single tumor, there was no TS. 5 patients with TS had positive family history. Partial tumor regression was observed in 6 patients. Gestational age range at diagnosis was 24 to 37 weeks. Conclusions: The result suggests that multiple cardiac tumors diagnosed in utero is associated with TS.

310

Diagnosis and outcome of fetal ductal constriction

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Fetal ductal constriction (FDC) is usually associated to maternal use of non steroid anti-inflammatory drugs (NSAID). FDC in the absence of NSAID usage is uncommon and its incidence is unknown. The main consequence of FDC is RV pressure overload and increased muscular thickness of pulmonary arterioles and pulmonary hypertension. The purpose of this study was to describe the etiology, echocardiographic diagnosis and neonatal evolution of a series of fetuses with FDC. Thirteen pregnancies with echocardiographic

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

diagnosis of FDC were reviewed. Diagnostic criteria were ductal systolic flow velocity (DSV) > 140 cm/s, diastolic flow velocity (DDV) > 50cm/s and pulsatility index (PI) < 1.9 or absent ductal flow. Patients with congenital heart disease or high DSV with normal or high PI were not included. Mean gestational age was 33 w (27–37w), mean SDV was 2.17m/s (1.75–3.0 m/s), mean DDV was 0.96 m/s (0.96–1.5 m/s) and mean PI was 1.28 (0.52–1.83). Two fetuses showed complete ductal occlusion. Two thirds of the patients had increased RV diameter. Tricuspid and pulmonary regurgitation were present in all patients and functional pulmonary atresia was present in 3. Seven mothers have used NSAID (indomicacin/5, AAS/2, Diclofenac/2). Flow returned to normal 4–48 d after withdrawal of the drug. Six patients had no risk factors for FDC. One 1 ductal flow normalized during fetal life. 3 parents had normal neonatal outcome(1 precluded ductus)and 2 had pulmonary hypertension and needed ventilatory support and prolonged hospitalization. Diagnosis of FDC is essential to good management of these pregnancies and can modify the prognosis in neonatal life.

311

Heterotaxy syndrome in the fetus: is prenatal diagnosis accurate?

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Heterotaxy syndrome (HS) is often associated with complex congenital heart defects. Prenatal identification of critical lesions such as ductus dependent pulmonary or systemic circulation, or total anomalous pulmonary venous return (TAPVR) is important for postnatal management and may reduce the high morbidity/mortality associated with HS. We sought to assess the accuracy of fetal echocardiography (FE) in the diagnosis of HS. Retrospective review of our FE database from 6/95–11/00 identified 18 pts with HS. Fourteen survived to term and had postnatal echocardiograms available for comparison. Sensitivity (Sn) and specificity (Sp) of FE for detection of atrioventricular and/or ventriculoarterial abnormalities, systemic and pulmonary venous anomalies, systemic and pulmonary outflow obstruction, and ductal dependency were determined. Degree of atrioventricular valve regurgitation (AVVR) was also assessed. Of the 14 pts, 13 had atrioventricular and/or ventriculoarterial abnormalities of which 12 were accurately identified prenatally (Sn 92%). Interruption of the IVC was present in 8, 7 of whom were diagnosed prenatally (Sn 1.a left SVC was missed on FE (Sn 80%, Sp 100%). TAPVR was seen in 4 (infradiaphragmatic(1), supradiaphragmatic(2), mixed(1)), only 2 of whom were accurately diagnosed prenatally (Sn 40%, Sp 89%). Prenatal and postnatal diagnoses concurred with regard to pulmonary and systemic outflow (pulmonic/aortic), and ductal dependency in all 14 pts. No significant AVVR was seen prenatally. Eight had only trivial AVVR postnatally. Conclusion: FE can accurately diagnose HS. FE is sensitive for predicting ductal dependency while TAPVR is more difficult to diagnose. Early changes in severity of AVVR do not develop in HS, despite the hemodynamic alterations that take place at birth. Efforts should be focused on developing techniques for better identification of pulmonary venous anomalies in the fetus with HS.

312

Right and left ventricular long axis function in the fetus using angular M-mode

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Right and left ventricular long axis (LA) function is well established in adults as a means of assessing function of longitudinally orientated myocardial fibers. It has not been reported in the fetus. The aim of this study was to assess the feasibility of determining tricuspid and mitral valve ring movement throughout the cardiac cycle at different gestational ages. The four-chamber view of the fetal heart was recorded in a cine-loop to which M-mode guided M-mode echocardiography was applied using angular M mode (Aloka SSD 550). This allows for angle correction of the position of the cursor line to take into account variable fetal position and to allow correct placement of the M-mode line from cardiac apex to tricuspid or mitral valve rings. Data from 18 fetuses between 17 and 29 weeks of gestation was available for analysis. Right and left ventricular LA recordings were obtained in 18 and 14 cases respectively. Total right ventricular LA excursion was: mean = 5.2 ± 0.5mm (SD), range = 3.9 to 7.2mm and total left ventricular free wall excursion was: mean = 4.5 ± 1.1mm (SD), range = 3.0 to 6.8mm. For paired data, the difference between means of right and left LA excursion was -0.8mm (95% C.I. = -1.2 to -0.5mm). Studies of ventricular LA function in the fetus are feasible if M-mode angle correction is used. Total right and left ventricular LA excursions do not show a clinically significant difference. This technique offers new avenues to study ventricular function in the fetus.

313

Simultaneous pulsed wave Doppler of pulmonary artery and vein in the fetus: Reference interval and variability of the atrioventricular interval during sinus rhythm

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Simultaneous PW Doppler recordings of fetal pulmonary artery (PA) and vein (PV) resemble the sequential atrial (A) and ventricular (V) depolarization on the ECG. The AV interval corresponds to AV delay and may be useful for recognizing fetuses at risk of heart block or pre-excitation and in assessing mechanism of fetal arrhythmias. The aim of the study was to obtain reference ranges for the AV interval in the fetus and its variability across gestation. 152 fetuses were studied by PW Doppler at 15–40 weeks. Sampling was guided by colour flow Doppler and recordings made in the inner 2/3 of lung parenchyma during 'fetal apnoea'. Measurements were made off-line on digitally stored images. The AV interval was measured from the onset of the 'a' wave on PV signal (atrial systole) to the onset of the PA signal (ventricular systole) and averaged over 5 consecutive cycles. Heart rate was calculated. Measurements were repeated on a separate occasion. AV interval values were obtained in 148 (97%) cases. Mean(SD)= 147(16.5)ms, range = 110–192ms, 95%CI= 122–177ms. There was no relationship with gestational age (linear regression, p=0.4) and no correlation with heart rate (r=0.26). Repeatability and intra-class correlation coefficients were 15ms and 0.86 and 21ms and 0.5 for measurements taken on the same and different occasions respectively. Pulmonary vein-derived AV interval in the fetus is easily obtained. Its mid-cavity narrow confidence interval and moderate to good reproducibility across gestation allows it to be used in analysing the mechanisms of fetal arrhythmias.

314

Assessment of the thymus at echocardiography in fetuses at risk for 22q11.2 deletion

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An absent or hypoplastic thymus is common in patients with 22q11.2 deletion (22del). We sought to determine whether echocardiography permits assessment of the thymus in pregnancies at risk for fetal 22del. Over a 18 month period, we searched for the presence or absence of thymus prospectively in 6 and retrospectively in 7 fetuses with either a conotruncal lesion (12) or with maternal 22del (1). Karyotype assessment for 22del was performed pre- or post-natally in all. By 2D imaging, the thymus was identified in the anterior superior mediastinum as a white hyperechoic area (see figure). Of all 13 cases, 5 had 22del, including 1 with tetralogy of Fallot (TOF) and right aortic arch, 1 TOF with pulmonary atresia, 1 truncus arteriosus (TA), 2 interrupted aortic arch type B and 1 fetus with no cardiac pathology and maternal deletion. In none the thymus could be identified. However, in 1 case of TA and 22del retrospectively analyzed, the thymus was not seen by echo but was present at autopsy. Eight cases were without 22del, including 4 with TOF and left aortic arch, 2 TOF and right aortic arch, 1 TA and 1 posterior malalignment VSD. In all 8 without 22del, the thymus was identified, however in one it was only identified in the follow up studies. Our preliminary study suggests that echocardiography permits assessment of the thymus in most fetuses at risk for 22del. This additional information is useful in counselling given the delay in obtaining chromosomal results for 22del. Further prospective assessment is necessary to confirm our findings in a larger number of fetuses with and without 22del.

315

Prenatal diagnosis of cardiac abnormalities detected by transvaginal ecocardiography in early gestation in fetuses with increased nuchal translucency

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INTRODUCTION: This study was done to evaluate the accuracy of fetal transvaginal ecocardiography (TVE) in the detection of defects in fetuses with increased nuchal translucency. METHODS: In our department we have performed transvaginal fetal echocardiography between 12th to 16th weeks in fetuses who presented increased nuchal translucency (NT). Since pathological association between increased NT and cardiac defects has been previously described, we have performed a detailed fetal heart examination in all cases with NT >2.5mm (n=153). The examination was considered

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

reliable after a precise image of the four chamber view, outflow tracts, double-crossing of the aorta, pulmonary trunk, ductus and aortic arch. Serial TVE was performed weekly until all views were obtained. Measurements of the ascending aorta and pulmonary artery diameter has been done because literature has describe in pathological examinations a dilation of the ascending aorta with narrowing of the isthmus in trissomy 21 fetuses. RESULTS: We obtained the best view of the cardiac structures at 14th weeks. From these 153 fetuses, 31 (20.2%) had cardiac abnormalities, 3 of them not detected on early pregnancy. Transvaginal fetal echocardiography could properly diagnose 28 cases of cardiac abnormalities as follow: ventricular septal defect (n=10), hipoplasic left heart syndrome (n=7), double outlet right ventricle (n=2), A-V septal defect (n=2, 1 with complete A-V block), tricuspid displasia (n=2), Coarctation and aortic hipoplasia (n=3), tetralogy of Fallot (n=1), transposition of great arteries (n=1), tricuspid atresia and complete A-V block (n=1). In this group, abnormal karyotype was found in 60% of the cases. Unusual inversion of the size of the great arteries (aorta wider than pulmonary artery) was found in 30 fetuses (19,6%) and only 5 have shown abnormal karyotype (16,6%). CONCLUSIONS: Our findings have showed that despite the limitation of early fetal cardiac images, evaluation in this period is possible. As previously described in pathological examination, increased NT seems to be associated with wider ascending aorta consequent to the narrowing of the isthmus and cardiac defects but further studies must be performed in order to evaluate the real incidence of those associated findings.

316

Thirteen years' experience in fetal cardiology, the importance of the collaborative work between cardiologists and obstetricians to develop the educational screening program for congenital cardiac malformations in Brazil

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AIM: To report our experience with fetal echocardiography in the last thirteen years in a tertiary center in fetal cardiology and to correlate the degree of detection of suspicious of congenital heart disease by standard obstetric ultrasound after 1995 when an educational screening program was initiated. MATERIAL & METHODS: We reviewed our experience with 9600 fetal echocardiograms, from 1987 to 2000. The main reasons for referral was maternal factors (29%), fetal arrhythmias (20%), fetal anomaly (22%) and family history of congenital heart disease (16%), abnormal heart on obstetric ultrasound (9%). An educational screening program was created to improve the prenatal detection of heart malformations by the obstetric ultrasound and consisted in 1) training courses, 2) fetal heart watching tape for the obstetricians and ultrasonographers and 3) Campaign in ultrasound meetings with an educative poster proposing an easy method to understand the 3 basic views of the heart: four chamber, short axis view (diary view) and long axis view (ballerina's shoe view). RESULTS: Abnormalities were found in 972 fetuses (10.78%), divided in two groups. 1) Anatomic/functional abnormalities – 460 cases and 2) Rhythm Abnormalities – 512 cases. In group 1 we found 460 cases of congenital heart disease and 20 cases of functional disturbances (12 cases of restrictive ductus arteriosus and 4 cases of restrictive foramen ovale). The most common arrhythmias in group 2 were premature atrial contraction (n=342), supraventricular tachycardia (n=51), complete heart block (n=68), atrial flutter (n=12), premature ventricular contraction (n=10), and ventricular tachycardia (n=3). Concerning to the anatomic/functional abnormalities detected on this referral center, the number of referrals statistically improved as well as the abnormal heart on obstetric ultrasound was responsible for 90% of referrals in the last 2 years. This improvement was due to educational and training programs developed in this University. CONCLUSION: We conclude that fetal echocardiography allows a precise diagnosis with great impact on obstetric management. We emphasize the important role of the University on education and dissemination of experience to those primary diagnostician, obstetricians and radiologists, in order to improve fetal diagnosis and outcome.

317

Foramen ovale restriction: a cause of right to left fetal asymmetry by fetal echocardiography

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Unusual fetal right-left asymmetry (FRLA) with right cardiac structures much larger than left structures by fetal echocardiography (FE), remains unexplained if no left side obstruction is detected. This presentation illustrates a case of FRLA secondary to restrictive foramen ovale (FO). A 36 years old pregnant woman was referred at 36WG for FRLA detected by obstetrical ultrasound examination. FE evidenced disparity between right to left cardiac chambers: RV/LV 1.4 & PA/AO 2.4. No left or right obstructive lesions were detected. The FO was restrictive (right-left velocity of 0.7m/s) with premature decrease of FO flap motion. Regular follow showed no development of signs of cardiac dysfunction. Vaginal delivery occurred at 39 WG. Apgar score was 10/10 and clinical examination was normal. Postnatal echocardiography revealed normal anatomical findings with normal LV dimensions associated with a tiny patent foramen ovale (left-right). Conclusion: Restrictive foramen ovale should be looked for by operators of FO anatomy & velocity, particularly in cases of FRLA with no ultrasonic anomaly detected.

318

Changes in myocardial glycogen content and effect of glucose-insulin infusion during fetal tachycardia in a porcine model

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AIM: To assess metabolic changes in the fetal heart with tachycardia in a porcine model. BACKGROUND: Fetal tachycardia may cause hydrops and lead to fetal death. The pathophysiology of this condition is poorly understood and the existing treatment methods remain suboptimal. The fetal myocardium almost solely metabolizes carbohydrates and glycogen depletion is likely to play an important role in the deterioration fetal heart function during tachycardia. METHODS/DESIGN: Three pregnant sows at 13-14 (out of 15) weeks gestation were used. From each litter, 2 to 5 fetuses were included in the study. The fetal chest was exposed through a laparotomy and fetal sternotomy was performed, while the fetus remained oxygenated and anesthetized through the umbilical cord. Cardiac output was measured with an aortic flow probe. The fetal heart was paced via the right atrial appendage. Normal fetal heart rate for pigs is 120-140/min. The fetuses were randomized into four groups: 1) Baseline measurement at 0 minutes, 2) 90 minutes pacing at 150/minute, 3) 90 minutes pacing at 250/min, 4) 90 minutes pacing at 250/min with glucose-insulin infusion. After termination, the hearts were removed and immediately fixed in liquid nitrogen. Myocardial glycogen was subsequently measured. The study will include 3 additional pregnant sows before completion Spring 2001. PRELIMINARY RESULTS: [tbd] CONCLUSION: Myocardial glycogen stores are severely depleted during fetal tachycardia. This may be of important pathophysiologic significance and can be prevented by (intolerance) glucose-insulin infusion.

Session 29: Catheter Interventions

319

Follow-up results of transcatheter valvotomy in patients with pulmonary atresia and intact ventricular septum

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Aims: To report the follow-up results of transcatheter pulmonary valvotomy in patients with pulmonary atresia and intact ventricular septum (PA/IVS). Methods & Results: Between June 1995 and August 2000, 28 neonates with PA/IVS, tricuspid valve Z-value > -3 (ranging from -1.8 to 0.6, mean±SD = 0.3±0.8) and without significant interatrial communications underwent an attempted transcatheter pulmonary valvotomy. For perforation of the atretic pulmonary valve, a guidewire was used in 6 and radiofrequency guidewire (BA 120, Osypka) in 22. The attempt failed in 3 patients, 1 of whom had pericardial effusion requiring emergent drainage. Pulmonary valvotomy was successful in 25 patients, 4 with a guidewire and 21 with a radiofrequency guidewire. A subsequent balloon pulmonary valvuloplasty was performed in these 25 patients. The right ventricular systolic pressure decreased from a mean of 121±22 to 54±14 mm Hg ($p<0.01$). Prostaglandin E1 could be weaned within 4 weeks in 19 patients, of whom 1 died of heart failure and infection. Six patients had required a shunt with or without a right ventricular outflow tract patch. One underwent ligation of the ductus because of heart failure. A total of 24 patients were discharged smoothly with systemic O2 saturation above 75%. After a follow-up period ranging from 2 to 62 months, one who had underwent a shunt implantation died of heart failure. The most recent echocardiograms in 22 patients showed a mean gradient of 25±20 (range from 0 to 72) mm Hg. All 23 patients had systemic O2 saturation above 85% during follow-up. Conclusion: Transcatheter pulmonary valvotomy can be an alternative to surgery in selected PA/IVS patients with adequate right ventricular size.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

320

A novel combined surgical-interventional approach for establishing total cavo-pulmonary anastomosis using the Amplatzer(tm) stent graft. Results of a feasibility study

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Background. In children with functionally univentricular hearts, total cavo-pulmonary anastomosis (TCPC) is a frequently chosen palliation. In high risk patients, the univentricular circulation is often established in a stepwise approach with creation of an aorto-pulmonary shunt followed by a Glenn anastomosis before TCPC is completed at a later age. We developed a stepwise approach with surgical pre-conditionning and interventional completion which reduces the need for cardio pulmonary bypass, reoperations and intracardiac sutures. Methods. In 10 sheep (30–50 kg) a unilateral Glenn anastomosis was created surgically using a Gore-Tex tubing (diameter 12 mm), the SVC was banded above the cavoatrial junction. A Goretex tube was cut lengthwise and sutured around the intrapericardial part of the IVC to provice resistance for the Amplatzer graft. For interventional completion, a guidewire circuit was created from the right internal jugular vein to the femoral vein. The banding of the SVC was dilated and an Amplatzer stent graft (Diameter 16 mm) was implanted with its portion in the IVC placed cranial to the hepatic vein and its portion in the SVC placed inferior to the Gore-Tex tubing connecting the SVC to the pulmonary artery. The animals were observed for 2 hours after interventional completion before they were sacrificed. Results: All 10 animals survived the combined one-stage surgical-interventional procedure before they were sacrificed two hours postoperatively. The Amplatzer (TM)-Graft was successfully implanted in all animals without causing obstruction to the hepatic veins. No rhythm disturbances were encountered. Conclusion: Our results demonstrate that TCPC may be established by surgical preconditioning and interventional completion using a self-expandable Amplatzer (TM) stent graft. This procedure may be an attractive option to reduce the number of surgical interventions in children with congenital heart defects necessitating univentricular palliation.

321

Long-term results after implantation of biodegradable stents produced from corrosive iron-based alloys into the descending aorta of New Zealand white rabbits

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Purpose: To evaluate neointima formation, thrombogenicity and local inflammatory response 3 to 15 months after implantation of NOR-1 biodegradable metal stents into the descending aorta (DAO) of New Zealand White Rabbits. Methods: 20 adult New Zealand White Rabbits (3050–3300 g). After femoral arterial cutdown, angiography (angio) of the DAO was performed. Using inflation pressures of 12 atm for 30 s, NOR-1 stents (20 mm length) were implanted distally to the renal arteries. Balloon size was chosen to achieve balloon diameter : vessel diameter ratios approximating 1.2. After implantation, d,l-pseudamol (2 mg/ kg) and aspirin (5 mg/ kg) was administered for 3 months. Repeat angio was performed after 3, 6 and 12 and 15 months. After euthanasia the DAO was evaluated histologically for neointimal hyperplasia and inflammation the organs were evaluated for assessment of systemic toxicity. Results: Angio demonstrated patent vessels in all animals 3,6,12 and 15 months after implantation of NOR-1 stents into the DAO. No thrombotic occlusion was encountered. Neointimal proliferation was mild and did not lead to significant lumen narrowing in all animals. Histopathological evaluation demonstrated complete endothelialization of the stent. Only mild granulomatous reaction was observed adjacent to the stent struts and degradation products. Conclusion: NOR-1 biodegradable metal stents have a low thrombogenicity. Inflammatory response of the vessel wall is mild and neointimal hyperplasia moderate 3 to 15 months after the implantation.

322

100 stents in native or postoperative vascular lesions

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Introduction: Growth is a determinant factor for stent implantation in children with vessel stenosis. Large diameter stents are needed in our field.(Palma). Following Mullins technique, we have used 100 stents in 72 patients with 2 ±1 previous operations. Age 0.01–41, mean 10.6 ±6.6 years, weight 3.1±77kg , to treat stenosis in pulmonary branches 52 (61 previous palliative surgery) Coarctation (aorta) 4, recoarct 4. Operated Mustard 3 and Fontan 3. Pulmonary homograft 1 and RVOT in 2. Pulmonary vein stenosis 1. Peripheral PA stenosis 1. Material and Methods: 58 implants in the cathlab and 15 in the operating room. 20 patients needed 2 stents. Used stents: Palmaz 64 cases, Corinthian, 1, Covered Talent 1, Coronary 1 and large P-401 4 cases. Sheaths: 13 (5F, 6 / 12F; 3 /8F. 11F the rest. Medtech 5F-1 long was. Stents mounted in VACA 12/15 mm and Power Flex or Maxibal in last 15 cases. Results: 56 uncomplicated cases. Vessel diameter increased from 9.4±2.7 to 14.2±4.9 mm. Gradient diminished from 21.3±15.5 to 7.1±10.3 mmHg and RV pressure from 78±19 to 54±8 mmHg. 0-IV one restenosis found in 7 year follow up. Complications: Death: 1 (1.3%) (intraoperative stent) CNS damage 2 , 1permanent, 1 transitory. Stent migration 6 (10%), (3 reimplanted) and 3 (4.8%) required emergency surgery. Haemorrhage 5 cases. RPA thrombus needing fibrinolysis 1 and occlusion of upper lobe PA in 2. Conclusion: Stent is an essential tool in complex CHD treatment and remains as the treatment of choice for most congenital or acquired vessel stenosis. Mortality is possible as well as serious complications. Restenosis seems to be rare. Technical drawbacks are: non mounted and not flexible stents. Research in this field is essential. Implantation in the OR may reduce operation time.

323

Use of new flexible balloon expanding stents to treat right-sided obstructions in patients with congenital heart disease

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INTRODUCTION: The use of stents in patients with congenital heart disease have been performed since the late 1980's. However, due to the rigidity of the stents, larger sheaths are required for delivery limiting their use in infants and small children. Recently, flexible balloon expanding stents, which have improved trackability and similar strength characteristics to the traditional Palmaz stent, have been developed which can be delivered through smaller delivery systems. We describe our initial experience with the Cordis Bx (CIQ) and Intrafirst/genitus LD (ITLD) stents in the treatment of right-sided obstructions in pediatric patients (pt). METHODS: Each pt underwent cardiac catheterization to determine the severity of the right-sided stenosis. In all cases, the surgeon and pediatric interventionalist involved determined that intravascular stent placement would offer the best treatment option for the pt. RESULTS: In 4 pt. 5 CIQ stents were used to treat right-sided conduit stenosis (CS) n=1, right pulmonary artery (RPA) stenosis n=1 and left pulmonary artery (LPA) stenosis n=3. The CIQ stent could be deployed through 5 Fr sheaths and expanded up to 13 mm in diameter. The CIQ stents was primarily used in infants and smaller children (mean weight= 7.7 kg, range= 4.6- 11.1 Kg). In 8 pt. 8 ITLD stents were deployed to treat right-sided obstructions (CS n=2, LPA stenosis n=3, LPA stenosis n=2, innominate vein/superior vena caval stenosis n=1). The ITLD stents were able to be deployed through 7 Fr sheaths and can be dilated up to 22 mm in diameter. The ITLD stents were primarily used in larger pt. (median weight= 40 Kg, range= 9- 90 Kg). In each case, stent deployment was successful and surgery avoided. CONCLUSION: The use of new balloon expanding flexible stents, with the improved trackability, allows the interventionalist to use smaller sheaths for intravascular stent deployment. These stents can be safely used in both infants and children for the treatment of right-sided obstructions.

324

Modified technique of stenting the atrial septum after Fontan operation

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Objectives: To develop a modified technique for stenting the atrial septum in the treatment of patients with a failing Fontan operation. Setting: Tertiary referral centre. Study design: Progressive ex-vivo experimental and clinical study. Patients and Methods: A stent was mounted on a standard valvuloplasty balloon catheter which was constricted to a predefined diameter by the use of a loop created from a temporary pacing wire. Full balloon inflation created a dumbbell shaped stent configuration. The technique was employed in two consecutive patients to relief symptoms of a failing Fontan circulation (bronchial cast and protein losing enteropathy). Results: Ex-vivo studies

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

confirmed that a dumbbell shaped configuration of a stent could be achieved using the above technique. Stent placement was successful in two patients. Systemic venous pressure was reduced by 2 and 1.5 mm Hg and arterial oxygen saturations decreased by 12 and 6 %. Both stents are patent 26 and 9 months post procedure and are in a stable position. In one patient the stent size was reduced during subsequent catheterisation procedure. Both patients had significant clinical improvement. No early or late complications were encountered. Discussion: This new technique allows placement of a dumbbell shaped stent with a predefined diameter across the atrial septum. This increases stent stability, facilitates re-crossing of the stent during future catheter interventions, and should ensure long-term stent patency. The technique is likely to prove of value in the management of patients with a failing Fontan circulation or in those with end-stage pulmonary hypertension.

325

Transcatheter coil occlusion of the arterial duct: results of the European Registry

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To report the Association for European Paediatric Paediatric Cardiology Registry for transcatheter coil occlusion of the arterial duct we performed a retrospective study of submitted information to treat data from 20 European and Middle Eastern tertiary referral centres. A number of clinical factors were chosen and were analysed as possible predictors of a sub-optimal outcome. These included age, weight, minimum duct diameter, duct shape, type of coil, number of coils implanted, immediate and whether or not there had been a previous intervention to close the duct. Results: Since 1994, 1297 attempted coil occlusions of the arterial duct were reported on. 1258 patients. The median age at the procedure was 4 years (range 0.1–52 years) and the median weight was 29 kg (range 1.8–100 kg). The immediate occlusion rate was 59% and this rose to 95% at 1 year. An unfavourable outcome occurred in 129 occasions (10% of procedures) and was defined as coil embolisation, abandoned procedure, persistent haemolysis, residual leak requiring a further procedure, flow impairment in adjacent structures and/or re-canalisation. Of all the clinical factors only decreasing duct size [Odds ratio of 2.6, 1 (CI 2–3.2)] and the presence of a tubular shaped duct [Odds ratio 2.4: 1 (CI 1.4–4)] were positively associated with an unfavourable outcome. Conclusion: Results of the European Registry support the view that transcatheter coil occlusion of the persistent arterial duct is a safe and effective procedure. Unfavourable outcomes are more likely when attempting coil occlusion of larger and/or tubular shaped ducts. In these situations alternative occlusion systems should be considered.

326

Use of snare to augment pulmonary balloon valvuloplasty (PBV) in selected patients with pulmonary atresia and intact ventricular septum (PA with IVS)

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Some selected group of patients with PA with IVS and well developed right ventricular (RV) sinus may require only PBV as definitive intervention. One of the more technically difficult aspects of the procedure is safe atrial perforation of the pulmonary valve (PV) and crossing it with an initial balloon dilation catheter (BDC). Many different approaches have been reported earlier utilizing different wires and radio frequency catheters (RFC) to achieve the goal. The shortcoming in all of them is a lack of a real time visualization of the plane of the atretic PV during the most critical part of the procedure. The most common approach to alleviate the problem partially is use of a previous angiogram as a road map and/or use of echocardiogram (epicardial or transesophageal). We describe a new method to help visualize the plane of the PV in real time that we used in our patient. Following the hemodynamic portion of the catheterization, the 10mm snare (Microvena Corp.) was advanced retrograde through a patent ductus and positioned in proximal main pulmonary artery where it was opened to the size slightly less than that of the diameter of the PV and then lowered to rest on the pulmonary surface of the atretic PV. The guiding catheter was then advanced to the RV outflow tract through which a straight 0.035 in teflon coated wire (Cook Corp.) was used for the perforation of the PV. The open snare seen on the fluoroscopy served as a

327

Initial experience with a new Amplatzer device to maintain patency of fontan fenestration

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Background: Creation of a fenestration in the lateral conduit in the extracardiac Fontan is a routine procedure. Spontaneous closure of fenestration may lead to Fontan circulation failure. We report the use of a new device which was successfully used to maintain patency of Fontan fenestration. Methods: The device was modified from the Amplatzer Septal Occluder, with a 4-mm central unexpanded waist. The loading, deployment and release were similar to the septal occluder. Patient 1 was an 8 year old with severe protein losing enteropathy and pericardial tamponade, chronic heparin therapy. Patient 2 was a 3 year old who had extracardiac Fontan. He had medically refractory persistent pleural effusion. In both patients, the medial wall of the lateral tunnel/conduit was successfully punctured with a transseptal needle and dilated with a 6-mm balloon. Under transesophageal echocardiography the device was deployed using a 7-French delivery sheath. Results: The deployment was successful in both patients on first attempt. The systemic saturation decreased from mid 90% to low 80% in both patients. Contrast injection revealed good flow through the fenestration. Echocardiographic examination at six month follow-up revealed good flow through the fenestration in both patients. Both patients had relief from their symptoms. Conclusions: The preliminary results suggest that the Amplatzer fenestration device can serve as a valuable tool in failing Fontan circulation and may help to avoid surgical intervention. More studies are needed to assess long-term efficacy of the device.

328

Open cell design stents for vascular obstructions in congenital heart disease: a comparison of InterStent versus Palmaz stents

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Slotted stainless Palmaz stents (PS) remain the most commonly used in congenital heart disease (CHD). Limitations of PS include rigidity, foreshortening and poor conformability on expansion, sharp edges with frequent balloon rupture, and jailing of side branches. Recently, stents with open cell design are most appropriate for congenital heart lesion have been introduced (Interstent, IS). We reviewed our initial experience with IS comparing performance with PS. METHODS AND RESULTS: Between 7/99 and 10/00, 21 IS and 23 PS were implanted in 40 patients aged 3m to 25y (mean 12y) in pulmonary arteries (22), venous baffles (7), Fontan fenestrations (2), coarctations (5), conduits (2), and collaterals (1). Stents ranged in length from 10 to 40 mm (median IS=16mm, PS=20mm, p=NS), and were dilated with balloons ranging from 4.5 to 20 mm diameter (median IS=9mm, PS=11mm, p=NS). Increase in lumen diameter (4.7 to 9.1 mm for IS vs 5.1 to 13mm PS) and gradient reduction (23 to 9mmHg IS vs 31 to 15mmHg PS) were comparable (p=NS). Other aspects of stent performance differed significantly (p<0.05): PS shortened more (mean 18% vs 3%), required larger sheaths (mean 9F vs 8F), and were more likely to be associated with balloon rupture on deployment (7/23 vs 0/22). IS conformed almost twice as well to vessel curvature as did PS. One IS was deployed across the origin of the right middle lobe pulmonary artery. The orifice of this vessel was dilated to 6mm through the side of the stent. CONCLUSIONS: Stents with open cell design foreshorten less are conformable, and allow access to 'jailed' branches. These characteristics may be beneficial for vascular obstructions in CHD.

329

Atrial septal defect closure with the Helex septal occluder device: the FDA phase I feasibility trial

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Background: The Helex is an ePTFE covered nitinol double circular disk device for transcatheter atrial septal defect (ASD) closure. We report preliminary data on device safety and performance. Methods: Between 4/00–12/00, all appropriate patients with secundum-ASD in two centers were enrolled in a prospective, non-randomized FDA phase-I feasibility trial. Procedures were performed using general anesthesia with transesophageal echocardiography guidance. Procedural success was defined as accurate placement of a device. Chest-x-ray and transthoracic echocardiography were

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

performed 24hrs, 1mos, 6mos and 1yr following deployment. Results: Thirty-eight patients, median age 13yrs (range 0.4–55yrs), underwent 40 catheterizations. Static balloon-stretched ASD diameter was 7.1–26mm (17–8.4). Device/balloon waist ratio was 1.3–4.2 (1.5–0.5). The procedure was successful in 34/40 catheterizations with 3/6 failures due to unavailability of the largest (35mm) device. One of these patients underwent closure with a 40mm CardioSEAL at initial catheterization and 2 returned for closure with a 35mm Helex device. The remaining 3 procedural failures were closed surgically. In all procedures, the device design allowed repositioning for optimal placement prior to release. There were 7 procedure-related minor adverse events with device embolization (unsuccessful retrieval) in 2-patients, transient arrhythmia in 3-patients and transient ST depression in 2-patients. Median fluoroscopy time was 23 minutes (including 4 patients with additional interventional procedures). Chest-x-ray and transthoracic echocardiography at 24hrs demonstrated a well-seated device in all patients and trivial/small residual leaks in 23/34(68%) of patients. Trivial/small residual leaks were present in 15/27(55%) of patients with 1mos data and 1/6(17%) of patients with 6mos follow-up. Conclusion: These data indicate that the Helex device is safe for secundum-ASD closure. The ability to easily reposition/retrieve this device may be an advantage. No patient has a significant residual leak and the incidence of trivial/small leaks decreases during follow-up.

Session 30: Arrhythmias, Electrophysiology, Sudden Cardiac Death

330

Azimilide inhibits outward potassium currents in cultured human fetal ventricular myocytes

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Objective: The purpose of the present study is to investigate the effects of azimilide, an NE-10064, on delayed rectifier potassium current (IKr) and transient outward potassium current (Ito) in cultured human fetal ventricular myocytes. Background: Azimilide is a new potassium channel antagonist. It blocks both slowly (IKs) and rapidly (IKr) activating components of IK. Azimilide has also been shown to significantly reduce the frequency of symptomatic arrhythmia recurrences in adult patients with atrial fibrillation and/or atrial flutter. However, relatively little is known about the effects of azimilide on IK and Ito in immature cardiac cells. Methods: The tight-seal, whole cell voltage clamp technique was used to investigate the acute effects of azimilide on IK and Ito in single cultured human ventricular myocytes. Results: We found that averaged cell capacitance of these cultured human fetal ventricular myocytes was 82.7±7.9 pF (n=8). Perfusion with 100 uM azimilide for 6–8 min inhibited the IK from 312±50 to 214±43 pA (n=6, p<0.05) at the clamping membrane potential of +40 mV. The current amplitudes were measured at the end of test pulse duration of 2 sec. The current-voltage relation of IK was not altered after perfusion of azimilide. We also found that averaged peak current amplitude of Ito in these cultured human fetal ventricular myocytes was significantly inhibited by 100 uM azimilide (from 599±194 to 152±133 pA, n=6, p<0.05, clamping membrane potential = +20 mV). In addition, azimilide seems to block both IK and Ito in a dose-dependent manner. Conclusion: The present study, for the first time, provides direct evidence that azimilide inhibits cellular membrane outward IK and Ito in cultured human ventricular myocytes. The results from the present study may have important clinical implications regarding azimilide treatment of pediatric patients with arrhythmias.

331

Dispersion of repolarization is not related to volume loading of the right ventricle in patients with Tetralogy of Fallot late after repair. A cardiac MRI study

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Background: Dispersion of repolarization as expressed by QT dispersion (QTd) has been proposed as an arrhythmic risk marker. Recent evidence has correlated QTd and total QRS duration in patients with repaired Tetralogy of Fallot (TOF) with arrhythmic events. We aimed to correlate ventricular volume, mass and function with: QT, JT and QRS dispersion. Methods: 30 consecutive patients with repaired TOF were included. A Picker Edge 1.5 Tesla MRI scanner was used for measurement of biventricular volume, mass and function (values indexed to BSA). ECG (at 50 mm/s) parameters were measured using electronic calipers. Correlations between the above MRI indices, age, time from operation and QT, JT and QRS dispersion were studied. Results: Patient characteristics were: mean age 31 ± 10, years from operation 23 ±7, baseline QRS 155±32ms, QTd 67.9 ± 26ms, JTd 64.2 ± 22 and QRSd 45.9 ± 16. There was poor correlation between QTd and Right Ventricular Mass index (RVMi) and Total Mass Index (r=0.4, p=0.049 and r=0.4 p=0.02 respectively). (See Figure) There was no correlation between RVEF or RVEF and QTd, JTd or QRSd. There was also no correlation between QRS duration and QTd. Stepwise backward linear regression analysis provided a model where RVMi, RVEF, RVESVi and RVSVi (RV mass, ejection fraction, end-systolic volume and stroke volume respectively) would predict QTd explaining only 48 % of its variation. Conclusions: QTd, JTd and QRSd are not correlated to the volume load of the right ventricle in TOF late after repair. The weak correlation with mass indices, may imply that hypertrophy is partially responsible for these repolarisation abnormalities, previously linked with arrhythmia generation.

332

Tip cooling techniques improve success for intra-atrial reentrant tachycardia (IART) ablation

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Background: Relatively low success rates and high recurrence rates for RF ablation of IART in pts with congenital heart disease have led to the introduction of high technology mapping and more powerful ablating tools. Methods: The impact of these changes were examined in 30 pts (ages 4–73 yrs, median 25 yr) who underwent 34 RF catheter ablation (RFCA) procedures for recurrent IART from 2/98 to 9/00. For 25 pt. (28 proc.) standard mapping was used, with ablation sites identified by classic atrial potentials, concealed entrainment and post pacing interval within 20 ms of IART cycle length. For 6 pt. (6 proc.) an electroanatomic mapping system (CARTO) was also used for mapping. When possible, RFCA was also used to bridge known anatomic barriers involved in the circuit. When RFCA was initially unsuccessful, a 6 or 8-mm tipped catheter (Marinr, Medtronic) or a cooled tipped catheter (Chili, Cardiac Pathways) was used. Follow up ranged from 2–30 months (median 18 months). Results: Overall procedure success was 91%, statistically regardless of the mapping technology used. However, many of the eventual successes would have failed without the addition of catheter tip cooling technology: 4 mm tip = 15/31 (48%) success vs 6/8 (mm tip = 8/8 (100%) vs Chili = 6/8 (100%) (p=0.01). Although the advanced mapping technologies had no quantitative effect on acute outcome, they did elucidate the anatomy and physiology leading to the impression of qualitative improvement. Overall recurrence rate was 19% with no clear technology effect (conventional mapping only 19%, electroanatomic 20%, 4 mm tip only 20%, 6/8 mm 25%, cooled tip 13%). Conclusion: IART can be successfully ablated with a combination of traditional and advanced mapping/ablation techniques. RFCA catheter tips which allow for application of increased power to the tissues are an important advance. However, recurrence rates remain significant. Electroanatomic mapping which allows for identification of anatomic critical barriers with cooled tip technology will hopefully improve late outcome.

333

School children's sudden death: cardiac death and cerebrovascular death

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Children's sudden death at school is a major problem in school health. In 1990–1999, the number of cases of sudden death at school in Aichi prefecture of Japan, were 115, where the population is about 6,000,000. In these cases, 39 cases had had congenital heart disease, arrhythmia, asthma, epilepsy or anaphylaxis and they exactly died of these diseases. In other 76 cases, any disease had not been found before the incidence. Of these 76 cases, however, the 23 cerebrovascular hemorrhage cases were found, and the 53 cases were diagnosed cardiac death. Regarding with 53 cardiac death cases, 16 cases were females and 37 cases were males. Cases were more in males than in females. Eighty percent of death cases occurred during exercise. Thirty-four percent of death cases occurred in the late morning, and 23 % occurred in

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

late afternoon. In contrast, the 12 cases of 23 cerebrovascular death were males. And 25 % of death cases occurred during exercise. Twenty-two percent of death cases occurred in the early morning, and 33 % occurred in the early afternoon. These results show that the inducements of sudden death are different between cardiac diseases and cerebrovascular diseases. We should consider various factors in school life which may become inducements of sudden death, i.e. gender, exercise, routine schedule in school health.

334

A cost-effectiveness analysis of project A.D.A.M. (automatic defibrillators in Adam's memory) for high schools in the Milwaukee public school system

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Introduction: Recent deaths of local high school athletes have focused attention on the potential benefits of the placement of automatic external defibrillator (AED) units in high schools. These deaths can be especially devastating, as they often occur to otherwise healthy teenagers who, upon autopsy, are discovered to have a previously undiagnosed but treatable congenital heart defect. Project A.D.A.M. is a joint effort between Children's Hospital of Wisconsin and other community agencies. While AED units are now the standard of care for emergency medical system response units, placement of AED units in locations accessible by and intended for use by the lay public is more controversial. Cost-effectiveness analyses for publicly placed AED units intended for use on adult populations have been reported but have not been reported in the pediatric population. Methods: A cost effectiveness analysis has been performed using the conceptual model shown in Figure 1. Costs for a school-based AED program as well as hospitalization have been addressed, as have quality adjusted life years (QALYs) for the affected population, which is as expected, high given the previously healthy nature of this population. Sensitivity analyses will account for ranges in the probability of an event, such as time to first intervention and the occurrence of ventricular fibrillation or asystole. Results: Five students suffering sudden cardiac arrests in Milwaukee Public School (MPS) system high schools between 1994 – 1999 have been identified, all of whom survived from the arrest. The expected cost for the implementation of Project A.D.A.M., including training, 50 AED units, and follow-up and evaluation for 5 years, is approximately $120,000.

335

Intracardiac electrogram fractionation – a potential predictor of sudden cardiac death after Mustard's or Senning's repair

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An increased risk of sudden cardiac death exists years after Mustard's or Senning's repair for transposition of the great arteries (TGA). Intracardiac electrogram fractionation is emerging as a technique for studying risk of ventricular arrhythmias and sudden cardiac death. The aim of our study was to determine whether abnormalities in electrogram fractionation exist following Mustard's or Senning's repair. METHODS: Fractionation studies were performed on 19 patients (age 12–22 years) with Mustard's or Senning's repair for TGA. One bipolar electrode catheter was positioned in the systemic atrium, 2 in the left ventricle and one in the right ventricle. Using each unipolar electrode catheter in turn as the pacing catheter and recording electrograms on the other ventricular catheters, the heart was paced AV sequentially at a basic cycle length of 500ms. A ventricular extrastimulus was introduced after every second beat, initially at an S1S2 interval of 450 ms, reducing the interval by 1ms on each subsequent beat to a minimum interval of 220ms. For each extrastimulus, the resulting electrograms were analysed for duration and fractionated components. RESULTS: - Major abnormalities in electrogram fractionation were evident in patients with Mustard's or Senning's repair. Even with ventricular extrastimuli at long S1S2 intervals, the resulting electrograms showed loss of early components suggesting that faster conducting fibres within the ventricle were being 'marked'. Similar findings are demonstrated in patients with long QT syndrome or with ischaemic heart disease and VF. Since starting the study, a 15 year old with previous Senning's repair died suddenly. He had shown marked loss of early electrogram components and an abrupt increase in electrogram duration even at long S1S2 intervals. The fractionation abnormalities were evident in the right but not the left ventricle. CONCLUSIONS: - Major abnormalities in electrogram fractionation exist after Mustard's or Senning's repair. The technique has the potential for evaluating risk of sudden cardiac death in this patient population.

336

Molecular autopsy identifies a KVLQT1 mutation in a 17-year-old male found dead in bed

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Background: Sudden unexplained death (SUD) claims over 4000 persons between the age of 1 and 22 each year in the United States. Nearly half of all cases have a normal autopsy and are dismissed without a definitive diagnosis. Methods: A previously healthy 17-year-old male was found dead in his bed in March 1999. No cause of death was established. Toxicologic screening was negative and the autopsy was unremarkable. The decedent's mother sought medical evaluation for her surviving 13 year old son to determine whether or not he was at risk for premature sudden death. Non-invasive clinical testing was performed. Blood samples and archived paraffin embedded autopsy tissue were obtained to screen DNA for cardiac ion channel defects predisposing to long QT syndrome (LQTS). Exon-specific amplification by polymerase chain reaction and direct manual sequencing of KVLQT1 (LQT1) was performed. Results: Echocardiographic and electrocardiographic evaluation of the decedent's immediate family was normal. Epinephrine provocation testing in the decedent's mother revealed paradoxical QT prolongation with epinephrine. Despite a low probability Schwartz score

337

Spectrum of cardiac pathology in infants who are victims of sudden unexpected death

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The goal of this study was to determine the importance and nature of cardiac pathology found at autopsy in victims of sudden unexpected death. All autopsies and coroner reports of infants who were victims of sudden death in infants older than a week and younger than 2 years in the province of Québec between April 1986 and March 2000 were reviewed. Age at time of death, sex, type of heart defect, duration of symptoms and whether cardiac disease was recognized before death were documented in each case. The majority(69%) of autopsies were performed in an academic center. A cardiac condition was present in 85 autopsies representing 8% of the total number of sudden deaths. Median age at time of death was 120 days. Males accounted for 61% of cases. A structural heart defect was present in 58(54%) of autopsies. These were classified as defects in cardiac septation(30), left sided obstructive lesions(14), cyanotic heart disease(8) and others(6). A non-structural heart condition was present in 35 cases(33%) including 17 cases of endocardial fibroelastosis, 13 cases of myocarditis and 5 cases of cardiomyopathy. Forty-four(52%) infants were found dead in their sleep with no preceding symptoms. In 23 cases death ensued within an hour of onset of symptoms. Only 16 infants(15%) had symptoms for more an hour before death. The underlying cardiac pathology was recognized pre-mortem in only 40% of cases. Among infants who died suddenly with a significant non-structural heart disease only 8% were previously known to the medical community. Significant cardiac pathology is present in 8% of infants who die suddenly. The victim is more likely to be male and to have been found dead or succumbed after a very brief clinical course. Structural and non-structural heart disease are common with the latter being rarely recognized prior to death.

338

Cardiovascular causes of sudden death in a pediatric population

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At difference from the adult population where the most common cause of sudden death (SD) is coronary atherosclerosis, the spectrum of pathologic substrates which may account for SD in children and adolescents is wider. Among 320 consecutive juvenile SDs (<35 yrs) collected in the Veneto Region, Italy, 80 (25%) occurred in the pediatric age (<18 yrs). They were 49 male and 31 female, mean age 12 yrs. In 9 cases, SD was unrelated to the cardiovascular system and was due to cerebral embolism (3) and asthma (6). In 71 cases (89%) SD was cardiovascular in nature and the pathophysiological mechanism of SD was mechanical in 3 (1 pulmonary embolism, 1 mitotic aneurysm and 1 dissecting aneurysm in the setting of bicuspid aortic valve) and arrhythmic in 68. Among the latter, the most common causes of SD were coronary artery anomalies (CAA=10, 15%), arrhythmogenic right ventricular cardiomyopathy (ARVC=9, 13%), myocarditis (9=13%) and hypertrophic cardiomyopathy (HC=7, 10%). 40% of SDs were due to congenital heart diseases. Nine cases remained unexplained even after careful histologic

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

examination and one of them had ECG documented long QT syndrome. Family history of SD was present in 73% of ARVC pts and 17% of HC pts. Previous syncope has occurred in 25% of ARVC and 33% of either CAA or HC pts. ECG changes were present in 75% of ARVC and 50% of HC pts. In conclusion, SD in people less than 18 yrs of age is mainly due to CAA, inherited cardiomyopathies and myocarditis. A congenital heart defect potentially detectable during life is present in nearly half of the cases (44%). Many of them should be suspected on the basis of prodromal symptoms, family history and ECG changes.

339
Does early pulmonary valve replacement following repair of tetralogy of Fallot change QRS duration?
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The incidence of late sudden death is 1–6% after repair of tetralogy of Fallot (ToF). QRS duration (QRSd) > 180 msec has been identified as a marker of ventricular dysrhythmias and sudden death. Increasing QRSd has been associated with greater degrees of pulmonary insufficiency (PI) and right ventricular (RV) dilation. We sought to determine if early pulmonary valve replacement (PVR) in repaired ToF would lead to a reduction in RV size and QRSd. Patients undergoing isolated PVR for symptomatic RV dilation post ToF repair were studied with ECG and echocardiogram. Studies immediately before PVR and on follow-up were assessed. QRSd was recorded. RV dimension on 2 D echo (RVED) was calculated from the short-axis view and normalized for body weight. Thirty patients were identified with a median age at ToF repair of 16.6 (1–94) mos., age at PVR of 13.6 ± 6.6 years and time to follow-up post PVR of 28 (range 1–69) mos. QRSd prior to PVR and on follow-up was 151.7–25.2 msec and 147.4–19.9 msec (p= .28). Only 2 patients had a QRSd > 180 msec. RVED and (normalized RVED) pre and post PVR were 34.1 ± 7.6 mm (1.0 ± .5 mm/kg) and 25.2 ± 7.8 mm (.5 +/- .3 mm/kg) respectively (p<.0001). QRS duration was greater in those patients with a longer time between ToF repair and PVR (r=.38 p= .04) but was not related to age at initial ToF repair. Despite a significant reduction in RV size following PVR, there is no change in QRS duration. QRSd >180 msec is unusual in young patients following ToF repair. Early PVR may prevent further RV dilation and QRS prolongation thus reducing the risk of ventricular dysrhythmias and sudden death.

340
QTc dispersion is increased after well repaired coarctation of the aorta: implication for sudden death
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Background: Long term data show a higher than expected incidence of sudden death in patients (pts) with repaired aortic coarctation (rCOA). The etiology for sudden death in this population has not been investigated. An increase in QTc dispersion (QTcD) has been considered as predisposing to serious ventricular arrhythmias or sudden death in various cardiac diseases. Purpose: To determine if QTcD is increased in pts with rCOA following a good operative result. Methods: Pts with rCOA were recruited with the following criteria: 1) arm-leg systolic pressure gradient < 20 mmHg; 2) normotensive at rest and 3) no aortic stenosis. Pts and age-matched healthy controls underwent electrocardiographic and 2D echocardiographic study with measurements for QTcD and left ventricular (LV) mass indexed to body surface area (LVM/m2). Results: The study contained of 40 pts (mean age 12.3±6.5 years) followed up after a successful rCOA (mean age at coarctectomy 4.8±5.1 years; follow-up after surgery 7.4±4.9 years). QTcD and LVM/m2 were significantly greater in pts vs control [65.4±21.6 vs 38.2±11 ms, p<0.0001 and 1.8±2.2 DS (Z-score), p<0.05, respectively]. No significant correlation was found between QTcD and LVM/m2 age at surgery and postoperative follow-up. Conclusion: Pts with rCOA and a good operative result show an abnormally increased QTcD. These data, combined with the observation that QTcD is not correlated to the LVM/m2, suggest that in these pts other factors (e.g., increased sympathetic tone and myocardial fibrosis) may be responsible for these electrophysiological abnormalities. Owing to the prognostic values of QTc dispersion a careful follow-up is warranted in these pts.

341
Chronological distribution of malignant arrhythmias in pediatric and congenital heart disease population
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Background: Recent studies have found variation in the frequency of acute cardiovascular events in the adult population, including life-threatening arrhythmias. These studies have suggested an increase in such events in the late morning; similarly, increases are seen in winter and early spring, compared with the remainder of the year. Pediatric patients represent a unique population, accounting for < 1% of all ICD implants. We sought to determine whether pediatric ICD recipients also have circadian and seasonal variability in shock frequency. Methods: We retrospectively examined our patients with implantable defibrillators to assess the timing of life-threatening arrhythmias. Our population consists of children and adults with congenital heart disease; a total of 70 patients who have had ICDs placed for previously identified malignant arrhythmias. Data from 1/1996 to 11/2000 was considered, with 20 patients receiving therapy for ventricular tachycardia or fibrillation, a total of 57 therapies. Results: We analyzed several variables including time-of-day, day-of-week, and month-of-year. Unlike in adult patients, very few events occurred in the morning (less than 2% of all events occurred between midnight and 6 a.m.), with the most therapies occurring between 6 p.m. and midnight (42.6). Therapy was required more frequently in fall and winter (Sept-Jan); these 5 months represented 60% of the total therapies given throughout the year. The day of the week also varied from a normal distribution, peaking on Mondays with 25% (expected rate = 14%), strikingly similar to adult ICD patient studies. Conclusions: The pediatric and adult congenital heart disease populations appears to show some seasonal and daily trends in ICD event rate, as seen in the adult populations. These findings suggest modes of arrhythmia vulnerability that may be in part unrelated to occupational, physical, and emotional stressors.

MAY 31 Time: 11:00—12:30

Session 31: Cardiac Imaging: Echo-2-D, 3-D, TEE

342
Atrioventricular septal defect: when is the left side too small for biventricular repair?
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In patients with atrioventricular septal defect (AVSD), it can be difficult to predict the adequacy of left heart structures for biventricular (2V) repair. During a ten-year period, 25 infants with AVSD presented for surgery. In 3, the defect was severely unbalanced, and Norwood procedures were performed. In 22, 2-D echocardiograms suggested candidacy for 2V repair. However, as only 15/22 was 2V repair possible, suggesting a low sensitivity for 2D prediction of adequacy for 2V repair. In an attempt to improve this sensitivity, transesophageal echocardiograms on these 22 infants were reviewed. From 4 chamber images, the distance between the hinge points of the left sided AV apparatus was measured, and expressed as a z-score. The maximal diastolic areas of LV and RV were measured, and expressed as LV/RV. Z scores ranged from -1.0 to +6.0. The mean z-score for those with unsuccessful 2V repair was +4.5, vs -2.75 for those with successful repair, p=0.001. However, 3 patients with z-scores of +4.0 and +4.1 had successful 2V repair, suggesting that z-score alone was inadequate to predict 2V repair. The mean LV/RV area ratio was 41.9% in those with unsuccessful repair, vs 96.5% in those with successful repair, p<0.001. When z-scores and area ratios were considered together, all patients with area ratio <54% and z-scores lower than -2.4 had unsuccessful 2V repair. Using these criteria in the subsequent three years, the sensitivity for prediction of successful 2V repair by 2D echo was 100%, commending the use of these measurements for this difficult problem.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

343
Doppler ultrasound evaluation of Blalock–Taussig shunt velocity profiles – assessment of pulmonary artery pressure and flow in infants with complex cyanotic congenital heart disease
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Objectives: To evaluate the potential utility of continuous wave Doppler velocity profiles obtained across modified Blalock-Taussig shunt (B-T shunt) in the non invasive estimation of pulmonary artery pressure and pulmonary blood flow. Background: Critically ill neonates with complex congenital heart disease frequently require a modified B-T shunt as the first step for surgical palliation. Early postoperative haemodynamic problems are frequently related to excessive pulmonary blood flow or elevated pulmonary vascular resistance. These two scenarios are often difficult to assess by the use of standard monitoring techniques. Previous studies have demonstrated the utility of atrial Doppler flow velocity profiles in estimating systolic, diastolic and mean pulmonary arterial pressures. Methods: In a prospective study of 12 children with complex congenital heart disease in whom a B-T shunt was the sole source of pulmonary blood supply, simultaneous cardiac catheterization and Doppler evaluation of shunt flow velocity profiles were carried out. Pulmonary artery pressure was estimated using the modified Bernoulli equation, and results were correlated with the catheter derived mean pulmonary venous wedge pressure. Using the time velocity integral of shunt flow, Doppler estimates of pulmonary blood flow were correlated with calculated pulmonary blood flow using the Fick principle. Results: There was a positive correlation between 1) The Doppler estimates for mean pulmonary artery pressure, using the diastolic flow velocity and the mean pulmonary venous wedge pressure (r=0.928, SEE=1.17mmHg, $p<0.001$) and 2) The Doppler derived and calculated pulmonary blood flow (r=0.903, SEE=0.19 liter/min; $p<0.001$). Conclusion: Continuous wave Doppler evaluation of B-T shunt flow velocity profiles provides an accurate non invasive and reproducible estimation of pulmonary artery pressure and pulmonary blood flow in patients with a B-T shunt as the sole source of pulmonary blood supply.

344
Assessment of pulmonary regurgitation in adults with repaired Tetralogy of Fallot – comparison between Doppler-echocardiography and MRI
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Background: Pulmonary regurgitation is a common complication after repair of Tetralogy of Fallot and results in chronic right ventricular volume overload, dysfunction, and arrhythmia. It has been recognized as a cause of morbidity and even mortality. Currently there is no gold standard technique for assessing pulmonary regurgitation severity in these patients. Methods: We studied 20 asymptomatic patients (aged 21±10 years, 23±5 years after initial repair, 8 females) using Doppler- echocardiography and compared with MRI. Right ventricular end diastolic dimension was taken from a frozen image of the parasternal view from the continuous wave Doppler trace, pulmonary regurgitation was classified as mild when pressure drop was maintained during diastole, moderate when equilibration between pulmonary artery and right ventricular pressures occured in late diastole and severe when in mid-diastole. Also, the ratio between pulmonary regurgitation duration and total diastole was used to assess the degree of regurgitation: a ratio =>85% for mild, 65–85% for moderate and =<60% for severe. Pulmonary regurgitant fraction was assessed using MRI, 0–15% for mild, 15–30% for moderate and =>30% for severe. Results: 8 patients were found to have severe regurgitation by the two techniques. Echo confirmed 3/4 patients with moderate regurgitation and the remaining had mild regurgitation by the two techniques (r=0.64, p<0.01). Doppler pulmonary regurgitation duration/diastole ratio correlated with MRI regurgitant fraction (r=-0.54, P=0.03). Echo assessed right ventricular end-diastolic dimension correlated with MRI end-diastolic volume index (r=0.68, P=0.005) and end-systolic volume index (r=0.61, P=0.02). Conclusion: Significant pulmonary regurgitation is commonly seen in asymptomatic patients with repaired Tetralogy of Fallot. This and its effects on right ventricular dimensions can equally be assessed by echo and MRI techniques.

345
Ventricular septal defects visualized by three-dimensional echocardiography
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Echocardiography is the major diagnostic tool in pediatric cardiology. Currently 3-D-echocardiography is increasingly available. We report our experience with 3-D reconstruction of intracardiac malformations, particularly VSDs. 3-D dataset acquisition was performed under sedation at the end of cardiac catheterization. We used a Hewlett Packard Sonos 5500 echomachine with the HP transthoracic transducer S3012 using a rotational scanning method for data acquisition. The video output was transferred to a Tomtec reconstruction system (Echo scan 4.2, Tomtec, Munich, Germany). 3-D images of various VSD types were reconstructed, ie. perimembranous VSD by LV en face views and new cutplanes, doubly committed VSD by LV and RV en face views showing the distance between the upper border of the VSD and both the aortic and the pulmonary valve rings, AV valve anatomy in patients with AVSD, VSD morphology in patients with tetralogy of Fallot. The results were compared with intraoperative findings. The learning curve for data acquisition dropped from 35% of the 2-D images not usable for 3-D reconstruction at the beginning of the study to 10% at the end. Intraoperative findings confirmed the results of 3-D echocardiography, particularly area measurement and shape visualization of VSDs and relation to other cardiac structures. In all patients 3-D echocardiography is a useful diagnostic tool in determining position in addition to 2-D scans, size and shape of different VSDs and their relation to cardiac structures in children with complex congenital heart disease.

346
Utilizing intraoperative transesophageal echocardiography to predict future neo-aortic valve function after the Ross procedure
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The Ross procedure (Ross) is an attractive alternative to mechanical prosthesis and homograft valves in the treatment of aortic valve disease in children and adolescents. The ability of the intra-operative transesophageal echocardiogram (TEE) to predict post-operative neo-aortic valve function after the Ross has not been evaluated. The purpose of this study was to determine how the intra-operative TEE correlated with the pre-discharge and follow-up transthoracic echocardiogram (TTE). All patients who underwent the Ross between 1/95 and 9/00 (n=88), who had an intra-operative TEE, a pre-discharge and follow-up TTE were eligible for inclusion. Eighty-six patients fit entry criteria. Median age at Ross was 9.7±2.6 yrs. Median time to pre-discharge TTE was 4 days (1–35 days) and median time to follow-up TTE 2.5 yrs (1 mo–5.5 yrs). No patient had more than mild neo-aortic insufficiency (neo-AI) noted on the intra-operative TEE. However, 4 patients (5.5%) had at least moderate neo-AI noted on discharge TTE, and 14 patients (16.2%) had at least moderate neo-AI at most recent follow-up TTE. During follow-up 6 of these 14 patients underwent replacement of the neo-aortic valve. All 4 patients with mild abnormalities of the pulmonary valve prior to the Ross required replacement of the neo-aortic valve, despite intra-operative TEE that suggested adequate function. Two of the 8 patients who had previous ventricular septal defect (VSD) repair required replacement of the neo-aortic valve due to distortion of the pulmonary annulus. Significant neo-AI on intra-operative TEE after the Ross is uncommon. Intra-operative TEE findings do not predict the degree of follow-up neo-AI. If the patient has abnormalities of the native pulmonary valve or annulus due to congenital defect or secondary to previous VSD repair.

347
Doppler evaluation of aortic regurgitation in children
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Doppler indexes have been used successfully to determine the severity of aortic regurgitation (AR) in adults but have not been evaluated in children. To determine the accuracy of Pulsed, Color, and Continuous-wave (CW) Doppler echocardiographic indexes in assessing the degree of AR in children, the correlation between the non-invasive measurements and angiographic grading of the regurgitant flow (1+ to 4+) was examined in 14 children (mean age: 11 ± 3 years) with chronic AR. Forward and reverse flows in the aortic arch were evaluated from the suprasternal notch using pulsed

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Doppler Aortic time velocity integrals (TVI) were measured during systole (forward flow) and diastole (reverse flow), and the ratio of reverse to forward TVI (%) was calculated. Doppler color flow mapping was used to detect and assess the severity of AR (which appears as chaotic turbulent signals extending in the left ventricular outflow tract during diastole) by using four color Doppler grades of severity. The envelope of the flow velocity pattern in diastole was recorded from the CW Doppler signal of AR with the transducer in the lower sternal border to determine the peak flow velocity and deceleration slope indexes. The ratio of reverse to forward aortic TVI and Color flow mapping grading showed strong correlation with angiographic grade (r= 0.89 and r= 0.87, respectively) but AR slope and peak flow velocity did not correlate well with the angiographic grade (r= 0.44 and r=0.72, respectively). We concluded that the severity of AR in children as determined by angiographic grading can be estimated with reasonable accuracy by noninvasive techniques based on color and pulsed wave Doppler. Use of these indexes may obviate the need for angiography to detect the severity of AR in Children.

348

Stress echocardiography and selective coronary arteriography in patients with transposition of the great arteries (TGA) and arterial switch (ASO)

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Since 1996 in our Department patients with TGA and ASO underwent Stress echocardiography and selective coronarography to identify myocardial ischemia caused by abnormalities of the coronary arteries. Forty-nine patients, mean age 6.1 years (range 4 months–21 years), mean weight 21.1 Kg (range 6–61), underwent Dypiridamole echo and selective coronarography under general anesthesia 4 months–17 years (mean 6.1 years) after operation. Dypiridamole echo was well tolerated without complications. In 47 patients (96%), including 2 cases with an intact Basal ECG and 13 with new abnormalities of segmental motility, no ischemic regional dysinergies were induced by Dypiridamole. In this 47 patients no coronary lesions were detected by coronarography. Dypiridamole echo was positive for ischemia in 2 pts: one patient did not show coronary stenosis. Dypiridamole scintigraphy in this patient demonstrated segmental perfusion alterations, probably caused by microcirculation abnormalities. In the other patient, occlusion of ramifications of the right coronary artery and severe stenosis of the anterior descending were identified. The sensibility and specificity of Dypiridamole echo were 100% and 98% respectively. To test the myocardial regional contractility under the effect of a pharmacological agent inducing ischemia with a different mechanism from Dypiridamole, 11 pts with negative Dypiridamole echo and coronarography, mean age 12±3 years (range 9–14), mean weight 41±16 Kg (range 27–63), underwent Dobutamine echo 4 years±6 months (range 3–5 years) after the previous evaluation. None of the 11 cases showed ischemic dysinergies induced by Dobutamine, that was well tolerated without complications. Our experience demonstrates the feasibility of Dypiridamole echo and Dobutamine echo in children and the good correlation of both tests with coronarography. Stress echocardiography is the method of choice for selection of candidates to coronarography in the follow up of patients.

349

Assessment of the atrioventricular junction in atrioventricular septal defect by three-dimensional echocardiography

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25 children with an atrioventricular septal defect were investigated by three-dimensional echocardiography. In each case there was a common atrioventricular junction guarded by a common valve, with four or five leaflets. In 15 cases, the superior and inferior bridging leaflets were separate structures with the common junction guarded by a common valve orifice. In 10 cases, a tongue of valve tissue joined together the facing surfaces of the bridging leaflets, producing separate valve orifices. In 5 of these, the bridging leaflets were attached to the crest of the ventricular septum so that only an interatrial communication was present. In 5 patients, the bridging leaflets and tongue attached to the underside of the atrial septum, giving rise to only an interventricular communication. In 2 patients with two valve orifices both interatrial and interventricular communications were observed. There were 2 patients in whom there was no interventricular or interatrial communication. The left ventricular component of the common valve had three leaflets in 22; in 3 cases the left mural leaflet was absent, giving the left part of the valve a bileaflet configuration. Four patients had a dual orifice left component caused by the joining together of adjacent leaflets by anomalous bridging valve tissue. Where three-dimensional echocardiography was compared with conventional echocardiography, the latter failed to demonstrate absence of the mural leaflet in 2 cases and a dual orifice arrangement in 2. Three-dimensional echocardiography may lead to the detection of additional abnormalities, which may have an influence on surgical treatment.

350

The impact of intraoperative transesophageal echocardiography on reinstitution of cardiopulmonary bypass following surgery for congenital heart disease

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Outcome following repair of congenital heart disease continues to improve. Intraoperative transesophageal echocardiography (IOTEE) is commonly used to assess the adequacy of repair of congenital heart disease. We sought to evaluate the impact of selective IOTEE on the decision to return to cardiopulmonary bypass (CPB) at our institution. From June 1998–Oct 2000, 1125 procedures using CPB were performed. IOTEE was requested in 474 (42%) cases. 170 (35%) septal defect corrections, 88 (19%) valve procedures, 86 (18%) Tetralogy of Fallot repairs, 28 (6%) outflow tract reconstructions, 25 (5%) corrections for transposed great arteries, 17 (3.5%) palliative procedures for single ventricle, and 36 (8%) other. Of those who underwent IOTEE, 60 (12.6%) had reinstitution of CPB. Reasons for returning to CPB, as identified by IOTEE, included residual septal defects in 12 (20%), valve regurgitation in 11 (18%), outflow tract obstruction in 10 (17%), ventricular dysfunction in 6 (10%), valve stenosis in 4 (7%), venous pathway obstruction in 1 (1.7%), prosthetic leak in 1 (1.7%), atrial baffle leak in 1 (1.7%) and malfunctioning in 13 (22%). Six patients required more than one additional course of CPB, 5 for severe valve regurgitation and one for ventricular dysfunction with mitral and aortic regurgitation. Four patients who did not return to CPB during their initial operation required additional surgery during the same hospitalization. In all four, IOTEE during the first procedure identified residual defects initially deemed acceptable (VSD in 2, and neo-aortic insufficiency in 2); however, the ensuing clinical courses prompted further surgery. In the current era, selective IOTEE plays a valuable role in identifying residual structural and functional defects in a significant number of patients (12.6%) and contributes to the overall excellence in outcome for repair of congenital heart defects.

351

Mechanisms of ventricular function/dysfunction during exercise in post-operative congenital heart disease

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Introduction: Patients with post-operative congenital heart disease (CHD) have been found to have a lower exercise capacity than healthy age-matched peers. Standard treadmill testing does not assess ventricular function. The purpose of this study was to examine the mechanisms of ventricular function and interaction in a group of 32 CHD patients using staged semi-supine cycle ergometry (SSCE). Methods: Twenty-two TET patients (9.8 years old, 8.2 years from surgery), 7 TGA (6 arterial repair/1 Rastelli (10.9 years old, 10.1 years from surgery)), and 3 Fontan (14.9 years old, 11.1 years from surgery) underwent SSCE testing. Two-dimensional echocardiographic imaging in parasternal long, short, apical four, long axis and 3 chamber views were performed during exercise to assess ventricular function and wall motion. Normal wall motion response was defined as increased contractility with exercise with movement in the appropriate direction. Results: Twelve TET patients had resting RV and septal wall motion abnormalities, but contractility improved with decreased RV area during exercise (Gp1A). Five showed decreased contractility with decreased RV area during exercise (Gp1B), while five showed no change in contractility or RV area during exercise (Gp1C). Two atrial repaired TGA patients had increased RV and septal contractility with increased RV area during exercise (Gp2A); 2 showed decreased contractility and area change (Gp2B), and 2 showed no change in contractility but increased RV area (Gp2C). The TGA Rastelli patient responded similarly to the Gp1A patients. The 3 Fontan patients responded similarly to Gp1C patients showing no change in contractility or RV area during exercise. Conclusions: Staged SSCE with echo-Doppler measurements can be used to demonstrate the presence of ventricular dysfunction, poor contractility, and wall motion abnormalities during exercise in children with post-operative CHD.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

352
Staged semi-supine cycle ergometry stress echocardiography in the evaluation of cardiac function in children with congenital and acquired heart disease
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Standard treadmill stress echocardiography (echo) does not assess hemodynamics or myocardial function during exercise. In order to accomplish this, we have developed a stress echo technique using semi-supine cycle ergometry (SSCE) with incremental exercise in 3 minute stages to volitional fatigue. Simultaneous blood pressure, ECG, and echo-Dopplers are performed before during each stage, and after exercise. Power output, BP, HR, LVEDD, LVES, posterior wall thickness (PWT), and peak aortic Doppler velocity are obtained. SF, MVCFc, stress at peak systole, VTI, ejection time, stroke volume index, and cardiac index are calculated. Segmental wall motion is assessed using ASE criteria. We have tested a total of 91 patients and 12 normal subjects using this technique. There were 32 patients assessed for cardiomyopathies (19 following anthracycline treatment, 4 dilated, 3 post viral, 2 with haemachromatosis, 2 hypertrophic and 2 miscellaneous). Twenty patients were assessed with valve disease, 6 had stenotic valves (5 aortic, 1 mitral) and 14 had regurgitant valves (6 aortic, 5 mitral, 2 Ebstein's, 1 mixed). Six transplant patients, 5 post-Mustard TGA patients, and 6 miscellaneous patients were also assessed. Normal values included workload=1740 joules•c-1; HR=188 bpm; a change in SF>10%; an increase and plateauing of SVI, and an increase in CI by 2–3 L•min-1 from resting value. In normals, wall motion increased in all segments with exercise. Echo was successful in more than 90% of patients. Abnormal responses in SVI, CI, contractility, and segmental wall motion were obtained in all patients. Significant stenoses were found in aortic and mitral valves and the SVC of post-operative Mustard patients. SSCE is an excellent technique for evaluating hemodynamics, myocardial function, wall motion, and can be used to unmask stenoses during exercise.

353
Impact of staged procedures on right ventricular size and function for hypoplastic left heart syndrome (HLHS): three-dimensional echocardiographic study
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The impact of staged procedures in HLHS on RV systolic function has been difficult to quantify. 2-D echo based measurements on inappropriate assumptions of RV morphology. 3-D echo allows accurate assessment of RV volumes and EF not dependent on geometric assumptions. We measured RV volumes and EF after staged surgical procedures by 3-D echo in patients with HLHS. 3-D echo RVEDV, RVESV and EF were performed in 30 HLHS patients on 35 occasions. 3-D echo sequential acquisitions were obtained by subcostal rotational scanning. 3-D measurements were performed on a dedicated 3-D echo system (DimEcho). Results expressed as mean +/- SD (Table). ANOVA of EDV and EF revealed no statistical difference between groups, p=0.19. However, EDV was different following stages I and III, p=0.03, with no difference in EF, p=0.1. Comparison of stage III and controls revealed no difference in EDV, p=0.12, but a significant difference in EF p=0.0002. 3-D echo demonstrates that patients with HLHS following Stage I have higher RVEDV than following Stage III, with no difference in EF. Patients following Stage III have similar EDV than normal controls, but lower EF. We speculate the changes reflect significant alterations in pre-load, and EF remains depressed from increased afterload and other presently undefined factors.

Session 32: Transplantation

354
Outcomes following repeat cardiac transplantation
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BACKGROUND: Cardiac transplantation is a safe and effective strategy for severe heart failure and congenital heart disease in children. Over time, accelerated graft coronary disease (CAD) and dysfunction may require repeat transplantation (RT). Information regarding the outcomes after RT is limited. The purpose of this study was to evaluate graft survival and morbidity in children following RT. METHODS: Data were reviewed from 388 pediatric heart transplant recipients from 11/85 to 11/00. RT secondary to CAD or hemodynamically significant graft dysfunction occurred in 17 patients. Seventeen age, sex, and era (date of transplant) matched patients who were undergoing initial transplant served as controls. All values are mean ± standard deviation. RESULTS: Mean age was 12.0 ± 2.9 yo for RT recipients and 12.1 ± 3.7 yo for controls. Six year Kaplan-Meier actuarial graft survival was nearly identical in RT vs controls (71% v 77%, p=0.46, see graph). No statistical differences were noted between the two groups in freedom from rejection, freedom from hospital readmission after initial discharge, and freedom from serious infection. Mean length of stay after RT was 19.4 ± 10.7 vs 14.6 ± 6.8 days for controls (p = 0.08). CONCLUSIONS: Repeat cardiac transplantation in pediatric patients for CAD or hemodynamically significant graft dysfunction demonstrates similar graft survival as initial transplantation. In addition, no increased morbidity is noted as assessed by rejection, serious infection, or hospital readmission.

355
Sustained elevated concentrations of cardiac troponin T during acute allograft rejection after heart transplantation in children
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The diagnosis of acute allograft rejection after heart transplantation is still heavily dependent on endomyocardial biopsy. In children, where the method may be technically difficult and associated with complications, a non-invasive technique would be desirable. The present study evaluated the myocardial damage marker cardiac troponin T as a marker for rejection in heart transplanted children. Serum from peripheral venous blood was collected at 124 endomyocardial biopsies in 14 heart transplanted children (1–20 years). Serum levels of troponin T was compared to histological rejection according to the International Society of Heart and Lung Transplantation (ISHLT (0–4). Nine episodes of rejection ISHLT 3 were found in 7 children. During rejection troponin T increased (from 0.05±0.07 (mean±SD) to 0.20±0.23 µg/l and remained elevated at 7 and 20 days thereafter (0.13±0.11 and 0.36±0.28 µg/l, respectively) before returning to normal 30–450 days after rejection. In surveillance biopsies, there was a considerable variation in troponin T at all rejection grades (ISHLT 0: 0.04 (0.01–2.04) µg/l (median(range)), ISHLT 1: 0.06 (0.01–0.67) µg/l, ISHLT 2: 0.08 (0.01–1.42) µg/l, ISHLT 3: 0.17 (0.01–0.93) µg/l). A receiver operating characteristics curve for troponin T versus biopsy ISHLT rejection grade revealed an area under the curve of 0.71 indicative of a moderate predictive value for troponin T. However, using a cut-off of 0.015 µg/l, yielded a specificity as low as 39%, with a sensitivity of 89%, while a cut-off of 0.1 µg/l resulted in a sensitivity of 53% and a specificity of 77%. Thus, we found that troponin T increased and remained elevated for at least one month during acute rejection. However, the diagnostic power for a single troponin T measurement was not sufficient to replace endomyocardial biopsy.

356
Pacing in pediatric heart transplantation recipients
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BACKGROUND: Electrophysiologic abnormalities are commonly noted after heart transplantation (HT) resulting in pacemaker implantation in 6–21% of adult HT recipients. This study examines the need for pacing in pediatric HT recipients. METHODS: Records were reviewed from 368 children who underwent HT at our institution from 11/85 to 11/00. Kaplan-Meier and Chi square analyses were performed. All numbers are mean ± standard deviation. RESULTS: 15/368 (3.4%) HT recipients (1.1± 2.3 y.o.) received permanent pacemakers. HT was performed due to cardiomyopathy in 4 patients (pts) and congenital heart disease in 9 (5 infants). 9/15 (70%) patients have survived for 3.5 to 13 years. No difference (p=0.6) was noted in 10 yr survival between patients with (78%) or without pacemakers (67%) (see graph). Pacemakers were implanted 2.8± 3.5 yrs post transplant. Indications for pacing included symptomatic sinus node disease in 7/15 (53.8%) & complete heart block (CHB) in 6/15 (46.2%). Two pts developed CHB after severe rejection & one after cardiac cath. Implants were epicardial in 6 pts and transvenous in 8 (one originally epicardial). Complications included: pace site infection in 3 pts & lead fracture in two. No association was noted for pacemaker implant with cold ischemia time, rejection, age at transplant, nor coronary disease. CONCLUSIONS: Pediatric HT recipients

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

have a lower incidence of permanent pacemaker implantation than reported in adult recipients. Need for pacing does not adversely affect survival in these children.

357

Mycophenolate mofetil (MMF) allows reducing dosage and nephrotoxicity of immunosuppressive therapy without leading to graft rejection

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Nephrotoxicity of cyclosporine (Cy) and tacrolimus (Tac) is a major problem after heart transplantation (HT). The purpose of our study was to determine whether introduction of MMF allowed a significant reduction of Cy or Tac dosage without leading to acute cardiac rejection. Seven patients with renal failure were included in the study 103 ±30 months after HT. MMF was started at 500 mg/m2 bid for one week, then increased at 600 mg/m2 bid, whereas azathioprine was discontinued and Cy (5/7) or Ta (1/7) dosage was reduced by 50%. Age at HT was 8.1±5.3 months. Data prior to inclusion were as follows: serum creatinine 111±38 µmol/L, inulin clearance 90 ±30 mL/min/1.73 m2 (N=106±17). Cy trough level 155±21 ng/ml. Serum creatinine was 94±26 µmol/L at 4.6±2.7 months after modification of therapy (p=0.02 vs creatinine prior to MMF). Gastrointestinal side effects were observed in 2/7, leucopenia in 2/7. Endomyocardial biopsy was obtained one month after the therapeutic modification in all patients, showing grade 0 in 5/7, grade 1 in 2/7. No changes in graft function were observed. Our data show that a fifty percent reduction in cyclosporine or tacrolimus dosage, associated with introduction of mycophenolate mofetil (MMF), decreases nephrotoxicity and does not result in acute rejection in heart transplant recipients.

358

Presurgical management of infants with hypoplastic left heart syndrome

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Background. Hypoplastic left heart syndrome (HLH) remains a difficult clinical problem. Survival to either cardiac transplantation or staged palliation has been problematic. HLH was defined as a condition with an inadequate left ventricle requiring duct-dependency. All patients were treated in an excise-to-transplant protocol. We reviewed all infants with HLH listed for cardiac transplantation from 1994 through 1999 in order to determine the survival to definitive treatment. Our approach has included inhaled nitrogen in patients with high oxygen saturations as well as a minimally invasive regime downplaying intubation, inotrope support, and length of ICU stay. Methods and Results: We conducted a retrospective chart review of all patients with HLH at The Children's Hospital of Denver listed for transplantation from June, 1994 through June, 1999. Thirty-eight of the children (84%) survived to transplantation. Orthotopic cardiac transplantations were performed at 6–166 (median 80.5) days of life. Inhaled nitrogen was used in 12 (93%) of the children, and very few patients ultimately ended up on supplemental oxygen. Improved oxygen saturation was achieved by late atrial septostomy in 13 (29%) of the patients at 5–204 (median 56) days of life. Less than 25% of the infants required inotropic support and 90% were extubated but only for 1–22 (median 3) days. Eighteen children (40%) were stable enough to be discharged home on a prostaglandin infusion prior to receipt of a donor heart. Seven patients died while listed for transplantation. Two of these deaths were attributable to late surgical intervention. Conclusions: These data indicate that infants with HLH can be successfully managed to the time of cardiac transplantation with minimal intervention.

359

Cardiothoracic transplantation for congenital heart disease

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Since 1980, 239 patients aged between 8 days and 52 years have undergone cardiothoracic transplantation (tx) at Harefield Hospital for congenital heart disease. Ninety underwent heart tx, 145 heart lung (H/L) tx and 4 bilateral lung transplant with repair of the underlying defect. Patients undergoing heart tx either had complex anomalies with a low pulmonary artery pressure (n=66) or myocardial failure after previous operation (n=24). Twenty of the patients undergoing H/L tx had complex pulmonary atresia and 123 congenital heart disease (CHD) with pulmonary hypertension (PHT). Four patients with correctable lesions had bilateral lung tx. The type of operation undertaken has been modified during the series to take account of the increasing scarcity of donors. Operative techniques are adapted according to the complexity of the anatomy. In-hospital mortality was 36/90 (40%) for hearts, 46/125 (37%) for H/L tx with CHD and PHT and 1n/20 (80%) for complex pulmonary atresia. There were no deaths in 4 patients undergoing lung tx and repair. Major risk factors for early death were age greater than 25 years and previous lateral thoracotomy in the H/L tx group. Actuarial survival in the heart tx group was 58%, 53% 44% at 1 year, 5 years and 10 years respectively and 60%, 45% and 36% in the H/L tx group. It is concluded that risk of early death is greater in patients with complex congenital heart disease undergoing heart tx than in patients with acquired heart disease. Thereafter survival in both groups of patients is similar to other patients undergoing heart and heart-lung transplantation.

360

Ten years of heart transplantation in children: long term renal function assessment and outcome

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Out of 102 children who underwent heart transplantation (HT) between Jan 1987 and DEC 1997, 42 have been followed for at least 3 years and have been assessed for long term renal function. The immunosuppression protocole included antilymphocyte globulins for 5 days, corticosteroids for 6 months, azathioprine and steroid. The mean age at time of transplantation was 8 +-5.2 years. Three patients died 3.5 and 12 years after HT and 4 were retransplanted. The mean duration of follow-up was 6 +- 3 years. Renal function was assessed by yearly determination of inuline clearance and urine concentrating ability following the administration of DDAVP. The mean inulin clearance was 85+-26 ml/min/m2 at 1 year, 81+-22 ml/min/m2 at 5 years and 58+-13 ml/min/m2 at 10 years while maximum urine osmolality was 750+-170 mosm/kg at 1 year, 669+-175 mosm/kg at 5 years and 612+-90 mosm/kg at 10 years. There was no correlation between the decrease of inuline clearance and the dose of cyclosporine received. The decline of renal function is associated with tubulointerstitial lesions with or without arteriolar lesions in 15 children who underwent renal biopsies. In conclusion, children with HT receiving cyclosporine show a progressive decline of renal function.

361

Plasma homocysteine levels in paediatric heart transplant recipients

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Background: Graft coronary artery disease (CAD) is a major determinant to long-term survival in paediatric heart transplant recipients. Elevated homocysteine (HCY) levels have been demonstrated to be an independent risk factor for CAD in the general population, and have been shown to be present in adult heart transplant recipients. To date, there is no published literature on plasma HCY levels in the paediatric heart transplant population. Methods: In this cross sectional study, 32 paediatric heart transplant recipients had fasting blood samples analyzed for HCY, vitamin B12 and folate. Associations were explored between HCY level, renal function, medications, and the presence of CAD. Results: Median age at transplant was 4.7yrs (1 day-17yr); Median time post-transplant at time of HCY level was 1.4yrs (7 days-5.9yr). Median HCY level was 8.7 µmol/L (4.4–40 µmol/L) with 14 patients (44%) having an elevated HCY level. Elevated HCY levels were associated with lower serum folate level (p=0.02), use of amlodipine (n=7, p=0.03), and use of nifedipine (n=9 p=0.04). There was no association with age at transplant, time post-transplant, serum B12, creatinine, or lipid levels, glomerular filtration rate, use of trimethoprim/sulfamethoxazole (n=26), or choice or dose of immunosuppressant agents. 6/31 (19%) had graft CAD by angiography. Kaplan Meier estimates of freedom from CAD were 96% at 1 year, 84% at 2 years, 76% at 3 years, and 51% at 5 years. 5/24 (21%) had an abnormal dobutamine stress echocardiogram (DSE). Neither the presence of CAD nor an abnormal DSE were associated with an elevated HCY level. Summary: HCY levels were elevated in 44% of paediatric heart transplant recipients. Though the cohort was small and there was no direct association with the presence of graft CAD, elevated HCY was prevalent and further study is required to determine the significance in this patient population.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

362
Dobutamine/atropine stress echocardiography: feasibility, safety and early results in paediatric heart transplant recipients
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Purpose: a) To assess prospectively the feasibility and safety of dobutamine/atropine stress echocardiography (DSE) in paediatric heart transplant recipients, and b) To review early results in comparison to the presence of coronary artery disease (CAD) by angiography and graft survival. Methods: In this prospective study, 44 DSEs were undertaken in 24 patients. Data collected included resting and peak heart rate (HR) and blood pressure, dose of dobutamine (DB), requirement for atropine (ATR), presence of changes on electrocardiogram (ECG), presence of CAD by angiography, and patient outcome. Results: Median age at transplant was 5.6 yrs (1 day-16.7 yrs). The median time from transplant to first DSE was 1.8 yrs (0.5–10.6 yrs). Looking at all 44 DSE studies, the mean dose of DB was 40+/-10 ug/kg/min. Atropine was required in 32 (80%). The mean double product at peak was 21633+/-4900 beats-mmHg/min (range 14820–30446). Target HR was achieved in 38 (86%). No ECG changes were detected in 41 (93%). 2/44 (5%) required termination: 1 for atrial flutter and 1 with asymmetric septal hypertrophy who developed a left ventricular outflow tract gradient of 100 mmHg. Looking at all 24 patients, 6/24 (25%) had an abnormal DSE with 3/5 (60%-1 pending) having CAD by angiography. 7/24 (26%) had wall motion abnormalities at rest with only 3/7 (43%) having an abnormal DSE. 7/24 (21%) had CAD by angiography with 3/7 (43%) having an abnormal DSE. All 3 deceased patients had normal DSEs and causes of death were unrelated to CAD. The 1 retransplanted patient had CAD and an abnormal DSE. 1/9 patients developed progressive worsening on serial study with progression of CAD by angiography. Conclusion: Technically adequate DSE may be performed safely in paediatric heart transplant recipients and serial DSEs may play a role in the assessment of graft CAD. Long term, sequential follow-up is required to truly evaluate the impact of DST on patient management and outcome.

363
Parental perception of quality of life following pediatric heart transplantation
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Heart transplantation (HTx) has become standard of care for children with end stage heart disease. As overall survival improves, the assessment of quality of life (QOL) in this population becomes critical. The Child-Health-Questionnaire (CHQ) was sent to the parents of 49 pts(3–26 yrs) who underwent HTx at a single center. The CHQ contains 50 questions regarding physical and psychosocial health status and generates summary scores for physical, emotional, psychological, behavioral and family functioning. The results were compared to age-matched U.S. normative data from healthy controls. Results: Thirty-two families (80%) responded. Median age was 10.5 yrs. There were 19 boys (54%). Median time since HTx was 7 yrs (range 4 mo-13 yrs). Nineteen pts were transplanted for cardiomyopathy and 16 for congenital heart disease. Twelve pts (35%) were off steroids. The parent-reported mean physical functioning and psychosocial summary scores for the HTx pts were significantly lower than those of healthy controls(44.3 vs 55.0,p<0.001 and 43.8 vs 51.2 p=0.002). Student's t-test revealed that 3 of 13 subscale scores were highly significant: (1)social-emotional subscale, a measure of limitations in school work and activities due to emotional and behavioral difficulties(74.6 vs 92.5, p<0.001), (2) parental emotional impact subscale, a measure of parental distress and worry (58.8 vs 80.1, p<0.001), and (3) family activity subscale, a measure of frequency in disruption in usual family life (66.9 vs 89.7,p<0.001). There was no association between QOL scores and time since HTx, age at HTx, age at survey, or preHTx diagnosis. Pts currently taking steroids tended towards a lower physical functioning score. Conclusion: Based on parent-reported data, child QOL as measured by physical functioning and psychosocial summary scores is significantly lower than that of healthy controls. The largest impact appears to be on the behavioral and emotional limitations of the child as well as the emotional distress on the family. Prospective studies are needed to control for pre-HTx QOL. Interventions aimed at supporting the emotional impact on the patients and their families are indicated.

364
Use of ECMO as a bridge to heart transplantation in children
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Heart transplantation is an accepted option for end-stage cardiac disease in children. Mechanical circulatory support is sometimes required prior to transplantation. Extra-corporeal membrane oxygenation (ECMO) is the only option available for mechanical support in most children. We reviewed the use of ECMO as a bridge to heart transplantation from November, 1994 to June, 2000 in one institution. During this period, patients were listed for heart transplantation 136 times resulting in 85 transplants. ECMO was used for circulatory support in 31 patients listed for primary transplant (re-transplantation excluded). Of these 31 patients, 6 were successfully weaned from ECMO, and survived to discharge. Of the remaining 25 patients, 12 were successfully bridged to transplant, while 13 developed complications rendering them non-candidates for transplant and died nor survive to discharge. Median ECMO duration for transplanted patients was 281 hours (range 2–1127) vs 123 hours (26–378) for the 13 who were not transplanted (p=0.04). Average weight for transplanted patients was 20 kg vs 4.6 kg for those not transplanted (p<0.001). Twelve month post transplant actuarial survival for patients bridged to transplant was 83% (n=12) compared with 95% for all primary heart transplants performed during this period (n=99, p=0.28). These data demonstrate the utility of ECMO as a bridge to heart transplantation in children. Many children can be supported until a donor heart becomes available, however, the morbidity and mortality associated with ECMO can render some patients ineligible for transplantation. Overall survival rates for children successfully bridged to heart transplant are comparable to non-bridged patients; a change in organ allocation policies so as to transplant these patients as soon as possible could result in improved outcomes for this critically ill group of patients.

365
Outcome of children listed for heart transplantation: a 10-year experience
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We reviewed a single institution's experience with the aim of describing the outcome, and potential determinants of outcome of children listed for orthotopic heart transplantation (OHT) in the highest status. Outcomes examined were survival to transplant, and actuarial survival after a first transplant. Potential determinants of outcome, including age, sex, year of listing, pre-transplant diagnosis (cardiomyopathy (CM) vs congenital heart disease (CHD)), and hemodynamics were evaluated by logistic or Cox regression. Between 1991 and 8/00, 132 children were listed for OHT as status 1 or 1A candidates. 12 of them responded to medical management and were removed from the list. Among the remaining 120 patients, 72 had CHD and of these, 24 were listed immediately after failed reconstructive surgery. 28 patients had pulmonary vascular resistance (pvr) > 4 wood units. Children with CHD listed for OHT immediately after failed reconstructive surgery had worse survival both before (odds ratio: 3.32,p=0.09) and after (odds ratio: 0.4,p=0.01) transplantation. There was a trend toward higher pre-transplant mortality among those with worse hemodynamics, including higher right atrial pressure (p=0.14) and higher pulmonary capillary wedge pressure (p=0.06). Median survival of the 88 children who received OHT was 9.8 months, with an overall survival of 73% at 1 year and 60% at 5 years. Actuarial survival after OHT was better among those with CM as compared with CHD (median survival > 125 months vs 42 months,p=0.047), even after excluding patients listed in the immediate postoperative period. In contrast to some previous reports, we found lower survival among children transplanted for CHD as compared with CM. Listing immediately after failed corrective surgery and worse hemodynamics were associated with increased pre-transplant mortality. No other pre-transplant characteristic, including elevated pvr, appeared to influence post-transplant survival.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Session 33: Pulmonary Hypertension, Pulmonology

366

L-arginine regulates apoptosis of pulmonary artery smooth muscle cells in rats with hypoxic pulmonary vascular structural remodeling

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To explore the impact of L-arginine on apoptosis of smooth muscle cells in pulmonary arteries of the rats with hypoxic pulmonary vascular structural remodeling, seventeen Wistar rats were randomly divided into hypoxia group (n=5), hypoxia with L-arginine group (n=5) and control group (n=7). Hypoxic challenge was performed by putting the rats into a normobaric hypoxic chamber with an oxygen concentration of 10%±0.5% for two weeks. L-arginine was administrated intraperitoneally at a dose of 500mg/kg/d. Pulmonary vascular morphometrie was measured under a light microscope. Apoptotic smooth muscle cells in pulmonary arteries were detected by TdT-mediated dUTP-biotin nick end labeling, and the expression of Fas protein by pulmonary artery smooth muscle cells was detected using immunohistochemistry technique. The results showed that pulmonary vascular structural remodeling developed after 2-week hypoxia. Meanwhile, the percentage of apoptotic smooth muscle cells to smooth muscle cells in pulmonary arteries was markedly decreased in hypoxic rats compared with normal controls (p<0.05). The expression of Fas protein of hypoxic rats was inhibited obviously. L-arginine ameliorated pulmonary vascular structural remodeling of hypoxic rats in association with an increase in the percentage of apoptotic smooth muscle cells to smooth muscle cells in pulmonary arteries and a strengthened Fas expression by pulmonary artery smooth muscle cells. The results suggested that L-arginine plays an important role in the regulation of development of hypoxic pulmonary vascular structural remodeling through promoting Fas expression and thereby strengthening apoptosis in pulmonary artery smooth muscle cells.

367

Pulmonary artery agenesis and contralateral pulmonary hypertension involution after surgical correction

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Aim: Pulmonary artery agenesis, right or left one, without associated cardiac defects generally presents a diversified clinical evolution due to the contralateral pulmonary hypertension (PH). We plan to relate 2 such cases in whom PH diminished in long-term evaluations after surgical correction. Material and Methods: Male patients, 22 and 11 months old with right cardiac insufficiency (RCI), peripheral oedema and cyanosis in the first patient and RCI, low weight gain in the second patient, besides PH signs in accentuated second heart sound, tricuspid insufficiency murmur, right sided heart overload in ECG, anterior cardiomegaly and dilated pulmonary trunk. Cardiac catheterization revealed systemic pressure at the contralateral pulmonary artery in right agenesis in case 1 and in left agenesis in case 2. There was a reserved cardiac shunt at the oval foramen in case 1. Results: Surgical connection between the pulmonary arteries was possible until the hypoplastic contralateral pulmonary hilum with Gortex tubes of 7 and 6 mm diameter, in both cases. Clinical and hemodynamic PH signs involution was evident at immediate and at long-term follow-up with 4 and 5 years of age respectively. Right / left ventricle relationship was 30 and 40% in both cases. Pulmonary blood perfusion by scintigram increased from 8 to 44% and from 8 to 23% in the two cases, at long-term period. Conclusion: This technique becomes the first approach choice for similar cases, rarely described in the literature, even having an accentuated PH and contralateral pulmonary artery hypoplasia.

368

The internal elastic lamina as a barrier to the migration of smooth muscle cells in secondary pulmonary hypertension: a confocal laser microscopy study in patients with congenital heart defects

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Pulmonary vaso-occlusive lesions appear in the evolution of patients presenting congenital heart defects with increased pulmonary flow. Continuous remodeling of arteries occurs, mediated by growth factors and local enzymes. We investigated the three dimensional morphology of the internal elastic lamina (IEL) in peripheral pulmonary arteries from patients presenting two different types of vascular lesions: isolated medial hypertrophy and intimal proliferation. Methods: Fourteen lung biopsies and 12 necropsy lungs from patients with congenital cardiac shunts and 6 controls were studied using the confocal laser scanning microscope. The mean ages were respectively 15.8, 30.0 and 14.7 months. Sections from paraffin-embedded tissue (30um-thick) were stained with Evan's blue to enhance the fluorescence of elastin. Pre-determined conditions of laser intensity (wavelength = 594 nm), brightness and contrast were used to examine pre and intra-acinar arteries. We obtained 24 serial Z-images for each artery at intervals of 0.5mm. In the image of the median slice we measured the thickness and determined the numbers of gaps of the IEL. Results: The mean thickness was significantly higher (p<0.04) and the number of gaps of the IEL was lower (p<0.02) in arteries larger than 100um in diameter from patients presenting with isolated medial hypertrophy when compared to those with intimal proliferative lesions and to controls. Comparing patients below and over 12 months of age, a significant difference was observed in the group with intimal lesions regarding the IEL thickness (lower values in the younger group) and number of gaps (greater values in the younger patients). Conclusion: These results suggest the IEL acts as a barrier to the migration of smooth muscle cells in some patients (isolated hypertrophy), while in others it suffers early remodeling thus allowing the development of intimal proliferative lesions.

369

Intravenous sildenafil (Viagra®) and pulmonary vascular resistance in children with congenital heart disease

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Background: Increased PVR remains a significant risk factor for perioperative morbidity and mortality in patients with CHD and has been attributed to the failure of the pulmonary endothelium to produce nitric oxide (NO). Inhaled NO and strategies to enhance endogenous NO production have previously proved effective, but their effects are variable. We examined the effects of the phosphodiesterase V inhibitor sildenafil on pulmonary vascular resistance during routine preoperative cardiac catheterisation in children with CHD and pulmonary hypertension. Methods: All patients were sedated, intubated and paralysed throughout the study. After routine haemodynamic measurements and before angiography, measurements were made during ventilation at low FiO_2, at FiO_2 0.6 (n=5), to which inhaled nitric oxide (20ppm) was added before and after the administration of intravenous sildenafil at 2 doses (0.35mg/kg/10 min, and 0.6mg/kg/10 min, see figure). PVR was measured according to the direct Fick principle using respiratory mass spectrometry for the measurement of oxygen consumption. Results: Sildenafil produced a greater fall in PVRI than NO (p<0.05). The effect was especially pronounced in patients with moderately raised PVRI. Conclusion: Sildenafil is a potent pulmonary vasodilator which may be an additional option in the treatment of pulmonary hypertension.

370

Pulmonary endothelial dysfunction (PED) after cardiopulmonary bypass in infants: impact on postoperative recovery

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Background: Overt or subclinical pulmonary endothelial dysfunction (PED) is demonstrable in most children after congenital heart surgery. We aimed to define the clinical impact of PED on postoperative recovery in infants after repair of ventricular or atrioventricular septal defect with cardiopulmonary bypass (CPB). Methods: Pulmonary vascular resistance (PVRI) was measured in 15 infants (age, median 0.31 years; weight, median 5.1 kg) in the immediate postoperative period using respiratory mass spectrometry during the following study protocol to evaluate PED: ventilation with high FiO_2, with sequential addition of infused L-arginine (L-Arg) and Substance P (sub P), and inhaled nitric oxide (iNO). Duration of mechanical ventilation was defined from the end of CPB to successful extubation. Results: PVRI at baseline was 11.7±5.6 WU*m2 and fell to 6.1±3.5 WU*m2. The ventilatory time was 0.86 – 34.9 days (median 1.75 days). The patient group with ventilation > 2 days (n=6) had significantly higher PVRI at all stages of the study protocol (see Figure), as compared with those ventilated < 2 days. Furthermore, there was a linear relationship between ventilatory time and lowest achieved PVRI (r^2 =0.59, p<0.05) and PVR after Substance P (r^2 =0.64, p<0.01). Conclusion: PED, either directly or as a surrogate for the global intraoperative insult, delays recovery after CPB surgery in infants.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

371

Nation-wide survey of primary pulmonary hypertension (PPH) in Japanese pediatric patients

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To investigate the epidemiology and clinical characteristics of pediatric PPH, we surveyed Japanese with PPH by sending questionnaires on 1453 hospitals. [Results] One hundred forty-one pts were reported, encompassing a treatment period from 1/1992 to 10/1997, and analyzed. The M/F ratio was 1:1.4. The mean age of onset was 8.2 y/o, mean age of admission 9.8 y/o, and interval from onset to admission 1.6 years. Incidence of familial PPH was 6.2% with a M/F of 1:1. The incidence of pediatric PPH is believed to be 1/1million/y. Initial symptoms included fatigue(28%), SOB(27%), syncope (4%), abnormal EKG (20%) and CXR(6%). The mean NYHA was 2.2 (I: 22%, II:45%, III:25%, IV:8%). Fifty-six pts died, and 65 pts survived, with a mean survival period was 3.5 y after onset. Causes of death included CHF (58%),sudden death (29%), CRF (5%) and hemoptysis (2%). Twelve pts unimproved after cont. PGI2 therapy.ANA was positive in 14%, anti-phospholipid Ab (13%). Plasma TXB2 470pg/ml, 6-keto-PGF1a 52pg/ml, TXB2/6kPGF1a ratio 3.5 1, ET-1 222pg/ml, hANP 262pg/ml and BNP 428pg/ml. Cardiac cath data revealed (surviving vs deceased) mRAP 6.0 vs 6.3, mPAP 62 vs 71 mmHg, CI 3.0 vs 2.9 L/m/m2, TPR 13 vs 22 unit. Regarding medical treatment, Lasix (74%), Aldactone (63%), oral PGI2 (Beraprost) (60%), home oxygen (52%), digitalis (40%), nifedipine (40%), warfarin (26%), ACE-I(17%), NO inhalation(7%) were administered. There were 17 pts <1 y/o (M/F 12:5, 1). Age at onset was 2.2 mo. Precocial syn. RVP was 70 mmHg, PFO was found in 6 pts. Syn. PAP/AoP was 0.9, TPR ranged 2.7 - 15.4 unit. Ten pts died (mean age of death 7.2 mo, 5.7 mo after onset.) [Conclusion] The natural history of pediatric PPH remains unsatisfactory. Oral PGI2 was some of effect, but is not promising. I.V. PGI2 should be started for NYHA III-IV pts.

372

Dexfenfluramine and the development of pulmonary hypertension: the complexity of mechanisms unmasked in a rat model study

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Dexfenfluramine (DEX), an appetite suppressant and inhibitor of serotonin reuptake, has been associated with pulmonary hypertension (PH). Elastase plays a pivotal role in the development of experimental PH. We therefore hypothesized that DEX induces PH by increasing elastase. We assessed the effect of DEX (2 mg/kg/day) for 15 days in obese, lean, and normal Sprague-Dawley (S-D) female rats under control conditions or following endothelial injury induced by the toxin monocrotaline (MCT) (60 mg/kg). Pulmonary artery (PA) pressure was measured, ratio of right ventricle to left ventricle plus septum (RV_w) was calculated, and % medial wall thickness of muscular arteries and muscularization of peripheral arteries were assessed as indices of pulmonary vascular disease (PVD). Although DEX reduced body weight gain in obese and S-D rats ($p<0.05$), it did not induce PH. The PH, RVH, and PVD observed in S-D but not in obese or lean rats after MCT injection, were paradoxically ameliorated by DEX ($P<0.05$), an effect not attributable to reduced expression of nitric oxide synthases. Neither DEX nor serotonin increased elastase activity in cultured PA smooth muscle cells but in rats, PA elastase was induced by DEX and by MCT with additive effects ($p<0.05$). DEX failed to induce PH in obese, lean or S-D female rats. It prevented rather than aggravated PH after MCT in S-D rats, despite inducing a further increase in elastase. It is possible that DEX blocks a downstream effect of elastase, the generation of which could cause malignant PH. Current candidates for such protective factors include genes involved in the pathway mediated by a bone morphogenetic protein receptor, the mutants of which are associated with primary PH.

373

Improvement of acute hemodynamic response to 100%oxygen after long-term continuous intravenous prostacyclin (PGI2) in patients with primary pulmonary hypertension (PPH)

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Continuous intravenous PGI2 has been shown to improve hemodynamics and QOL in patients with advanced PPH. However, it remains unknown why chronic PGI2 therapy shows beneficial long-term effects irrespective of acute hemodynamic results of PGI2 provocation. Hence, the acute response to 100%oxygen (O2) for 15 minutes were evaluated in terms of changes in pulmonary vascular resistance (PVR) 3 months and 1 year after the initiation of PGI2. Twelve patients with PPH (age 13±6 years, 6 females) in NYHA class III (n=5) and IV (n=7), who have been on continuous intravenous PGI2 for > 3 months, were enrolled in this investigation. Significant (SR), partial (PR) and poor response (NR) to 100%O2 were defined as >20%, 10–20%, and <10% decrease in PVR, respectively. NYHA class improved at 3 months in all but one (class II: 5, class III: 5, class IV: 1, n=12). At 3 months, PVR was 26±11 U•M2, and mean decrease in PVR was 13% (SR: 5, PR: 2, NR: 5). NYHA class has further improved at 1 year (class I: 3, class II: 2, class III: 2, n=7). PVR also fell to 21 ± 11 U•M2, and the mean decrease in PVR was 18% (SR: 3, PR: 3, NR: 1). Furthermore, plasma brain natriuretic peptide (BNP) level and the distance walked in 6 minutes (6MWD) at 1year after PGI2 significantly improved compared to those at the initiation of PGI2 (BNP: 268±278 vs 511±411 pg/ml, $p<0.05$, 6MWD: 410±114 vs 208±129 m, $p<0.001$, respectively). We conclude that long-term continuous intravenous PGI2 may attenuate remodeling of pulmonary arteriopathy resulting in improved acute response to 100%O2, in addition to improvement of hemodynamic parameters with mean air and symptoms in patients with PPH.

374

von Willebrand factor in pulmonary hypertension: what have we learned?

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We have been involved with several aspects of von Willebrand factor (vWF) in the context of pulmonary hypertension. Since findings have potential pathophysiological and clinical implications we planned this report as an attempt to summarize observations. Studies involved 52 patients (37 females) aged 1.2 to 58 (median 29) years with primary (PPH: 12 pts) or secondary (SPH: 40 pts) precapillary pulmonary hypertension, including 29 associations with congenital heart disease (CHD-PH). Analysis of vWF included measurement of plasma antigenic (vWF:Ag) and biological (ristocetin cofactor) activities and assessment of multimeric as well as subunit structure (Western blotting). vWF:Ag was increased in patients vs controls ($p<0.001$) with exceedingly high levels in PPH ($p=0.003$ vs SPH) but unrelated to age or gender group. In contrast, biological activity was decreased in parallel to a profound defect in the multimeric structure ($p=0.004$). Besides, hypoxia was associated with heightened vWF:Ag ($p=0.014$). Improvement of arterial oxygen saturation was followed by a decrease in vWF:Ag levels and partial correction of multimeric defects. Also, lowering of hematocrit by means of hemodilution in patients with Eisenmenger syndrome (CHD-PH) had no effect on arterial oxygen saturation, but did provoke a significant reduction in vWF:Ag levels ($p=0.021$), albeit structural abnormalities persisted. Subunit analysis showed that altered multimeric structure was mainly due to increased proteolytic degradation of vWF in vivo ($p=0.033$) probably associated with decreased sialic acid content of carbohydrate components ($p<0.05$). Finally, high vWF:Ag levels ($p=0.0087$) and the multimeric abnormalities of vWF ($p=0.0276$) were significantly correlated with decreased one-year survival in both PPH and CHD-PH groups. Thus, vWF is involved in a complex network of pathophysiological phenomena in pulmonary hypertension, including endothelial dysfunction, hypoxia, hypercoagulability and proteolysis. Abnormalities, which have impact on short-term prognosis may be used as indexes for therapeutical decision.

375

Chronic prostacyclin therapy in patients with primary and secondary pulmonary hypertension, survival analysis at a single institution

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Introduction: Pulmonary hypertension is a challenging problem in pediatric patients. Prostacyclin (PGI2) has been shown in some studies to improve quality of life and prolong survival. In this study, we analyze our own experience with pulmonary hypertension and chronic PGI2 infusion. Methods: The charts of all patients with pulmonary hypertension evaluated at St. Louis Children's Hospital between January 1990 and October, 2000 were retrospectively reviewed. Patients with left sided obstructive lesions or intrinsic

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

lung disease were eliminated. Remaining patients were divided into two subgroups based on whether they did (group I) or did not (group II) receive PGI2. Results. 82 patients with primary (n=23) or secondary (n=59) pulmonary hypertension were identified. Kaplan-Meier analysis showed no difference (p=0.75) in survival between group I (n=28) and group II (n=54). Analysis of patients with primary pulmonary hypertension alone showed no difference (p=0.41) in survival between group I (n=14) and group II (n=9). Analysis of patients with secondary pulmonary hypertension alone showed no difference (p=0.43) in survival between group I (n=14) and group II (n=45). Analysis of baseline demographic and hemodynamic data showed no significant differences between groups I and II in age at diagnosis (p=0.98), mean pulmonary artery pressure (p=0.3), time to lung transplantation (p=0.57), or time to death (p=0.44). There was a significantly lower cardiac index in group I compared to group II (mean 2.45 vs. 3.3 l/min/m2; p=0.025) and a trend towards greater indexed pulmonary vascular resistance in group I compared to group II (mean 27.7 vs. 26.9 wu, p=NS.0533). Analysis of hemodynamic data in group I at baseline and after initiation PGI2 showed a significant increase in cardiac index (mean 2.53 vs. 3.59 l/min/m2, p=0.035) and a trend towards decrease in indexed pulmonary vascular resistance (mean 14.6 vs. 12.3 wu, p=0.073). There was no difference in mean pulmonary artery pressure after initiation of PGI2 (p=0.18). Conclusion. Retrospective analysis of patients evaluated at our institution with pulmonary hypertension showed no improvement in survival when treated with PGI2. The disparities in baseline hemodynamics may indicate a select patient population chosen to be treated with PGI2 and may have contributed to the lack of difference in survival between the two groups. While there is apparent improvement in hemodynamics associated with PGI2, a prospective, multi-institutional study is indicated to fully define the clinical significance of treating pulmonary hypertension patients with chronic PGI2 infusion.

376

Improved insights in pulmonary hemodynamics by assessment of inducible changes in pulmonary blood flow velocity

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Pulmonary hypertension is characterized by a complex process from impaired endothelial function to irreversible structural changes of the pulmonary vasculature. Therefore assessment of endothelial dependent and independent vasodilation and endothelial biochemical pathways like nitric oxide synthesis may allow more individualized therapeutic strategies. Patients and methods: Pulmonary hemodynamics were examined in 18 children aged 0.5 – 19 years, with established pulmonary vascular disease (6 pts. with primary pulmonary hypertension, 6 pts. with congenital heart defects) or pulmonary hypertension due to increased pulmonary flow (6 pts.). In addition to conventional hemodynamic evaluation, pulmonary blood flow velocity (PBFV) was measured by 0.014-inch intra-arterial Doppler wire (FloWire, Cardiometrics, CA). After graded local infusions of the endothelial dependent vasodilator acetylcholine at adjusted intrafemoral concentrations of 10^{-8} to 10^{-4} M. Nebulized iloprost (1.5 µg/kg, maximal dose 25 µg) was applied to assess preferentially endothelial independent vasodilation. Arginine, citrulline and ornithine were measured by ion-exchange column chromatography from arterial blood samples taken before pharmacological testing. Results: Pulmonary blood flow velocity increased up to 614 % of baseline velocity after acetylcholine infusion. There was an inverse linear correlation between the maximum increase of PBFV and the ratio of systemic-to-pulmonary resistance ($r = -0.565$, $p < 0.005$) with complete lack of endothelial-dependent vasodilation in 6 children with established pulmonary vascular disease. Acetylcholine-induced PBFV was highly predictive for the response of mean pulmonary pressure to nebulized iloprost ($r = 0.72$, $p < 0.005$). In contrast to iloprost induced changes of pulmonary pressures, the ratio of citrulline to ornithine as a marker for intrinsically stimulated nitric oxide synthesis was inversely related to the maximum PBFV increase ($r = -0.43$, $p < 0.05$). Conclusions. In pulmonary hypertension, assessment of PBFV and biochemical endothelial pathways may provide significant information on the integrity of the pulmonary vasculature.

377

Plasma endothelin-1, homocysteine, and nitric oxide values of patients with left-to-right shunt

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Pulmonary hypertension (PH) is associated with increased endothelin-1 (ET-1) levels that correlate with the severity of the disease. There is also diminished expression of endothelial nitric oxide synthase, the enzyme responsible for generating nitric oxide (NO) in patients of the same disease. Homocysteine found in the plasma of patients with coronary heart disease, induces vascular smooth muscles cell proliferation. We studied the effect of pulmonary blood flow (PBF) and PH on plasma ET-1, NO, and homocysteine in patients with left to right shunt: lesions both having PH and normal pulmonary arterial pressure (NPAP). We studied also a group of patients operating because of same disease. Plasma ET-1, homocysteine, and NO were measured. n 44 patients (group 1) with left to right shunt having NPAP (Q_p/Q_s: 2.3); 65 patients (group 2) with left to right shunt and PH (Q_p/Q_s: 2.4); 20 normal control subjects (group 3), and 17 postoperative patients (group 4). Mean concentrations of ET-1 were 1.0/±0.51 pg/mL in group 1, 26.86±7.051 pg/mL in group 2, 11.82±1.9251 pg/mL in group 3, and 151 pg/mL in group 4. ET-1 levels were significantly higher in group 2 than group 1 ($p<0.039$). Mean concentrations of NO were 22.28±15.57 micromol/l in group 1, 30.46±44.94 micromol/l in group 2, 7.24± 45 micromol/L in group 3, and 12.74±9.35 micromol/L in group 4. NO values are significantly higher in group 1 than group 3 ($p<0.005$). Mean concentrations of homocysteine were 8.16±2.85 micromol/l in group 1, 12.65± 4.85 micromol/L in group 2, 10.69±4.27 micromol/L in group 3, and 8.4±2.99 micromol/L in group 4. Plasma homocysteine levels were significantly higher in group 2 than group 1 and 3 ($p<0.001$ and $p<0.01$, respectively). We concluded that an increase PBF alone does not result in an increase in plasma ET-1 and homocysteine levels. But ET-1 and homocysteine levels are increased in patients having PH. NO levels are higher in both groups having PH and NPAP with left to right shunt. In postoperative patients ET-1, NO, and homocysteine levels are lower than those of preoperative patients having PH, but it is not significant because of small sizes postoperative patients.

Session 34: Surgical Management and Results: Abnormal Venous Return, Left Ventricular Outflow Obstruction/Aortic Stenosis, Pulmonary Atresia

378

Outcomes of biventricular repair in atrioventricular septal defects with small right ventricles

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Objectives. We reviewed outcomes of biventricular repair (BVR) in children with a small RV and unbalanced atrioventricular septal defect (uRV/AVSD). Methods: Between 1985 and 1999, 31 children were identified with uRV/AVSD. 24 of whom underwent BVR. Down syndrome was present in 20 pts. Moderate-severe left atrioventricular valve (AVV) regurgitation was present in 4 pts (BVR=2, non-BVR=1). Prior PA banding was performed in 7 pts. Pts in the non-BVR group had a lower ratio of right AVV to total AVV area (mean .37± .05 vs. BVR .43±.04, p< 04), lower RV/LV length ratio as measured from the AVV annulus to the apex (.64± .13 vs BVR .79±0.2, p<.001) and lower median ratio of right AVV to total AVV diameter (.57 (.24–.9) vs BVR .8.5 (.32–.8), p<.005). BVR included 2 patch technique (n=18), coronary sinus drainage into the RA (n=18) and residual restrictive ASD in 5. Results: There were 3 deaths, with Kaplan-Meier survival estimates at 5 years of 95% for BVR vs. 71% for non-BVR pts (p=0.066). In the non-BVR group, 3 pts received fenestrated Fontan operation. Compared to the non-BVR group, pts in the BVR group had lower CVP on arrival to the ICU (10+/-3 mm Hg vs. 17+/-6 mm Hg, p=.005) and 24 hrs later (13+/-7 mm Hg vs. 22+/-12 mm Hg, p=.03). Pts in the non-BVR group had higher median peak lactates (2.7 (1.8–12.4) vs. 2.7 (1.6–6.7), p= .0019). In the BVR group, O2 saturation at 24 hrs correlated with absolute right AVV annulus size (r= 0.47, p=.0102) and RV length (r = .42, p=.04). On last follow-up, pts in the BVR group had higher O2 saturation (93+/-6%) compared to non-BVR pts (80.1+/-14%, p=.056). Conclusions: Outcomes for BVR in patients with AVSD and small RV are good. In children with diminutive RV dimensions, a residual ASD contributes to successful BVR.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

379

Long-term results of the lateral tunnel fontan procedure in patients with atrial isomerism

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OBJECTIVE: Atrial isomerism is often associated with complex congenital heart defects requiring single-ventricle repair, but abnormal atrial and venous anatomy can complicate the creation of a Fontan circulation. Intra- or extracardiac fenestrations are frequently used, but in many patients construction of an intra-atrial lateral tunnel (LT) is also an option. However, long-term results remain to be determined. METHODS: Twenty three patients (age 7 months – 20 years) with left- or right atrial isomerism underwent a lateral tunnel Fontan procedure between 11/87 and 7/98. Current follow-up information was obtained for all patients (mean follow-up = 11.3+/-7 years). RESULTS: All patients had anomalies of systemic and/or pulmonary venous connections. Four patients (17%) had moderate AV valve regurgitation, and 12 (52%) had preoperative supraventricular arrhythmia. Nine patients required lateral tunnel modifications due to the atrial anatomy, and 12 patients had a baffle fenestration placed. Bilateral superior vena cava-to-pulmonary artery anastomoses were constructed in 7 patients. There were 2 early (8.7%) and 2 late (9.5%) deaths, with a 10-year survival of 82±13%. During follow-up, 2 patients (10%) developed new bradyarrhythmia and 4 patients (25%) new supraventricular tachyarrhythmia. Overall, freedom from any form of supraventricular arrhythmia (including pre-existing arrhythmia) was 25±12%, and freedom from new postoperative arrhythmia was 62±14%. No patient had evidence of protein-losing enteropathy, but 2 patients (10%) had thromboembolic events. Functional status was good in 14 (74%) and satisfactory in 5 (26%) patients. CONCLUSIONS: The lateral tunnel Fontan operation results in good long-term palliation in patients with atrial isomerism. Although atrial isomerism did not prove a risk factor for postoperative arrhythmia when we examined our overall experience with LT Fontan, the prevalence of postoperative arrhythmia is markedly higher than that seen in other patients with a lateral tunnel Fontan.

380

Right ventricular dysfunction and the role of pulmonary valve replacement after correction of tetralogy of Fallot

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Background Correction of tetralogy of Fallot(ToF) often leads to pulmonary regurgitation, sometimes warranting pulmonary valve replacement(PVR), for which indication and timing to achieve are not yet clear. This retrospective study describes follow-up and interventions in our ToF population. Methods Review of all consecutive patients operated for ToF between 1977 and 2000. Included are date and type of repair, Dopplerechocardiography (2D-echo), ECG's, re-operations and physical condition. Results Total repair was performed in 270 patients, mean age 1,9 +/- 2,5 yrs, 82 were excluded because of follow-up abroad. Right ventriculotomy was used in 92%, transatrial VSD closure in 8%, 69% received a transannular outflow-patch. Pulmonary atresia required a pulmonary graft in 13(8%) patients. Overall 20-years-survival was 88%. Last follow-up: ECG's show RBBB in 67%(QRS complex 129+/-29,3 msec). RVOT aneurysms were detected in 16% 2D-echo demonstrates mild pulmonary insufficiency(PI) in 40%, severe in 21%, dilated RV in 76%, both increasing with post-repair age. In 39% RV dimensions are equal or even exceed LV dimensions. 45% show tricuspid insufficiency and the RA is enlarged in 14%. Reintervention was necessary in 39/185 pts, this included angioplasty for residual stenosis and PVR (22/39 homografts, 6 pts of PA group) at a mean age of 11,2 yrs after correction. In 7 the RV returned to normal dimensions and symptoms disappeared, but in 3 severe dysfunction developed. Eleven others still present with RV dilatation and/or PI. In total 75% were free of reintervention at first ten years. The right atrial approach diminishes severe RV dilatation and prolonged QRS duration (p=0.001 and 0.007). Early correction reduces the risk of re-operation(p=0.011). Conclusion Severe RV dilatation(39%) and PI(51%) secondary to outflowtract repair in ToF are frequently occurring sequels developing slowly over time. Early transatrial repair slightly favors the outcome.

381

Medium term outcome with atrial-arterial switch for congenitally corrected transposition (ccTGA) in twenty-nine patients

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The morphologic right ventricle(RV) often fails following conventional repair of ccTGA. Atrial-arterial switch restores the morphologic left ventricle(LV) to the systemic circulation and may provide better functional outcome. Methods: 29 patients underwent atrial-arterial switch for ccTGA between 1991–2000. Median age was 2.4 years (range 0.1 –24.5 years) and median weight 13kg (range 3–52kg). 18 patients had ventricular septal defect(VSD)of whom 16 underwent pulmonary artery banding(PAB). 9 of these had moderate to severe tricuspid regurgitation(TR). 8 patients had intact ventricular septum of whom 6 underwent PAB to train the LV. The remaining 3 patients were in severe heart failure following previous conventional repair. Surgery involved the Senning repair without supplementation in all cases and arterial switch, with the Lecompte maneuver in 12 cases. Tricuspid valve repair was not required. Median cardiopulmonary bypass time was 160 min, pulmonary artery 40 min and aortic cross-clamp time 125 min. Results. Median follow-up was 39.7 months (range 2–102 months) and was 100% complete. Preoperative mean NYHA class was II with 9 patients in NYHA III and 6 patients in NYHA IV. There were 2(7%) early deaths and 2(7%) late deaths. Moderate neoaortic regurgitation developed in 4/25(16%) and severe in 1/25(4%). Two went on to receive aortic valve replacement. LV function was good or mildly impaired in 21/25(84%), moderate in 3/25(12%) and poor in 1/25(4%). No patient had worse than mild TR. Two patients required angioplasty for systemic venous obstruction. Conclusion: Atrial-arterial switch can be advocated in ccTGA to avoid development of RV failure. In the presence of RV failure and TR before or after conventional repair atrial-arterial switch is the procedure of choice.

382

Analysis of risk factors for outcome in cases atrial correction of double outlet right ventricle

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In a retrospective review of factors influencing surgical outcome in patients undergoing biventricular repair for double outlet right ventricle (DORV). Between 1989 to 2000, 350 patients with diagnosis of DORV and its variants underwent various surgical procedures. The median age was 3 yrs. The VSD was subaortic in 56.9%, sub pulmonic in 19.6%. Doubly committed in 6% and non-committed in 26.6% patients. 22.6% had associated pulmonary arterial hypertension, and 39.9% had other associated cardiac defects. 10% were previously palliated with systemic pulmonary artery shunt. 240 patients (69.6%) underwent biventricular repair through transatrial approach using Gore-tex patch, continuous suture technique. In 7.5% cases the VSD was restrictive and had to be enlarged before closure. Trans annular patch was used in 31% patients. 11% underwent Rastelli operation with cryopreserved aortic/pulmonary homografts and 6% patients with Taussing Bing anomaly underwent arterial switch operation. In hospital mortality was 15% (36/240). The incremental risk factors for early mortality by univariate analysis were sub pulmonic VSDs, associated IAH, severe PS, additional surgical procedures, CPB time > 100 minutes and AoX time > 50 minutes. By logistic regression, risk factors for early mortality were young age, year of operation before 1995, previous palliation, subpulmonic, doubly committed and subaortic VSDs, abnormal relationship of great arteries, arterial switch operation, Rastelli's procedure, CPB time > 100 minutes and AoX time > 50 minutes. Of the survivors 141 patients (58.8%) followed up for a median period of 5.1 years (range 1 month – 12 years). 18 patients (8.8%) required redo surgery for residual defects and there were 2 hospital death (11.1%) among them. Sixteen patients (7.8%) required catheter intervention. Majority of the DORVs can be repaired transatrially. Early and mid term results are satisfactory.

383

Complete atrioventricular septal defect repair, results and risk factors in the current era. A study in thirteen European units

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Atrioventricular septal defect (AVSD) is often complicated by additional risk factors, which decrease survival. Reported mortality varies from 2%-16%

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

As most published series have 'publication bias' in common, in that successes rather than failures are reported, average mortality might well be higher. The purpose of this multicenter European study therefore, was to accumulate information on non-selected results of surgery of complete AVSD in the current era. The study covers the years 1998/9. Sixteen surgical units from 12 European countries included all 290 patients (range: 5–43 patients) with a complete AVSD where the interventricular component was closed with a patch. Median age was 166 days (6 26 years); median weight was 5.0 kg (2.38–65 kg). Down's syndrome was present in 142 (66.2%). Median bypass time was 138 min (45–480 min), median aortic crossclamp time was 80 min (29–206 min). Circulatory arrest was employed in 42 patients with a median duration of 38 min. (2–91 min). The two patch technique was used in 248 patients (85%), single patch in the rest. The 'cleft' was closed completely in 219, partly in 54 and left open in 15 patients. Pulmonary artery banding was performed in 25 patients (8.9%), shunting procedures were done in 5 patients (1.7%). Twenty patients died within 30 days (6.9%); 11 patients died late (3.8%), thus total mortality was 10.7% (range between hospitals 0% 37.5%). Logistic regression analysis showed that hospital, reoperation for bleeding/tamponade (7) and preoperative peritoneal dialysis (10) were risk factors for early death. Hospital, delayed sternal closure (24) and early reoperation (29) were risk factors for late death. In conclusion, hospital is a consistent risk factor within hierarchically levels room for improvement and any necessary reoperation increases risk significantly.

384

Endothelin-1 levels positively correlates with outcome in infants undergoing open heart surgery

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Objective: To determine serum endothelin-1 (ET-1) levels in infants submitted to open heart surgery for complete repair of congenital heart defects immediately pre and during the first 24h post-operatively. Methods: We enrolled 32 infants (>28day-old and <1 year-old) who underwent open heart surgery with cardiopulmonary bypass (CPB) from June 1998 to April 1999. Blood samples were withdrawn at six time points: 1) during induction of anesthesia, 2) 15 min after beginning CPB, 3) 15 min after warming off CPB, 4) at arrival in cardiac PICU, 5) 6h after arrival in cardiac PICU and 6) 24 h after surgery. Each blood sample was centrifuged and frozen at -16°C. ET-1 was measured by Elisa radioimmunoassay technique. For statistical analyses we used repeated measures ANOVA and values were considered statistically significant at $p<0.05$. Results: We analysed 32 infants, 19 (59%) of whom were male, the mean weight was 5.6 + 2.8kg, with a mean age of 5.35 + 2.76 month. Nine (28%) infants were cyanotic and 23 (72%) acyanotic. Five (16%) infants died and 27(84%) were discharge alive. There was an increase in ET-1 levels following open heart surgery in infants ($p=0.0006$). There was a positive association between ET-1 levels and post surgical ($p=0.002$) Fig 1. There was not enough statistical power to demonstrate a difference in ET-1 levels between cyanotic and acyanotic patients across different time points ($p=0.08$) Fig 2. Conclusion: ET-1 levels increase after open heart surgery in infants and high levels are associated with poor outcome. The trend to higher ET-1 levels in acyanotic patients is probably related to increased pulmonary blood flow, and thus contributing to higher pulmonary artery pressure and resistance. Key words: endothelin-1, cardiopulmonary bypass, cardiovascular surgery.

385

Ascending aortic replacement in pulmonary atresia – is it justified?

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Background: Pulmonary atresia causes in utero remodeling of the combined ventricular output through the aorta, which is already markedly enlarged at birth. Little is known about the long term fate of these aortas, often reaching diameters raising concern about the danger of rupture. There is also uncertainty if the wall changes of these dilated vessels resemble those of true aneurysms. Methods: Since 1995 we have replaced the ascending aorta as an additional procedure in 6 patients with pulmonary atresia (age: 5,6,14,23,32,33 years). The aortic diameters ranged from 4.0 to 6.5 cm. Twice the distal anastomosis was performed in the proximal aortic arch under deep hypothermic circulatory arrest. Concomitant procedures comprised: bipulmonary Glenn anastomosis including pulmonary artery plasty (3), staged unifocalisation with VSD closure (2), and RVOT conduit change (1). One insufficient aortic valve was replaced with a composite graft. The aortic prosthesis had diameters of 18–26 mm. Results: All patients survived. Four showed definite histological changes typical for media degeneration, two marked intimal thickening and fibrosis. Two corroborated the degenerative changes found by us in the aortic wall biopsies of 5 neonates with pulmonary atresia undergoing central shunt procedures. CT-scans at follow-up were unremarkable after 9 to 30 months. One patient has since undergone total cavo-pulmonary connection. Conclusion: Replacement of an enlarged ascending aorta in pulmonary atresia may be indicated to facilitate other procedures, especially upon the central pulmonary arteries, as well as to avoid aortic valve insufficiency or possible aortic rupture. Degenerative aortic wall changes seem to develop very early in life.

386

Single stage complete unifocalisation and repair for VSD, pulmonary atresia and MAPCAs – early and mid term results

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In an earlier period these patients were treated with multistage unifocalisation through thoracotomies. Recently an aggressive approach has been adopted to treat these patients with single stage unifocalisation. From June 1997 to April 2000, 40 patients were treated with a single stage complete unifocalisation and repair. The age ranged from 6 months to 23 years (median 3 years). Median sternotomy approach was used in all cases except patients in whom earlier thoracotomy had been performed. These patients were approached through a clamshell incision for unifocalisation. 126 MAPCAs were unifocalized. Tissue to tissue anastomosis was achieved in all patients except one in whom polytetra fluoroethylene tube graft was used. All patients had complete unifocalization. Twenty three (58%) patients had final repair, 13 (25%) patients had RV to PA homograft conduit (VSD left open) and 6 (15%) patients had central shunt. There were 6 (15%) deaths. The follow up ranged from 7 months to 42 months. Four patients had successful completion of VSD closure and one patient had reconstruction of pulmonary stenosis. There were 3 late deaths out of which two patients died with progressive pulmonary vascular disease and one patient died with endocarditis. Single stage unifocalisation reduces the number of operations and more number of patients undergo final correction at an early age. Early results are encouraging, mid term follow up showed progressive pulmonary vascular disease in certain patients and on going follow up will help us to identify which patients will benefit from surgery.

387

Surgical management for pulmonary atresia with intact ventricular septum associated with sinusoidal communications

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Objective: Late outcome after surgical treatment for pulmonary atresia with intact ventricular septum associated with sinusoidal communications with or without right ventricle-dependent coronary circulation (RVDCC) remains poor in most reported series. The aim of this study was to evaluate surgical outcome of this entity. Methods: A retrospective charts of 26 patients with pulmonary atresia with intact ventricular septum associated with pinnipedal coronary sinusoids between January 1990 and August 2000 were reviewed. Results: Of 51 patients with pulmonary atresia with intact ventricular septum, 26 patients had sinusoidal communications and 7 patients had a RVDCC. A systemic-pulmonary artery shunt was performed in 17 patients, with 1 hospital death and 3 late death. A right ventricle outflow tract reconstruction and central shunt was performed in 1 patient. Modified Fontan operation was performed in 12 patients and biventricular repair was performed in 1 patient without any mortality. Actuarial survival including both in-hospital and late deaths, by Kaplan-Meier method in the patients with RVDCC and the patients without RVDCC revealed 54% and 89% survival rates at 3years. The actual survival rate was significantly lower in the patients with RVDCC than in the patients without RVDCC by Wilcoxon test ($p=0.048$). Conclusion: Results of the definitive operation for the treatment of patients with pulmonary atresia with intact ventricular septum associated with sinusoidal communications were satisfactory. When right atrial cavity was obliquely partitioned and tricuspid valve left open at the modified Fontan operation for, oxygenized blood supply the coronary artery from right ventricle. Prevention from ischemic complication was most important factor in the patients with patients with pulmonary atresia with intact ventricular septum with RVDCC.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

334

Long-term results after management of critical aortic stenosis in infants and neonates

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Introduction: Critical aortic stenosis (CAS) in children necessitates urgent intervention for patient survival. The optimal treatment, however, continues to be controversial and has still high morbidity and mortality. The study examined the late outcome after treatment of CAS in infants and neonates. Methods: Sixty-seven children (24 boys and 43 girls) underwent surgery for CAS between 1967 and 1999. Procedures performed include closed transventricular valvotomy (CTV) (n= 53), open aortic valvotomy with cardiopulmonary bypass (OAV) (n=14). The mean age at the last intervention was 24.8 days ± 18.4 days (ranges from 1 to 140 days). Associated cardiovascular anomalies were in 35 (49%) patients. Results: The hospital mortality was 14.9% (10/67). The mean duration of follow-up for the hospital survivors was 7.2±3.8 years. The actuarial survival for the hospital survivors was 94.2%±3.9% at 5 years and 88.7%±6.4% at 10 years, whereas event-free survival (reintervention, endocarditis, or early death) was 87.7%±6.6% at 5 years, 79.9%±8.2% at 10 years. At last follow-up (mean 7.8±5.1 years), 45 of the long-term survivors (n=55) were in functional class I and 10 were in functional class II. 42 patients in CTV group the ejection fraction (EF) was 50.8±17.6% and left ventricular end-diastolic volume (LVEDV) = 52.4±20.6 ml/m2. 13 patients in the OAV group the EF was 46.2±21.5% and LVEDV 50.2±10.8 ml/m2 by echocardiography-Doppler. Two patients have died and 13 patients have required aortic valve replacement during the follow-up period. Conclusions: Critical aortic stenosis in children is a difficult problem with a high initial mortality. Late survival and functional class are initial hospitalization, but most require further intervention within 10 years.

339

Combining stent implantation in the arterial duct with bilateral pulmonary artery banding – a new option for newborns with left heart obstruction

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Objectives: Stent implantation in the arterial duct (AD) is proven to be effective for patency in pts with ductal dependent circulation. We describe the combination of stenting the AD with bilateral pulmonary artery banding (BPB) for first step palliation in newborns with severe left heart obstruction until further decision of palliative or corrective surgery is made. Methods: 1998–2000: 13 pts had stent placement in the AD combined with BPB. Diagnosis were hypoplastic left heart syndrome (HLHS) (n=10), aortic atresia in L-TGA (n=1), interrupted aortic arch (IAA) (n=2). Balloon expandable stents were placed transvenously or transarterial through 4–5F sheet introducer (6a-b). In 3 pts a second stent was necessary. BPB was performed 1–5 days after the catheter intervention to reduce mean pulmonary artery pressure (PAP) to 20mmHg and arterial saturation to 75–80%. Results: There were no procedural deaths. The AD was augmented to a final diameter between 7–10 mm and held open for up to 331 days. 2 pts with HLHS had successful heart transplantation, corrective surgery was possible in 2 pts with IAA. 4 pts with HLHS were operated as single step combined Norwood and II operation, with Glenn anastomosis and aortic reconstruction at the age of 3–6 months with excellent neurological outcome. 4 pts are still waiting for the mentioned combined Norwood procedure at home. Conclusion: Maintaining ductal patency by stent implantation combined with BPB allows recovering of the heart, especially in pts with cardiac shock who are otherwise not considered for the classical Norwood procedure. Because of low PAP, waiting time for a heart donor is not limited in case of transplantation, and a combined Norwood procedure stage I and II beyond the neonatal period may improve outcome in long-term follow up.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

GENERAL POSTER SESSIONS

MAY 28 Time: 11:00–12:30

Session 1
Surgical Management and Results: Abnormal Venous Return, Left Ventricular Outflow Obstruction/ Aortic Stenosis, Pulmonary Atresia

P1

The experience of Modified Fontan operation in Hong Kong

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A retrospective study of the Modified Fontan operation in a single center in Hong Kong.
From October 1982 till December 1999, 110 patients received modified fontan operation in Grantham Hospital, Hong Kong. There were 65 male and 45 female patients. Thirty seven (33.6%) patient received Fontan operation below the age of 4 year old. The hospital mortality was 17.3% and the late death rate was 5.5%. Eighty three (75.5%), 13(10.9%), 7(6.4%) and 7(6.4%) of patients with their Fontan circulation connected as atriopulmonary direct anastomosis, extracardiac, lateral tunnel and total cavopulmonary using an atrial flap respectively. Majority of patients suffered from Univentricular heart (90.9%) and Tricuspid atresia (40%). Seventy five (68.2%) patients received palliative procedures with 56 patients had modified BT shunts and 15 patients underwent pulmonary artery banding. Eight (7.3%) patients had CVA, 9(8.2%) patients had postoperative bleeding that needed exploration. However, 39(35.5%) patients needed peritoneal dialysis and 31(28.2%) patients had deranged liver function tests immediately after the operation. Additional procedures carried out at the time of Fontan operation included 6 AVV replacement, 3 correction of TAPVD, 2 modified Damus and Kay procedure, 4 patch repair of left pulmonary artery, 1 aortoplasty of ascending aorta and 4 thoracotomy for haemostasis. Among the 85 survivors, 61and 11 patients were in NYHA I and II respectively. Survival at 1 year 81.7%, 5 year 76.5% and 10 year 71.6%. In conclusion the Fontan operation offers a good palliation for patient.

P2

Regional low flow perfusion in complex infant aortic arch reconstruction

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Background. Hypothermic circulatory arrest (HCA) has long been considered unavoidable during complex infant aortic arch reconstruction. Because of concerns regarding permanent neurologic effects of HCA, we have developed a technique of regional low flow perfusion (RLFP) that provides cerebral circulatory support during aortic repair. Methods: RLFP was employed in 18 infants diagnosed with aortic arch hypoplasia or interruption from August 1998 to August 2000. Using near-infrared spectroscopy (NIRS), RLFP flow rates were adjusted to maintain baseline (as measured on full bypass) cerebral blood volume (CrBVI) and oxygen saturation (CrSO2). Results: Single ventricle repair was performed in 12 patients (group A) and biventricular repair was achieved in 6 patients (group B). The average age and weight at operation was 4.9±2.8 days and 3.3±1.0 kg in group A and 17±22 days and 2.8±0.8 kg in group B (p=0.07, 0.24). The mean RLFP flow index was 0.26±0.1 L/m2/min in group A and 0.56±0.4 L/m2/min in group B (p=0.02). Mean bypass time was 158±40 minutes in group A and 256±152 minutes in group B (p=0.02). The duration of RLFP was 49±11 minutes in group A and 60±52 minutes in group B (p=0.5), with only 8±4 and 10±13 minutes of HCA, respectively (p=0.7). CrBVI was maintained at baseline values in both groups. CrSO2 was measured within 2.1±6.1% and 3.0±8.3% of baseline in groups A and B, respectively (p=0.05). Operative survival was 89% (16/18). There were no adverse clinical neurologic outcomes. Conclusions: RLFP is a safe and effective technique that reduces the need for HCA during complex aortic arch reconstruction in infants undergoing both single ventricle and biventricular repair. Cerebral malperfusion is avoided using real time NIRS data. RLFP should reduce the risk of HCA associated neurologic deficits.

P3

Medium and long-term follow-up after thoracoscopic ductal interruption

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Video-assisted thoracoscopic surgical (VATS) interruption of a patent ductus arteriosus (PDA) is safe and effective, but little medium or long-term follow-up data are available. We present our effective out-patient experience from 5/95 to 8/00 of 51 patients (age 4.4 +/- 4.8 yr, range = 0.2–17.1 yr, weight 17.3 +/- 12.8 kg, range = 4.5–55.2 kg) with various PDA sizes by echocardiography (3.4 +/- 1 mm, range = 1.5–6 mm). In 48 patients immediate ductal closure was documented by exam and echocardiogram. The procedure was not completed in 3 patients: 1 converted to open ligation for bleeding, 1 for inadequate ductal visualization, and 1 too large for clip interruption. One patient had a tiny re-opened duct on post-op day 4. Long-term follow-up was available in 41 patients, none of which had clinical evidence for a PDA at an avg. time of 33 +/- 20 mo post-op (range 3–82 mo.). Twenty-seven patients had transthoracic echocardiograms at an avg. time of 7 +/- 6 mos. post-op (range 0.5–25 mo.), none of which demonstrated a PDA. Complications included 1 patient with a self-limited pericardial effusion, 1 with mild bleeding requiring minimal extension of a surgical port wound for exploration, and 1 with hoarseness for 6 weeks. VATS PDA interruption is safe and efficacious with wide patient application and good medium and long-term results.

P4

Combined technique coarctation repair: forward or reversed subclavian flap with end-to-end anastomosis

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Objective: Left subclavian flap angioplasty and resection with end-to-end anastomosis are accepted as surgical treatments for coarctation of the aorta. There are delicate cases that require the use of modified techniques combining both subclavian flap and resection with end-to-end anastomosis. To determine the safety and efficacy of two forms of this combined technique we retrospectively reviewed the records of our experience from 1972 through 1999. Methods: Twenty patients (4.5% of the total number undergoing coarctation repair) were treated using one of two combined techniques. Reversed left subclavian flap with end-to-end anastomosis (Group A) was used for 11 (55%) patients mainly due to associated significant transverse aortic arch hypoplasia. A combination of forward subclavian flap with end-to-end anastomosis (Group B) was used for 9 (45%) patients due to an unusually long coarctation segment. The median age at repair in Group A was 8 days and was 3.4 months in Group B. Eleven (55%) patients had associated intra-cardiac lesions, 7 in Group A and 4 in Group B. Results: There were no mortality or complications in either group. The mean hospital stay was 7.8 days (range 3 – 28 days) for both groups. In 17 patients, intraoperative pressure gradients were measured at the conclusions of the repair, and there was no gradient in any of these patients. The mean follow-up period was 5.5 years for group A (range 93–8002 days) and 2.36 years for group B (range 3 – 2698 days). All patients were asymptomatic and none have required re-intervention. Conclusion: The combined technique of forward or reversed subclavian flap angioplasty plus resection with end-to-end anastomosis yields excellent results at intermediate term follow-up. These techniques represent safe and effective surgical options for the repair of various forms of coarctation of the aorta.

P5

Anatomic correction of ebstein's anomaly

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Objective: Anatomic correction of Ebstein's anomaly restores tricuspid and right ventricular function. The early results of this procedure are reported. Methods: Between December 1997 and April 2000, 16 consecutive patients (Male 7, Female 9, age range 3 to 32 years, mean 12.) underwent anatomic correction of Ebstein's anomaly. Eleven patients had associated congenital malformation (atrial septal defect in 4, patent foramen ovale in 4, ventricular septal defect in two, patent ductus arteriosus in one, double outlet right ventricle in one). Two patients were Carpentier type A, three type B, 11 type C. The operative technique was following: tricuspid to triangle excision of attached chamber, sheetening tricuspid annulus, detachments of the septal and posterior leaflets from the displaced annulus and reimplantation their papillary muscles. Simultaneous correction associated congenital malformation

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Results: All patients survived, recovered uneventfully and were in sinus rhythm; their New York Heart Association class improved. Preoperative examinations revealed tricuspid incompetence (8 severe, 8 moderate); postoperative echocardiography showed that tricuspid incompetence disappeared in 13 and was mild in 3. tricuspid valve leaflets were at normal level, right ventricle reduced in size remarkably, and stenalized chamber vanished. In our follow-up study (1 to 17 months, mean 7 months), their exercise tolerance improved to normal. Echocardiography indicated that tricuspid incompetence disappeared in 15 and mild in 3. Conclusion: our technique allows anatomic correction of Ebstein's anomaly, even in case usually reserved for primary valve replacement, with satisfactory early results.

P6

The arterial switch at 25 years old; a follow-up study

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Background: The arterial switch operation (ASO) is used to correct transposition of the great arteries since 1976 and has replaced atrial palliations. This study examines the long-term outcome of the arterial switch. Methods: Included in the study are 195 patients after ASO from 1977–2000. Patients were evaluated for functional class, pulmonary stenosis, left ventricular function, arrhythmias, aortic sequellae, and coronary pathology. Results: Perioperative mortality was 15%. In the last 5 years, mortality was 4 %. Of 151 survivors two died late, 1 of pulmonary hypertension and 1 of ventricular fibrillation after coronary pathology. 145 patients are in NYHA class I and 4 patients are in class II. The most frequent complication was pulmonary stenosis requiring 45 re-interventions in 26 patients. Left ventricular dysfunction was noted in 5 patients. Arrhythmias were seen in 5 patients: 2 patients developed ventricular fibrillation, 1 died and 1 required a defibrillator implantation. 1 developed sick sinus syndrome, 1 developed atrial flutter and 1 had a single attack of supraventricular tachycardia. No or trivial aortic valve incompetence was seen in 146 patients. 3 had mild, 1 had moderate and 1 severe incompetence. Coronary sequellae were found in 5 of the 61 patients who had angiography. Conclusion: Long-term clinical outcome of ASO is good and peri-operative mortality is low now. Morbidity is dominated by pulmonary stenosis, infrequent aortic regurgitation and coronary artery pathology with potentially lethal arrhythmias. ASO is the preferred method of treatment in patients with transposition.

P7

Median sternotomy for modified Blalock-Taussig shunt

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<Purpose> We investigated the merit of sternotomy approach against the lateral thoracotomy for modified Blalock-Taussig(m-B-T) shunt. <Patients and Methods> Twenty-five patients underwent m-B-T shunt through median sternotomy(S group) between 1995 and 2000. Forty-four patients underwent m-B-T shunt through lateral thoracotomy(T group) between 1991 and 2000. The operative results and the complications were compared between the 2 groups. <Results> The hospital death was 1/25(5%) in S group and 0/44 in T group. Shunt size was 4.1±0.1mm in S group and 4.4±0.1mm in T group. Early shunt failure was 2/24(8%) in S group and 2/44(5%) in T group. Late shunt failure within 2 years was 0/24(0%) in S group and 2/44(5%) in T group. 2 cases of phrenic nerve palsy, 3 cases of mediastinitis, 1 case of chylopericardium and 2 cases of pulmonary overflow needed the treatment for those complications in S group, whereas only 1 case of phrenic nerve palsy was diagnosed in T group. At the cardiac repair 4 cases (9%) of T group and a case (4%) of S group needed the repair of PA deformity induced by B-T shunt. ICU stay was longer in S group than in T group (7.0±1.6 vs 2.6±0.4days, p<0.05). Postoperative hospital stay was also longer in S group. <Conclusion> Median sternotomy approach for m-B-T shunt has the advantage of reduced frequency of longterm shunt failure and PA deformity, but it has the disadvantage of early postoperative complications and long hospital stay.

P8

Anatomically corrective repair of complete atrioventricular septal defects and major cardiac anomalies

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Thirteen patients with balanced forms of complete atrioventricular septal defects (AVSD) and associated major cardiac anomalies underwent anatomically corrective repair. Eight of 13 patients had atrial isomerism (right in 1, left in 7) with double outlet right ventricle (DORV) in 5 and associated cardiac anomalies in 3. Four had DORV with trisomy 21, and one had tetralogy of Fallot. A ventricular septal defect with subaortic extension was present in 7 of 9 with DORV. Ages at operation ranged from 5 months to 10 years (median 4.7 years). AVSDs were treated through the atrium in all patients and an additional right ventriculotomy in 8 patients. Two-patch technique was performed in 10 patients. Atrial separation for isomerism with an additional prosthesis was accomplished in four patients. The interventricular septum was enlarged cephalad in two and muscular subaortic stenosis was resected in two. Two hospital deaths (15%) and one late death (7%) occurred only in the isomerism group. Three reoperations were required solely in the left isomerism group: one replacement of the valved external conduit concomitantly with reconstruction of the left ventricular outflow obstruction (LVOTO), one mitral valve replacement for severe regurgitation of left AV valve and one relief of progressing LVOTO in the patient with intermediate AVSD. The results of anatomically corrective surgery for AVSDs with major associated cardiac anomalies in the isomerism group were excellent. The optimal surgical options for isomerism heart, however, were still controversial now.

P9

A case of organising the special paediatric cardiac centres: improved surgical outcomes at the latvian state cardiology centre for children

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To optimise care for children with congenital heart diseases in Latvia was established The Latvian State Cardiology Centre for Children in 1994. Until that year the paediatric cardiac care was part of adult cardiac centre. The main tasks of organising the full-time echocardiographic examinations, paediatric cardiac anesthesiologists, perfusionists and cardiac surgeons, education of surgical nurses and required preoperative postoperative critical care cardiology teams. To assess the efficacy of this program in improving results we compared surgical services for three years prior to initiation of the program with the subsequent three years. Group I (January 1, 1994 – December 1996): only 21 repairs under CPB for patients in the first year of life (postoperative mortality rate 23.5–5/21). Group II (September 4, 1997 – August 2000): 240 repairs under CPB for patients in the first year of life (mortality rate 9%). We conclude that a focused congenital heart management team significantly improves surgical repairs. We believe that data have important implications supporting the development of regional paediatric cardiac centres.

P10

Long-term results of repair of complete atrioventricular canal defect in infancy

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Background: Early and long-term results of repair of complete atrio-ventricular canal defect (complete AV canal) in infancy were examined. Patients and methods: Sixty three patients with complete AV canal underwent total correction in infancy with two patch method at the Fukuoka Children's Hospital between 1990 to 1999. There were 22 patients with Down's syndrome. Results: Early operative mortality was 4.5% (0/13 in Down and 6.5% in non-Down, N.S.) The reoperation free rate and actuarial survival rate at 15 years were 89.9 ± 4.6% and 92.0 ± 3.4%, respectively. The reoperation free rate and actuarial survival rate at 10 years were similar between the patients with and without Down's syndrome. While the body weights of the patients with Down's syndrome after 2 years of surgery were significantly smaller than those of non-Down's syndrome (81 ± 12 % of normal average value vs. 89 ± 9 %) although preoperative values were similar in both group. Conclusions: Repair of complete AV canal in infancy can be done safely and its long-term results were satisfactory. The Down's syndrome does not affect the long-term results except for the postoperative gain of the patients body weights.

P11

Median sternotomy approach to correct late complications after surgery for coarctation

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Introduction: Recurrent obstruction and aneurysms have been reported

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

following coarctation repair. We present our experience with reoperation through a median sternotomy with use of CPB. Patients and methods: Fourteen patients underwent surgery for recoarctation (mean age 7.9 years [3 days–18 years]). Repair consisted in end-to-end anastomosis (9), patch plasty (2), subclavian flap technique (2) and tube interposition (1). Correction of TGA with VSD (2) and VSD (1) was performed simultaneously. After a mean interval of 11.4 years (3 months – 20 years) three patients were reoperated for aneurysm (3) and/or obstruction of the arch (11). In 6 patients balloon dilatation had been performed previously. Reoperation consisted in resection with end-to-end anastomosis in 5, patch augmentation in 7, tube interposition in 2 and replacement of ascending aorta and arch in 2. All patients were operated through a median sternotomy with hypothermic cardiopulmonary bypass. A mean circulatory arrest period of 21 (10–35) minutes was used in 5 patients. Antegrade cerebral perfusion was used in 9 patients. In 4 patients concomitant cardiac anomalies were corrected. Results: No mortality occurred. Temporary paralysis of the left recurrent laryngeal nerve occurred in one patient. After a follow-up of 31 (1–72) months no residual obstruction was observed in any of the patients. All postoperative mean gradients remained less than 10 mm Hg. Conclusions: A median sternotomy approach with the use of CPB and circulatory arrest or antegrade cerebral perfusion is safe and effective to correct obstruction or aneurysm of the aortic arch following repair of coarctation.

P12

Staged correction of pectus excavatum and cardiac anomalies in young Marfan patients

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Introduction: Patients with Marfan's syndrome may present with both pectus excavatum and cardiac anomalies. Cardiac surgery may be difficult or impossible because of the chest deformity. We present our experience with a staged approach. The chest deformity is first corrected, followed by cardiac operation some months later. Patients and methods: Five patients with a mean age of 13 (6–16) years had a severe pectus excavatum and atrial septal defect (1), mitral valve insufficiency (1), aortic dissection (1) or aortic root aneurysm (2). The pectus was corrected by resection of costal cartilages, turning of the perichondria and sternal osteotomy. After a median interval of 29 (6–60) months the cardiac anomalies were operated through a median sternotomy. Three Bentall-gore rotors, 1 mitral valve repair and 1 atrial septal defect closure were performed. Outcomes were studied by retrospective analysis of patient records. Results: No early mortality occurred. Pectus excavatum was corrected with good functional and cosmetic result in all patients. The cardiac operations were uncomplicated in all patients. Mean follow-up was 35 (16–76) months. Chest deformities did not occur after cardiac surgery. Conclusions: A two stage approach is safe and effective in patients with Marfan's syndrome who present with a need for surgical correction of a pectus excavatum and a cardiac anomaly. The cardiac operation may be performed as soon as 6 months after the correction of the chest deformity. A median sternotomy can then be used to approach the heart in a safe way.

P13

Surgical treatments for complications of transcatheter procedures in congenital heart disease

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Transcatheter interventions have assumed an important role in the management of congenital heart disease. The purpose of this study is to present our experience with twelve such complications. Methods: Twelve patients who required operation after a transcatheter intervention between 1992 and 1999 are described. Seven patients were emergency operation and five were semi-emergency operation. Four patients were underwent RVOT repair and Bjork operation due to rupture of RVOT for ballon dilatation procedure. Three patients were underwent to remove the device and ASD closure due to troubles for ASD device closure. Three patients were underwent to repaire due to the troubles for ballon dilatation of coarctation of aorta (one patient was dissection of descending aorta and two were aneurysm of descending aorta). Two patients were the troubles for coil embolization for PDA. Three patients were opened their chest in the catheter room. Results: All patients survived. One patient had a brain complication for brain infarction by camponade. Two patients underwent Glenn operation after 6month later for catheter intervention. Another patients are doing well with no further need for catheter intervention or operation. Conclusions: If complications of catheterintervention are occurred, the patients condition will be very bad immediately. Emergency operation will be needed. Time condition. When operation is required, results are typically very good.

P14

Surgical treatment of complete atrioventricular canal with tetralogy of Fallot

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Purpose: Associated complete atrioventricular canal with tetralogy of Fallot (CAVC+TOF) is a rare form of congenital heart defect, usually occurring in Down's syndrome. The aim of this report is to review the surgical experience of the patients with CAVC+TOF. Methods: Between 1982 and 1999, ten patients with the mean weight of 14.9 kg (8.5–31.5) underwent surgery at the mean age of 71 months (7–226). Nine children (90%) had Down's syndrome. Associated anomalies included: atrial septal defect (ostium secundum type) (40%), patent ductus arteriosus (10%), right aortic arch (20%) and anomalies of the urinary tract (10%). Four children (40%) underwent primary repair and 6 patients (60%) palliative systemic pulmonary shunts and later total correction. In all patients CAVC was corrected using 2-patch technique. The right ventricular outflow tract obstruction was relieved by transannular patch in 6 children (70%) and by valvulotomy in 3 (30%). The mean cardiopulmonary bypass time was 104.3 minutes (70–145). The mean aortic cross-clamping time was 59.4 minutes (45–79). The mean ventilatory support time was 109.3 hours (6–624), while inotropic support was employed in 7 patients for 1–13 days. The mean stay at intensive care unit was 14.7 days (1–75). Results: No early postoperative death occurred. One child (10%) died 59 days after total correction due to multiorgan failure and sepsis as a result of persistent low cardiac output state. Another patient underwent successful reoperation for residual ventricular septal defect. The mean follow-up is 58 months (7 patients are in the New York Heart Association class I and 3 in class II). Conclusions: These results demonstrate that CAVC+TOF can be repaired early with low mortality and morbidity. A complete repair can be successfully performed without a prior palliative procedure.

P15

Congenital discontinuity of the pulmonary arteries

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Congenital discontinuity of the pulmonary arteries (CDPA) is defined as an anatomical isolation of one or both pulmonary arteries from each other, and from antegrade ventricular flow. We sought to investigate the outcome of CDPA and look at the impact of intracardiac anomalies on survival and residual pulmonary arterial stenosis. We reviewed our medical, surgical, and pathology databases and identified 31 pts diagnosed with CDPA. Six pts had normal intracardiac anatomy (group I) and 25 pts had intracardiac anomalies (group II): 11 with tetralogy of Fallot (TOF), 6 with TOF/pulmonary atresia (PA), 7 with Heterotaxy syndrome (all with PA), 1 additional pt had RV aorta/PA. There were 11 deaths (35%), all in group II (p= 06 Fisher's exact). Three deaths occurred prior to creation of pulmonary artery confluence, and one child low her pulmonary artery with PDA closure. Pulmonary arterial continuity was therefore surgically created in 27 pts. There were 5 per operative deaths (< 30 days), leaving 21 patients for whom follow up (range 7 mo to 10 yr, median 3.5 yrs) was possible. Of these 15 (71%) required 28 ther interventions for subsequent stenosis including: 15 balloon dilations, 5 stent placements, and 10 further surgeries. Late intervention for stenosis was more likely in group II pts vs group I (p<0.05). Conclusions: Mortality is high and the development of late pulmonary stenosis is common in pts with CDPA and associated intracardiac anomalies. Poor outcome may be due in part to concomitant impediment of normal antegrade blood flow; despite creation of pulmonary arterial continuity. We speculate that aggressive complete repair of intracardiac defects with creation of pulmonary arterial continuity may offer the best chance at normalization of pulmonary blood flow and successful outcome in these pts.

P16

Autologous reconstruction of the pulmonary artery in Ross procedure

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Background. Pulmonary autograft replacement (Ross operation) became popular in aortic valve surgery, especially, for children and young adults

Prosthetic replacement of the pulmonary valve and its consequences is one of the main disadvantages of the Ross procedure. Methods: Between May 1996 and November 2000, 34 consecutive patients with mean age of 15±6.8 years, the range from 5 months to 30 years, underwent Ross procedures. Pulmonary valve reconstruction in 30 patients was performed using autologous pericardium. This technique was performed by three different modifications: the straight connection of the distal pulmonary artery to the right ventricle hole with pericardium monocusp; glutaraldegid treated autologous pericardium tube with autologous pericardial monocusp; straight connection of the distal pulmonary artery to the right ventricle with monocusp from anterior wall of the main pulmonary artery. Clinical evaluation and Echo-investigation were performed in early and late postoperative period. Results: The hospital mortality was 13,3%. The follow-up period was from 6 months to 5 years. Two patients died in late period (1 – homograft related endocarditis death and 2 – non-cardiac death – duodenal ulcer bleeding). No major complications were noted in the post-operative period in survived patients. Mild pulmonary insufficiency was noted without hemodynamical problems in all patients. In mid-term follow-up all patients were in Class I NYHA. The monocusp function was noted in all patients no longer than six months, except two patients, where monocusp functioning was continued for 8 and 15 months respectively. All the patients had mild pulmonary regurgitation without right ventricle dilatation. Conclusion: Autologous pulmonary artery reconstruction in the Ross procedure can be performed with satisfactory early results and good mid-term right ventricle functioning.

P17

Outcomes of repair of supravalvar aortic stenosis: impact of left ventricular outflow characteristics

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OBJECTIVE: To determine the effect of the anatomic ratio for the aortic annulus (AA) and sinuses (MS), sinotubular junction (STJ) and ascending aorta (AAO) before and after repair of supravalvar aortic stenosis (SAS) on late valve function. METHODS: We reviewed outcomes of 54 children having repair of SAS between 76 and 99 at a median age of 4.6 yrs (3 mo – 18y), of whom 20 had associated other left obstructive lesions. The repairs were: 1 sinus patch (35), 2 sinus patch (22), other (2). Echocardiographic measurements were obtained pre- and post-operatively, and at late follow-up. The measurements were normalized to the aortic annulus diameter at a point, and then related to early and late valve function and reoperation. RESULTS: Kaplan Meier freedom from reoperation for aortic disease was 94%, 79% and 73% at 1, 5 and 10 yrs; freedom from death was 97% at 20 yrs. Small subaortic ratio was predictive of an increased late gradient (p=0.009). Patients with moderate AI tended to have a higher STJ ratio. A 1 or 2 sinus patch was not predictive of outcomes measured. Prior resection repair or open aortic valvotomy were more likely to have late AI (p=0.06) and higher LV outflow gradient (p=0.03). CONCLUSIONS: Outcome of surgical repair for isolated SAS is good. A small subaortic ratio at prior correction repair or aortic valvotomy was associated with a higher late outflow gradient and/or AI. Operative technique did not affect measured outcomes.

P18

Outcome of children with critical heart disease undergoing extracorporeal cardiac life support

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OBJECTIVE: To determine early and late survival and neurologic outcome in patients undergoing extracorporeal life support (ECLS) for myocardial failure over a 10 year period. METHODS: Retrospective analysis of clinical, operative, perfusion data and results of neurologic imaging and autopsy findings was performed to determine the incidence and the predictors of poor survival and adverse neurologic outcome. RESULTS: Ninety children (median age 9 months, 1 day to 18 years) underwent 97 runs of ECLS for myocardial failure between 01/1990 and 06/2000. ECLS was instituted preoperatively for resuscitation in 22 children (24%), intraoperatively for post cardiotomy myocardial dysfunction in 48 (53%) and for postoperative low output state in 20 (22%). Thirty four (37%) patients had cardiopulmonary resuscitation (CPR) prior to ECLS. While on ECLS 15 children received a heart transplant (2 died) and 13 had intracardiac repair (11 died). Mean duration of ECLS was 107 +/- 78 hrs. Median red cell transfusion per patient was 9.8 liters/m2/day (range 0.9 – 57.8 liters) with median of 77 (4 – 521) donor exposures. Hospital survival was 34%. Kaplan Meier survival at 1 month, 1 year and 5 years was 38%, 33% and 28% respectively. Hospital survival for the congenital group was 28% vs 58% for myocarditis/cardiomyopathy group (p= 0.015). Age, duration of ECLS and the need for pre-ECLS CPR did not affect survival. Neurologic imaging (US/CT/MRI) or autopsy findings were available in 45 patients, 29 of whom (64% of those imaged and 32% of entire group) had a definite abnormality in brain structure. CONCLUSIONS: ECLS is an effective modality for salvaging critically ill children with cardiac disease. However, the associated mortality and comorbidity are high. Survival may be improved with early, aggressive triage to heart transplantation, especially in patients with postcardiotomy myocardial failure.

P19

Problems with the bidirectional glenn shunt (BGS) in high risk situations

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To present our experience with the palliative BGS with emphasis on the problems experienced in high risk patients. Total patients (n=64), 1990 through 2000. Ages: 8 months to 10 years. Mean 4.8. Tricuspid atresia (n=31), single ventricle complex (n=28), other/complex (n=15). Previous surgical procedures (n=31). Results: no early hospital mortality, major complications – superior vena cava (SVC) syndrome with excessive pulsatility (n=1), complex post-operative course (n=1). Effusions (n=3). (One late death). Thrombosis (n=1). Obstruction right SVC, but patent left SVC (n=1), venous collateralization (n=2). Although the BGS is more forgiving than a Fontan procedure and no early mortality occurred, significant complications were encountered when the criteria for a BGS were loosened.

P20

Direct surgical implantation of anomalous origin of right pulmonary artery (RPA) or left pulmonary artery (LPA) from ascending aorta into main pulmonary artery

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Retrospective study to present local experience during the 24 year period 1976 through 2000. 13 patients were operated, male (n=5), female (n=8). Ages: 6 weeks to 25 months. Mean 8.6 months. RPA with left arch (n=12). LPA with right arch (n=1). Isolated RPA / LPA (n=11). Tetralogy of Fallot (n=2); one of the tetralogies also had an absent pulmonary valve. Patent ductus (n=4). Only detaching the artery (n=7). Aorta transected (n=5). Early mortality (n=3). Failure to terminate bypass with severe pulmonary hypertension (n=1). Post-op pulmonary hypertensive crisis (n=1) and extensive post-op lung problems (n=1). There were no late deaths. All survivors functionally did well and the post-operative pulmonary artery pressure decreased significantly. In 4 re-investigated patients mild stenosis of the re-implanted artery was noted (gradient less than 20mmHg).

P21

Palliative Blalock-Taussig shunt procedures in the neonatal and infant period

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Palliative systemic to pulmonary shunts are mostly performed in emergency situations in cyanotic patients of the neonatal and infant age groups. Prognosis of patients who will undergo this procedure is affected by preoperative diagnosis and surgical technique. Systemic to pulmonary shunt was performed on 62 patients in the neonatal or infant period in our department in the last five years. 24 patients were neonates. Out of these 24 patients, 13 were operated by Blalock-Taussig anastomosis due to having intact ventricular septum and pulmonary stenosis. In the neonatal period, grafts having 3 – 3.5mm diameters were used. In the infant period grafts having 4 – 5mm diameters were used. 70% of the patients were not heparinized in the post op period. Before all anastomoses were completed the graft had no contact with blood. Pulmonary artery anastomoses were constructed closer centrally. In the post operative period, 5 neonates and 1 infant died. Five patients developed cardiac insufficiency due to excessive blood flow. Mortality and morbidity are affected by preoperative diagnosis, graft size and pulmonary artery anastomosis positioning in systemic shunts done in the neonatal and infant periods.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P22

Cavopulmonary anastomosis: immediate and long-term results comparison in presence or not of atrial fenestration

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Aim: Cavopulmonary anastomosis became the most used Fontan variant for functional correction of congenital cardiac defects not amenable for anatomical one correction. Material and Methods: 62 patients were operated on between 1988 and 1999 being atrial fenestrations performed on 41 of them (Group I), remaining 21 without atrial fenestration (Group II). Tricuspid atresia predominated in G-I (23–56%) and single ventricle in G-II (14–66%). Median ages at operation and at long-term period were 7.9 and 7.6 years in both groups and 10.6 and 12.8 years, respectively. Results: The overall mortality were 7.3% in G-I and 4.7% in G-II. Asymptomatic pleural effusion occurred in 41.4% of G-I patients and in 22.8% of G-II and accentuated pericardial effusion in 29.2 and 14.7%, respectively in both groups. The central venous pressure was discretely accentuated in G-II – 17.7 cm H2O in relation to 15 cm H2O in G-I and the overall hospital post operative stay was similar, 26.3 and 21.8 days. Cyanosis and arterial desaturation were present in 5 patients and FC-II in 4 patients, all of them belonging to G-II. Fifty-eight patients (93.5%) are in FC-I. Sinus rhythm are present in 94% and pulmonary perfusion were similar in both groups. Physical tolerance was good in 11 patients submitted to maximal cardiopulmonary exercise test. There was not statistical significant difference between both groups. Conclusion: Atrial fenestration did not change favorably the immediate and long-term evolution of patients submitted to cavopulmonary anastomosis, being dispensable in the majority of cases.

P23

Transplant approach for repair of the congenitally malformed mitral valve

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A right atrial incision, prolonged across atrial septum and left atrial roof, was evaluated in the treatment of children with congenitally malformed mitral valve. Method: from January 1998, 9 patients were operated on employing the described approach. Mean age and weight at surgery were 39 mos (range 7–217) and 29 kg (5.7–64). Mitral regurgitation (4 pts) was secondary to Marfan disease (2), repaired cAVSD (1), isolated mitral cleft (1), floppy valve (1), endocarditis (1), cardiomyopathy (1), mitral stenosis was associated with aortic stenosis, coarctation and PDA (1), repaired DORV (1). Mitral repair was performed along the techniques of Alfieri, Carpentier and David; a flexible ring was adopted in 5 pts. Associated procedures (4 pts) included ASD closure PMK (1), commissurotomy (1), Batista procedure (1), aortic valve endoscopy (1). CPB and aortic cross-clamp time averaged 148 and 77 min. Results: one pt died of intracerebral hemorrhage, she had a recent history of septic brain embolism. In a mean follow-up interval of 11 mos (1–35), mitral regurgitation was judged absent in 1 pt, mild in 2, moderate in 1. mitral stenosis was markedly reduced in 2 pts (mean gradient dropped from 16.5 to 8 mmHg). Two pts required reoperation: heart transplantation after Batista procedure (1), mitral valve replacement after cAVSD repair (1). Six of 8 survivors remained in sinus rhythm. Two pts had a permanent PMK implanted prior to atrial repair, one of whom retained normal sinus activity thereafter. Conclusion: The transplant approach" provides unsurpassed exposure of the malformed mitral valve. Sinus rhythm is preserved despite the extensive atrial incision."

P24

The midterm and long term results of surgical repairment of tetralogy of Fallot

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To evaluate the midterm and long term results of surgical repairment of tetralogy of Fallot, 109 patients were studied before and after surgical repairment by using Doppler echocardiography and color flow mapping. 81 male and 28 female in aged 5 – 156 months. One-staged reparative operation was performed in 105 cases, while two-staged reparative operation in 4 cases in which only a transannular patch was given in the initial stage. Right ventricular outflow tract reconstruction contained transannular patch in 79, right ventricular outflow patch in 28 and artificial conduit in 2 cases. The follow-up period by echocardiography was 1 – 72 months. Results: The postoperative residual shunt occurred in 20 cases (18.3%). Although the diameter of the residual VSD was 0.1–0.2 cm in 70% and had no left-sided heart dilation, there was 1 case of bacterial endocarditis. Residual obstruction occurred in 7 cases (6.4%). Pulmonary regurgitation occurred in 85 cases (77.9%), of which the occurrence of right ventricular dilation was significantly higher than those without pulmonary regurgitation (92.9% vs 20.8%, p<0.001). Follow-up data showed that pulmonary hypertension occurred in three of the four cases after the initial procedure. Conclusion: The midterm and long-term results of surgical repairment of tetralogy of Fallot are generally satisfactory. Relatively high occurrence of pulmonary regurgitation deserves further investigation. Intensive surveillance is advocated in the patients with residual shunt or having undergone palliative procedures.

P25

Traction techniques for improving accessibility in minimally invasive pediatric cardiac surgery

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In minimally invasive pediatric cardiac surgery (pMICS), a deep, narrow field makes safe surgical procedures especially difficult for less practiced surgeons. We report 12 consecutive recent cases of simple cardiac defects in which pMICS was performed, with traction on the skin, pericardium, right atrial appendage, aortic root and both venae cavae to improve safety. The following surgical technique was used: A skin incision (5% of patient height) below the nipple level, and a partial sternotomy splitting the sternum in the mid-line below the third intercostal level to form an inverted Y shape, were performed. The skin at the cranial end of the incision was drawn cranial with a mechanical retractor. The edges of a vertical incision in the pericardium were drawn caudad to drag the ascending aorta into view. The right appendage was drawn caudad with a purse-string suture to expose the area around the aortic root. Two traction sutures (traction suture technique of aortic root) were secured in the fatty and epicardial tissue of the aortic root, drawn caudad and fixed, to make the aorta more accessible for safe cannulation. For aortic cannulation, the Seldinger method is preferable for its simplicity. After establishment of a cardiopulmonary bypass, the venae cavae were raised with tourniquet tapes and fixed, facilitating intracardiac procedures. No intraoperative or perioperative complication occurred, and no blood transfusion was needed. Because aortic cannulation was initially transcostal, three cases required aortic side-clamps, and the aortic traction sutures facilitated their application. Our traction techniques facilitate exposure of the surgical field for safe pMICS. Allowing direct inspection, our procedure is simple, and is helpful for those working toward technical mastery.

P26

Ascending aortic aneurysm due to takayasu's arteritis.

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Takayasu's arteritis is an inflammatory arteriopathy, which affects aorta, its main branches and the pulmonary arteries. We report the case of a 7-years old child with an ascending aortic aneurysm due to Takayasu's arteritis. The transthoracic echocardiographic evaluation of the patient who was followed since birth showed ascending aortic aneurysm with a diameter of 4.8 cm. In the operation ascending aorta was resected, and replaced with a dacron graft. Histopathologic examination of the ascending aorta showed findings of Takayasu's arteritis. Ascending aortic aneurysm due to Takayasu's arteritis is a rare event in literature. Our case is the youngest patient who has ever been reported.

P27

Single stage unifocalization with median sternotomy using the pericardial roll

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The operative strategy for VSD, PA, and MAPCA is still controversial. After multistage unifocalization procedures the difficulties of total correction are apparent. Hence, recently single stage unifocalization and correction procedures under median sternotomy have been proposed. In our department out of the 13 patients who underwent unifocalization, 4 were executed using median sternotomy with pericardial roll. These patients who had native pulmonary arteries less than 2 mm or none at all, had their MAPCA's under median sternotomy prepared between the aorta and the superior vena cava. After pericardial roll the MAPCA's on the right were anastomosed to one end

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

of the pericardial roll, and the ones on the left, to the other end. A BT shunt was done to the middle of the pericardial roll from the innomino brachiocephalicus. In two case, valved conduit was used between right ventricel pericardial roll. One patient out of the 13 who had undergone unifocalization was lost due to low cardiac output and hypoxia. Compared to procedures with multiple thoracotomies, single-stage unifocalization with median sternotomy revealed to be more effective.

P28

Evolution and outcomes of the fontan procedure in a small centre

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Background. Surgical approaches to single ventricle variants have evolved from one stage to three-stage fenestrated Fontan procedures. This study compared outcome with these modifications of the Fontan operation in a small centre. Methods. Perioperative risk factors and results were reviewed in 85 patients (pts) undergoing the Fontan procedure between 1985 and 2000. Diagnoses included tricuspid atresia (n=26), double inlet left ventricle (n=14), and complex anomalies (n=37). A palliative procedure was required in 68 pts (80%). A one stage Fontan procedure was carried out in 28 pts at a median age of 64 months. A staged Fontan procedure was performed in 57 pts with a bidirectional Glenn anastomosis at a median age of 9.2 months and the second stage fontan operation at a median age of 45 months, respectively. A fenestration was added in 56 pts (67%). Results: The mortality of the entire cohort was 8.2% (7/85). The mortality decreased from 35% in the period between 1985–89 to 2.4% in the period between 1995–2000 (p<0.0001). The most common postoperative complications were low output syndrome (n=27), pleural effusion (n=28), infection (n=22), and arrhythmias (n=7). The fenestration was closed with a device in 33 pts, others have either closed spontaneously or remain clinically insignificant. Follow up complications included arrhythmias (n=10), severe ventricular dysfunction (n=11), and protein losing enteropathy (n=4). One patient underwent transplantation and 2 pts required Fontan conversion. The actuarial survival at 1 and 5 years is 88.9% and 85.3%. Complex anomalies and early period of repair were risk factors for death (p<0.0004). Conclusion: The Fontan modifications have led to significant improvement in early survival. Longer follow-up is needed to assess the clinical impact of the latest Fontan modifications.

P29

Is there a role for staged repair in surgical correction of tetralogy of Fallot?

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Background: Although primary repair of tetralogy of Fallot (TOF) in infancy has been the favoured surgical technique over the past decade, a staged operation may still be used in selected patients (pts). Methods: Between June 1976 and November 1999, 193 children underwent repair of TOF. Children with pulmonary atresia were excluded. Indications for staged repair were young age (<2 months) and complex anatomy. Results: A staged repair (Group 1) was performed in 62 pts at a median age of 19.5 months (5–179). Median age at palliation was 3.2 months (0.03–63). The interval between palliation and repair was 15.9 months (3.4–169). Primary repair (Group 2) was performed in 131 pts at a median age of 12.4 months (2.9–281), with 87 pts less than 12 months and 29 less than 6 months of age. A transventricular patch was required in 50% of Group 1 and 44% of Group 2 pts. The median RV/LV pressure ratio was 0.57 (0.3–1.0) and 0.5 (0.2–1.0) in Group 1 and 2 pts, respectively (p=NS). The preoperative hemoglobin, cross-clamp time, days of ventilation, days of inotropic support, ICU length-of-stay (LOS) and hospital LOS were similar in both groups. Early and late mortality was 6.7% and 8%, respectively. Postoperative complications included low output syndrome in 16%, respiratory failure in 15% and bleeding in 3% of pts. A permanent pacemaker was implanted in 3 pts. Pulmonary valve implantation for severe insufficiency and right ventricular dysfunction was required in 22 pts (12.4%) at a median interval of 9.6 years (8–15). The median length of follow-up was 92.9 months (3–282). Actuarial survival rates at 5 and 10 years were 91% and 90%, respectively in Group 1, and 94% and 91%, respectively in Group 2 (p=NS). Freedom from re-operation at 5 and 10 years was 88% and 77%, respectively in Group 1 and 91% and 83%, respectively in Group 2 (p=NS). Conclusion: A staged repair is an appropriate approach in selected patients. Our results suggest that early primary repair is not associated with higher risk.

P30

Development of pulmonary arteriovenous fistula after total cavopulmonary connection in patients with left isomerism

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Objective: To investigate potential risks of pulmonary arteriovenous fistula (PAVF) in patients with left isomerism undergoing total cavopulmonary connection (TCPC). Methods: Since 1997, TCPC has been employed in 18 patients with the particular atrial arrangement, 8 of these undergoing previously partial right heart bypass by bidirectional cavopulmonary anastomoses. IVC was interrupted in 10. SVC was bilaterally present in 10. Results: Two patients postoperatively died of abundant systemic-to-pulmonary collaterals. Of the 16 survivors, PAVT obviously developed after TCPC in 5 patients with interrupted IVC. PAVF was present within either the right (in 2) or the left (in 1) lung, with hepatic venous effluent directed exclusively to the contralateral lung. Abnormal communications were also found between the hepatic veins and the pulmonary vessels. Latest arterial oxygen saturation proved to be 63%, 64%, and 77%, in these 3. In the other 12, hepatic venous drainage joined pulmonary flow to both the right and the left lungs, oxygen saturation being 94±2 (A)>96) %. The presence of bilateral SVC provided a better circumstance to drain the hepatic veins bilaterally in a balanced fashion. Catheterization 34±2 months after TCPC in 14 demonstrated, pulmonary resistance 2.0±0.9 (0.9–3.6) units*m2, and cardiac index 3.0±0.6 (2.0–4.0) l/min/m2. Conclusion: PAVF may develop after TCPC in patients with left isomerism. When constructing connections between the systemic veins and PA, attention should be paid to orientation of these anastomoses to include hepatic venous drainage within pulmonary flow to the both lungs.

P31

Surgical implication of coronary arterial anatomy in patients with discordant atrioventricular connections

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Objective: To determine precise anatomy of the coronary arteries, and its surgical relevance, in patients with discordant atrioventricular connections. Methods: Coronary arterial patterns were investigated in 55 patients with this particular lesion undergoing biventricular repair, on the basis of findings on angiography and during the surgical procedure. Results: Two patients had a solitary stem. In the other 53, dual orifices were present at the facing sinuses. In 45 of these, the anterior interventricular artery arose from the right artery which reached the atrioventricular groove between the morphologically RA and LV, with the posterior interventricular artery from the left circumflex artery. Both interventricular arteries originated from the dominant right artery in 7, the left artery being hypoplastic. In reverse pattern was seen in 2. In the remaining one with concordant ventriculoarterial connections, coronary arterial branching was comparable to normal. Of 4 patients undergoing the arterial switch for anatomic repair, high take off from the aorta was seen in 2 affecting the surgical maneuver. An incision to RV would have been restricted by the presence of the substantial coronary arteries in 17. Such circumstances were found, in reality, in 4 of 24 undergoing anatomic repair by intraventricular rerouting. Similarly, a left ventriculotomy for functional biventricular repair would have been limited in 29 by a characteristic course of the right artery possessing a considerable distance between its aortic origin and the atrioventricular groove. Conclusion: Coronary arterial anatomy is an important issue to be recognized when achieving either anatomic or functional biventricular repair in this setting.

P32

Surgical approach in patients having double outlet right ventricle with subpulmonary vsd and obstruction of the aortic arch

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Objective: To determine the optimal strategy in patients having DORV with subpulmonary VSD and the obstructed aortic arch. Methods: Since 1989, 16 patients have undergone surgical interventions in this setting. In 5 infants, the malformations were repaired as a single-staged fashion. In the other 11, the aorta was initially reconstructed with the pulmonary trunk banded, because of perioperative shock in 4, multiple VSDs in 4, intramural courses of the coronary arteries in 2, and small RV in 1. The total procedure was carried

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

out at the age of 15±12 (3–41) days. Results: The 5 patients undergoing primary repair are doing well in the longer terms. The other 15 patients also survived the initial palliative procedure. Subsequent biventricular repair was successfully employed in 4 with previous ductal shock. They could have undergone primary repair had such a neonatal episode been avoided. Of the 4 with multiple VSDs, biventricular repair was attempted in 2, with one succumbing and the other dying of low cardiac output. In the remaining 2 with mesh-like multiple VSDs, we have proceeded to either the bidirectional Glenn procedure or the palliative arterial switch so as to improve cyanosis and to aim towards a Fontan-Kreutzer procedure. Of the 3 patients with other impediments to primary repair, biventricular repair was attempted in one who postoperatively died of mediastinitis. Conclusion: Morphologic spectrum should be perfectly noted in this subset of malformations, since primary repair, or even staged biventricular repair, might be unsuitable because of the structural features.

P33

Aggregated ultrafiltration in paediatric open heart surgery

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Background: Conventional (CUF) and modified ultrafiltration (MUF) have been shown to improve haemodynamics, pulmonary function and blood conservation. Since 1997, we have adopted ultrafiltration in our paediatric cardiac surgical practice, combining CUF and MUF since 1998. In this retrospective review we examined their effects on clinical outcome. Methods: This review included 251 patients operated from 1995 to 2000. Ninety-five patients operated before 1997 were used as historical control. Twenty-seven patients had CUF only, whereas eighty-nine had both MUF and CUF. MUF was used in children less than 10kg, carried out for 20 mins and extracting volumes of up to 20ml/kg. Median age at surgery was 10.9months(0.4–70.8), 8.2m(0.3–48.8) and 4.6m(0.3–51.9) respectively for control, CUF and CUF+MUF. Mean weight was 6.4±2.9kg, 5.1±2.1kg and 4.9±2.0kg for control, CUF and CUF + MUF. Result: Early mortality was 8.9%, 22% and 9% for control, CUF, CUF/MUF respectively. Blood loss (ml/24hrs) was 125±107 in the CUF/MUF versus 193±166 in controls and 166±93 in CUF (P=0.015). Platelet transfused (units/24hr) was 1.5±0.6 in CUF/MUF versus 2.5±0.2 in control and 1.3±0.5 in CUF(P=0.002). CUF/MUF achieved a final haematocrit of 32.4±5.9 as compared to 31.5±4.9 in control and 29.2±4 in CUF (p=0.05), as well as reduction of central venous pressure to 7.1±3.1 versus 9.3±3.3 in control and 5.4±3.0 in CUF(p=0.006). There were as complished without affecting systolic blood pressure in subgroups with complex congenital heart disease(p=0.48) or in patients weighing less than 5 kg(p=0.15). Ultrafiltration did not shorten days on ventilation (p=0.07) and days of stay in the intensive care unit (p=0.167). Conclusion: Aggregated ultrafiltration has improved haematocrit and initial haemodynamics, but it does not affect overall clinical outcome.

P34

Restenosis and aortic aneurysm late after repair of coarctation of the aorta

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We focused on the incident of restenosis and aortic aneurysm after repair of coarctation of the aorta in the long follow-up period. Patients and Methods: The subjects were 87 patients with coarctation of the aorta who survived the repair from January 1970 to May 2000. Using a prospective database, we analyzed late mortality, the incidence of restenosis and aortic aneurysm, reoperation, and risk factor for restenosis and aortic aneurysm. Results: Mean follow-up interval was 15.9±7.8 years. Two patients died during follow-up period, One after mitral valve replacement for congenital mitral insufficiency and the other of acute pneumonia. Actuarial survival rate was 95.4% at 20 years. Restenosis was recognized in 7 patients (14.9%): one in 18 patients with Subclavian flap angioplasty (SFP), one in 9 with End-to-end anastomosis (EE), 4 in 13 with Patch angioplasty (P), and one in 7 with other procedures. Aortic aneurysm was recognized in 3 patients (6.4%), all of whom underwent Patch angioplasty. Overall actuarial rate of freedom from both restenosis and aortic aneurysm was 68.0% at 20 years (92.9% in SFP, 83.3% in EE, and 39.8% in P). Reoperations were performed on 4 patients with restenosis by bypass graft technique and on one patient with both restenosis and aortic aneurysm by aortic arch graft replacement. Patch angioplasty was risk factor for late restenosis and aortic aneurysm. Conclusion: Although the postoperative outcome of coarctation repair was good, patch angioplasty demonstrated a higher incidence of restenosis and aortic aneurysm late after surgery. Therefore, careful observation of patients with patch angioplasty is necessary.

P35

Systemic venous flow evaluation of total cavopulmonary connection in asplenic hearts. Should the reconstruction technique individualized?

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Objective: Outcomes after univentricular repair for patients with asplenia syndrome remains unsatisfactory, not only because of clinical difficulties in patient selection, but also secondary to technical difficulties in the separation of the systemic and pulmonary circulations, especially with the remaining technique for the inferior systemic vein. Method: Between February 1995 and May 2000, 14 consecutive patients with asplenia syndrome underwent a bidirectional cavopulmonary connection with obliteration of additional pulmonary blood flow followed by a total cavopulmonary connection. The remaining technique for the inferior systemic venous blood flow was individualized to maximize flow smoothness, the pathway prosthetic load, and suture load on the atrial wall. The lateral tunnel or extracardiac conduit technique was used in an extra-, intra-atrial or intra-atrial fashion. No fenestration was applied. Results: There was no hospital mortality. Systemic venous flow was evaluated using magnetic resonance angiography, which revealed no signs of obstruction, turbulence, or stasis in and near the reconstructed pathways, irrespective of the remodeling technique. Postoperative catheterization revealed favorable hemodynamics including an inferior vena cava pressure of 13 ± 2 mmHg and arterial oxygen saturation of 93.4 ± 3.5% at room air. All patients have been free of symptoms, while one patient died of acute sepsis complications 3.5 years after the procedure. Conclusions: The complexity of cardiac anomalies in asplenia syndrome warrants individualization of the total cavopulmonary connection technique tailored to reconstruction of the inferior systemic venous pathway. Creating a maximally smooth flow in the pathway should be a priority. A staging approach allows the proper selection of candidates for univentricular repair.

P36

Long-term follow-up of management of coarctation of the aorta, surgery vs balloon angioplasty

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Background: Long-term results of surgical repair and balloon angioplasty for native coarctation of the aorta in infants and children are evaluated and compared. Methods: Surgical repair (group A: 273 patients, age 1.6 ± 2.9) and balloon angioplasty (group B: 36 patients, age 4.8 ± 4.6) for native coarctation of the aorta were performed from 1977 to 2000. Per- and post-intervensional pressure gradients were analysed with Student's t-test. Kaplan-Meier curves were constructed to compare the intervention free probability in both groups. Results: Resultant reduction in peak-to-peak systolic pressure gradient was 19.0 ± 5.6 mmHg (t-test, p<0.001). All-over mortality was 7.3% in group A. In group B no mortality occurred. Hospital stay ranged 6–22 days in group A and 48 hours in group B. Mean length of follow up measures 13.0 ± 7.3 years, ranging 0.6 to 28.4 years in group A and 0.1 to 9.1 years in group B. In group A restenosis occurred in 48 patients (18%). 31 patients were treated with balloon dilatation, 3 with end-to-end re-anastomosis and 14 with patch angioplasty, of which one developed a re-recoarctation, managed with balloon angioplasty. In group B recoarctation developed in 4 patients (12.5%). 2 were re-operated using end to end anastomosis, in 2 patients balloon angioplasty was performed. Aneurysm formation was encountered in 6 patients in group A (2%). No aneurysms were encountered in group B. No significant statistical difference was found between the intervention free probabilities in group A and B. Conclusion: Balloon dilatation as primary treatment of coarctation of the aorta yields comparable results to surgical management, immediately as well as in long term follow up and is therefore a justifiable choice for patients over 3 months of age and a focal/tubular type of coarctation.

P37

Fontan conversion to total cavo pulmonary connection (TCPC) associated with accessory-tachycardias ablation: early results

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https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Ten failing Fontan patients (age 20.9±6.7 yrs) underwent TCPC arrhythmias ablation and AAI-PM implantation 15.6±2.5 yrs from Fontan repair. Giant right atrium was present in all patients, refractory arrhythmias in 9, ventricular dysfunction in 4. One early postoperative death occurred, due to pneumonia. Mean follow up was 11.8±4.8 months. Stable sinus rhythm was present in 7/9 patients (medically restored in 2, after recurrence of atrial tachycardia). Myocardial scintigraphy showed reversal of rest and/or exercise dyskinesis in 4 patients and improvement of systemic ventricular function in 5. Basal EF varied from 37±6.9% to 46.1±6.9% (p=0.049) and EF on effort from 39.9±11.4% to 49.5±7.7% (p=0.013). Our data show that TCPC associated with arrhythmias ablation may restore a stable sinus rhythm, allows a better control of arrhythmias and improves ventricular function in patients with failing Fontan.

P38

Mitral valve anomalies in pediatric age

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Purpose of the study: diagnosis and surgical results of the mitral valve (MV) anomalies observed in an 8-yr period. Relevant MV anomalies were found in 218 pts (1.4%) among 2000 paediatric patients undergoing heart surgery at our centre in the last 8 yrs. Mean age 5.8 yrs (range 2 mo-14 yrs). 20 pts had MV insufficiency (I) and 8 MV stenosis (S). Anatomical lesions in MVI were: results of surgical repair of A-V septal defect in 9 pts, congenitally dysplastic valves in 9 and post rheumatic in 2. The MVS cases were parachute MV in 5, post rheumatic in 2 and supravalvar ring in 1. Thirteen pts had other surgical procedures in addition to the MV plasty. Diagnosis was accomplished in all cases but 3 by echo color doppler (TT and TE) techniques. Operative techniques included: resection of supravalvar rings, splitting of papillary muscles, plasty of anterior MV leaflet valve annuloplasty, suture of cleft and repair of residual a-v septal defects, quadrangular resections, artificial chordae, shortening of papillary muscles. Results: there were 2 hospital deaths (7%). At a mean follow-up of 50 mo (2 mo-8yrs) there were no deaths. Three pts (11.5%) underwent MV replacement at a mean period of 18 mo (2 mo-3 yrs) following the mitral surgery. Twenty pts with native valves are in NYHA class I and 3 in class 2. On echo only 1 pt with native valve has moderate to severe MVI. Conclusions: 1) conservative surgery on MV is possible in pediatric age with low operative mortality. 2) echo and Doppler evaluation is the best diagnostic modality; 3) the complexity of the lesions and the presence of associated anomalies make the results of the plasty on the valve less predictable than in the adult population.

P39

The Ross operation: surgical technique and mediatm term results

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Purpose of the study: evaluate the surgery in the early and medium-term outcome of some technical modifications applied to the Ross operation during our 6 yrs experience. Method: 35 pediatric patients (pts) (mean age 9, range 2–16 yrs) received a Ross operation for aortic insufficiency 28, aortic stenosis 7 pts. Eight pts had received a transcatheter and / or a surgical procedure at a mean time of 8 mo (2–72 mo) prior to the Ross operation. Technical modifications were: 1) Dissection of the autograft and homograft wrote with bearing heart; 2) Reinforcement of the aortic annulus with autologous pericardium; 3) In older pts banding of the autograft with a Gore-tex membrane and / or with the native aortic wall. Results: no early nor late mortality. Mean cross-clamp time has decreased from 130 ± 18 m to 80 ± 9 m comparing the early with the more recent series (1994 – 1997 versus 1998–2000). No differences were found in hospital stay duration and post-operative complications. Clinically the whole series of pts have improved significantly passing from a mean NYHA functional class 3 to 1.2 (p < 0.001). Follow-up at a mean F-up of 38 mo (1–65 mo) there was 1 reintervention for iatrogenic aortic pulmonary window. One pt with rheumatic heart disease has moderate to severe aortic insufficiency 2 yrs post-operatively. Three pts in the early series have moderate to severe dilation of the ascending aorta. Conclusions: 1) Our results with Ross operation in pediatric age show good early and mediuca term results; 2) There are no major complications; 3) The technical modifications adopted decrease cross-clamp time and perhaps ectasia of the ascending aorta.

P40

Repair of coarctation of the aorta using left heart bypass for spinal cord protection: increased risk or benefit?

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Background: Repair of coarctation of the aorta carries the inherent risk of paraplegia due to cross clamping of the descending aorta. Cardiopulmonary bypass performed as left heart bypass may potentially eliminate this risk. The aim of the study was to analyze the clinical course and outcome of all patients who had repair of coarctation using left heart bypass. Methods: We retrospectively analyzed the clinical outcome and late follow up of all patients who were operated on since 1997. Of special interest was the influence and side effects of cardiopulmonary left heart bypass techniques. Results: Since 1997 repairs of coarctation of the aorta beyond infancy (n=10) were performed using left heart bypass by cannulation of the left atrium and descending aorta. The median age was 31.3 years (range 2,1 to 36,4 years). The median bypass time was 24 min and median operative time was 115 min. No complications related to the use of left heart bypass were observed in the complete follow-up of 9–50 months late. Even in small children cannulation of the descending aorta is feasible. Conclusion: Repair of coarctation of the aorta using left heart bypass is a safe procedure and potentially precludes the risk of paraplegia. It can be performed even in small children (< 10 kg) by cannulation of the left atrium and descending aorta. Total operative time is reasonable.

P41

Corrective operation for ventricular septal defect in a developing country: the impact of severe malnutrition and lung infection on outcome

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Objectives: We sought to examine the effects of severe malnutrition, preoperative pneumonia, and age at operation on outcome following VSD closure in a large referral center in south India. Methods and Results: We analyzed the records of 100 consecutive infants (age 7.4±3.3 months) with large VSDs who underwent surgical closure at our institution from July 1998 to June 2000. Primary outcome variables were postoperative mortality, duration of mechanical ventilation, ICU stay, hospital stay and infectious complications. Preoperative variables analyzed included age, weight and length z-scores, and presence of pneumonia. Nutritional status at surgery was poor (weight z-score -2.8±1.5, range -5.6 to 2.3; length z-score -1.9±2.0, range -6.7 to 3.7). Preoperatively 35 infants had pneumonia and 4 required mechanical ventilation. Six patients died. None of the preoperative variables was associated with death. Duration of mechanical ventilation, ICU stay, and hospital stay were longer for younger patients (spearman rank correlation for ventilation -0.25, p=0.02; for ICU stay -0.33, p< 0.001; for hospital stay -0.27, p=0.007) and for those with preoperative pneumonia (median duration of mechanical ventilation 46 versus 24 hours, p<0.001, median ICU stay 7 versus 4 days, p<0.001; median hospital stay 15 versus 8 days, p=0.002). Patients with postoperative infections (n=16) were younger than those without infections (median age < 5 months versus 7 months, p=0.01) and more likely to have experienced preoperative pneumonia (50% versus 20%, p=0.02). Preoperative weight and length z-scores were not associated with any of the outcome variables. Conclusions: Poor nutritional status, preoperative pneumonia, and younger age do not increase mortality following successful VSD repair. Younger age at surgery and preoperative pneumonia are associated with longer in-hospital recovery times. Repair of large VSDs should not be delayed because of these preoperative characteristics.

P42

Predictors of complicated post-operative course following repair for tetralogy of Fallot: perspective from a developing country

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Background and objectives: There is limited information regarding post-operative course after intra-cardiac repair (ICR) for tetralogy of Fallot (TOF) from developing countries where late presentation of congenital heart disease is common. This study examines the perioperative course of a large population of patients with TOF from a large referral center in south India and attempts to identify determinants of a complicated post operative course. Methods and Results: Between July 1998 and July 2000, 177 patients (age 3 months –48 years) underwent ICR for TOF at our institution.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Complicated post-operative course was defined as presence of one or more of the following parameters: death, hypotension requiring norepinephrine infusion, mechanical ventilation for >14 days and, elevated liver enzymes (serum alanine transaminase > 1000 IU/liter). Forty four patients had a complicated post-operative course as defined above and this group included 16 deaths (9%). The influence of various pre-operative and operative variables on the post operative course was analyzed (table). Conclusions. Based on this retrospective study from a developing country, younger age, low body weight, smaller size (low body surface area) poorer nutritional status (lower weight Z scores) need for trans annular patch and longer cardiopulmonary bypass times appear to predict a complicated postoperative course following intra-cardiac repair for TOF

P43

Late clinical outcome of the fontan procedure in patients with tricuspid atresia

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Objective: Evaluate the late clinical outcome of the Fontan procedure in patients with tricuspid atresia. Methods: We analyzed retrospectively the late follow-up of 25 out of 26 patients that underwent the Fontan procedure or one of its variants for tricuspid atresia between August 1980 and January 2000 at the Heart Institute of Rio Grande do Sul. Four patients were submitted to the classic Fontan procedure, 7 to Kreutzer variant, 6 to Bjork, 9 to fenestrated total cavopulmonar anastomosis and 5 to non-fenestrated total cavopulmonar anastomosis. The mean age at the time of surgery was 5.4 + 3.1 years and mean weight was 15.8 + 6,1Kg. Male sex was predominant (60,5%). Results: The 25 patients were evaluated on outpatient clinic with a late mean survival time of 5,5 + 4,2 years (from 30 days to 17,4 years) and a late mortality rate of 8%. Arterial saturation varied from 77,2 + 18,8 % at the preoperative period to 91 + 6,7 % at the last clinical visit (p=0,05). Sixty-seven percent of patients were asymptomatic and 87% tolerated physical efforts based on subjective response at the last clinical review. Ten patients (40%) had suffered some complications such as cardiac arrhythmias, cyanosis, protein-losing enteropathy, neurologic events, right heart failure, effusion, intolerance and reoperation. Conclusion: These results suggest that, beyond the immediate postoperative period (in which the organism adapts to the new circulatory physiology), patients with tricuspid atresia submitted to the Fontan procedure go on well through the late postoperative period, although with few, but significant morbidity.

P44

Long term results of reconstructive mitral surgery in rheumatic mitral valve insufficiency in a young population

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Background: It is well recognized that reconstructive mitral valve surgery is superior to valve replacement. However there is a lack of informations concerning the long term evolution. This study is the evaluation of one center using the same surgical technique for rheumatic mitral valve insufficiency. Methods: From 1970 to 1994 among 951 patients operated on for an isolated mitral (MV) insufficiency from rheumatic origin, 442 were under 19 years old. Aortic valve disease were excluded. The age was between 4 and 19 years (mean 12 y). Functional class (NYHA) were 2 in 43% (190 pts), 3 in 47% (208 pts) and 4 in 10%(44 pts). 87% of the patients were in sinus rhythm. Acute fever was present in 33pts(7%). Patients were classified according to leaflet motion: Type I pure annular dilatation 6% (26 pts), Type II prolapsed leaflet 45%(197pts), Type III restricted leaflet motion 20% (92 pts), Type IIIa/IIIp prolapsed anterior leaflet and restricted posterior 29%(127 pts). Carpentier techniques were used in all pts with the use of a prosthetic ring in 94% (414 pts). Prolapsed leaflet was treated with chordae shortening. Commissurotomy was associated in 22% (99 pts). Leaflet extension was used in 41 pts (11%) in order to use a large prosthetic ring. Mean follow-up was 11,9±5.11 years (max. 29 y) with 3569 pt/y. 7% of the pts were lost from follow-up. Results : Operative mortality was 1% (6 pts). Survival at 5, 10, 15 and 20 years were: 95, 90, 88 and 86%. Postoperative functional class (NYHA) was 1 : 65%, 2 : 28%, 3 : 2% and 4 : 1%. Sinus rhythm was present in 95% of the survivors. Thromboembolic events were very rare with 6 events : actuarial rate 0.2 % pt/y. Freedom of reoperation at 5, 10, 15, 20 years were: 95, 90, 88 and 86%. The majority of reoperation were due to fibrosis. A restrictive prosthetic ring was changed in 2 patients. Patients operated on at the acute phase of the rheumatic fever had an operative mortality of 4%, a survival rate at 20 years of 60% and 55% were free of reoperation at 20 years follow-up. Conclusion : Reconstructive surgery of rheumatic mitral valve insufficiency carries a very low incidence of operative mortality and thromboembolic events. Reoperations rate were decreased when all the functional mitral val

P45

Transsiphoid approach for congenital heart defects

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Interest in minimally invasive cardiac surgery (MICS) for cardiac disease continues to increase because it causes less surgical trauma and produces a better cosmetic appearance. We introduced the transsiphoid approach without sternotomy for the correction of congenital heart defects. During a period of 18 months, fourteen children (5 boys and 9 girls) whose ages ranged from 18months to 108m months underwent closure of atrial septal defects. The approach consisted of a 4- or 5-cm low midline incision with division of the xiphoid only with bicaval cannulation and ascending aortic arterial cannulation through the incision. To improve exposure of the cardiac lesion during MICS, we developed a new venous cannula. All the patients survived the operation and did not require blood transfusion. Postoperative thrombosis, echocardiography showed good left ventricle function and the absence of residual shunts. Transsiphoid approach is a safe, feasible alternative procedure for selected congenital heart defects and should be considered as a less invasive technique.

P46

Unidirectional valved patch technique for closure of left to right shunt defects in borderline Eisenmenger syndrome

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Background: Patients with severe pulmonary hypertension(PH) due to prolonged left to right shunt are frequently labelled as inoperable because of high operative mortality and risk of irreversibility of the pulmonary vascular disease. The unidirectional method for closure of these defects increases operative safety in these borderline patients giving them the chance of improvement in pulmonary vascular disease if it has still some degree of reversibility. Method: This technique in which we use a double patch with unidirectional valve function stops any left to right shunt but allows the right side to decompress into the left in case its pressure exceeds early post-operatively and at least theoretically decrease mortality. Between 1997 and 2000, 16 patients with very severe PH (13 VSDs, 1 ASD, 2 A-P windows) were operated with this method. 8 of them were marked as inoperable since many years before. Mean pulmonary artery(PA) to aortic pressure ratio: 1.04, mean Qp/Qs: 1.15, mean TR gradient: 78 mmHg, mean mean Oxygen saturation: 73%. Results: One patient died (3 months old baby with A-P window). Most of the other patients showed significant decrease in PA pressure (mean PA/Aortic pressure: 48%). In 3 patients PA pressure remained high in early post-op period ,in two of which right to left shunt through the patch could be confirmed by echocardiography. Cardiac catheterization (mean 1.9 years post-operatively) showed even more improvement in PA pressure in most patients(mean PA/Aortic pressure: 0.43, mean O2 saturation: 93%). Conclusion: Although the number of cases is still small, the quite acceptable early results and promising mid-term study shows that:1)The mainly catheter based criteria of inoperability may need some more discussion and clinical condition should be strongly considered before labeling patients as inoperable. 2)With unidirectional patch technique a larger number of those labeled as borderline Eisenmenger Syndrome patients may have the chance for successful operation.

P47

Effect of modified Blalock Taussig shunt on growth of pulmonary arteries

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We reviewed our experience with patients undergoing BT shunt for cyanotic CHD. The effect of BT shunt on growth of PAs was studied. Eighty three patients underwent BTs over a 2 year period between Jan 98–Mar 2000 for TOF(62.6%) and other lesions(37.4%). Followup data was available in 56 pts for a mean period of 8.6 mo (6–30 months). Shunt size ranged from 4–6 cm. Median age at shunt was 18 mo (1day–9 years). There is Mean SaO2 increased

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

from 65–84% after the shunt and hematocrit decreased from 64.9–40.1%. Mean RPA and LPA sizes increased from 6.2±1.5 and 5.6±2.4 to 7.5±1.9 and 7.0±2.1 mm respectively ($p<0.001$). 82.1% underwent Rt and 17.9% Lt BT shunt. Both PAs grew equally in 67.9% while in 23.2% there was preferential RPA growth after a Rt BT shunt. This was not seen with L BT shunt ($p<0.01$). No change in PA size was seen in 8.9% pts. Maximum change in PA size was seen in the first 3 months after surgery in 64% of patients. There was no correlation between age at surgery, prior PA size, presence of a PDA on PA growth after BTs. Distortion of PAs and stenosis at BTs insertion was seen in 3.9%. Early shunt blockage occurred in 3 pts (all < 3 mo age). Conclusion. There is significant and uniform growth of both PAs after a BTs in majority of patients. Maximum growth occurs in the first 3 months post-op. Distortion of PAs and stenosis was uncommon.

P48

Bentall and Yacoub's procedures in children: a series of 14 patients

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Fourteen patients (median age 5; range 1–17 years) underwent the surgical replacement of the ascending aorta with or without aortic mechanical valve substitution. Six had a Marfan syndrome, 1 a Ehlers Danlos type 3, 1 Turner syndrome, and 6 had dysmorphic aneurysms of the aorta with coarctation in 2. Three patients had undergone a previous cardiac surgery: 1 pulmonary artery aneurysm, 1 coarctation repair, 1 Ross procedure for aortic insufficiency. Indication for ascending aorta replacement was acute aortic dissection in one and rather major dilatation of the aorta or rapid progression of the aortic diameters in the other 13 patients. The median rate of progression of the aortic diameter was 4.5 mm/year. The median ascending aorta diameter at surgery was 48.5 mm (42–63). Eight patients had a Yacoub's procedure and 6 a Bentall operation. Concomitant procedures were aortic arch replacement ($n=1$), coarctation repair ($n=2$), mitral valve and tricuspid valve repair ($n=1$), aortic valve commissurotomy ($n=1$). One patient operated on (Bentall) at 1 month of age for a neonatal Marfan syndrome died postoperatively of massive mitral valve regurgitation. At last follow up (mean 18 months, range 3–60 months), 13 patients are alive and doing well. Two underwent a subsequent procedure (mitral valve and tricuspid valve repair in one, aortic valve replacement after a Yacoub's procedure in one). In conclusion, aortic aneurysms with or without aortic regurgitation, although very rare in children, can undergo elective Bentall's or Yacoub's procedures with low operative risk and excellent survival rate with low morbidity.

P49

Surgical results in coarctation of the aorta with the extended end to end anastomosis

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The purpose of this paper is to show the results in 76 patients (pt) operated with the extended end to end anastomosis (EEA) from January 1989 to October 2000. In the EEA the usually hypoplasic aortic arch is widely opened, the fibrous ring is excised and the descending aorta is mobilized and being widely opened, is connected to the proximal segment. The patients were divided in three groups (G) according the age and weight: G I: 30 p from newborn to 59 days (d) of age (x:28d) weighting 2,2 to 3,9 Kgs (K) (x:3.130 K), GII: 38 p 2 to 30 months (m) (x:7m) and 4 to 12 K (x: 5.2K) GIII:22p. 2.5 to 21 years (y) old (x: 6.9 yr and 14 to 88 K (x: 29K) Twenty patients had associated lesions (14 p from GI and 6 from GII), such as interrupted aortic arch (7), VSD and pulmonar hypertension (5), severe aortic stenosis(4), Taussig-Bing sindrome (2) single ventricle (1) and DORV (2). The total early surgical mortality was 9.2% (7/76): five of these had severe associated lesions which were either cianotics or palliated (mainly banding of the PA); and had a 6.6% mortality, 2 (2.6%)had isolated CoAo . All patients belonged to GII, seven pts of this group had late death (9.2%) due to other causes than the well corrected aortic coarctation. Follow up from 1m to 13.7 m (x:30m) was obtained in 70 p. Two p remained, one 20 d old was reoperated 1m later and the other had balloon angioplasty 6 y later. The rest of pts had good femoral pulses and 22 p ; had light gradients with Doppler (10–20 mmHg.). The EEA is a good surgical choice to correct even in very small children not only the fibrotic tissue causing the aortic coarctation, but the hypoplasic aortic arch with low incidence of arch recoarctations. Mortality is related with associated lesions.

P50

Right-sided maze procedure for atrial tachyarrhythmias associated with congenital heart disease

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Atrial fibrillation and flutter are commonly associated with congenital cardiac anomalies that cause right atrial dilatation. Preliminary results indicate the right-sided maze procedure reduces atrial tachyarrhythmias when combined with cardiac repair in these patients. It is unknown if these results are durable. We retrospectively reviewed the records of all patients who had the right-sided maze procedure for atrial tachyarrhythmias on our surgical service between 1995 and October 1, 1999. Forty-four patients underwent a right-sided maze procedure during the course of repair of other congenital cardiac lesions. Patient age ranged from 9 to 72 years (mean, 40). Atrial tachyarrhythmias were paroxysmal in 32 (73%) and chronic in 12 (27%). Cardiac pathology included Ebstein's anomaly (n=31, 70%), isolated atrial septal defect (n=4, 9%), non-Ebstein's congenital tricuspid regurgitation (n=2, 5%), and other various lesions (n=7, 15%). There was one early death from ventricular arrhythmia. Morbidity included permanent pacemaker for tachy/brady arrhythmia (n=1) and reoperation for delayed tamponade (n=1). Discharge rhythm was sinus (n=33, 77%), junctional (n=5, 12%), atrial fibrillation (n=4, 9%), or atrial flutter (n=1, 2%). Rhythm follow-up was complete in 34 (79%) patients and ranged from 1 to 66 months (mean, 17 months). Follow-up rhythm was sinus (n=29, 85%), junctional (n=2, 6%), atrial fibrillation (n=1, 3%), atrial tachycardia (n=1, 3%), or paced (n=1, 3%). There were no late deaths or reoperations. The inclusion of a right-sided maze procedure with cardiac repair of congenital anomalies that cause right atrial dilatation eliminates most associated atrial tachyarrhythmias. The results are durable at mid-term follow-up.

P51

Aneurysm in silent persistent ductus arteriosus: a case report

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Ductal arteriosus aneurysm formation is a possible complication described in cases of persistent ductus arteriosus (PDA), a potentially fatal condition. We report a case of ductal aneurysm formation in a patient without previous cardiologic diagnosis. A one year old boy, with congenital multiple arthrogryposes, had a craniotomy in order to drain a subdural hematoma, complicated with staphylococcal septicemia. He was treated with appropriate antibiotics. EV. Returned to hospital few days after being discharged with a new onset of systolic murmur 5+/6, tachycardia and run-off pulses. Chest X-ray showed a normal size heart and enlarged mediastinum. Echocardiogram revealed ductal aneurysm measuring 3cm X 2.5cm, shunt left to right, normal heart function. The patient was submitted to surgery immediately, with no complications. The surgical results were excelent, with the post operative echocardiogram showing no residual lesions. The initial reports of PDA indicated a high mortality rate, mainly due to infectious endocarditis, but the risks seem to be reduced in the present time. The aneurysm dilatation, as a complication of endarteritis in a ductus was already described in the literature. Its real frequency and pathogenesis are uncertain. The surgical treatment is always indicated in these cases. There were no reports about these complications in clinically silent ductus as we describe. There is no consensus in the literature regarding the best management of clinically silent PDA. Many authors believe that the risks are not important enough to indicate closure of all silent PDA. Others believe that, as in the patient reported, the risks of endarteritis and aneurysm are significant. Our tendency is to follow the conservative approach, having in mind the possible complications.

P52

Vsd repair with fresh autologous pericardium, 10 years experience

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VSD repair with fresh autologous pericardium, 10 years experience. Paul C. Cartier, Zhitao Qi, Jean Perron, Christine Houde, Alain Cloutier, Jean-Marc Coté, Georges Delisle, Jean-Michel Guay. Hôpital Laval, Ste-Foy. The objectives of this study were to determine the incidence of VSD patch aneurysm, residual VSD and its evolution, when autologous pericardium was used to close a VSD. Preoperative, early and late postoperative echocardiographic data

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

for each patient were retrospectively reviewed. From February 1986 and March 1997, autologous pericardium was used to close VSD in 266 patients, mean age of 2.9±5.6 years and mean weight of 12.9±13.9 kg. Of the 252 early survivors, 237 (94.1%) were followed for up to 11 years (mean 4.3±2.7). Patch aneurysm was found in 2 (<0.8%) of the 237 early survivors followed. Residual VSD was detected in 103 (38.7%). The residual VSD were smaller than 3 mm or too small to be measured in 92% of the 103 patients and multiple in 10%. Three reoperations (1.1%) were needed to close hemodynamically significant residual VSD. Of the 93 patients with residual VSD whose follow-up data were available, residual VSD closed spontaneously in 61(65.6%) patients from 7 days to 9 years (mean 1.6±2.0 years) postoperatively. Even a stepwise logistic regression procedure only VSD size to body weight ratio was a significant risk factor for residual VSD (odd ratio 1.04). VSD closure with fresh autologous pericardial patch is a safe procedure. Residual VSD is frequently noted, but most of them are too small to be measured. They are likely to close spontaneously or become smaller. Reoperation for patch aneurysm or residual VSD is rare.

P53

Population-based study of tetralogy of fallot with absent pulmonary valve

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Purpose: To define the prognosis of Tetralogy of Fallot with absent pulmonary valve (TetAPV) in a population based study and to determine if fetal echocardiography can predict pregnancy. Methods: We retrospectively reviewed our surgical database and medical records. Results: Between May 1976 and November 2000, 12 children with Tetralogy of Fallot with absent pulmonary valve (TetAPV) were diagnosed and treated at the only pediatric tertiary care center in British Columbia, population 3.7 million. The median age at diagnosis was one day (range 19 weeks gestation to 5 weeks postnatal). Four were diagnosed antenatally. One patient died on the first day of life of respiratory failure. One awaits surgery, and the remaining ten have all had surgical repair. One required a modified Blalock-Taussig shunt and one required pulmonary artery banding. Of the ten patients who have been repaired, eight received homograft conduits and two had transannular patches. Median age and weight at repair were 27.5 months and 11.9 kg, respectively. There was no operative mortality, but one late death due to related airway obstruction. The two patients who died had both been diagnosed antenatally, but there were no echocardiographic features to distinguish them from the two that survived. Follow-up of the ten surviving patients is complete with a median age at last visit of 61 months (range 5–174 months). One adolescent patient has carried a successful pregnancy. Nine patients are classified as New York Heart Association Class I and one patient with severe pulmonary insufficiency is Class II. Conclusion: The prognosis for TetAPV is generally favourable, with no surgical mortality in our series. Although in our study 2 of 4 antenatally diagnosed infants died, no fetal echocardiographic features were predictive of prognosis.

P54

Surgical outcome of congenital valvar aortic stenosis

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Objectives: valvar aortic stenosis is a common congenital heart defect for which surgical procedures can be done with low risk except the infants whose conditions are seriously compromised. The risk of sudden death in patients without operation varies from 1% to 19% though for severe stenosis probably exceeds 7%. Reports of surgical results have indicated that reoperation will be necessary in a significant number of patients. The purpose of this report is to present our experience with the results of surgical aortic valvotomy for congenital valvar aortic stenosis performed at our hospital. Methods: the study group composed of 29 patients, 4 females and 25 males, with ages ranging from 19 days to 26 years (median age 12 years), who underwent aortic valvotomy for valvar aortic stenosis. The case records of all the patients were retrospectively reviewed and 24 survivors, who were followed for 1–10.5 years (mean 5.92±2.38 years) after relief of aortic stenosis, were scheduled for evaluation. Resultsfive patients had died, one (neonate) at operation, 2 early and 2 late postoperative deaths. Four of the five deaths had occurred in infants, and a 13 year old girl had developed infective endocarditis and died 1.5 years after the operation. No sudden deaths occurred in this series. Sixteen of the 24 surviving patients were recatheterized and 15 were found to have (93.7%) aortic regurgitation on angiography. Peak systolic pressure gradients (mean±SD) were 65.9±19.5mmHg before and 36.7±14.6mmHg (p<0.05) after the operation. Of the surviving patients 45.8% had a new postoperative diastolic murmur. Twenty patients (83.3%) had residual stenosis and 3 (12.5%) had recurrent stenosis. Two patients (8.3%) had undergone reoperation (6–7 years after the initial aortic valvotomy and most of the others will require reoperation in the future. Conclusion: timely relief of obstruction prevents sudden death and produces symptomatic improvement in valvar aortic stenosis, but aortic valvotomy is only a palliative measure.

P55

Congenital aorto-right atrium communication/tunnel. a surgical case.

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Congenital aorto right atrium communication/tunnel is a very rare pathology with almost no cases described in the literature. A 21 yrs old male patient with a congenital aorto-right atrium communication was admitted for surgical closure. The 'tunnel' began in the left valsalva coronary sinuses (diameter approx. 0.5cm) very close to the ostia of the LAD and Cx, and represented in the right atrium (RA) (diameter approx. 2.5cm). In a first approach it was only possible to close the RA opening. Although an initial attempt was made to close the aortic side, segmental alterations of anterior/lateral ventricular wall appeared and it was decided to leave it opened. Twelve months later the patient was admitted repeatedly with pulmonary thromboembolism. The TEE then showed an enormous mass posterior in the RA and a 'tunnel', of approximately 1 cm in diameter, exiting from the left coronary sinuses. The angiography revealed that the LAD and the Cx arose from this tunnel. The patient was re-operated, a mobile mass (diameter approx. 2cm) was found in the RA and was excised. In the Ao side, the left valsalva coronary sinuses was dilated, its lowest part continued with a 'tunnel' that was totally filled with old and recent clots. LAD and Cx emerged near the aortic extremity of this tunnel. Due to the new enlarged dimensions of the communication it was now possible, (although not very simple) to close its aortic opening. There were no operative complications and the patient was discharged with oral anticoagulation.

P56

Postoperative long term follow-up of pediatric patients with corrected transposition (CTGA) and other anomalies with atrioventricular discordance

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Results from a series of pediatric patients with CTGA operated at one institution prior to 21 years of age were reviewed for early and late morbidity and mortality. 111 patients (70 female, 41 male) [68 CTGA, 43 CTGA with DORV] were operated between July 1971 and January 1996 at a mean age of 8.9 years (94 days–20 years). Sites abnormalities were more common in the DORV group (29/43 vs 9/68). 19 patients had 76 previous operations (pacemaker excluded). Overall early mortality was 16% decreasing to 3% for patients operated after 1986. Factors associated with early mortality were VSD patch closure (p<0.008) and earlier calendar year of operation (p<0.022). Thirty-day survivors had 5 and 10 yr survival of 90% and 80% respectively (mean length of follow-up 11.4 yrs). Previous operation (p<0.001), higher NYHA class (p<0.033) and preoperative rhythm other than sinus (p<0.035) were associated with higher late mortality. Reoperations were common (41%): conduit replacement, systemic ventricle (SV) and pulmonary ventricle atrioventricular valve (AVV) replacement or repair, residual VSD repair, aortic valve replacement or repair and cardiac transplant. Survival free of pacemaker at 10 yrs among the 68 late survivors without pacemaker was 89%. Though reduced ventricular function was not found to be a risk factor for early and late mortality, there was significant difference (p<0.031) between the mean preoperative and postoperative SV ejection fraction. There was a tendency for the SVAVV insufficiency to become more significant during follow-up. Despite recent low early mortality, patients with CTGA need frequent visits to monitor changes in ventricular function and AVV status. A longer follow-up will be needed to assess long-term outlook.

P57

Successful surgical repair of aortic arch obstruction in the Neonatal Intensive Care Unit

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https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

A 970 g premature infant with coarctation of the aorta and hypoplastic aortic arch underwent resection and end to end anastomosis repair at the Neonatal Intensive Care Unit. The surgery was performed in the neonatal unit because of extensive caudal sepsis and we were reluctant to risk cross contamination in our operating theatre. The initial surgical recovery was uneventful though the child required balloon angioplasty of a moderate recoarctation some two years later. Now aged 3 years the child is asymptomatic, normotensive and not on any medication.

P58

Persistent Pulmonary Hypertension after Closure of Ventricular Septal Defect with Down Syndrome: Influence of Coexistent Patent Ductus Arteriosus.

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BACKGROUND: Down syndrome (D) is known to have pulmonary vascular problems and pulmonary hypertension (PH) ensues in some patients with VSD and D even after VSD closure in infancy. We hypothesized that coexistent patent ductus arteriosus (PDA) may have a role for residual PH after VSD closure and investigated pulmonary hemodynamics before and after VSD closure in patients with D and PDA. METHODS: Thirty-seven patients with simple large VSD repaired in infancy were divided into four groups according to the presence of D and PDA. All patients underwent VSD closure at less than one year of age. Cardiac catheterization was performed before and late after the VSD closure. Pulmonary (PAm) and aortic mean pressure (Aom) were measured. Pulmonary flow and resistance indices (Qp/Qs, Rp/Rs) were calculated. RESULTS: There were no statistical differences in operative age, postoperative period, and preoperative PAm or Aom among four groups. Patients without D nor PDA (D(-)PDA(-)) had higher Qp/Qs and lower Rp/Rs than the other three groups preoperatively. After VSD closure, only patients with D and PDA (D(+)PDA(+)) had persistent PH while PAm of the other three groups returned to normal range. (Results are shown as mean +/- SD. *: $p<0.05$ vs. the other groups) CONCLUSIONS: Pulmonary vascular resistance in simple large VSD in infancy was elevated especially with D or coexistent PDA preoperatively. Pulmonary hypertension in patients with both D and PDA remained after the VSD closure. We conclude that both D and PDA may contribute to the residual PH after VSD closure in infancy. These results indicate that much earlier VSD closure may improve postoperative persistent PH in patients with D and PDA.

P59

Cardiopulmonary bypass (CPB) management in congenital heart surgery using nitroglycerine and blood cardioplegia

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Our experience during the last 22 months with a new approach to CPB management is reported. Methods: 129 consecutive patients with congenital heart defects were operated on using CPB (median age 10 months, DI: 197, median weight 7.4 kg, 2.3–65). 38 patients were <1 month, 32 patients were between 1 and 12 months and 59 patients were older than 1 year. Dexamethasone (3 mg/kg) was given at the beginning of anaesthesia to children < 1 year. At the start of CPB, nitroglycerine was infused at a rate of 1–3 µg/min/kg for the duration of CPB. Cardioprotection was established by blood cardioplegia, which was either applied antegrade or retrograde every 20 min. In patients < 10kg blood cardioplegia was infused by hand on minimal priming volume. Results: 52 cases were simple (ASD, VSD, ect), 44 cases were complex (TOF, AVSD, TGA, ecc) and 48 cases were very complex (Glenn, TCPC, Norwood, ecc). CPB-time ranged from 26–427 min (mean 144 min). Aortic cross-clamp time ranged from 0–344 min (mean 57 min). Circulatory arrest was required in only 3 children, because selective cerebral perfusion was used for Norwood-type repair and other arch procedures. Perioperative and 30 day mortality was 0 %. Perioperative inotropic support was required in 21 % (mainly low-dose dopamine up to 5 µg/kg/min). Postoperative ventilatory support ranged from 1–428 hours (median 8 h). Conclusions: Complex congenital heart lesions can be surgically treated with low mortality and morbidity. Our approach to CPB management results in good tolerance of the operative procedure. We speculate that the use of the nitric oxide donor nitroglycerine and Dexamethasone during CPB may lower the inflammatory reaction in response to CPB.

P60

Norwood-type surgery with continuous cerebral and myocardial perfusion

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Objective: The Norwood procedure and its modifications are commonly performed in circulatory arrest. The degree of neurological injury from circulatory arrest is directly related to the duration of arrest time. We report a technique of selective cerebral and myocardial perfusion with the aim to reduce ischemic damage to brain and heart. Methods: We performed a modified Norwood procedure on four neonates (4–29 days) with single ventricle physiology, coarctation, and hypoplastic aortic arch. In all cases the ascending aorta was of adequate size for arterial cannulation. We cannulated the ascending aorta and clamped the aortic arch distally to the innominate artery. Aortic arch repair was performed in moderate hypothermia with the heart beating, while both the brain and the heart were selectively perfused. Results: The cross period of selective cerebral and myocardial perfusion was 35, 42, 50 and 61 min. All children recovered uneventfully without neurological or myocardial complications. Conclusion: Norwood-type procedures can successfully be performed without total circulatory arrest. Protection of both brain and heart is achieved by continuous, selective perfusion.

P61

Left Pulmonary Artery coarctation in cyanotic patients: difficult diagnose to make?

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Coarctation of pulmonary artery branches is a recognized complication of cardiac surgeries' procedures or ductal construction. Its diagnosis can be unmasked by a number of factors. We report two cases of severe desaturation following early and long-term postoperative course caused by left pulmonary artery (LPA) coarctation with total exclusion of that lung perfusion. The cardiac malformations of the patients were dextrocardia, discordant AV and VA connections, pulmonary atresia and VSD. They were submitted to a systemic-pulmonary shunt (from the ascending aorta to the right pulmonary branch), through a right lateral thoracotomy in the neonatal period. The first patient was discharged home one week following the surgery with a peripherical saturation of 82% in room air. The second patient had a complicated recovery due to sepsis and remained on ventilatory support with a saturation of 80% (O2 40%). Three weeks later this patient could not be weaned from ventilation due to persistent low O2 saturation. A lung perfusion scan revealed a discrepancy between right and left lung perfusion and angiography showed a flow interruption to the left pulmonary branch just beyond its origin. The other patient, who were at home for five months, arrived at emergency room very desaturated and had similar findings on his lung perfusion scan and angiography. Both patients were operated through a midline sternotomy. The pulmonary trunk and its branches were dissected, there was a kink in the left pulmonary branch and a coarctation at the insertion site of the ductus arteriosus. The ductus was ligated and divided, the segmental coarctation was resected and the branch was reconstructed with a 6mm Gore Tex tube. Lung perfusion scan after the surgery depicted reperfusion on the left lung of both patients. We suggest that LPA coarctation should be considered as a possible cause of persistent cyanosis following systemic to pulmonary shunt and that repair, even in the late post-operative period, should be encouraged.

P62

Ventricular performance after anatomic repair in children with discordant atrioventricular connections

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Objective: To determine efficacy of the postoperative circulation in patients with discordant atrioventricular connections undergoing anatomic biventricular repair. Patients: Cardiac performance was examined by catheterization in 29 out of 30 children with this particular malformation undergoing the definitive procedure since 1989. Patients with left isomerism were excluded. For reconstruction of the channel from the morphologically left ventricle to the aorta, intraventricular rerouting had been needed because of the presence of pulmonary stenosis or atresia in 26 patients, while the arterial switch was feasible in the other 4. Results: Catheterization carried out 1.3 ± 0.6 years after

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

the procedure showed that LVEDV and RVEDV were 119 ± 37 (64 – 226)% and IDI = 54 (51 – 166)% of the anticipated normal values, with ejection fractions 55 ± 10 (35 – 77)% and 55 ± 9 (32 – 74)% respectively. LVEDP was 10 ± 4 (3 – 26) mmHg, RA pressure 6 ± 3 (1 – 12) mmHg, and mean PA pressure 16 ± 6 (9 – 37) mmHg. Cardiac index was calculated as 3.3 ± 0.6 (2.2 – 4.5) l/min/m2. Perimembranous inlet VSD, impaired coronary arterial perfusion, as well as tricuspid enlargement of VSD appeared to be unfavourable factors. In patients with pulmonary obstruction undergoing reconstruction of the RV outflow tract without use of an external conduit, RVEDV and RA pressure were slightly greater than others. Catheterization in the longer terms 6.4 ± 1.4 years (in 10) and 8.6 ± 0.8 years (in 3) after repair showed no significant differences in the parameters. Conclusions: Anatomic biventricular repair can provide excellent postoperative circulation in this setting. The surgeon, however, should note that some impediments could exist.

P63

Early repair of Tetralogy of Fallot (under 6 months of age) – own experience

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Early repair of Tetralogy of Fallot (under 6 months of age) became a dominant strategy in surgical treatment of this kind of defect. The results of surgery are subjected the correct study. Between 1998 and 2000 seventy repairs of tetralogy of Fallot were performed. 35 children (50%) underwent surgery at the age of under 6 month of life. 32 children (18 male – 56%, 14 female – 44%) were in follow-up. Surgery criteria for correction were based on echocardiografic and angiographic parameters. Morphology and localization of the obstruction of the right ventricle outflow tract were considered: subvalve obstruction – 3 pts (9%), valve obstruction – 9 (28%), mixed - 20 pts (63%). Total gradient in RVOT ranged from 60 to 120 mm Hg. Hypertrophied muscular bands were missed and partially removed in all children by atrial approach. VSD was closed with PTFE patches. Incision of pulmonary annulus was performed in 11 pts (34%). Ventriculotomy in front across the pulmonary valve annulus was made in 21 pts (66%). The pericardial (31 pts-97%) or atrial (1 pt – 3%) transannular patch was applied to extend RVOT. Gradient across RVOT, PV and TV regurgitation were assessed in postoperative period. Values below 20mmHg were revealed in 27 pts (84%). Gradient ranged from 20–40mmHg was present in 5 pts (16%). Four of them were extended RVOT by transannular patching. In one case RVOT was extended by pulmonary tract incision and commissurotomy. Pulmonary valve regurgitation graded 2/4 was present in 16 pts (50%). Tricuspid valve regurgitation graded 2/4 was revealed in 3 pts (9%). Early repair of tetralogy of Fallot is effective and safe method. There were no deaths in postoperative period in our series. Postoperative gradient across RVOT was hemodynamically non-significant. No medicament supply was required.

P64

Complete atrioventricular canal (CAVC): two-patch repair in early infancy (under 3 months of age)

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From January 1998 to October 2000 forty-two infants (from 1 to 13 months old) underwent repair of CAVC, which was done using a two-patch technique. We evaluated 17 patients (88%), who were less than 3 month-old (mean 2 months). There were 19 boys and 20 girls. Down syndrome was present in 25 infants (68%). Weight ranged from 2.2 kg to 4.8 kg (mean 3.4 kg). CAVC was associated with tetralogy of Fallot (in 1 case) and double outlet right ventricle (in 1 case). The early mortality was 8%: two patients died of sepsis and one of undiagnosed subaortic stenosis. Three patients (8%) required reoperation: two had mitral valve repair with good result and one successful mitral replacement. All patients were postoperatively in normal sinus rhythm and in NYHA class I or class II. At postoperative echocardiographic examination mitral regurgitation was mild or moderate in 34 and severe in 3 patients (before operation: 32 and 5, respectively). The residual regurgitation often observed post operation was clinically well tolerated. The mean LVEDD/BSA% ranged within normal values, preoperatively (98% of the predicted high borderline) and postoperatively (73%, respectively); the difference was not statistically significant ($p > 0.05$). The mean value of RVEDD/BSA% was moderately enlarged, prior to surgery (95% of the predicted high borderline) and after correction (85%, respectively), but no significant regression was found ($p > 0.05$). The early result of the two-patch repair of CAVC in infants less than 3 month-old (mostly weighing less than 4 kg) has been satisfactory. Thus we advise total correction of CAVC and additional abnormalities in very small infants.

P65

Left ventricular function after Ross procedure-mid-term results

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Objective: Aortic valve replacement with pulmonary autograft became an acceptable surgical treatment for aortic valve pathology in children and young adults. It's a difficult operation with long aorta cross-clamping time. Methods: Between 1995 and 2000 25 pts at the age ranged from 6 months to 21 years underwent Ross procedure due to complex aortic valve disease. This study includes 14 pts at the age from 8 to 18 years. The aim of the study is to evaluate left ventricular function after surgery. The follow-up ranged from 6 months to 5 years (mean 18 months). Assessment was based on clinical examination, transthoracic echocardiographic evaluation (LVEDD, IVSd and LVPWd were measured before and at least 6 months after surgery).

P66

Management and outcome of mitral valve cleft with ventriculo-arterial discordance

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Congenital mitral valve (MV) cleft causing left-ventricular outflow obstruction (LVOTO) and/or mitral regurgitation (MR) occurs rarely in patients with ventriculo-arterial (VA) discordance and may complicate biventricular repair. We reviewed the clinical features, management and outcome of 16 patients with MV cleft, VA discordance and 2 well-developed ventricles. In all cases, echocardiography showed the cleft dividing the anterior leaflet of MV, with attachments of its components to or through the ventricular septal crest. Three patients had LVOTO whereas 2 had more than mild MR. The mean follow-up period was 3.4 years (0 to 10.5). Nine patients underwent biventricular repair, with 2 early deaths. Single ventricle palliation was preferred in 7 patients, with no postoperative deaths. Twelve patients were in NYHA class I at last follow-up. In conclusion, biventricular repair is not always feasible in patients with MV cleft and VA discordance. However single ventricle palliation can be achieved with good results in these patients.

P67

Management and outcome of isolated cleft mitral valve.

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We retrospectively studied 14 patients with isolated cleft mitral valve (ICMV). All the patients with atrioventricular canal (canal), hypoplastic ventricles and ventriculo-arterial discordance were excluded. The median age of diagnosis was 0.2 years (range 0 to 4.8). Echocardiography demonstrated the ICMV on the anterior (aortic) leaflet in all patients, with attachments of its components to the ventricular septum. A more than mild mitral regurgitation (MR) was present in 7 patients. Associated cardiac lesions were perimembranous ventricular septal defect (5), atrial septal defect (2), patent ductus arteriosus (1), left-ventricular outflow tract obstruction (3) and tetralogy of Fallot with absent pulmonary valve (1). The mean follow-up period was 3.7 years (1 to 10.5). Surgery with mitral valve repair was indicated in 5 patients. One newborn with severe subaortic stenosis died prior surgery. At last follow-up, the remaining 13 patients were all in NYHA class I. Echocardiography showed a more than mild MR in 2 cases whereas one patient had mild mitral stenosis. In conclusion, ICMV is a correctable cause of MR with a good outcome.

P68

Coronary sinus drainage to the left or right atrium in Fontan procedure: Long-term clinical implications.

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Coronary sinus drainage to the left or right atrium in Fontan procedure: Long-term clinical implications. A. Cloutier, Z. Qi, J.M. Cote, C. Blinde, G. Delule, J.M. Guay, J. Perron, P.C. Camu. Quebec. The Classical Fontan procedure elevates venous pressure in the coronary sinus, secondary to increased systemic venous pressure. It remains undetermined if this elevation could

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

impair coronary perfusion and ventricular myocardial function, and thereby influence long-term evolution. The purpose of this analysis was to compare long-term clinical evolution of patients with coronary sinus drainage to left or right atrium after a Fontan-type operation. From 03/81 to 03/98, 45 patients aged 2.9 to 26.6 years (median 6.5±5.7) underwent a Fontan-type operation. The coronary sinus was in left atrium (group L) in 25 patients, right atrium (group R) in 19 and undetermined in 1 patient. There were 2 early deaths (4.3%) and 7 late deaths (17.8%). Of the 2 early-deaths, 1 was due to poor cardiac output, while the other was due to volvulus and malrotation. The 7 late deaths were related to Fontan failure. Actuarial survivals at 1, 5 and 10 years were 95.5%, 85.5% and 73.4% in the whole group. Group L / Group R (N 25/19). Age (yr) (median) 5.3±6.2 / 6.8±5.6. Follow-up (years) 5.0±3.8 (0.5–12.4) / 7.9±4.0 (0.3–14.4). Survival 1 year 95.6% / 89.5%, 5 years 89.6% / 76.6%, 10 years 89.6% / 61.0%. NYHA class I–II 85.7% / 62.5%. Ventricular dysfunction 4.8% / 25.1%. Antiarrhythmics 0 / 21.4%. Other drugs** 71.4% / 76.9%. *p<0.05, ** including vasodilators, cardiac glycosides and diuretics. Directing coronary sinus drainage to the low-pressure left atrium could favor improved long-term survival after Fontan-type operation.

P69

Sinus venosus atrial septal defect: Long-term follow-up of 115 repaired patients

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BACKGROUND: Sinus venosus atrial septal defect (SVASD) differs from ostiumsecundum atrial septal defect by its location in the atrial septum and association with anomalous pulmonary veins (APV) which may increase the risk of surgery. METHODS AND RESULTS: The data on 115 patients with isolated SVASD with or without APV who underwent repair from 1972–1996 at age 34±25 years were analyzed. Preoperative symptoms were present in 66 patients (57%). atrial fibrillation/flutter in 20 (17%) and sinus node dysfunction (SND) in 8 (7%). 109 patients (95%) had a superior SVASD and 6 (5%) an inferior/posterior type. A persistent left superior vena cava was found in 17 patients (15%). An anomalous connection to the junction was done in 23 patients (20%). Perioperative mortality was 0.9%. SND (in 21%) at hospital discharge was more common in patients with persistent left superior vena cava ($p=0.001$) and APV from the whole right lung ($p=0.002$) but independent of the type of repair. Postoperative follow-up (133±102 months) was possible in 108 patients (94%). Improvement in symptoms was found in 93 patients (77%) which was not different in preoperatively asymptomatic patients (67%, p=ns) or in patients >40 years at repair (86%, p=ns). SND for >6 months was present in 32% but not related to the type of repair or associated anomalies. Atrial fibrillation was found in 23 patients (21%). No reoperation was necessary during follow-up. Sixteen patients (7%) died at age 69±19 years. Survival was similar to expected. CONCLUSIONS: Despite the complex anatomy, SVASD repair is associated with low morbidity and mortality even if repaired at older age. SND is common and may be due to intrinsic sinus node disease. Postoperative improvement is independent of preoperative symptoms and age.

P70

Quality assurance of paediatric cardiac surgery at a single institution.

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To develop effective quality assurance methods to monitor outcomes following paediatric cardiac surgery at a single institution. All patients undergoing cardiac surgery were enrolled prospectively on admission to intensive care. Patients were stratified by complexity of surgical procedure into five groups, with Category 5 being the most complex procedure. Outcome measures are listed death, length of admission and morbidity from complications. From Sept 1995 to Sept 2000, 1558 patients underwent 1680 surgical procedures. 1221 (72.7%) were open procedures, and 437 (26%) were complex (Category 3 and 4) procedures. Mean patient age was 2.1 years (range, 1day-20years) and patient weight 15.1kg (range, 700g-90kg). 51 patients (3.0%) died during the study period, with 11 of these deaths occurring intraoperatively. During the study surgical practices changed with an increased incidence of complex surgery (21% in 95/96 to 30% in 99/00). The annual surgical mortality ranged from 1.8%–5.0%, but when the mortality logistic regression model, is adjusted for complexity of surgery and body surface area, there was no significant yearly variation in the mortality rate ($p=0.5$). Despite increased complexity of surgery, mean ventilation time decreased from 82.1 to 34.3 hours ($p=0.003$), with rates of failed extubation falling from 7.9% to 2.2% ($p=0.0116$). The incidence of significant neurological complications fell from 3.9% to 2.5% ($p=0.015$) and the incidence of arrhythmias from 8.7% to 3.5% ($p=0.06$). Infection rates ranged from 1.5% to 5.9%. Analysis of individual surgeon's results showed no significant difference in the mortality rate by complexity of surgery performed ($p=0.62$). Stratifying complexity of surgery proved valuable in monitoring surgical outcomes and detecting differences in performance over time as large subgroups were created for analysis.

P71

Blood lactate measurements as one of several predictors of early outcome following neonatal complex congenital heart surgery

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Post-operative serum lactate levels, thought to reflect oxygen delivery and tissue hypoxia, have been used to predict outcomes after neonatal congenital heart surgery. Controversy exists regarding the utility of this measurement in lieu of other risk factors. Purpose: To evaluate the association of serial blood lactate levels and other risk factors with neonatal outcome after surgery for complex congenital heart disease (CHD). Methods: A retrospective review of 42 selected infants from a 3-month period who underwent CHD surgery at our institution. Data included intra- and post-operative variables, and outcome to discharge. Poor outcome was defined as need for mechanical support or death during hospitalization. Results: Seven of the 42 patients had an adverse outcome, including 4 deaths. Initial lactate level > 6 mmol/L within the first 6 hours after cardiopulmonary bypass (CPB) was associated with a 7.5 fold increased risk of poor outcome (95% CI: 1.4 – 40.2; p=0.02). The negative predictive value was high (92%), the positive predictive value was low (36%). Lactate levels were significantly higher, stratified on outcome, in the first 36 hours after CPB. Other factors associated with a poor outcome included lower weight (< 3.5 kg; OR: 11.5; p=0.03) and CHD with ductal-dependent systemic blood flow (DD-SBF) (OR: 6.4, p=0.04). An elevated lactate level was more likely with younger age (OR: 12.4, p=0.002) and DD-SBF (OR: 8.9, p=0.004). Inclusion of either of these factors in a multivariate model with lactate level negated the significance of lactate levels as a predictor of outcome. Conclusions: While blood lactate levels do differ stratified on outcome, the positive predictive value was low; therefore lactate levels may only be a marker of higher risk group (young age, low weight, DD-SBF).

P72

Two-stage repair of truncus arteriosus associated with left diaphragmatic hernia

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A 2.5 kg female infant with prenatal diagnosis of left congenital diaphragmatic hernia (CDH) associated with truncus arteriosus type II was born at term after in vitro fertilized fecondation. In the 4th day she underwent repair of CDH by means of PTFE patch through a left subcostal approach. Surgery was done at bedside in neonatal intensive care, while the patient was ventilated with high frequency oscillation (HFOV). Despite maximal medical therapy (HFOV, vasodilators and inotropic support) she could not be weaned from the respirator. At that time primary correction of truncus was considered a too high risk procedure and we considered to palliate the patient by banding only the right pulmonary artery (RPA) since the left pulmonary branch was hypoplastic. A 12 mm teflon band was placed around the proximal RPA through a limited superior median sternotomy. The infant was weaned from assisted ventilation one week after. At 7 month of age a cardiac catheterization showed a well positioned band and a mean pressure of 17mm hg in the distal RPA. Soon after she underwent debanding of RPA and repair of truncus arteriosus by means of a 15mm xenograft. Prolonged assisted ventilation was required. Patient was discharged home after 40 days in good general condition with 95% saturation. In presence of pulmonary hypoplasia due to CDH, banding of the contralateral pulmonary artery may be considered a useful palliative procedure.

P73

Outcome and intermediate- term follow- up after surgical correction of atrioventricular septal defects in a 10 years era in 2 university centers

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

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To evaluate our results and surgical techniques we studied all patients operated for atrioventricular septal defect between 1/1986 and 1/1996. Two hundred thirteen pts were operated, nearly 40% < 6 months. Hospital mortality 12 pts (5.6%). Two hundred one survivors were included in outpatient follow-up. Operated at an age 2.8 yrs (+/- 3.7 SD). Follow-up ranged from 2.9 to 13.5 yrs, mean 5.6. Associated lesions operated (62 pts) included persistent ductus arteriosus (37), Fallot's tetralogy (8), ASD II (6), other VSD (9), double orifice left av-valve (7), hypoplastic left ventricle (2), aortic stenosis (5), and coarctation (3). At operation the defect was closed with no patch in 2 pts (1%), one patch in 84 (39.5%), two patches in 118 (55.4%) and 3 in 5 pts (1.4%). During the follow-up period 16 pts (7.5%) died, because of endocarditis, respiratory or chronic heart failure or non cardiac. Twenty-nine pts (13.6%) were reoperated, mainly for left av-valve regurgitation. Five pts (2.5%) had a prosthetic valve implantation at the left side. 185 survivors now are in good clinical condition, 11% use supportive medication. Left av-valve regurgitation minor 108 (59%), moderate 42 (23%), severe 10 (5%) pts. Right av valve regurgitation minor 63 (34%), moderate 6 (3%), severe 2 (1%). Reoperation correlated with early postoperative regurgitation of the left av valve and not closing the left-sided cleft. No other indication for morbidity and mortality were found, although endocarditis was a major cause for death (pts). We conclude that atrioventricular septal defects can be operated at young age. After surgery av-valve regurgitation is a subject of concern for the future.

P74

Cardiac surgery and risk stratification: one year multicentre study

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Aim: to standardize data collection of congenital heart defects and to develop a risk stratification criteria in cardiac surgery. Methods: one year multicentre study was carried out on hospitalized children for congenital heart defects in three Italian pediatric hospitals (July 1 1996 – June 1 1997). Clinical data were coded in a specific questionnaire. A preliminary review of the ICD 9 CM coder was performed. For surgical cases a matrix of clinical severity was created by matching diagnosis and procedures. Each surgical condition was stratified into three levels of surgical complexity (SC) according to agreed criteria. A total of 85 Surgical Complexity Profiles were identified: 32 in SC I, 31 in SC II and 17 in SC III. On the basis of the SC stratification, 48.9% of the surgical patients (n=329) was classified in SC I, 29.6% (n=199) in SC II, and 21.2% (n=140) in SC III. Urgency admission, emergency surgery and death rate increased with SC score. The age distribution was markedly different in the three SC score: in SC I only 15.0% were aged less than 1 month vs 50.0% in SC III. Lenght of stay significantly increased with SC score not only in presence but also in absence of complications (P<0.0001). The presence of complications was strongly associated with the SC score (P<0.0001), with the presence of associated cardiac defects (P<0.0001) and clinical condition (P<0.004). Conclusion: the surgical complexity matrix can be a reliable system of risk stratification in pediatric cardiac surgery, particularly useful for standardize health care profile and weight for the health service programme. Matrix is now under revision and a new data collection are being collected.

P75

Anomalous origin of left coronary artery from pulmonary artery. Results of surgical correction in 5 infants

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Five infants operated for anomalous origin of left coronary artery from pulmonary artery are retrospectively analysed. The mean age at surgery was 12+/-6.7 weeks [3.5 to 20 weeks] and mean weight at surgery was 4.471+/-0.68kg [3.7 to 5.27kg]. All babies presented in infancy with LV failure. Three had evidence of ischaemia with LV strain and two had Q waves in anterolateral leads. Cross sectional echocardiography showed dilated LV with poor contractility in all babies with ES of 15.8+/-4.02% [12%to20%], moderate MR was seen in all. All babies underwent surgery as soon as diagnosis was made. Four had direct reimplant of LCA into the aorta while one had a Tunnel repair. IABP was used for hemodynamic instability in one baby and prophylactically in remaining four babies post operatively for 115+/-26.7hrs [72 to 144hrs]. All had delayed closure of the chest. There was no operative mortality. One patient was reoperated for mitral stenosis and relief of pulmonary stenosis 4 months after the primary repair. All patients are followed up for 172 patient months and show improved EF. Early surgery ,IABP for LV support and delayed sternal closure are the key to good results.

P76

Efficacy of intra-aortic balloon pumping in neonates and infants with refractory heart failure to conventional medical treatment

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From August 1994 to November 2000 intra-aortic balloon pump was used in 18 children in our institution. Eight patients were under 6 months of age. Average age was 10.56+/-8.8 weeks [1st 24 weeks] (CI [6.6+/-5.95]. They weighed from 3.5 to 5.8 kg with average weight of 4.264+/-0.72kg. Three were males and 5 were females. Five patients underwent surgery for AS,CAPA,2 had arterial switch and one had bilateral cavopulmonary shunt. All patients were on pharmacological support all on dobutamine and enoximone, 4 were on adrenalin, 5 were on GTN. Average duration of intra aortic balloon pump use was 100.87+/-62.85hrs. There were 2 deaths and 6 survivers. One patient died of major myocardial infarction and second due to RV failure. Long term survival was 62.2%. There was no IABP related complication. Use of intra aortic balloon pump in neonates and infants less than 2 months of age with refractory cardiac failure to conventional pharmacological treatment is safe, effective and improves long term survival.

P77

The 'chubby' infant with Tetralogy – a distinct morphological entity?

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Aim: To confirm a clinical observation that amongst infants undergoing surgical correction for tetralogy of Fallot (TOF) in our centre those who were unusually 'chubby' seemed to have a higher morbidity and mortality. Methods: 90 consecutive patients less than 2 years of age undergoing corrective surgery for TOF were analyzed for risk factors for morbidity and mortality. Results: In hospital mortality was 4.90 (4.4%). 14 infants were noticeably “chubby” in appearance (in the Indian context – where malnutrition is the norm). Operative mortality in this group was significantly higher (2/14 [21.4%]). Likewise morbidity in terms of prolonged ventilation, seizures and low output syndrome (11/14) was significantly higher. Distinctive morphologic features in this sub-group included a severely deformed RV outflow (11/14), significant aortopulmonary collaterals (6/14), a high or doubly committed VSD (7/14) and increased LA return (5/14) at surgery. Recognition of this entity as a possible high risk group half-way through this series prompted us to de-viate from our current norm of echo based surgery and perform aortography on all 'chubby' infants and embolize significant aorto-pulmonary collaterals. Post-operatively ventilatory and inotropic support was extended electively till sustained hemodynamic stability was achieved. This strategy helped eliminate mortality in the last 8 patients. Conclusion: Unusually chubby infants with TOF appear to have a distinctive morphologic combination of a very deformed RV outflow, a high VSD and significant collaterals. Unless recognized and managed in a proactive manner this combination appears to contribute to increased morbidity and mortality.

P78

Complete atrioventricular septal defect, Down syndrome, management strategy and surgical outcome

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We evaluated outcome and incremental risk factors for death or reoperation after repair of complete atrioventricular septal defect (CAVSD). Excluding those with atrial isomerism or those in whom a biventricular repair was not thought feasible, 147 consecutive children underwent repair between January 1986 and December 1998. 106 had Down syndrome (DS), 37 normal chromosomes and 4 had other syndromes. 108 underwent primary repair, 19 had prior pulmonary artery banding and 20 additional Tetralogy of Fallot. The median age at primary repair was 4.1 months. A two patch technique was used for repair in 68%. The total 30 days mortality was 15% (70% CL 12 -19%). Analysis of incremental risk factors showed presence of a double orifice atrioventricular valve (DOAVV) to be a highly significant risk factor (P= 0.002) with 6 of 11 patients dying. If DOAVV patients are excluded the total mortality falls to 12% (70% CL 9–15%). There was no difference in the

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

mortality between DS and chromosomally normal children, but the latter more commonly required reoperation p=0.006. The reason appears to be presence of a dysplastic left atrioventricular valve (DLAVV), (24% vs 3% in Down children, p<0.001). A Cox's regression model controlling for the two variables was constructed and showed (DLAVV) to remain a significant risk factor for reoperation (Hazard ratio 3.80, 95% CI 1.37, 10.59). The presence of DS remained protective although no longer statistically significant. That presence of DOAVV was an unforseen risk factor for death had not been foreseen prior to our retrospective analysis. We should perhaps have a lower threshold for primary valve replacement in this group.

P79

Ross procedure in the pediatric population, a 10 years experience

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Pulmonary autograft has been our procedure of choice for aortic valve replacement in the pediatric population for the past 10 years. Growth potential and freedom from anticoagulation are the main advantages. However for pulmonary valve substitute a still the Achille tendon of the procedure. From 1990, 41 children less than 18 years of age underwent a Ross procedure in our institution. 181 adults underwent the procedure during the same period. Mean age was 10.1 (sd 5.6). 85% were male. The main etiology (85%) was congenital aortic stenosis. 70% had undergone at least one previous surgery. Patients were followed yearly by echo doppler. Mean follow up is 6 years (sd 3.2). Mean gradient for the pulmonary autograft was less than 1 mmHg. Mean regurgitation was less than 1+. Mean gradient for the pulmonary Homograft used for pulmonary valve replacement was 20 mmHg compare to 8 mmHg in the adult group. Five patients required reoperation for pulmonary Homograft replacement (12%). Three within one year and two at 5.5 and 7 years post-op. During the same time only four adults needed their Homograft replaced (2.2%). Surgical analysis revealed that older donor, older patient and larger size valve reduced the risk of Homograft stenosis. Method to decreased the immunologic reaction to the homograft are still to be found. In conclusion, Ross procedure is a good alternative for AVR in children, however pulmonary valve substitute is still problematic.

P80

The fate of small-diameter (<15 mm) homografts in pulmonary position

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Background: Implantation of homografts to establish right ventricular to pulmonary artery continuity is an accepted surgical treatment for some cardiac lesions in neonates and young infants. With growing numbers of total repair in very young patients and the increasing rates of implantation of homografts of very small diameter it seemed timely to investigate the longevity of the latter. Methods: Between July 1987 and August 2000, small diameter homografts (<15 mm) were implanted to establish continuity between the right ventricle and the pulmonary arteries. There were 59 aortic and 33 pulmonary homografts. The cardiac anomaly corrected were truncus arteriosus (n=57), pulmonary atresia and VSD (n=10) or tetralogy of Fallot (n=4). One newborn underwent a Ross operation. The patients were analysed in 3 groups according to homograft diameter: group I: 8–9 mm (n=44); group II: 10–11 mm (n=15) and group III: 12–13 mm (n=41). Results: Overall survival was 87.3 % at one year and remained constant afterwards. Eight patients required conduit replacement for outgrowth, one because of infection without mortality. Freedom from reoperation (FFR) for homograft replacement after 1, 5 and 10 years was respectively 97 %, 82 % and 73 %. Aortic and pulmonary conduits showed no difference in longevity. The FFR for homograft replacement was significantly smaller in group I (41 % at 2 years) comparing with the group II (100 %) and group III (95 %) (p<0.01). Conclusion: Very small diameter homografts (8–9 mm) have to be replaced early, usually in the first two years following implantation. Homografts sized 10 mm or more, regardless of type, have a remarkable longevity and remain excellent valved conduits for reconstruction of right ventricular to pulmonary artery continuity in neonates and infants. Homograft replacement can be done with very low risk.

P81

Impact of complex in comparision to regular coronary anatomy on the arterial switch operation

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Background: The outcome after arterial switch operation especially depends on the complexity of coronary artery origin and epicardial course. During a two year period we reviewed the surgical results with special focus upon different coronary artery pattern and technique of reimplantation. Methods: From 9/98 until 11/00 seventeen newborns were operated. Patient age was 11 ± 7 days and body weight 3.7 ± 0.6 kg. Descriptive coronary artery anatomy was normal in eight (group N) and complex in the other nine patients (group C), including Cx from RCA in five, LAD and Cx with separate ostia in two and inverted coronary arteries in two patients, respectively. Four patients (all C) had additional ventricular septal defect closure and one of them additional IAA Type A repair. Results: Regarding the coronary arteries extensive mobilization especially in patients with complex anatomy was performed. In group N before re-implantation excision of the neo-aortic sinuses was performed whereas in group C trap-door technique was applied. Cross-clamp-duration was 93 ± 11 (N) versus 112 ± 26 (C), p = 0.05. Primary chest closure was achieved in 5 patients each whereas secondary closure was performed after 2.3 (N) versus 3.3 (C) days. Patients were intubated for 4 (N) versus 4.5 (C) days and ICU stay was 8.5 (N) versus 9.5 (C) days, respectively. There was neither early nor late mortality. Control coronary angiography revealed no perfusion abnormalities in the complex group. Conclusions: The arterial switch operation can be safely performed as regular as well as in complex coronary anatomy. Meticulous dissection of the coronary artery buttons as well as sufficient epicardial mobilization in complex coronary artery anatomy together with trap-door re-implantation technique is essential.

P82

Fate of Intramural Coronary Arteries after Arterial Switch Operation

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Purpose: Intramural course of coronary arteries is an anatomical variant in patients with transposition of the great arteries (TGA). Coronary transfer is performed under consideration of the missing tunnel at the coronary ostium and the course of this coronary artery with the commissure of the valve. The aim of this study is to evaluate the relevance of intramural coronary arteries for the peri- and postoperative course after arterial switch operation. Material and methods: Between 1981 and 1999, 245 patients with D-TGA underwent arterial switch operation. Five patients (1.4%) had an intramural course of the left coronary artery. In three of these patients the left main stem and in two patients the left anterior descending coronary artery showed an intramural course. The coronary transfer was performed with a collar-coded dissection of the commissure with prior longitudinal splitting of the intramural section. Results: None of these patients died; intraoperative course was uneventful. Postoperatively, three patients developed extracardiac complications (cerebral bleeding, capillary leak), myocardial ischemic changes were not observed. Outpatient follow-up investigations showed no signs of myocardial ischemia. In three patients follow-up cardiac catheterisation after 5, 16 and 50 months revealed occlusion of the intramural coronary ostium. Exercise electrocardiogram and myocardial scan-riscanning proved myocardial ischemia. Two of these patients have undergone successful A. mammaria interna-bypass. Conclusion: Intramural course of coronary arteries in patients with D-TGA is rare and does not cause increased mortality and myocardial infarction rates. However, risk of coronary occlusions seems to be higher. Therefore, selective coronary angiography and exercise investigations in clinically uneventful patients are necessary. Revascularisation with an A. mammaria interna-bypass may be indicated.

P83

Early complete correction for right ventricular outflow tract obstruction

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Background: In children with complex right ventricular outflow tract (RVOT) obstruction the optimal surgical strategy remains controversial due to potential risk for re-interventions in patients with hypoplastic pulmonary valve (PV). Aim of the study was to evaluate the results following complete correction in infancy and to analyze different surgical techniques of RVOT reconstruction. Methods: Since 05/98 19 infants were included. Tetralogy of

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Fallot in 15, pulmonary atresia in 3 and double outlet right ventricle in 1 patient, respectively. 30 patients received primary total correction whereas 9 patients with critical hypoxic spells underwent staged correction with neonatal modified Blalock-Taussig shunt. Mean age at total correction was 7 ±5 months. Follow-up was complete in all patients after an interval of 8 ± 6,3 months. Results RVOT reconstruction was performed using a transannular pericardial patch with pericardial monocusp PV reconstruction (sandwich technique) in 8, a two pericardial patch technique (RVOT and main pulmonary artery patches) saving the native PV in 6 and a valved conduit (pulmonary homograft or Xenograft (Contegra(tm))) in 5 patients, respectively. There was neither in-hospital nor late mortality. Aortic cross-clamp was 59 ±17 min and endotracheal incubation 9.5 ±3.6 hours. All patients were discharged home after 12 ±3.9 days. At follow-up all pulmonary valves were competent. 7 patients had residual moderate pulmonary stenosis at a maximum gradient of 30 mmHg. There was no need for any re-intervention. Conclusions RVOT reconstruction using an autologous pericardial monocusp valve during infancy is a reliable therapeutic strategy yielding good results at intermediate follow-up. Resorption of PV function is improving recovery and protecting right ventricular function early postoperatively. Due to the functionally good results the pericardial monocusp valve is superior to homograft implantation in this age group.

P84

Coarctation of the aorta in neonates: simplified surgical management

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Background Different surgical techniques have been used for repair of coarctation of the aorta (CoA). Aim of this study was to evaluate the growth potential of the anastomose after using two different techniques for CoA repair in newborns. Methods 24 patients with CoA were operated in a two year period. In 11 patients the obstruction was proximal to the left subclavian artery (LSA) including hypoplastic aortic arch (group A). 13 patients had the obstruction distal to the LSA (group B). Mean age was 13± 3 (A) versus 14 ± 3 (B) days, respectively. Two anastomotic techniques were used. Descending aorta to transversal aortic arch in group A and end – to – end anastomosis after complete resection of ductal tissue in group B. Additional procedures were VSD closure in 3 patients (A) and pulmonary artery banding in 6 patients (A), respectively. Results There was no early and one late (A) mortality due to multiple organ failure despite regular aortic blood flow profile. Average clamp time for extended anastomosis (A) was 14.7 ±5 minutes versus 11.7 ±3 minutes in group B. Using echo doppler postoperatively unobstructive aortic arch flow pattern was diagnosed. Maximum flow velocity was 2.2 ±0.3 m/s with no diastolic flow. At mean follow-up of eight months the flow pattern remained non obstructive in all patients. Average isthmus or descending aorta diameter ratio was 0.39 preoperatively, 0.71 postoperatively and 0.95 at follow-up. Comments These results clearly demonstrate that after complete resection of ductal tissue there is sufficient proportional growth of the whole aorta. Our approach simplifies surgical management into two procedures which achieve safe, effective and reliable relief for all types of CoA. Sacrifice of subclavian artery or implantation of prosthetic material is never required.

P85

Thirteen years experience with tricuspid valve repair, replacement in Ebstein disease

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Thirteen years experience with tricuspid valve repair, replacement in Ebstein disease. P Carrier, G Deslate, A Cloutier, C Houde, JM Coté, M Guay, Québec. We review our experience with tricuspid valve repair, replacement in Ebstein disease. Since 1987, Nineteen TVR or repairs were done on 12 patients. Porcine bioprosthesis was used in most of the first operation. Repair was attempted in all but failed in 10 out of 11. The last patient had had a TVR twenty years previously. No patient was lost to follow-up. Clinical and echocardiographic follow-up was achieved yearly. One patient had his valve replaced the same day of his valve repair. Six out of 11 who had their first TVR in our institution needed their prothesis to be replaced. Mean time between surgery 8,8 years. Main symptoms were recurrence of supraventricular arrhythmia. Causes for replacement were valve stenose in 5 and valve insufficiency in one. All valves but one were replaced with pericardial valve. The last one was replaced with a mitral Hemograft. There was no operative death or late death in this group. All are either NYHA functional class I or II. Discussion Tricuspid valve repair in Ebstein anomaly is the procedure of choice, unfortunately it often failed. Bioprosthesis is the replacement of choice since there is no need for anticoagulation. In our group, despite the fact that the largest size valve available was implanted, the failure mode was stenosis in most of them. Pericardial valve might achieve better long-term result, only follow up will tell.

P86

Early indicators for changes in the quality of surgical performance.

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Introduction Increased mortality led to an abrupt halt and subsequent restart of surgery for congenital heart disease in Utrecht. This study compares mortality and morbidity rates in the periods 1991–1995 and 1996–1999 in which different surgeons operated in the hospital in order to define early indicators for surgical problems. Methods Data of all patients undergoing paediatric heart surgery since 1991 (1209 patients) were collected retrospectively. Perioperative mortality, ICU-stay, delayed sternal closure and paralysis of the phrenic nerve were included. The patients were divided into 2 subgroups for different time eras: group 1: 1991–1995 (504 patients), and group 2: 1996–1999 (565 patients). Hypoplastic left heart (HLH) patients were reviewed separately. Results The overall mortality rate for the 2 periods were respectively in group 1: 9.9% (+HLH) and 8.6% (-HLH) and in 2: 4.8% (+HLH) and 3.6% (-HLH) ($p=0.0001$). The mean ICU-stay was in group 1: 7.3 days and 7.1 (-HLH) in 2: 4.9 and 3.7 (-HLH). Delayed sternal closure occurred in group 1: 10.3% and 10.9% (-HLH) and in 2: 3.8% and 2.9%. Paralysis of the phrenic nerve was encountered in group 1: 2.2% and 2.1% (-HLH) and in 2: 0.8%. Conclusions In the evaluation of surgical results mortality rates are of predominant importance, but variables like ICU-stay, the requirement of delayed sternal closure and phrenic nerve paralysis suggest to be more sensitive indicators and are likely to predict surgical deterioration earlier.

P87

Safety and efficacy of ministernotomy in congenital heart surgery.

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Purpose: Safety and efficacy in minimal access congenital heart surgery is still unclear. To assess this we reviewed our experience with minute sternotomies. Methods: From July 1998 to November 2000, 45 patients (35 pediatric, 8 adults) underwent repair of congenital heart defect via lower ministernotomy. We prospectively followed operative data, incision length, ICU and hospital stay and complications. All pediatric patients had postoperative echoes. Results: 1) Pediatric Group – Median age was 4.3(5–16 years). Operative procedures were for 25 ASDs, 2 subaortic stenoses, 3 VSDs, 3 partial AVSDs, 1 CAVSD and 1 supra-aortic stenosis. None required conversion to full sternotomy. Mean pump and cross clamp times were 47.4 +/– 20.7 min and 35.3 +/– 13.1 min respectively. Mean length of incision was 5.8 +/– 1.3cm. The mean ICU stay was 1 +/– 0.2 d with a hospital stay of mean 2.5 +/– 1.9 d, median 2 d (2–13). There were no deaths and 3/35(9%) had postoperative complications (pleural effusion, pericardial effusion, transient complete heart block). No residual defects were seen on postop echoes. 2) Adult Group – Ten patients underwent ASD repair. Two required conversion to full sternotomy (aortic cannulation site bleeding, scoliosis). Of the remaining 8 patients, the median age was 29 yr (19–65). The mean pump and cross clamp times were 92.4 +/– 10.4 min and 73.5 +/– 7.4 min respectively. The mean ICU stay was 1.0 +/– 0 d with a hospital stay of mean 3.4 +/– 0.5 d, median 3 d (3–4). There were no deaths and 1/8 had a complication (atrial fibrillation). Conclusion: Ministernotomy can be used safely and efficaciously for certain types of congenital heart surgeries, with no mortality and acceptable morbidity. We believe the hospital stay is reduced, likely due to less postoperative sternal pain and earlier mobilization.

P88

The conduit of choice for the reconstruction of the right ventricular outflow tract: a 26 – year experience with valved and nonvalved conduits

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Objective: The reconstruction of the right ventricular outflow tract (RVOT) in congenital heart disease often requires the implantation of a valved or nonvalved extracardiac conduits (ECC). Early results of reconstruction of the

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

RVOT were excellent. However, later results revealed failure of these EC due to stenosis and valve insufficiency. We compared the long-term durability of all conduits in the RVOT over a 26-year period. Methods: Between 2/1974 and 7/2000, 287 patients (mean age 12.8 years, range 4 days to 60 years, mean weight 32.6 kg, range 1.8 – 126.8 kg) with congenital malformations received a conduit (243 valved and 44 nonvalved). The conduit size ranged between 8 and 33 mm (mean size, 20.1 mm). Results: There was 10% early mortality (28/278). Long-term follow-up data were available for 253 (88%) patients. Seventy-two conduits (28.5%%) required replacement: 27% for valved conduits (65/243) and 16% for nonvalved conduits (7/44). The interval between first and second surgeries was between 3 months and 16 years (mean 4.4 years). Stenosis was the main mode of failure (66/72: 92%). For patients with valved conduit the mean interval from freedom of conduit exchange was 6.2 years. For patients with nonvalved conduit the mean reoperation-free interval was 3.5 years (P<0.04). Comparing two groups, we found no difference in patient survival probability (P=0.7), but there was significant difference between xenografts (n=116) and homografts (n=248) as valved conduit group (P<0.021). At 10 years, the freedom from reoperation for EC obstructions was 73% for valved EC and 84% for nonvalved EC (P=0.1). Conclusion: Although an ideal long term study seems promising, we conclude that pulmonary homograft use in reconstructing RVOT has been the conduit material of choice for the patients with RVOT pathology.

P89

Elective Repair of Tetralogy of Fallot in Early Infancy: Five-Year Experience

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Background: Between 1/1995 and 4/2000, 195 patients with tetralogy of Fallot (TOF) underwent primary repair at our institution. Of these, 95 had severe cyanosis and/or additional anatomic features (31 pulmonary atresia, 12 absent pulmonary valve) which necessitated non-elective repair or were referred at >6 months of age for surgery. The remaining 100 patients who underwent elective repair within the first 6 months of life form the basis of this study. Methods: Retrospective case series utilizing chart review. Results: The median age at repair was 3.2 months. The median weight was 5.5 kg (range 2.7–9.6kg). Trisomy 21 was present in 9. The VSD was closed via ventriculotomy in 59 patients, via transatrial approach in 35, and via combined approach in 6. A transannular patch was used in 77, with additional left pulmonary artery augmentation in 47. A potential atrial level right to left shunt was intentionally left in place or created in 70. The median bypass time was 70 minutes (range 31–208 minutes). Circulatory arrest was used in 47 patients (median 37 minutes, range 13–55 minutes). There were no hospital or late deaths. Postoperative complications included indications for: four reoperations (residual VSD [1], thoracic duct ligation [1], sternal debridement [1] and right ventricular outflow tract obstruction [1]); tetanies (n=4); junctional ectopic tachycardia (n=5); and acute cardiorespiratory failure requiring ECMO (n=2). The median stay in the cardiac intensive care unit was 3 days (range 1–62 days) and in the hospital 5 days (2–62 days). The duration of hospital stay was 1 week or less in 84 patients. Five patients (5%) have undergone late reoperations (residual VSD [2], RV outflow tract obstruction [3], ASD / partial anomalous pulmonary venous connection repair [1]). Conclusion: Elective repair of TOF can be performed at less than 6 months with no mortality, a short hospital course for a great majority of patients, and a low incidence of post-operative complications.

P90

An individualized approach to the Fontan operation improves early morbidity & mortality

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Re: only of all patients who underwent the Fontan operation from 6/1995 to 9/2000 (n=75) were reviewed & divided into groups: non-Fenestrated (n=25) & Fenestrated (n=50). The impact of age, underlying anatomy, need for staging & operative modifications were analyzed. Results: Hospital mortality was 0%. Fontan failure was 0%. Mean age 3.6 y (range 1–27y). Mean Pulmonary artery pressure 13.6 mmHg (range 6–32), Pulmonary vascular resistance 2.2 Wood unit (range 0.4–4.5) & ventricular end diastolic 10.6 mmHg (range 5–22). Moderate Atrioventricular valve (AVV) regurgitation (n=7). Ventricular function by intra-operative Trans-esophageal echocardiography (good n=54, fair n=19, & poor n=2.). Selective use of Staging, Intra-fenestrate & extracardiac conduit (EC) was performed based on anatomy & hemodynamics. The Fontan operation consisted of Lateral tunnel, (n=67) or EC (n=8). Additional procedures: Pulmonary arterioplasty (n=14), AVV repair (n=3), pacemaker implant (n=6) & radiofrequency ablation (n=5). Hospital length of stay was 7 & 8 d respectively (p=ns). O2 saturation was 97 & 91%. Persistent pleural effusion and arrhythmia were not different in both groups. Age was the only significant variant between both groups (p=0.009). Conclusion: Selective utilization of Fontan baffle fenestration, Staging & Extracardiac modification provides an excellent outcome. The presence of a fenestration does not predictably decrease the occurrence of pleural effusion.

P91

Unsupported valvuloplasty for congenital mitral regurgitation – long-term results

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Purpose – Valve repair is always preferred to replacement, particularly in children with mitral regurgitation. This paper intends to review long-term clinical results of valvuloplasty techniques without rings for annular support, in congenital mitral regurgitation, in patients under 18 years-old. Methods – A nonselected, consecutive, series of 32 patients submitted to valvuloplasty from 1977 to 1999 were clinically assessed late post-operatively. Surgical techniques included Wooler annuloplasty, cleft closure, chordal shortening, and leaflet resection, all employed alone or in association. No rings or prosthetic bars for annular support were implanted. Mean age: 8.6 ± 5.1 years (10 months to 17 years). Follow up was from 1 to 22 years. Patients included in this series had mitral regurgitation as the only or the main lesion when associated with other cardiac defect. Results – There were 1 early (3.2%) and 2 late deaths (2.4%). Six patients (18.8%) were reoperated late post-operatively for a new repair or valve replacement. There were no cases of endocarditis as well as no cases of thromboembolism in this series. At the last clinical evaluation, functional class (NYHA) was I in 24 (80%), II in 5 (16.7%) and III in 1 (3.3%). In a concomitant group of 50 rheumatic patients, submitted to mitral repair, mortality was absent early and 10% (5) late, with 8% (4) endocarditis, 4.0% (2) thromboembolism and 20% (15) reoperations. Conclusions – The results in this series substantiate the concept that a stable and reliable valve function may be achieved with mitral repair without ring (annular) support. Techniques without prosthesis material may be advantageous in children with severe mitral regurgitation.

P92

Surgical treatment of transposition of the great arteries based on long-term results comparison after neonatal atrial and arterial switch operations.

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The aim of presentation is to compare long-term results of atrial (Senning operation) and arterial switch (Jatene operation) corrections performed within the first 4 weeks of life for newborns with simple transposition of the great arteries (TGA). METHODS: Thirty eight children mean age 6,1 years (range from 1 up to 15 years) who underwent neonatal correction of TGA were reviewed. Group I consists of 18 patients mean age 8,5 years (range from 1 up to 9 years) who underwent arterial switch operation and group II – 20 patients mean age 8,5 years (range from 3 up to 15 years) after atrial switch operation. Types and frequency of occurrence of complications after corrections have been studies as well as their influence on the comfort of life and the development of patients. RESULTS: Tricuspid regurgitation occurred in 5,6% of the sample in group I and in 15,0% of group II (p<0,1). Right ventricle outflow tract stenosis occurred in 16,7% of patients only in group I (p<0,08). Arrhythmias were diagnosed in 43,7% of children in group II only (p<0,06). All the patients belong to New York Heart Association functional class I. 33,3% of children in group I and 25,0% in group II are underweight (p>0,1). CONCLUSIONS: Development of patients in group I and group II is similar. Complications after both procedures did not affect the comfort of life of any patient.

P93

Double outlet left ventricle

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BACKGROUND: Double outlet left ventricle is the rarest type of ventricu-

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

loarterial connection. There is great variability among patients with DOLV in pathologic morphology, associated cardiac defects, and types of surgical repairs. METHODS: Six patients (age 1 day – 2 years) with DOLV underwent surgical repair between 1997 – 2000. Four patients were preoperatively diagnosed and 2 were diagnosed intraoperatively. Important morphologic features were L-TGA in 3/6 and D-TGA in 3/6 patients, PS in 5/6 patients, PA and dextroversion in 1/6 patients. Ebsteins anomaly in 1/6 patients. PDA in 2/6 patients, and coronary artery anomaly in 1/6 patients. Palliative surgeries included Rashkind procedure in 2/6 patients, systemic-PA shunt in 4/6 patients, bidirectional Glenn in 2/6 patients, PDA ligation in 2/6, and pulmonic conduit in 1/6 patients. In 4/6 patients surgical repair consisted of IVC-PA valved conduit. In 2/6 patients translocation of PV from LV to RV was performed thus avoiding conduit. In all patients VSD was baffled to aorta. Additional surgeries included takedown of systemic-PA shunt (3), creation of PFO (2) and infundibular muscle resection (3). RESULTS: There were no early/late mortalities. Early morbidity included seizures in 1/6 patient. Late morbidity included SVT in 1/6 patient controlled with Inderal. Later reoperations included LPA angioplasty and conduit valve balloon dilation in 1/6 patients 5 years after initial repair, and conduit change 2 years later. 1/6 patients had conduit change 6 years after initial repair. At follow-up 3 – 96 months after repair, all patients are asymptomatic. Only one patient has mild conduit obstruction. CONCLUSIONS: Morphologic features of DOLV can be quite heterogenous. With tailored surgical techniques, excellent early and midterm results are achieved. Conduit can be avoided in patients with normal semilunar valve by PA translocation.

P94

Midterm result of aortic regurgitation and pulmonary stenosis after arterial switch operation for d-transposition of great arteries

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Background: Late development of AR and PS are major surgical concerns after arterial switch operation (ASO) for dTGA. We have utilized trapdoor technique and fresh autologous pericardial patch reconstruction of PA. The purpose of this study was to review early and midterm results, especially focusing on AR and PS. Method and Results: For the last 9 years, 52 infants (median age: 12 days) underwent ASO. One of 52 patients, 13 had VSD. Mean aortic cross clamp time was 75.5 +/- 27.0 min. There was 1 operative death, who had non-coronal coronary died from preoperative myocardial infarction. For 51 survivors, mean follow up period was 49.3 +/- 31.5 months. There was 1 late death, possibly due to myocardial ischemia. Doppler echocardiography demonstrated that 12 patients had no AR and 21 had less than mild AR. Median peak pressure gradient between RV and right PA / left PA were 14.5 +/- 21.6 / 14.3 +/- 21.1 mmHg, respectively. Two patients (4.0%) required reoperation due to PS at 12 and 54 months after surgery respectively. Both survived reoperation. Conclusion: ASO for dTGA can be performed with very low operative mortality and good midterm survival. Trapdoor technique can prevent development of AR and reconstruction of PA using fresh autologous pericardium can achieve low reoperation rate for late PS.

P95

Systemic pulmonary shunt through median sternotomy

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There is still controversy regarding systemic pulmonary (SP) shunt for the palliation of cyanotic lesions in neonates or early infants. We performed consecutive thirteen cases (14 times) of systemic to pulmonary shunt through median sternotomy. Median sternotomy approach has several advantages, this approach enables construction of the anastomosis at just distal bifurcation of the pulmonary artery and does not require lung compression. We select SP shunt through median sternotomy in all cases associated with reduced pulmonary blood flow. Two central shunts were performed for pulmonary coarctation and the other 11 patients received modified Blalock-Taussig. The median age was 3.5 (0-20) months old and median weight was 3.4 (2.4-5.7) kg. Median anesthetic time was 174 (44-468) hours and ICU stay was 2 (1-27) days. There was no mortality and no morbidity, and no stenosis in pulmonary artery was showed on the preoperative evaluation by cardiac echo and angiography. The demerit of the procedure is adhesion at the reoperation. In our thirteen patients, five patients underwent reoperation. The reoperation has not been associated with an increased incidence of complications. Systemic pulmonary shunt through median sternotomy has several advantages and is useful.

P96

Surgical management of simple and complex aortopulmonary window

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Objective: Aortopulmonary window (APW) is an uncommon malformation, frequently associated with other cardiovascular defects. We evaluated our experience with APW to determine the impact of associated lesions on management and outcome. Methods: Fifteen patients with APW underwent repair between 1982 and 1999, at a median age of 57 days (range 5–2184). Patients were divided into Group C, with complex associated anomalies (n=8), and Group S, without associated anomalies (n=7). We retrospectively reviewed patient charts to determine the perioperative course of the groups. Results: Anomalies in Group C included coarctation (2), type A interrupted arch (2), pulmonary atresia/ventricular septal defect (1), d-transposition of the great arteries (1), ventricular septal defect (1), and absent left pulmonary artery (1). All patients were taken to surgery with intent to perform complete repair, which was successful in 14. The patient with absent left pulmonary artery could not be weaned from bypass, the repair was taken down. Eight years later lung transplantation was performed with closure of the APW. Circulatory arrest was more common in group C (4/8) than in group S (0/7), p<0.001. Post-operative length of stay was significantly shorter in Group S than in Group C (7.9+/-1.8 vs. 16.9+/-8.1 days, p=0.016). There was no early mortality. One late death occurred, in the lung transplant patient, of overwhelming sepsis 4 months after transplant. The APW was closed in 14 patients using prosthetic material. In the pulmonary atresia case autologous pulmonary artery was used to close the defect. Follow up echocardiography confirmed complete APW closure in all patients. Conclusions: Repair of simple and complex APW can be accomplished with extremely low risk. Associated lesions increase the complexity of repair, prolong hospital stay, but do not increase mortality.

P97

The outcome of Fontan operation in the isomerism patients

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The fontan operation is the surgery of choice for single ventricle. The surgical results improved recently as the surgeons understand the special physiology about fontan circulation. However, in isomerism patients, the results are not good till now. We reviewed the Fontan operation in the isomerism patients. From June 1994 to June 2000, 58 (M:F=34:24), age:50.9±60 (month) patients diagnosed right or left atrial isomerism. Among these patients, 52 patients received initially the Glenn procedure(16), systemic to pulmonary shunt(9) and total cavopulmonary connection (Kawashima procedure 4, Fontan operation 23). 6 patients was not performed operation. 22(44%) of 52 patients received Fontan operation. The most common associated cardiac anomalies are atrioventricular septal defect (100%), common atrioventricular valve (89%), pulmonary outflow tract obstruction (100%), ventricular hypoplasia as single ventricle (79%), Total anomalous pulmonary venous drainage (69%) and bilateral SVC (77%). Most of associated anomalies were corrected at the Glenn procedure. There were 8 perioperative death (15%) only in the right isomerism. Of these patients, 4 received Fontan operation (mortality of Fontan: 27%). The results of fontan operation were much higher than the results of Fontan operation for the patients without isomerism. The isomerism patients have less developed lung parenchyma, which is more damaged by associated anomalies. We prefer the corrective surgery for associated anomalies at the early Glenn procedure and the more strict indications was applied to the isomerism Fontan candidates.

P98

Anomalous coronary arteries in tetralogy of fallot: pre-operative aspects and surgical management

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In patients with Tetralogy of Fallot (TF), the release of proximal right ventricular outflow tract (RVOT) is sometimes difficult, specially with hypoplastic pulmonary annulus. In presence of anomalous coronary arteries (ACA) crossing the RVOT, the surgical approach becomes harder, with higher morbidity and sometimes need of prosthetic conduits. We reviewed 652 patients with TF operated on from January 1990 to July 2000 and 94 (1.38%) of these presented ACA. Mean age was 52.8 months and 66.7% were male.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Diagnosis of ACA was not established pre-operatively in 22.2%. The most common pattern of coronary anomaly was the left anterior descending artery coming from the right coronary artery (66.7%), crossing the RVOT. a left coronary artery originated from the right aortic sinus, a higher and posterior take off of the right coronary artery and a very well developed infundibular branch crossing the RVOT were also found. Mean pre-operative RVOT gradient was 78.1 mmHg by Echo, and 62.3 mmHg by angiographic study. The obstruction was valvar and subvalvar in 77.8%. Surgical approach was transatrial-transventricular (22.2%), transatrial-transpulmonary (11.1%), transatrial-transpulmonary plus right ventriculotomy (66.7%). Pulmonary valvotomy was performed in 4 patients (44.4%) and in other 2 children (22.2%) the restrictive annulus was enlarged using a monocuspid valve attached to a transannular patch, positioned superiorly to the ACA. In another case the patch was placed under the ACA, that was dissected and isolated. In one case (11.1%) an extracardiac conduit was used, from RV to pulmonary artery. No surgical deaths were observed. One patient died in the 17th day in consequence of multiple system organ failure. The post-operative RVOT gradient by Echo was 27.4 mmHg and all 8 patients remained asymptomatic in a mean follow-up period of 28.3 +/- 27.8 months. One patient developed a RVOT aneurysm 5 years after surgery and is listed to repair. In conclusions, the correction of TF with ACA crossing the RVOT was possible with usual surgical techniques, with no increase in morbidity. Satisfactory release of RVOT obstruction was obtained in all cases.

P99

Interrupted aortic arch: one stage or two stage repair?

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INTRODUCTION: Interrupted aortic arch is a complex congenital heart defect which is uniformly fatal if left untreated. Surgical treatment has evolved from a two-stage repair to a one-stage correction. Controversy exists regarding the optimal management strategy. METHODS: We undertook a 20 year review of our experience with treating interrupted aortic arch to document operative risk, late complications and changes in management strategy which have evolved with time. RESULTS: Fifty-six patients were operated for interrupted arch. There were 19 type A, 35 type B and 2 type C interruptions. Median age was 9 days. Ten patients with complex intracardiac anomalies underwent single-stage repair using circulatory arrest with a 30% mortality. Three of 7 survivors required arch revision. Forty-six other patients had staged repair. There was a 6.5% mortality at the first stage and 0% mortality at the second stage. No deaths have occurred in the last 20 cases in the group. Median age at 2nd stage was 9 months, but is lower in more recent times. Additional procedures in the group included replacement of conduit (9), resection of subaortic stenosis (5) and band-related problems (6). DISCUSSION: We believe that staged repair is a safer option for treatment of interrupted arch. Single-stage correction may be reserved for type C interruptions and lesions requiring late banding (eg AP window truncus). A significant proportion of patients with staged repair will require further surgery for conduit change and associated problems but the apparent disadvantage compares with the frequent need for arch revision in single stage correction.

P100

Transverse plication of posterior wall of main pulmonary artery due to aneurysmal dilatation after total correction of tetralogy of Fallot

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We can frequently meet the aneurysmal dilatation around right ventricular outflow tract (RVOT) after total correction of tetralogy of Fallot(TOF), especially in the case with the stenotic pulmonary tribute. It may develops on the situation of pulmonary stenosis, high right ventricular pressure and/or pulmonary regurgitation. For more laminar flow, we placated dilated posterior wall around pulmonary valve transversely. Since Apr. 1997, 12 patients (recent TOF 8, primary total correction of TOF 3, redo PA 1) received surgery due to RVOT dilatation and stenosis without mortality. Between the origin of pulmonary valve and the end of the main pulmonary artery or end of pulmonary sinus, concave space was found in the posterior wall. This space is obliterated with the absorbable continuous suture from the origin of pulmonary valve to the distal end of concave space. Preoperatively, the echocardiography shows the even diameter through main pulmonary artery. The pressure ratio of right ventricle and left ventricle changed from 0.82 to 0.43 and the pulmonary regurgitation was decreased, however, the size of distal pulmonary artery did not increase. The adjuvant transverse plication of posterior wall of main pulmonary artery may be helpful for complicated stenovalvular RVOT at redo-correction of TOF because the flow in RVOT will be more laminar.

P101

Surgical repair of complete atrioventricular septal defects in infancy with surgical modification: Ten-year experience

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The operative mortality of complete atrioventricular septal defects decreased over the past 20 years. Our history of patients with atrioventricular septal defects presenting to our institution in infancy between July 1989 and November 1999 were reviewed with the purpose of analyzing the long-term surgical results. 37 consecutive patients (M:36, F:34, age range, 1 to 12 months, mean body weight 5.9 ± 1.4kg) underwent primary intracardiac repair of complete atrioventricular septal defect. Down's syndromes were present in 19(51.4%). The 25 valves were postoperatively classified as Rastelli type A(67.6%), 2 classified type B(5.4%), and 9 classified type C(24.3%) with 1 not clearly classified. Mean follow-up time was 36.3 months. All 37 patients underwent biventricular repair. The two-patch technique was applied in all patients. A modified simple suture technique was applied in 26 (70.3%) and reversed-shape of patch for ventricular septal defect was applied in 16 (43.2%). Early operative mortality was 13.5% (5 patients) and overall mortality was 21.6% (8 patients). Causes of death were right heart failure (3 patients), sepsis (2 patients) and weaning failure from cardiopulmonary bypass (1 patient). Mean operative time, mean total bypass time and mean aortic cross clamp time were 260.4±65.1, 130.5±44.2 and 101.2 ±25.2 minutes. 4 reoperations underwent in 3 patients(8.1%). 3 of 4 were the replacement of left atrioventricular valve, remaining 1 was the repair of left atrioventricular valve. Postoperative complications developed in 14 patients (37.8%). 5-year actuarial survival, including operative mortality, was 82.3% and freedom from reoperation was 95.8%. Repair of atrioventricular septal defects in infancy with surgical modification has acceptable early mortality, long-term survival rate and a low incidence of reoperation.

P102

Repair of complete atrioventricular septal defects with tetralogy of Fallot

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Between 1984 and 2000, 10 patients with complete atrioventricular septal defects with tetralogy of Fallot underwent surgical repair. The mean age at operation was 9.5 years, and the mean body weight 26.5 kg. Five patients underwent 5 systemic-pulmonary shunts respectively. Five patients had Down's syndrome. The ventricular septal defect was closed through a right atrial and right ventricular approach in seven patients, and through a right atrial approach in three patients, using a comma-shaped prosthetic patch. The atrial septal defect was closed with a separate patch. The left anterior and posterior leaflets were sutured together using multiple interrupted sutures. Right ventricular outflow obstruction was relieved by a infundibular patch (one case), monocusp equine pericardial patch (two) or monocusp autologous pericardial patch (seven). There were two hospital death (20%), caused by low cardiac output in both cases. There was no late mortality in the long-term follow-up study(0.7–16.1 years, mean 5.1±5.6). right ventricular outflow tract gradient ranged between 0 and 41 mmHg (mean, 19.6±13.1). Two patients had mitral regurgitation, 1 needed reoperation. It was concluded that our technique for this lesion contribute to satisfactory immediate and long-term results after repair.

P103

Initial conclusions with pediatric minimally invasive cardiac surgery

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The objective of this study is to show the experience with pediatric minimally invasive cardiac surgery (PEDMICS) to treat different congenital cardiac defects. From aug/1996 to aug/2000, 58 children (33 female) were operated on by minimally invasive access; the age ranges from 25 days to 19 years (36 months) and the weight from 3.4 to 52 kg (15.5kg). 44 children (29 female) had patent ductus arteriosus (PDA) and 14 had atrial septal defect (ASD). In the PDA group, a small incision (2.5cm) was performed in the posterior elbow, in left 4th intercostal space. After lung retraction, the PDA was

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

exposed and a double clip ligation was performed, no chest tube was placed. In ASD group, all children were operated on by a mini-sternal access with no sternal opening. CPB was implanted by the same incision and in all cases the ASD was closed by continuous suture. 60,1% of the children were extubated in the OR. No immediate mortality was observed and the most frequent complication was systemic hypertension detected in 66,9% of PDA cases. All children were discharged from hospital between 4th and 6th PO day, despite one child who died in 45th PO day due to pulmonary infection complications. Post-operative Echo showed no residual defects. In conclusion, the initial experience with PEDMICS showed safe and efficient with good results. The hospitalization was short and no major complications were observed. The closure was an isolated event, with favorable evolution in all the other children. The cosmetic aspect of the incision was considered very satisfactory.

P104

Midterm result of transatrial repair of tetralogy of Fallot – non-neonatal repair Blalock-Taussig shunt

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Summary. Between November 1990 and September 2000, we performed transatrial repair of tetralogy of Fallot (TOF) in 117 patients and 22 staged palliation for repair of TOF. Patients age was ranging from 2 months to 14 years in repair group and 17 days to 7 years in palliation group. Total repair was consisted of transatrial approach utilizing extra-transannular patch in necessary cases. Palliative procedures were 20 modified Blalock Taussig shunt and 1 branch PA plasty. All patients has been followed up in outpatient clinic. There was only one non-cardiac mortality in repair group (0.8%) from fulminant hepatitis. No operative mortality nor morbidity in shunt group. In repair group there was no surgically created AV block. No patient necessitated RV-PA conduit because of anomalous coronary artery on RVOT. No episode of life-threatening arrhythmia was noted. There were four reoperation after repair (3.4%). All of them were needed for RVOTA± branch PS. All patients, more than 1 year after repair, does not have any exercise restriction. Conclusion: We achieved excellent result in non-neonatal transatrial/pulmonary repair of TOF allowing staged approach if necessary. Staged approach did not add any risk in our series. Considering excellent midterm outcome of patients hemodynamics and exercise tolerance, we consider transatrial repair as a surgery of choice for Tetralogy of Fallot instead of transventricular approach, even if which can be performed.

P105

Tracheobronchial compression (obstruction?) after arterial switch operation

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A rare complication following arterial switch operation (ASO), tracheo-bronchial compression (obstruction?) is reviewed through our cases. From 1990 to 2000, 37 patients with transposition of great arteries (TGA) underwent ASO, and 19 patients survived. Among 19 survivors, 2 patients developed tracheobronchial compression after surgery. Patient 1, with TGA and multiple VSDs, underwent ASO by Lecompte's maneuver and VSD closure on day 47. Three months later, he showed up with dyspnea, and a chest X-ray showed rightward shifted trachea as well as hyperaerated left lung. A magnetic resonant imaging revealed severely obstructed left main bronchus by ascending aorta. A flexible fiberoptic bronchoscopy showed pulsatile compression of the left main bronchus. He underwent surgery on 4 months of age. The ascending aorta was suspended toward the sternum, and his symptoms related to airway obstruction improved dramatically. Patient 2, with simple TGA, underwent ASO by Lecompte's maneuver on day 45. After operation he developed atelectasis of his left lung. So he required respiratory support physiotherapy in one month. His CT scan showed rightward shifted trachea and compression of the left main bronchus by ascending aorta. As ASO by Lecompte's maneuver allows the aorta lies beneath the pulmonary artery, the aorta can compress the left main bronchus, and push the trachea toward right side. Therefore, this potential complication, tracheobronchial compression must be considered in case any respiratory distress develops following.

P106

Management of PDA for extremely low birth weight infants with pulmonary atresia with intact ventricular septum following Brock operation

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Brock operation is thought to be the only surgical procedure for extremely low birth infants (ELBWIs) with pulmonary atresia (PA) with intact ventricular septum (IVS). Even for a patient with well-developed right ventricle suitable for the Brock operation, management of PDA is important for ELBWI during both pre- and perioperative period. Between 1991 and 2000, 2 ELBWIs underwent Brock operation. Patient 1 was critical PS with birth weight of 878g and patient 2 was PA with IVS with birth weight of 752g. In patient 1, anterior cerebral blood flow decreased since 6th day of life under continuous infusion of lipo PGE1. After Brock operation on 10th day, PGE1 infusion was discontinued. The diameter of PDA was evaluated as 2.0 mm with UCG. On 14th day, diastolic blood flow was detected in celiac, superior cerebral, renal, and mesenteric arteries. Blood supply through these arteries was thought to decrease. Meferamor acid on 16th and indomethacin infusion on 25th and 43rd day induced spontaneous closure of PDA. Abdominal aortic blood flow was detected as reverse on the third day of life under lipo PGE1 infusion in patient 2. Peritoneal drainage was performed on her 8th day for necrotizing enterocolitis. After Brock operation on 11th day, indomethacin infusion did not make any effect on PDA closure. The diameter of PDA was 3.3 mm with UCG. To keep systemic blood flow well, we treated PDA on 13th day. Brock operation should be performed early in ELBWI with PA, IVS before decrease in systemic blood flow due to PDA occurs. In cases with enough pulmonary blood flow through the pulmonary valve after Brock operation, early PDA closure after the operation may be required.

P107

The effects of Blalock–Taussig shunt on the pulmonary artery growth and ventricular function in patients with single right ventricle

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To identify the preoperative factors (except for main ventricular type) that influence the growth of pulmonary artery and ventricular function after Blalock- Taussig shunt (BTS), sixty-three patients with functionally univentricular heart of right ventricular type who underwent a BTS operation were reviewed. After BTS, satisfactory growth of the pulmonary artery and preservation of ventricular function were recognized in most patients. However, there were several preoperative risk factors that significantly influenced the pulmonary artery growth. The growth of the pulmonary artery was greater in younger age group (<1 year old) when the development ratio was calculated as the ratio of postoperative to preoperative pulmonary artery index [post PA/pre PA] (<1 year old: 189.9+/-113.0% vs >1 year old: 142.2+/-62.2%, p=0.002). Patients with common AV valve (CAVV) or heterotaxia had significantly higher incidence of AV valve regurgitation after BTS (p<0.01). Postoperative pulmonary vascular resistance (Wood units) was higher in the group (CAVV: 2.2+/-0.2 vs. others: 1.7+/-0.1, p=0.05), and highest in asplenia/polysplenia: 2.6+/-0.3 vs others: 1.9+/-0.1, p=0.02). Our results suggest that BTS in early infancy is not recommended in patients with single right ventricle for later Fontan-type operation. However, there may be undetermined factors that influence the development of pulmonary artery and AV valve regurgitation in patients with CAVV and/or heterotaxia.

P108

Management of tetralogy of Fallot and double outlet right ventricle with pulmonary atresia associated with coronary artery anomaly

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Although only 2–9% of patients with tetralogy of Fallot (TOF) have important coronary artery anomalies, these often have an important impact on the timing and technique of operative repair. 437 patients with TOF and 38 patients with DORV who were operated on between June 1989 and October 2000 were reviewed retrospectively for coronary anomaly. 67 out of 475 patients had coronary anomalies diagnosed during catheterization, selective coronary angiography or surgery. Incidence of coronary anomalies was 13.4%. The most common anomaly was left anterior descending artery (LAD) to accessory LAD originating from right coronary artery (RCA) in 28 (31.3%) and single coronary artery originating from left sinus valsalva in 16 (24%). The course of RCA was anterior to aorta in 6 patients with single

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

coronary artery and these arteries crossed the RVOT. Long one or two conal arteries arising from RCA were diagnosed in 38(27%) of 139 patients. These conal arteries crossed the RVOT. A total of 45 coronary arteries crossed the RVOT. 58% of coronary anomalies were diagnosed in first cardiac catheterization. 20% of angiograms were insufficient for diagnosis of coronary anomalies. After surgical diagnosis of coronary anomalies all angiograms were reexamined. We were still unable to diagnose coronary anomalies in 15% of the angiograms. As a total, anomalies/one angiogram ratio was 11.6%. Total correction was succeeded in 43 patients without palliation and in 7 patients with palliation. Transannular patch was used in 27 (54%), conduit was used in 9 patients (18%). 14 (28%) were operated without transannular patch. Right to left ventricle pressure ratio was 0.52±0.10 (0.37–0.77) at total correction group. Surgical mortality was 5.9%. Doppler gradient was 16±9 mmHg(6–57) in last echocardiographic examination. In conclusion the incidence of coronary anomaly was found higher than in other series if not searched for carefully the diagnosis would be impossible. Staged repair with early palliation may be required in a quarter of this patient group. Although most patients were operated on using only transannular patch, conduit repair may be necessary in a small group.

P109

The Fontan procedure in adults

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[Purpose of study] A retrospective clinical study was performed to document the course of adult patients undergoing the Fontan procedure. [Method] Between 1981 and 1999, 13 adults aged 18 to 36 years (mean age, 25.4 years) underwent a Fontan procedure. Anatomic diagnosis was an upper atresia in 2, double inlet right ventricle in 1, and various single ventricles in 10. Other complex lesions were TAPVC in 1, bilateral SVC in 4, hemiazygos continuation of the IVC in 1, asplenia syndrome in 2. The Eight patients had undergone prior palliation. Preoperative examination were mean PA index 379, mean mPA pressure 11.4 mmHg, mean Rp 1.45 units, and mean mean ventricle EF 63.1%. Ten underwent a right atrio-pulmonary artery connection (APC), 3 had total cavapulmonary connection (TCPC). [Results] The operative mortality rate was 7.7% (1/13). The remaining 12 patients have been followed for a mean of 9.2 years. During follow up period, two patients have required reoperation and one of the two patients died of postoperative MOF. There has been 3 other late death, which were probably due to ventricular arrhythmia. All survival cases were in NYHA class I, and have had development of atrial and/or ventricular arrhythmia requiring medication. The actuarial survival rate at 10 years was 58.5 %. [Conclusion] These results indicate that properly selected adults can undergo the Fontan procedure with low mortality. However, late-developing arrhythmias and decreasing ventricular function are serious problems that mandate careful follow-up.

P110

Vascular arch anomalies and tracheo–esophageal compression in children

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The anatomical types, clinical presentation, associated conditions, investigations, treatment and outcome of children who were diagnosed to have symptomatic vascular arch anomaly over a 15 year period was reviewed. 10 subsequent patients (6 males, 4 females) who underwent surgical treatment were analysed retrospectively. The median age was 6 months (3–36 months). Four patients had double aortic arch, 3 had right sided arch descent with left ligamentum, one aberrant right subclavian artery, one pulmonary artery sling, and one had right aortic arch with retroesophageal descent. Eight children presented with respiratory symptoms and 2 with dysphagia. The associated conditions were cleft lip and palate (1), tracheobronchial stenosis (1), presence of diaphragm (1) and Di George syndrome (1). The associated cardiac anomalies were bicuspid aortic valve, ventricular septal defect and aorto pulmonary window. Chest radiography, barium meal, 2-dimensional echocardiography and a computerised tomograph (CT) scan were performed in all patients. Barium meal proved to be the most reliable investigation to detect the presence of a vascular anomaly with a consistent feature of either anterior and/or posterior indentation of the esophagus. Nine patients had a division of the vascular anomaly in which 2 had aortopexy and one had reimplantation of the pulmonary artery. Two infants died. One due to septicemia post surgery and one during a subsequent surgery for correction of aorto pulmonary window. Six patients had rapid improvement in symptoms within 1–2 weeks after surgery with the remaining patients improving over a 3–6 months period. Vascular arch anomalies are rare and needs a high degree of suspicion for diagnosis. But non-invasive investigation for early recognition.

P111

Atypical coarctation of the aorta with multiple saccular aneurysms

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Single Ventricle (SV) with Subaortic stenosis (SAS) is difficult subset of patients to achieve successful Fontan operation. Between June 1993 and November 2000, Fifteen patients with SV/SAS were enrolled Fontan protocol. 9 patients underwent PA banding +- CoA repair (Group P) and 6 patients underwent Damus-Kaye-Stansel (DKS)/Norwood operation at first palliation (Group D). Indication for DKS/Norwood operation is diameter of Ascending Ao or LVOT less than [Body weight +1]mm and/or subaortic stenosis potentially treat SAS rapidly. LVOT/AoV/Asc Ao diameter (mm) were 3.8+-0.5/5.3+-0.6/ 6.0+-1.0/ in Group D and 7.6+-2.2/7.9+-1.8/9.3+-5.0/ in Group P respectively. Age at first palliation were 14+-15 days in Group D, 97+-8 days in Group P. In Group D, 1 patient with Cor triatriatum died due to progressive PVO. 4 patients proceeded with bidirectional Glenn (BDG) operation at age 6.8(3.1 months and 1 patient is waiting for BDG. Fontan operation was performed in 2 patients and another 2 patients are waiting for Fontan as good candidate. In Group P, there was no early and late mortality. 8 patients underwent DKS/BDG operation at second palliation and 1 patient underwent DKS/Fontan without second stage. Mortality rate for entire protocol was 6.6%. Comparable Fontan result could be achieved in Fontan protocol for SV with SAS utilizing DKS/Norwood or PA banding at first palliation and early conversion to DKS/BDG before developing SAS to the patients after PA banding.

P112

Surgical correction for double-chambered right ventricle

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From 1998 to 2000, 10 patients with double-chambered right ventricle (DCRV) underwent surgical correction. The diagnosis of DCRV was established by echocardiography, cardiac catheterization, and recently 3- dimensional computed tomography (3-D CT) (Fig 1). Age at operation ranged from 1 to 6 years (mean 2.8 ± 1.9 years) in 6 pediatric patients and ranged from 30 to 58 years (mean 44 ± 13 years) in 4 adult patients. The mean pressure gradient between the right inflow chamber to pulmonary artery was 47 ± 18 mmHg in the pediatric group and 92 ± 25 mmHg in the adult group. Surgical correction consisted of resection of abnormal muscle bundles and closure of the ventricular septal defect if present through a right ventriculotomy. All patients but one survived. One patient died in the early postoperative period of right heart failure. No residual right ventricular outflow obstruction was detected with mean pressure gradient of 12 ± 6 mmHg. At a mean follow-up of 4.1 years, there were no late death and no reoperation and all survivors were in NYHA class I. Aortic regurgitation (AR), not detected preoperatively, developed in 2 pediatric patients (20%) 5 and 6 years after operation respectively. The causes of AR were subaortic fibrous ridge and unknown. In conclusions, surgical correction for DCRV was performed in 10 patients including 4 adults with satisfactory outcome results. Recent 3-D CT was very useful for understanding of anatomy of RV outflow. Development of AR might be future sequela in some pediatric patients with DCRV.

P113

Clinical pathway of first track recovery to school activities in children after minimally invasive cardiac surgery

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Background: Minimally invasive cardiac surgery (MICS) for simple congenital heart disease has been introduced and become a standard method. As first track recovery to school activities is important for children to enjoy normal school life as normal children, we have made clinical pathway of first track recovery to school activities in children after MICS. We assessed how the clinical pathway was achieved in children with ASD and VSD after MICS. Methods: Fifteen children in school age who underwent repair of atrial and ventricular septal defect (ASD and VSD) through lower midline sternotomy

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

were investigated. The median age was 10.1±7.8 years. Clinical pathway of these children was followed: They were extubated in OR. After postoperative echocardiography at 5POD, they were discharged at 7POD. They visited first outpatient clinic 5 to 7 days after discharge and were allowed to go school and to do lower body exercise within two weeks, and normal all gymnastics within 6 weeks after MICS. Results. Postoperative hospital stay was 8.7±1.1 days and they re-started going school 11.2±3.1 days after discharge. The length until participating gymnastics was 41±11 days. 12 of 15 (80%) children were able to recover according to our clinical pathway. Conclusions. Minimally invasive cardiac surgery can be performed safely in all children in the study. In addition to improve cosmetic results, this technique can reduce the length of hospital stay and recovery length until going school. Future advances in technology should minimize the impairment of postoperative school activities in the majority of children undergoing cardiac surgery.

P114

Surgical treatment of cardiac tumors in early infancy

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Objective: primary cardiac tumors in infancy are rare, and they are characterized by heterogeneous nature and clinical features. Surgical treatment is advocated when symptoms or hemodynamic impairment are present. Methods: from October 1989 to April 2000, 6 infants (2 males, 4 females) with diagnosis of primary cardiac tumor were addressed to surgery. Ages ranged from 5 to 456 days. There were cardiac fibroma (2 pts), hamartoma (1 pt), multiple rhabdomyomas (1 pt), teratoma (1 pt), left atrial myxoma (1 pt). Diagnosis was made by bidimensional echocardiography in all, while CT scan and cardiac catheterization were added in 2 patients, MRI in one. All patients underwent midline sternotomy. Complete resection of tumor was possible in 5 patients, while orthotopic cardiac transplantation was performed in one. Results: there were no hospital deaths. One patients died late for cerebral neoplasm 2 years after cardiac transplantation. No reoperation was required. At a mean follow up of 39 months (range 8–58 months) all survivors are asymptomatic and well. Bidimensional echocardiographic data show good ventricular function in all, with mean EF of 74%. Conclusion: surgical excision of obstructive neoplastic masses is safe and feasible even in infancy; cardiac transplantation is indicated in case of large tumors which extensively invade myocardial tissue and compromise cardiac function irreversibly. Two dimensional echocardiography is in most instances a reliable diagnostic tool, seldom requiring other complementary diagnostic imaging techniques. Complete and correct diagnosis of cardiac tumor still deserves histopathologic characterization.

P115

A report of 2 cases with palliative Jatene –practical strategy for staged radical operation

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[Background]. Jatene procedure is the definitive method for d-Transposition of Great Arteries (d-TGA). However, we have an opportunity to use the Jatene procedure as a palliative operation. We have 2 cases of a Palliative Jatene procedure for complexed congenital heart defects. [Patient 1]: Two-year-old-boy was diagnosed as d-TGA (typeO2) with restrictive VSD and hypoplasia mural valve, and performed ASD creation, VSD enlargement, and PAB just after birth. We hesitated a radical operation using Jatene procedure because of hypoplastic mitral valve like parachute. Therefore, the patient was received a Jatene procedure as a palliative operation in 14-month-old for progressing cyanosis, and radical operation in 24-month-old. [Patient 2]: Four-year-old-boy had Single LeftVentricle (SLV) and right sided atrioventricular valve atresia with d-malposition of great arteries. This patient was performed a PAB at 10-days of age. However, PAB was not enough to against the pulmonary hypertension because of subaortic stenosis. Therefore, the patient was received the Palliative Jatene and re-PAB in 15-month-old, and Fontan operation in 23- month-old successfully. [Summary]: The first patient was improved cyanosis, and the second patient was prevented from exacerbating pulmonary hypertension after Palliative Jatene procedure. The quality of life of the patients with palliative Jatene procedure was much better than that prior to the palliative operation because the Palliative Jatene was effect on controled pulmonary blood flow. Moreover, the radical operations were also performed more smoothly. [Conclusion]: We propose that the Palliative Jatene procedure is one of practical ways bridging to radical operation of complexed congenital heart de

P116

Cardiac echinococcosis: results of combined surgical and medical treatment

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Cardiac echinococcosis is very uncommon, especially in children, but it is associated with a high risk of potentially fatal complications, anaphylactic shock, embolic events, intracardial inlantation(MI). We present a retrospective analysis of the management of 5 patients with cardiac echinococcosis, operated between February 1998 and April 2000. Echocardiography has proved to be the diagnostic method of choice. MRI was performed in 2 patients, CT-scan in one. Serologic tests were positive for echinococcosis in 3 and negative in 2 of the cases. Three patients had a single cyst and 2 patients had 2 or more cysts. The size ranged from 1 to 10 cm in diameter. The localization was subepicardial communicating with the RA and compressing RCA and LAD with subsequent MI, RV-wall, apical IVS and pericardial in 2 cases. The patient with intraseptal localization was treated preoperatively with Albendazol for 99 days, but growing of the cyst with structural changes was noted on EchoCG and he was sent for surgery. In one case preoperative echocardiography and MRI showed 2 large formations" resembling residual cysts, but the descriptions coincided exactly with the intraoperative findings, so we concluded that this impression was due to the pericyst layer. The patient with epicardial localization and MI had a rupture of the cyst in the RA with anaphylactic shock just before the operation and died on the 4th postoperative hour. The other 4 patients underwent successful surgical enucleation and the follow-up is uneventful. Albendazol was administered postoperatively for all the patients. Conclusions: 1. Echocardiography is the diagnostic method of choice and can provide the exact diagnosis alone or together with MRI, CT-scan and serologic tests. 2. Surgical enucleation remains the treatment of choice. 3. Pericardial localization seems not so rare. 4. We think Albendazol should be administered routinely postoperatively."

P117

Atrial septal defect with left main coronary trunk compression by dilated main pulmonary artery

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We report a case of a twelve-years-old girl with narrowing of the left main coronary trunk, which was compressed by the dilated pulmonary artery, associated with the atrial septal defect and severe pulmonary hypertension. Cardiac catheterization revealed the following: pulmonary/systemic flow ratio (Qp/Qs) 1.97, pulmonary/systemic arterial systolic pressure ratio(Pp/Ps) 0.54, pulmonary resistance index(Rp) 5.67. The coronary angiography demonstrated localized narrowing of the left main coronary trunk, but normal shape was observed. the following CAG. Treadmill exercise test disclosed ischemic change on the electrocardiogram without symptoms. A central type of atrial septal defect was closed with a Dacron patch. Additionally, coronary artery bypass grafting to the left anterior descending artery with the left internal mammary artery and lung biopsy were performed. The postoperative catheterization revealed that the Pp/Ps decreased to 0.40, and the coronary angiography showed the improvement of stenosis of the left main coronary trunk. Treadmill exercise test at discharge did not disclose obvious ischemic change. The pathological examination of the lung revealed the growth of the vascular smooth muscle and elastic fiber, the occlusive change of the pulmonary vessels and the possibility of the postoperative residual pulmonary hypertension. We conclude that the dilated pulmonary artery due to high resistance of pulmonary artery might compress and stretch the left main coronary trunk and cause severe stenosis of it, and the coronary artery bypass grafting with closure of the atrial septal defect might be required in the case of high pulmonary r

P118

Short term follow-up after the Ross operation in children

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Aortic root dilatation, early homograft stenosis and left ventricular dysfunction due to coronary problems are major concerns of the pulmonary homograft replacement of the aortic root in children. The purpose of this study was to

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

assess the short-term follow-up in children after the Ross procedure. The charts of 13 consecutive children, with severe aortic regurgitation who underwent the Ross procedure in our institution were retrospectively reviewed. Median age at operation was 10±5.4 years, median weight 31.8±17 kg. All patients survived (median hospital stay 14±5.8 days). The neo-aortic valve showed no or trivial insufficiency in all but 1 child (grade II). Median follow-up was 25±18 months. Analysed parameters were LVEDD, diameters of the aortic valve annulus and aortic sinuses and paired t-test was used for statistics. LVEDD decreased significantly after surgery ($p<0.0001$). LV shortening fraction (SF) decreased from 32±5 to 23±7 ($p<0.003$) but improved to normal within 6 months in all but 1 patient who had a decreased FS preoperatively. Postoperatively there was no significant increase of the neo-aortic valve diameter ($p<0.089$) but a significant increase in diameters at the level of the sinuses at 12 months ($N=7$; $p<0.041$) and 24 months ($N=6$, $p<0.026$) without aortic valve impairment. In a 2-year-old patient the homograft in pulmonary position had to be changed due to severe stenosis 5 months after the Ross procedure. The Ross procedure in childhood appears to be a safe procedure and leads to a prompt reduction in LVEDD. Reduced movement of the IVS might cause transient reduced SF. There was evidence that dilation of the neo-aortic sinuses occurs. The significance of these findings regarding coronary filling and aortic valve competence must be assessed in long term studies.

P119

Minimal invasive approaches in pediatric cardiac surgery

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Full midline sternotomy is the standard approach for correction of congenital heart defects in pediatric patients. However minimal invasive approaches will more and more be favoured due to their advantages regarding postoperative recovery and cosmetical results. Between July 1999 and November 2000, 22 infants and children (7 male, 15 female, mean age 5.6 year (2 months-13 years), mean weight 19.1 kg (4.2–63 kg) were operated using minimal invasive approaches. Nine children (8 girls and 1 boy) were operated via a limited right anterolateral thoracotomy for ASD II (n=7), PAPVR (n=1) and intermediate type AV canal (n=1). Thirteen patients (7 girls, 6 boys) had partial inferior sternotomy using a special retractor for ASD II (n=8), PAPVR (n=1), ASD I (n=1), VSD (n=2) and TOF (n=1). Access for CPD was gained through the same incision in all patients to avoid groin incisions with the risks of peripheral vascular lesions. There were no perioperative complications. Mean operation time was 3.2 h (2–5.15 h), mean CPB-time was 72.4 min (21–149 min) and mean aortic cross-clamp time was 38.1 min (15–92 min). Eleven patients were extubated in the OR, the other 11 patients were ventilated for a mean of 13h (2 – 47h). Mean ICU-stay was 1.6 days (1–4 d). There was one postoperative complication (pneumothorax on the 5th postoperative day). Mean hospital stay was 8.5 days (6–14 d). Follow up was uneventful in all patients despite a secondary wound infection in one case 4 weeks after discharge. All parents considered the cosmetic result excellent. Surgery for the correction of some congenital heart defects can safely be done in pediatric patients via minimal invasive approaches without femoral cannulation. Cosmetic results are excellent.

P120

Anomalous Left Coronary Artery from the Pulmonary Artery – Variable Presentation with Excellent Surgical Outcome

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Since 8/91, 16 patients have been diagnosed with anomalous left coronary artery from the pulmonary artery (ALCAPA) at the Children's Hospital of Wisconsin. Nine of the 16 patients presented in infancy (mean age 4.7 mo, range 1.5–11 mo) with congestive heart failure and echo findings of a severe dilated cardiomyopathy (mean LVEF 23%). The remaining 7 patients were older at the time of diagnosis (mean age 7.5 yr, range 6 mo–18 yr), and 6 of these 7 patients were asymptomatic, presenting with a murmur and/or cardiomegaly on CXR. One patient presented at the age of 18 yr with an episode of sudden death, and was resuscitated. The most common echo findings in the older group were marked right coronary dilatation and septal coronary collaterals in 7/7 patients and endocardial fibroelastosis of the mitral valve apparatus with prolapse and significant mitral valve insufficiency (MI) in 4/7 pts. The echo determination of the coronary insertion correlated well with surgical findings. 12 had anomalous insertion into the main pulmonary artery while 3 had insertion into the right pulmonary artery. Fifteen of the 16 pts have undergone surgical correction with no surgical or late mortality. Nine pts had direct reimplantation of the coronary, 6 pts had a tunneling procedure because of a left lateral insertion of the coronary. The older pt with sudden death also had a resection of a LV aneurysm and placement of an AICD. The infant group had a longer hospital stay (mean 33.5 days vs. 6.3 days), with 2 of the 9 infants requiring post-operative ECMO support. Follow-up echocardiographic studies in the infant group have revealed normalization of ventricular function in all pts, with mild to moderate residual MI in 6/9 pts. Follow-up echocardiographic studies in the older group reveal normal EF in all pts except for the pt with sudden death, and improvement in the degree of MI in 2/4 pts. In conclusion, despite the strikingly variable presentation of ALCAPA, surgical outcome is excellent. The infants with severe LV dysfunction may require intensive post-operative support, but recovery of ventricular function is expected. Residual mitral valve dysfunction is common in many pts, and warrants long-term follow-up.

P121

Repair of Double-Outlet Right Ventricle with Taussig-Bing by Arteries Switch Operation

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Objective: Double-outlet right ventricle with subpulmonary ventricular septal defect (Taussig-Bing) was repaired by arteries switch operation. Methods: Five patients with Taussig-Bing have been repaired by arteries switch operation. The operative age was 2.7 months, the weight was 4.5–6.3kg. Two patients had pneumonia and heart failure, they were repaired by emergency operation. The operative procedure consisted of intraventricular tunnel repair for VSD to pulmonary artery and arteries switch operation, underwent deep hypothermia and circulation arrest or low-flow perfusion. One patient had abnormal coronary artery. Result: One patient died of heart failure and low urine output postoperation. All four patients had an eventful recovery and discharged postoperative 10–24days. Conclusions: Taussig-Bing was repaired by arteries switch operation to prevent postoperative left ventricular outlet tract obstruction, avoid used of conduit in right ventricle, decrease operative mortality and create advanced operative results.

TeleHealth

P122

Impact of telemedicine on pediatric cardiology practice 1987–2001

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Introduced in 1987, telemedicine has transformed patient care and education in our tertiary pediatric cardiology service. 1 High resolution local hand transmission of echocardiograms live from remote sites has allowed 24 hour access to pediatric cardiology expertise by 6 regional hospitals. During 13 years over 650 studies have been transmitted with excellent quality. Avoidance of travel costs more than offsets the operating costs of the network. 2 Since 1995 teleconsultations (384 kbps) have been performed for 7 selected patients with arrhythmias or syncope and 19 pre-op counselling sessions. Both families and health personnel adjusted quickly to the teleconference and expressed satisfaction. Families appreciated avoiding the cost and time of travel for in-person visits. 3 Cardiosurgical teleconferences with a referring cardiologist have used 384 kbps bandwidth to transmit echo or angiographic images with suitable resolution for conferences. 4 Monthly resident teaching teleconferences have linked 6 training programs across Canada. Expertise has been shared in cardiac pathology, arrhythmias and MRI. Thus, telemedicine has greatly expanded both access to care and teaching for pediatric cardiology in our regional center.

P123

Impact of telemedicine on medical and financial outcomes in neonates with congenital heart disease

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Telemedicine improves ability to diagnose congenital heart disease (CHD) expeditiously. We hypothesized remote supervision of echocardiograms by

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

telemedicine would decrease time to diagnosis and more effectively triage patients for transport. In a prospective cohort study, nine pediatric cardiology centers participating in the Multicenter Telemedicine Collaboration Group entered patients into a web site database. Inclusion criteria for the study are: 1) infants < 6 weeks old, 2) referring diagnosis cyanosis or murmur, 3) echocardiograms performed. The experimental and control groups were diagnosed using telemedicine or traditional means (infant transport, cardiologist travel, or videotapes sent) respectively. To date, 363 telemedicine and 304 control patients have been entered into the database. As expected, mild heart disease (PDA, PFO, small VSD) predominated in the infants transported from level 2 or 3 nurseries to cardiac centers. For patients with mild disease, one in the telemedicine group and 33/46 in the control group were transported. Significant heart disease (left or right heart obstruction, D-TGA, truncus arteriosus, TAPVR or other) occurred in 35/363 (9.5%) of telemedicine patients. Median time to diagnosis of serious disease was significantly less for the telemedicine group (140min) compared to control (378min) ($p < .005$). In the telemedicine group with serious disease 3/35 (8.6%) died and 2/35 (5.7%) sustained cardiac arrest, compared to controls where 5/50 (10%) died and 3/50 (6%) arrested (p=NS). We conclude that telemedicine technology significantly decreases time to diagnosis in infants with serious congenital heart disease. Telemedicine also significantly decreases the need for transport of infants with suspected congenital heart disease when serious heart disease can be excluded.

P124

The mobile pediatric cardiology team of Thailand

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The team is organized by The Pediatric Cardiology Foundation of Thailand under the Patronage of H.R.H. Princess Galyani Vadana (The Elder Sister of H.M.King Bhumipol of Thailand). The expenses are supported by public donation. The Team is comprised of about 6–10 pediatric cardiologists, 4–6 pediatric cardiology fellows, 1–2 CVT Surgeons and 10–15 Foundation Officials. It has been organized and dispatched to up-country of destinations once or twice annually for the past 8 years. The purposes are to examine and plan to help children with heart diseases and their families. The Simple and uncomplicated defect such as PDA could be carried out at site by our surgeons with the cooperation of the local teams.

The Adult with Congenital Heart Disease, Pregnancy/Delivery for the Women with Congenital Heart Disease

P125

Anomalous origin of the left coronary artery (LCA) from pulmonary trunk with systemic collateral supply to LCA

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We present the case of 15 year old asymptomatic girl referred to our institution with the diagnosis of mitral valve prolaps and a suspicion of coronary artery fistula. Detailed diagnostics (echo + cardiac catheterization + aortography) revealed Bland-White-Garland syndrome (BWG) with coronary aneurysmatic right coronary artery (diammeter of 9mm). In addition, on surgery, multiple collateral vessels between right and left coronary artery were found crossing over pulmonary trunk and right ventricle. Because of fragile and calcified anterior wall of main pulmonary artery we had to elect Hamilton rather than takeuchi technique. The procedure was aggravated by continuous blood outflow from enlarged LCA ostium suggesting systemic collateral supply. Weaning from cardiopulmonary bypass and perioperative period was uneventful. We were able to find only two published reports on systemic collateral supply to LCA in patient with BWG.

P126

Outcome of pregnancy after the mechanical prosthetic valve replacement: Japanese nationwide survey

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Background & Methods. Uncertainty is still expressed concerning the risks associated with pregnancy in women with mechanical prosthetic valves as well as regarding the choice of anticoagulants. To clarify the current situation of this problem, questionnaires were sent to all major hospitals in Japan. Responses were collected from 498 hospitals. Results: Ninety-three pregnancies in 72 women with mechanical valve replacement were reported. Their ages ranged from 20 to 43 years (mean 29.8) at the time of pregnancy. Replaced valves were mitral (n=12), aortic (n=20), mitral+aortic (n=7), and other (n=3). The Saint Jude Medical valve was used in 79 lesions, Bjork-Shiley in 25, Starr-Edwards in 11, Carbomedics in 6, and others in 4. Thirty-one pregnancies were terminated, and the remaining 62 pregnancies were continued. Warfarin (2–6 mg/day) was being used in 55 of the 62 pregnancies at the time of awareness of pregnancy, and treatment was switched to heparin in 10 of the 55 patients in the first trimester. Vaginal delivery occurred in 29 infants. Seventeen infants were delivered by Cesarean section because of fetal indications. There were 4 stillbirths at 25 to 29 weeks gestation, 3 of the 4 fetuses had intracranial hemorrhage. Maternal complications were reported in 12 pregnancies. Two patients who refused anticoagulant treatment died due to prosthetic valve thrombosis. Bleeding requiring blood transfusion occurred in 5 patients, progression of heart failure occurred in 3, an upper limb paralysis occurred in 1, and sepsis occurred in 1. No fetal embryopathies related to warfarin administration were reported. Conclusions: Although life-threatening complications are not rare, and the rate of fetal loss increases in pregnancies in women with mechanical prosthetic valves, a high rate of successful outcomes may be expected with suitable anticoagulant management and education.

P127

Double-chambered right ventricle (dcrv) in the adult patient

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Double-chambered right ventricle (DCRV) is a frequently misdiagnosed cardiac anomaly in adults. Between 1977 and 2000, we studied 42 patients with DCRV (mean age 49.1 years, range 22 to 63). Seven patients had a referral diagnosis of VSD and 6 patients had a diagnosis of valvular PS. All patients were treated surgically and there was no perioperative mortality. Post-op TTE showed mild residual RVOTO in 1 patient and no obstruction in the remaining 41. Nine patients were followed from 1 to 270 months. Five patients had a follow-up duration greater than 12 months (mean 10.9 years). During this period, 1 patient had moderate residual RVOTO on TTE and was followed clinically. All other patients had no residual RVOTO. DCRV is a frequently misdiagnosed congenital cardiac anomaly in adults. Patients can be successfully treated surgically with significant relief of the RVOTO, improvement in their clinical condition, and excellent long-term results.

P128

Management of pregnancies in mothers with congenital heart disease

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Today congenital heart disease represent 30–50% of cardiac disease in pregnancy and it will getting more, because of advances in medical treatment and surgical management. The incidence of congenital heart disease in the normal population is 0.8%. Fetal echocardiography and heart catheterization are the most common screening methods. Pregnancies of women with congenital heart disease require a special management. The explanations about the physiologic adjustments during pregnancy, the genetic risk and the ante-, intra- and postpartum management including an endocarditis- or thromboticprophylaxis are part of this management. In this study the outcomes of 55 pregnancies/babies in 37 patients were studied. 34 patients had an acyanotic and 3 patients a cyanotic congenital heart disease. Exactly cardiologic examinations and a gradation according to their ability-index ante/intra/postpartum were undertaken. There was a deterioration of cardiac function (EF) in the 2nd trimester in 5 mothers (1 untreated Aortic stenosis), 1 edema (TOF-repair). Ability index decreased in 4pts permanent from II°-III°, while it was reversible in 5. 55 pregnancies resulted in 41 live-births (74%), 9 spontaneous abortions (17%) and 5 therapeutic abortions (9%). 25 deliveries (54%) were vaginal, 6 deliveries with forceps, and 10 were by secerian section. The birth weight of the 19 infants, 2 were preterm, born to acyanotic women was between 1970–4030g. The birth weight of 2 infants born to surgically corrected cyanotic women was average. There was a 0% incidence of congenital heart disease in the offspring. Our study shows that women with congenital heart disease should be followed closely throughout their pregnancy to be able to deliver in good cardiac state healthy babies.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P129

Psychosocial competence and intellectual skills in adolescents with congenital heart disease

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An increasing number of patients with congenital heart disease are reaching adolescence and adulthood due to corrective surgery. They are mainly in good hemodynamic state and good physical condition. We tried to assess also the intellectual skills and psychosocial state. Questionnaires like Meyer-Probst Encephalopathy-Q, Youth-Self-Report (YSR), Child behaviour checklist (CBCL) were distributed to the parents and patients and a Hamburg-Wechsler-Test (HAWIK) and Test of variables of attention (TOVAC) were performed. There was a control group (I, n=20) consisting of patients with innocent murmurs, or mild CHD without any necessity for treatment. In Group II (n=21) were pts after corrective surgery for cyanotic CHD and in group III (n=20) those after corrective surgery for cyanotic lesions. Groups were comparable for the socio-economic state of the family, and age and sex distribution (mean age 11.8 years). HAWIK: There was no difference in the overall intellectual skills and the verbal part. Significant differences could be detected in the mathematical part (I were better than II p<0.01), activity skills II and III both worse than I (p<0.01) as they were in the visual part (p< 0.006). No significance was found in the CBCL and TOVAC, although in all test group I was best and III better than II. To explain the higher results of group III compared to group II we suspect a more intensive training of the pts in group III as they are more often cared for for their cardiac problem too. Problems in social competence and logical thinking are also influenced by the family background. Overall we found our adolescents very well adapted to their situation. Further studies for the long-term problems – job, pregnancy – are to be carried out.

P130

Monitorization of placental and fetal perfusion during surgical management of aortic coarctation in a pregnant woman- case report

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Coarctation of aorta is an unusual cause of hypertension in pregnancy. It is poorly tolerated during pregnancy and may cause maternal and fetal mortality. There is limited data regarding the outcome of pregnancy in patients with aortic coarctation. A 19-year-old woman presented at 16th gestation week with the complaint of headache. At physical examination, heart rate was 80/min and blood pressure was 170/100 mmHg. Systolic ejection murmur was detected at left sternal border. Femoral pulses were absent. Left ventricular hypertrophy was present at ECG and echocardiographic examination was revealed severe aortic coarctation distal to left subclavian artery (65 mmHg gradient) and bicuspid aortic valve. No fetal pathologic finding was detected at antenatal fetal examination and fetal echocardiography. Resection of coarctation and graft interposition by using 18 no dacron tube graft was performed under general anesthesia. Topical hypothermia (33°C) was employed during cross clamp period. Throughout the operation, radial and femoral artery pressures, and fetal heart rate were monitored continuously. Splanchnic perfusion was measured by using gastric tonometry catheter. Pulsatility index (PI) of maternal uterin artery, PI of umbilical artery and PI of fetal median cerebral artery was measured to assess fetal and maternal perfusion. Transient decrease was detected in fetal heart rate from 138/min to 90/min during cross clamp and fetal hypothermia. Femoral artery pressure decreased to as low as 52 mmHg. The patient had uneventful postoperative course with normal fetal findings at control USG examinations. At term, the patient had a healty baby via vaginal delivery. In this case, feto-maternal circulation was not adversely affected during correction of aortic coarctation. We think that surgical management is appropriate for pregnant patients with severe coarctation of the aorta with acceptable maternal and fetal risk.

P131

Does hemodynamic worsening of congenital aortic stenosis during pregnancy correlate with clinical progression?

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Women with bicuspid aortic valve stenosis (AS) tolerate pregnancy although the hemodynamic gradient worsens as pregnancy progresses. To compare hemodynamic progression of AS with clinical course 15 women were studied before and during 17 pregnancies clinically and by serial Doppler Echocardiography. Subjects had mild to severe AS prior to pregnancy and most had aortic insufficiency. Aortic gradients increased or were unchanged during pregnancy. Peak gradient before pregnancy was 40.8 ± 19.5mmHg compared to 50 ± 21mmHg during pregnancy (p<0.005). The mean gradient increased from 25.3 ± 12.6mmHg to 35.7 ± 15.1mmHg (p<0.005). No clinical deterioration occurred. Apparent hemodynamic worsening of AS during pregnancy must be interpreted with clinical status. These data suggest that 'expected' increases in the aortic valve gradients result from increases in stroke volume and do not signify progression of disease or predict poor pregnancy outcome.

P132

Direct stenting of aortic coarctation in adults

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To report our results and short term follow up of direct-stenting of aortic coarctation in adults. Five patients, (3 male) mean age 48±15 y (range 26–68), with native coarctation (3), end to end anastomosis (1) and synthetic interposition graft + proximal aneurysm (1). Mean pre dilatation gradient 43±16 mm (range 25–65). Associated pathology: moderate left ventricular hypertrophy and dysfunction (3), severe 3-vessel coronary artery disease (2), severe aortic regurgitation + 2 restricted VSD's (1) and aberrant right subclavian artery (1). Under general anesthesia, J&J stents (4014, 5014) mounted on Owens balloon catheters, were advanced in retrograde approach through long 12F sheaths and dilated to 18–20 mm by hand inflation. Post procedure gradient was satisfactory in all patients, mean 8±7 mm (range 0–17) and angiographic result was optimal in 4 and suboptimal in one with very distorted and severe native coarctation. No complications were encountered. All patients were well over month following dilatation. The oldest patient, who also had IHD, died suddenly two months later, one with known IHD needed CABG surgery 5 months later, and one got pregnant one month post dilatation and delivered successfully at term, blood pressure remaining normal throughout pregnancy.

P133

Pregnancy and outcome in 235 women with congenital heart disease

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Introduction. We evaluated the incidence pre- and perinatal morbidities according to the type of congenital heart defects in 235 women with CHD. Patients and Methods: 235 pregnant women with CHD who admitted to the clinic of congenital heart disease between January 1990 and November 2000 were included in this study. The patients include following CHD: ASDII n=82, PAPVD n=10, VSD n=16, mild AV-insufficiency n=16, moderate-severe AV-insufficiency n=5, SVD n=10, ASD I n=5, PS n=10, AS n=11, coarctation n=2, , Marfan Syndrome n=4, Ebstein anomaly of TKV n=4, others n=16, CAVSD n=7, TOF n=10, DORV n=3, L-TGA n=4, d-TGA with Senning-OP n=4, CUCM n=5, Blalock-White-Garland n=3, TAC n=1, PA-VSD n=4, DILV n=1, d-TGA-VSD n=1. The 294 pregnancies in these patients resulted 245 (83%) immature live born infants, 28 (9.4%) spontaneous abortion, 10 (3.4%) premature birth, 4 (1%) fetal death at term. CHD were detected only in 6 (1.5%) of the offspring. Conclusion. Pregnancy and spontaneous birth in women with CHD is possible and depend mainly on the maternal oxygenation and hemodynamic status. The highest incidence of maternal and fetal morbidity was found in the those women with cyanotic uncorrected CHD. Interdisciplinary Cooperation before, during and after pregnancy between the pediatric cardiologist and perinatologist is necessary to achieve a favorite outcome at fetus and mother.

P134

Abnormal lung perfusion after surgery for Fallot's tetralogy does not impair cardio-pulmonary function

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To examine the effect of lung perfusion abnormalities on cardiopulmonary function in the long term operated Fallot's tetralogy patients. Fifty patients (17 female) age range 17–55 years (mean 28.9±11.2) after surgical repair of Fallot's tetralogy were examined. All had a chest radiogram, pulmonary

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

perfusion scintigraphy, spirometry with diffusion capacity, echo-Doppler cardiography and cardio-pulmonary exercise test on a bicycle ergometer. Pulmonary perfusion was considered abnormal if right lung perfusion was less than 40% or more than 57% of the total. Pulmonary perfusion was normal in 32 patients and abnormal in 38. There was no difference between groups in any of the cardio-pulmonary exercise parameters as well as clinical parameters except for the mean age at operation: 6.3 vs 12 years in the abnormal and normal perfusion groups respectively. Exercise performance in both groups was inversely related to cardio-thoracic ratio, right ventricular pressure, and the gradient across the right ventricular outflow tract. Abnormal lung perfusion, usually the result of branch pulmonary artery stenosis, does not affect exercise performance long term after surgical repair of Fallot's tetralogy.

P135

Transcatheter closure of patent ductus arteriosus using glanzucco coils in adolescents and adults

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We present the short and intermediate term results of transcatheter closure of patent ductus arteriosus with Gianzucco coils in adolescents and adults. During a 4.4 years period, 46 patients (36 females and 10 males) with ages ranging from 14 to 52 years (median 23) underwent attempted transcatheter closure of patent ductus arteriosus with the Gianzucco coils. The diameter of narrowest segment of the ductus ranged from 0.8 to 7.6 mm (3.2±1.3 mm). Group I consisted of 7 patients with a ductal diameter <2 mm, group II consisted of 22 patients with a ductal diameter ≥2 mm but <4 mm, and group III consisted of 17 patients with a ductus ≥4 mm. Four to five loops Gianzucco coils were used, which were deployed via retrograde arterial route. Multiple coil technique was generally applied in group II patients. Balloon occlusion technique in combination with multiple coil technique was generally used in group III patients. Seven patients had pulmonary hypertension (mean >20 mm Hg). Deployment of coil was successful in 43 patients (93%) but fail in 3. The success rate of coil deployment in group I, II and III were 100%(7/7), 95% (21/22) and 88% (15/17), respectively. A mean of 2±0.7 coils was deployed in per patient. The 3 patients with unsuccessful coil deployment of whom 2 had a large ductus (diameter >7 mm) and 1 had associated aortic stenosis, underwent ligation of the ductus. Distal embolization of 14 coils occurred in 7 patients (1 in group II and 6 in group III), from whom 13 coils were retrieved with a gooseneck snare and 1 coil was removed during surgery. The mean diameter of ducts in the 7 patients with distal embolization was significantly larger than that in those without (5.6±1.5 vs 3.7±1.1 mm, $p<0.01$). Among the 43 patients with successful coil deployment, immediate complete closure was achieved in 17 (40%), while trivial-to-mild leak was present in 25 (60%). No significant complications were encountered. After a follow-up period ranging from 3 to 42 months, 3 patients had a small residual shunt: 6 months following the initial procedure and all 3 underwent a second intervention with complete occlusion. None had left pulmonary artery stenosis documented with Doppler echocardiography. Transcatheter closure of patent ductus arteriosus with the Gianzucco coils is safe and feasible in adolescents and adults.

P136

Transcatheter closure of residual post surgical asd defect

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We report 5 patients with previous surgical closure of ASDs, who underwent successful transcatheter occlusion of residual defects. Between November 1997 and June 2000, 315 patients underwent transcatheter occlusion of secundum type ASDs. Among these, 5 female patients (age range 28 – 55 years, mean 40.6±9.2) were found to have a residual shunt at atrial level after previous surgical closure of their ASD at the mean age of 12.3±8.5 years. Four of them had direct suture and the 5th patient patch closure of the ASD. The exact diameter of the ASD was evaluated by balloon sizing under TEE monitoring. The mean Qp/Qs ratio was 2.3±0.96. Four out of 5 patients were judged suitable for transcatheter closure. In the last patient the residual defect was very large with an absent postero-inferior rim. The mean ASD size was 11±3.9 mm on TTE, 12.6±4.9 mm on TEE and 14.6±3.5 mm at balloon sizing. The mean Qp/Qs ratio was 2.3±0.98. CardioSeal-Starflex devices were implanted in 3 patients and Amplatzer ASD occlusion device in 1. Complete occlusion was achieved in all patients. In one patient the device (Starflex 28 mm) embolized in the right atrium before deployment and was successfully retrieved. A 33 mm device was then implanted without complications. No patients experienced late complications. In our opinion a residual post-operative ASD should be approached as native ASDs following the same criteria for patient selection.

P137

Conversion to an extracardiac conduit with a limited right atrial maze procedure for the failing fontan with atrial tachycardias.

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Background: Atrial arrhythmias are a frequent late complication of Fontan procedures. Reduction of right atrial pressure and wall tension by conversion to an extracardiac conduit combined with reducing right atrial size should improve hemodynamics and reduce the development of tachyarrhythmias. However, atrial tachyarrhythmias may still persist and more effective control may be achieved by interrupting atrial arrhythmia circuits and inserting pacemakers at the time of the Fontan revision. Methods: Since 1997, we have performed this operation in 5 patients aged 14 to 25 years (mean 20.6) at an average of 13.5 (7 – 14) years after their original Fontan procedure. All of the patients had medically uncontrollable atrial tachyarrhythmias and grossly dilated right atria with markedly reduced exercise tolerance. One had a large right atrial thrombus. An EP study was performed on all patients preoperatively. Along with extracardiac conduit insertion, each patient underwent a limited RA maze procedure using a combination of cryotherapy and incisions in addition to RA reduction. All patients had epicardial pacemakers. Results: All the patients survived with an average hospital stay of 15 days (7 – 38). Exercise tolerance has improved in all 5 and atrial tachyarrhythmias have either decreased (2) or disappeared (3). Only one patient is on antiarrhythmic medication other than digoxin. Follow-up is a mean of 20.4 months (6 – 35). Conclusion: Right atrial maze, size reduction and pacemaker implantations are worthwhile additions to simple conversion to an extracardiac conduit in the failing Fontan. Without compromising safety one gives a better chance of long-term relief from debilitating and persistent atrial tachycardias.

P138

Diameter of the thoracic aorta throughout life

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Introduction: Helical CT is increasingly used for evaluation of the thoracic aorta in adults with congenital heart disease. Proper interpretation can only be achieved on the basis of age-related normal values, which are not available yet. Methods: In 70 adults (24 female, 50.2 ± 16.5 years, 73.1 ± 15.7 kg, 172.4 ± 8.1 cm) we analysed the aortic diameters with helical CT (Somatom Plus, Siemens) at 7 intrathoracic levels. All was indicated for various reasons, none of the patients had cardiac or vascular disease. Multiple regression analysis was performed to evaluate influence of weight, height, body surface area, sex and age. Results: Regression analysis revealed no influence of body size. Males had significant larger diameters on levels 2, 5 and 7. Influence of age was highly significant at all investigated levels. Age-related means can be calculated from the regression line parameters shown in the table. Age-related mean [cm] = increase x age [years] + intercept. Conclusions: Our data present normal diameters for the thoracic aorta in spiral CT in relation to age. Increase in diameter during adulthood must be considered to assess dilatation, stenosis or hypoplasia, especially in patients with coarctation or connective tissue disease.

P139

Management of the grown – up congenital heart (guch) patients in moravia

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Due to development of diagnostic and therapeutic methods in children's cardiology, generation of the Czech system of complex care of children with a heart defect and its interconnection with primary sphere, led to development of completely new, minority group of people of productive age in our population with diagnosed or corrected congenital heart defect in childhood. Quality of life of majority of these children was strongly changed. However, complex and systematic monitoring followed during their childhood, in consequence of natural development of the heart defect with or without correction, such patients require lifetime monitoring because of different degree of threat of development of various later complications, including sudden death, depending on the individual morphology of the defect and results of survived intervention. Authors present introduction of an identical, complex, multidisciplinary, systematic, and team care of 350 adult patients with this location in the region of

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Moravia. They compare of critical heart defect in newborns, re before and after establishment of cardiocentres in our republic.

P140

Health related quality of life (QOL) on adults with congenital heart disease (CHD)

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As a result of continuing improvements in cardiac surgery and perioperative care, most children with CHD now have the potential to survive to normal adulthood. Since 1995 adult patients with CHD visited our interdisciplinary outpatient department combining both adult and pediatric cardiologists. Methods: 173 patients (mean 29.4 years) were divided into 4 groups: Complex heart disease (31%), septal defect (32%), outflow obstruction of the right or left ventricle (27%) and coarctation of the aorta (11%). These groups were compared to each other and to an age-related collective of 102 healthy persons. QoL was measured with the validated questionnaire SF-36 Health Survey. The SF 36 is a morbidity measure that features a profile of 8 dimensions, including clinical, social, emotional and professional status. Results: The best QoL was determined in coarctation. These patients had similar QoL scores as healthy adults ($p>0.05$). Patients with septal defects and congenital outflow obstruction presented lower QoL scores on the dimensions emotional role and mental health ($p<0.05$). The worst QoL was found in complex congenital heart disease, especially in patients with a tetralogy of fallot. The dimensions general health perception, social functioning, physical limitation and vitality were highly restricted on patients with complex heart disease ($p<0.05$). The dimensions physical role and physical pain were without restrictions on adult patients with congenital heart defects. Conclusion: The QoL showed a considerably variation in adults with CHD. Except: patients with coarctation, all adults with CHD had a significant low of QoL in one dimension at least. These patients could benefit from a special care, managed in cooperation of the special facilities of adult and pediatric cardiologist, supported by social workers and clinical psychologists.

P141

Inquiry about adults with congenital heart disease in a children's hospital in

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Background. Our hospital was founded in 1970 just for children, so it's a problem how to deal with patients overgrowing child age. There are many problems as no physician for adults, no special clinic and no ward for them. Therefore, an inquiry was made to know what these patients think about their physical and social condition and what they hope for our hospital. Results: Now we have 183 adult outpatients (20–31 years old) with congenital heart disease who have had an operation. We contacted them directly at clinic or by letter and 42 patients replied. Half of them are cyanotic heart diseases. Twenty-one patients (50%) have steady jobs. Twenty four (57%) think they are in good condition, 13(29%) feel tired with hard works and 4(10%) feel tired only with light works. Some of them told they felt difficulty in physical training. Eighteen (43%) do not worry about their diseases, but 22(52%) do sometimes. Thirty-nine (93%) come to children's hospital without reticence, but some feel shy among younger patients. Moreover 34(93%) hope to keep coming to the children's hospital and 38 do not want to move to hospitals for adults. However, when they need consultation about their diseases, 10 of 12 patients talk to their family and only 2 talk to medical staffs. Conclusions: Most adult patients live in good condition except when they do hard works. They hope to keep coming to children's hospital because they think the hospital staffs they have known from their childhood know well about their physical conditions. However we have no system for adults, for example, specialists, wards and consultants for adults. From this inquiry the problems we have and what we should do for.

P142

Pacemaker experience in adults with congenital heart disease

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To describe the character sticks of patients (pts) with congenital heart disease (CHD) and implanted pacemakers (PM), a retrospective cohort study including adults (age >16 yrs) with CHD seen in 1998 and in 1999 was performed. Out of 316 pts with CHD, 39 pts (11% mean age 32.7±14.5 yrs, 22 males) had a PM. Principal diagnoses were: atrial switch procedure due to complete transposition of the great arteries (d-TGA, 13), tetralogy of Fallot (TOF 5), congenitally corrected TGA (l-TGA, 4), aortic valve anomaly (2), Ebstein anomaly (2), Eisenmenger syndrome secondary to patent ductus arteriosus (2), AV-septal defect, double outlet right ventricle, complex pulmonary atresia, tricuspic atresia. Shone complex and ventricular septal defect in one each, others (5 pts). Mean age at the time of first PM implantation was 23.7±16.0 yrs (median 20.6 yrs), mean pacing time 7.4±6.6 yrs (median 5.1 yrs). In 10/39 pts, PM implantation was done within 20 days after cardiac surgery. 18/39 pts required at least one subsequent PM related procedures. Expected end-of-life of the battery (1 pt), lead dysfunction/displacement (10x) were most frequent. Transvenous leads were present in 30 at the time of last follow up. Primary indications were: Sick sinus syndrome (SSS) in 9 pts with d-TGA (5 x DDD) and in 2 others (DDD in both), AV-block in 4 pts with d-TGA (VVI in 4), in 4 pts with l-TGA (DDD in 2), in 5 with TOF (3 DDD), in 15 others (13 DDD). Two infected PMs were removed. Adults with d-TGA, l-TGA and TOF are the most common population with a PM. Main indications for PM-implantation were SSS in pts with d-TGA and high degree AV block in pts with l-TGA and TOF. Many pts require subsequent PM related procedures.

P143

Quality of life is good after cardiac surgery in childhood. Investigation in 1949 adult patients 9 to 46 years after operation

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In order to find out whether the correction of congenital heart defect in childhood results in good quality of life we studied all the 4246 patients undergoing pediatric cardiac surgery between 1962 and 1989 at the Hospital for Children and Adolescents, University of Helsinki. Ninety-seven percent of the patients could be traced. A questionnaire was sent to 2517 adult patients. The response rate was 77%. The mean follow-up time was 20 (9 – 46) years and the mean age at follow-up 28 (18 – 59) years. The defects are presented in table 1. The patients were satisfied with their life: 75% described their condition to be good, 23% moderate and 3% poor. 96% of the patients were estimated to be in NYHA classes I or II, 3% in NYHA III and only five (0.2%) patients in NYHA IV. The estimation was based on patients' own classification (normal, poorer than normal, poor) and description of exercise capacity. Most patients (71%) were occupied. Wide range of occupations were presented, from professor to ordinary workman. Altogether 75% of patients had more than comprehensive education and 12% were still studying. 10% of patients had an university degree. Mental retardation was the main cause of disability to work among the disabled patients (7%). Only 6% of patients were unemployed while within the general population the unemployment rate was 11%. Women were married more often than men, 63% vs 53%, and had more often children, 59% vs 40%. The use of a family did not differ by gender, but the patients had bigger families than the general population. We conclude that the vast majority of patients after cardiac surgery in childhood are satisfied with their health and live normal life.

P144

Closure of congenital VSD in adult patients

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The classic indications for ventricular septal defect (VSD) closure have been congestive heart failure, pulmonary hypertension, aortic valve regurgitation, and prior endocarditis. Less clear remains the indication for congenital VSD in adults when the VSD is small, the right ventricle pressure is normal, the Qp/Qs is less than 2:0 and there is no aortic valve involvement. This review examines our results of VSD closure in pts older than 16 yrs with particular attention to the indications. From January '85 to August 2000, 31 pts underwent closure of VSD in our Institution. The mean age was 32 yrs (range 16–70 yrs). All patients underwent pre-op echocardiographic evaluation and 23 cardiac catheterization. Five pts were post-surgical residual VSD. Associated cardiac malformations (ACM) were: aortic insufficiency in 10, tricuspid insufficiency in 4, subaortic stenosis in 1. 29 pts underwent surgical closure of VSD and 2 transcatheter device closure. Associated surgical procedures were: tricuspid valve plasty in 2, aortic valve plasty in 2 aortic repacement in 3, subaortic resection in 1. There were 2 (2/31 = 6.4%) early deaths in 70 and 58 yrs old pts operatively for endocarditis. In conclusion we can affirm that there is no controversy over the indication of closure of a VSD hemodynamically important or with ACM. Risks of development of bacterial endocarditis and / or aortic

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

regurgitation, new possible transatrium approach and low surgical risks can justify the closure of restrictive VSD in pts with a Qp/Qs of 1.5 or less.

P145

Reoperations in adolescents and adults with congenital heart disease: early results

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Objective: Only a minor proportion of patients with congenital heart disease (CHD) are definitely cured following primary repair during infancy. A significant number will require reoperations during adulthood because of long-term problems. Methods: To assess perioperative mortality and morbidity we retrospectively analysed 56 grown-up patients with CHD, needing reoperation between January 1980 and September 2000. At the last operation mean age was 28 ± 12 (range 14 – 64) years, mean BSA 1.63 ± 0.34 (0.94–2.1) m2 and preoperative NYHF 56 ± 21 (20–92) %. Dyspnea (27 patients), angina pectoris (11), arrhythmias (4) and recurrent cyanosis (5) were the most frequent preoperative symptoms. Primary congenital cardiac pathology was ASD (5), VSD (7), TOF (9), complex d-TGA (4), LVOT pathology (15), aortic coarctation (8), Marfan syndrome (6), single ventricle (1), truncus arteriosus (1), with congenital syndromes (4) and situs inversus (2). Indication for reoperation was valve dysfunction (n=13), recurrent stenosis (n=12) or shunting (n=12), heart failure (n=5), conduit-related problems (n=4) or others (n=21). Results: Reoperation included correction of late complications (n=26) or residual defects (n=20), conversion after palliation (n=8), cardiac transplantation (n=3) or other not to the primary cardiac defect related procedures (n=8). Early mortality was 9.5% (5 patients with d-TGA, pulmonary atresia or Marfan). Serious preoperative complications occurred in 20 patients (36%): low cardiac output (5), respiratory failure (7), neurologic dysfunction (3), renal failure (2), complete av-block (6), arrhythmias (10), MOF (3) and re-exploration (1). Conclusions: Reoperations in GUCH patients are challenging with an important perioperative risk due to the complexity of the underlying cardiac pathology, the diversity of long-term problems and the surgical difficulties frequently encountered. Correction of late complications and residual defects remain the most frequent types of reoperation.

P146

Long-term outcome in adults with severe congenital heart disease: health-related quality of life and medical status

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Objectives: Aims of this study were to evaluate health-related quality of life (HRQoL) and medical status and to determine the relation between these parameters in adults with non-anatomically-corrected congenital heart disease. Background: Nowadays many patients with severe congenital heart disease survive beyond childhood. Long-term medical complications are to be expected. Outcome of HRQoL and its relation to medical status is unknown, but required for optimal counseling of the individual. HRQoL is defined as the patient's own appraisal of health status, which is specified in 3 scales: physical, social and psychological functioning (World Health Organization), that all consist of different dimensions. Methods: Eighty-one patients with non-anatomically corrected congenital heart disease (aged 18–32 years) were randomly selected from our database. A validated questionnaire (TNO-AZL Adult Quality of Life; TAAQOL) was used to measure HRQoL on the 3 scales mentioned above, reference data are available. Medical status was determined with the NYHA-class and the Somerville-index. Results: HRQoL in severe congenital heart disease patients was significantly worse compared to that of the general population on the dimensions Gross Motor Functioning ($p<0.01$) and Vitality ($p< 0.01$). Mean scores for the medical indices were: Somerville-index 1.41 and NYHA-class 1.83. Correlations of the HRQoL-dimensions with the NYHA-class and Somerville-index were only significant and relevant for the dimension Gross Motor Functioning. Conclusions: Patients with non-anatomically-corrected congenital heart disease experience limitations only on the physical scale of HRQoL (not on the social or psychological scale). This stresses the need for attention to physical aspects. Routinely used indices such as the NYHA-class and Somerville-index do not sufficiently predict HRQoL. Therefore, dedicated questionnaires for HRQoL should complete the medical supervision as in a study.

P147

Medical follow-up, health-related quality of life and social limitations in adults with a minor congenital heart disease

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Objectives: To study efficiency of medical follow-up, social limitations and health-related quality of life (HRQoL) in adults with a minor congenital heart disease. Methods: Patients (aged 18–32) were randomly selected from the Archives of Pediatric Cardiology. Most recent information was traced for at hospitals, general practitioners or local authorities. All patients had a minor defect in childhood, not needing an operation (for example, restricted septal defect=VSD-, pulmonary or aortic stenosis). Participants, who had been physically examined longer than one year ago, were re-examined. All received a questionnaire on social limitations and HRQoL (defined as the person's own appraisal of health status, specified as physical, social and psychological functioning). Results: Eighty-two patients agreed to participate (response 91%). Sixty-five participants (59%) had been discharged before. 19 (23%) had neglected medical supervision and 18 (22%) were still under medical care. Update medical examination changed the diagnosis in 13 participants (16%) and had consequences for antibiotic prophylaxis in 9 (11%; 5 could stop & had to restart prophylaxis). Four of these participants had neglected medical supervision. In 6, a VSD had closed (of 26 formerly VSD-patients). Nine participants (11%) had experienced rejection or impediments for sports, education, medical examination for work and sperm, mortgage, health insurance or life insurance. Outcome of HRQoL in mild patients was not significantly different from the general population. Conclusions: HRQoL in mild congenital heart disease patients is good. However, a substantial number of them had experienced social limitations. Furthermore, a large percentage of patients neglected their routine medical check-up and update medical examination changed their diagnosis and antibiotic regime. Based on these results, we suggest that doctors should impose upon these patients to have a routine medical examination at least every 5 years.

P148

Correlates of educational, occupational, and psychosocial outcome in adolescents and adults with congenital heart disease.

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While advances in the treatment of congenital heart disease (CHD) have dramatically increased survival rates, little is known about the factors that mediate good versus poor quality of life outcomes in adolescents and adults with CHD. The goals of this correlational study were to: 1) identify factors related to good versus poor academic or occupational achievement in adolescents and adults with CHD and 2) identify factors underlying good versus poor psychosocial adjustment in adolescents and adults with CHD. Approximately 200 patients were recruited from the Toronto Congenital Cardiac Centre for Adults and the Hospital for Sick Children over a period of 2 years. In order to maximize the homogeneity of the sample with regard to the physiological and psychological impact of disease, subject selection was limited to patients with either Tetralogy of Fallot or Transposition of the Great Arteries. Outcome variables included educational achievement, occupational status, social relations, self-esteem, anxiety, and depression. Predictor variables included: attributional style; beliefs about personal cardiac health and lifestyle restrictions; knowledge of cardiac condition; expectations for academic/occupational success; and achievement motivation. Control variables included physical functional capacity; IQ; socio-economic status; and actual disease severity. Results confirmed hypotheses that a more pessimistic attributional style, poor knowledge of one's heart disease, perceptions of severity and its imposed restrictions, and lower expectations for success were related to poorer quality of life outcomes independent of actual disease severity and actual restrictions. On average, this population demonstrated average range intellectual ability, thus this was not a limiting factor to academic and occupational achievement. These findings have important implications for interventions with this population in that their quality of life appears to be significantly affected by factors that are very amenable to modification.

P149

Early repair of tetralogy of Fallot without outflow patch revealed favorable influence on late arrhythmias and sudden death >27 years follow-up study in uniform surgical approach.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

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(Background) Although long-term survival of patients with correction of Fallot (TOF) has been reported to be excellent, late sudden death remains a serious complication after TOF repair. To identify risk factors of late death, it is important to study the patients group repaired with uniform approach. (Patients and Methods) Survival pattern of 167 patients, who survived 30 days after complete repair of TOF in 1966–75, was studied prospectively examining hospital records (50), interviews (63), and death certificates (54). All patients were repaired at mean age of 6.4 years old (median 4.6) without outflow patch by a single surgeon. Among 167 patients, the status of 99 patients was identified from hospital records and interviews. Current hemodynamics of 50 in 167 patients was assessed using ECG and/or Echocardiography. (Results) The overall 29-year actuarial survival rate was 86%, which was same as the previous reports (NS). Incidences of late sudden death (4%), sustained ventricular tachycardia (2%), and atrial tachyarrhythmia (2%) were as low as those of the previous reports (NS). QRS duration (148msec) and incidence of moderate/severe pulmonary regurgitation (36%) were similar as those of the arrhythmia-free group reported by Gatzoulis MA et al (Lancet. 356:975–981) (NS). Incidence of moderate/severe tricuspid regurgitation (0%) was lower than that of the report ($p=0.02$). Mean left ventricular ejection fraction was 59.2%. Mean aortic root dimension was 26mm. (Conclusion) Although long-term survival pattern and functional status of this group are similar as those of previous reports, amount of pulmonary regurgitation and right ventricular dilatation were less than those of the previous reports. Furthermore, left ventricular systolic function and aortic root dimension were well preserved. Subsequently mortality and morbidity of the group will be good.

P150

Predictive Value of Thromboelastography In Adult Cyanotic Cardiac Surgery

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Thromboelastography (TEG) is a method of monitoring blood viscosity that can be used to evaluate various components of the coagulation system. The efficacy of using TEG analysis in CABG surgery is proven in many studies. However, there have been no published reports of use of TEG in adult cyanotic patients undergoing CPB. This study was aimed to determine the predictive value of TEG in such patients. Twenty-eight adult cyanotic patients undergoing CPB at our hospital were evaluated using TEG as a sole guide for transfusion of blood products in the perioperative period. The first TEG was performed 20 minutes after initiation of CPB and the blood products were ordered based on the result obtained. The blood products were transfused following protamine administration in the operation theatre. The second TEG test was done in ICU after shifting the patient to assess the coagulation and determine any further need for transfusion. The prediction was deemed accurate if the following criteria are met: 1. Chest drainage of < 4ml/kg in first 24 hours. 2. No reexploration because of residual cause. 3. Return of normal TEG after transfusion of blood products. Twenty-six out of twenty-eight patients had accurate prediction based on the criteria mentioned. Two patients bled 12.5ml/kg and 15 ml/kg respectively in first 24 hours after shifting to ICU. We conclude that TEG is an accurate predictor of the requirements of blood products in adult cyanotic patients undergoing CPB for intracardiac repair."

P151

Reduced baroreflex sensitivity in adults with repaired tetralogy of Fallot

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Background: Sustained ventricular tachycardia and sudden cardiac death remain the most devastating late complications after surgical repair of Tetralogy of Fallot. Whilst reduced baroreflex sensitivity (BRS) is depressed and a strong predictor of sudden cardiac death in patients after myocardial infarction and in chronic heart failure, little is known about BRS in patients after Tetralogy of Fallot repair. Methods. We measured BRS and HRV in 19 Fallot patients (15 male, age 17±3y, 25±2y after repair, mean±SE) and 19 age-matched normal controls (15 male, 37±3y). Subjects underwent 20min of resting measurements of heart rate (by ECG) and non-invasive beat-to-beat blood pressure (using a Finapres). A 5min period of 0.1Hz controlled breathing was also recorded. BRS was evaluated: i) by calculating the α index as the square root of the ratio between R-R interval and systolic blood pressure (SBP) spectral powers in the low frequency (0.04–0.15Hz, α-LF) and high frequency (0.15–0.4Hz, α-HF) bands, ii) as the ratio of the average amplitude of oscillation in R-R interval and the average amplitude of oscillations in SBP during controlled breathing (BRS.Cbr) and iii) the sequence method (linear regression slope of R-R interval vs SBP; BRS.Seq). Results. The results are presented in the following table: (See attached file) Conclusions: Tetralogy of Fallot patients, late after repair, have significantly reduced BRS when compared to normal age-matched controls. There is, therefore, unbalanced autonomic control in these patients, which may contribute to the mechanism of sudden cardiac death.

P152

Right ventricular myocardial mass after the Mustard operation: severe hypertrophy reflects impaired adaptation to systemic pressure load

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Background. The Mustard operation for transposition of the great arteries leaves the right ventricle (RV) supplying the systemic circuit. We have previously shown impaired RV myocardial perfusion reserve in these patients, associated with wall motion abnormalities. We postulated that the RV is a less efficient systemic pressure pump than the left ventricle (LV); a greater degree of hypertrophy would therefore be required when the systemic ventricle is of right ventricular morphology, placing extra demands on coronary arterial supply. Methods. MRI was performed using a 1.5T Picker Edge Cardiac scanner on 15 Mustard patients (median age 27 years) and on 16 normal controls (median 30 years). A series of 10mm transverse cuts was taken through the ventricles from the base of the heart to the apex. Mass was calculated from myocardial volume, assuming density to be 1.05g/cm3. We compared systemic RV mass in the Mustard group with the systemic LV in the control group, and also compared total myocardial mass in the two groups. Results. RV mass index was significantly greater and LV mass index significantly less in the Mustard group: RV 99±25g/m2 vs 56±6 ($p<0.0001$); LV 60±9 vs 78±15 ($p=0.0001$). The mass index of the systemic RV (99±26) was significantly greater than the systemic LV (78±13) ($p=0.004$). Total mass indexed mass index is greatly increased in the Mustard group (159±28 vs 115±17, $p<0.0001$). Conclusion: This study supports the hypothesis that the RV myocardium is less well adapted to functioning at systemic pressures, necessitating a greater hypertrophic response. This degree of hypertrophy is likely to affect myocardial function and will also increase total myocardial oxygen consumption and demands on coronary arterial supply. ACE-inhibitors and/or β-blockers may be beneficial for these patients.

P153

Exercise capacity in adults late after correction of Fallot repair. Relationship with biventricular mass volume and function

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Background. Exercise capacity has been shown to be affected by pulmonary regurgitation in paediatric patients after TOF repair. We investigated the relation of cardiopulmonary exercise stress test parameters with biventricular volume, mass and function as assessed by MRI. Methods. We used a Picker Edge 1.5 Tesla MRI scanner to measure biventricular volume, mass and function (values indexed to BSA). Patients were subjected to a modified Bruce exercise stress protocol, with concurrent measurement of oxygen consumption. Correlations between MRI indices, age, time from operation and peak oxygen consumption and other exercise parameters (heart rate, blood pressure, exercise duration) were studied. Spearman's correlation was used for statistical analysis. Results. 20 patients were included. Patient characteristics were: age 32 ± 12, years from operation 24 ± 8, pulmonary regurgitant fraction (PRF) 27 ± 15%, RVEF 51 ± 10%, LVEF 66 ± 9%, RVEDVi (end diastolic volume index) 112.9 ± 36 ml/m2, RVESVi (end systolic volume index) 58 ± 30 ml/m2, RVMi (mass index) 52 ± 17 g/m2, peakVO2 31 ± 8 ml/kg/min which corresponded to 66 ± 17% of the predicted peakVO2 and exercise duration 12.8 ± 2.8 minutes. PRF and RVESVi correlated negatively with the percentage of peakVO2 achieved versus peakVO2 predicted ($r=-0.58$ $p=0.028$ and $r=-0.47$ $p=0.05$ respectively). Of note is that only one patient was in NYHA III class, the others being in class I. Conclusions. Pulmonary regurgitation late after TOF repair has a deleterious impact on exercise capacity as assessed by

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

cardiopulmonary exercise stress testing. This seems to be due to increasing RVESVI as a consequence of pulmonary regurgitation.

P154

Determinants of atrial tachyarrhythmias in adults with the Fontan operation: the influence of right atrial dilatation

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Introduction. Atrial tachyarrhythmias are a common complication after the Fontan operation. The causes are thought to be a combination of atrial scarring and progressive right atrial (RA) dilatation. We studied a group of adult Fontan patients in order to assess the influence of RA size on development of arrhythmias. Methods. We reviewed patients with atrio-pulmonary Fontans who underwent MRI at our centre between 1993 and 2000. Patients were grouped into those who had never had sustained atrial tachyarrhythmias (group 1), those with paroxysmal flutter / fibrillation (group 2) and those with permanent flutter / fibrillation (group 3). Maximum RA dimensions were measured from MRI in para-sagittal transverse and coronal planes. A RA volumetric score was calculated by multiplying these three dimensions (values given as mm3 x 103). Other MRI indices measured were: Fontan pathway obstruction, pulmonary artery or vein stenosis, ventricular function and AV valve regurgitation. The MRI reporter was handed no clinical data. Results: 29 studies were performed in 22 patients (10 female), median age 25 years at time of MRI. Median time since Fontan was 12 years. The RA volumetric score was least in group 1 (n=6), 359±94, significantly greater in group 2 (n=21) 529±251 (p=0.066) and greatest in group 3 (n=2) 815±166 (p=0.07); see figure. No other MRI indices correlated significantly with arrhythmic status. Time since Fontan was also not related to arrhythmic status. Conclusions. This study demonstrates that right atrial size is the most important determinant of atrial tachyarrhythmias in adult Fontan patients. This data provides support for an early strategy of converting atrio-pulmonary Fontan circulations to cavo-pulmonary connections.

P155

Improved results of the Fontan operation in the adult patient

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Reported results for adults undergoing the Fontan operation have been discouraging (5% mortality & 30% major complications). We reviewed the records of all adult pts > 16 yr who underwent the Fontan operation from 6/1995 to 9/2000 to determine perioperative course, mortality & modifications leading to improved outcome. Results: 11 pts were identified (6 first time & 5 redo). The indications for the first time Fontan included cyanosis/decreased activity (n=4) & arrhythmic events (n=2). Mortality both early & late was 0%. Mean age was 22.1 years (range 18–40). Mean Pulmonary artery pressure 14.9 mmHg (S.D 5.7). Pulmonary vascular resistance 1.9 mmHg (S.D 1.1) & ventricular end diastolic 10.5 mmHg (S.D 4.5). Atrioventricular valve regurgitation was mild in 5 & moderate in 2 patients. Ventricular function was good in 7, fair in 3 & poor in 1 pt by intra-operative trans-esophageal echocardiography. A non-fenestrated Fontan was completed in all patients [Lateral tunnel (LT) in 5, Pericardial (EC) in 1, Redo LT in 3 & Redo EC in 2]. Secondary procedures: Device annuloplasty (n=1), Cosgrove Ring (n=1), Atrial debulking (n=4) & pacemaker implant (n=5). Mean hospital length of stay (LOS) was 8 d (range 5–16d) & ICU LOS 2d (range 1–5). Complications included: new onset arrhythmia (n=3), transient Bradycardia (n=1) & persistent chest tube drainage (>4days) (n=1). Conclusion. Adult age does not preclude successful Fontan outcome. Fenestration of the atrial baffle may not be necessary in this population.

P156

Cardiac transplantation in adult patients with end-stage congenital heart defects: High risk procedure or therapeutical option?

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Introduction: Problems and results of pediatric cardiac transplantation (Htx) are well-known from the literature. Different palliative operations are performed to avoid Htx. A diagnosis which is not CAD or myopathy is a significant risk factor for 1-year mortality. To find out, whether Htx is a considerable therapeutical option for grown-up pts. with pretreated congenital heart diseases (CHD), data of 6 adult patients, who underwent Htx from 3/1989 until 10/2000 because of different CHD were retrospectively analysed and compared with the population of 995 adult (> 16 years) transplant recipients with different other indications. Results: From March 1989 until October 2000 6 pts (2 male/4 female) were transplanted because of CHD. 3 pts. have been preoperated, one female pt. twice. Mean age was 24.7 years (15.8 to 26.2 y.). Mean time on waiting list was 229 days (29–727 d.). Time of extracorporeal circulation was 105 minutes (68–154), donor heart ischemia-time 178–276 (mean:213) min. Mean postoperative mechanical ventilation-time was 8.75 hours (2.25–16 hrs). Time on ICU was 7.9 days mean (1–12 d.). 4 pts. needed postoperative blood-transfusions (mean: 4.5 units) and 5 pts. positive inotropic support (dopamin 2.5 -8 g). There was no case of acute renal failure. Length of stay was 30.6 days (22–40). 1 female pt. died 1.3 months postoperative because of intracerebral infarctation, 5 pts. are alive. Mean follow-up-time is 9.5 months (25.9 to 113.9), cumulative survival-rate is 83.3% vs. 88% in pts. with non-congenital indications. Conclusion. HTX in adults with CHD can be performed without an increased risk and with a good long term prognosis. Usually there is no fixed increase of pulmonary vascular resistance (< 2WE) and no need for postoperative pacemaker-therapy. Previous palliative operations do not effect the good outcome.

P157

Ventricular response to Dobutamine stress in patients with Mustard repair for transposition of great arteries

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Background: Adaptability of the right ventricle to systemic circulation remains a concern long term after Mustard operation for transposition of great arteries (TGA). Even in asymptomatic patients, exercise tolerance and cardiorespiratory response are limited. Methods: 15 patients (aged 29±10 years, 7 females) 24±6 years after Mustard operation (10 in NYHA 1 and 5 in 2) underwent dobutamine stress echocardiography for the assessment of ventricular function using M-mode and Tissue Doppler techniques. Resting values were compared with 15 age matched controls. Results: All patients but three had mild tricuspid regurgitation at rest. No patient developed symptoms at peak stress. Left ventricular outflow tract obstruction was present in one patient at rest and 5 at peak stress (>3.5 m/s). Conclusion. Dynamic left ventricular outflow tract obstruction occurs in some patients after Mustard repair for TGA. Subclinical systemic right ventricular dysfunction at rest and peak stress is present which may contribute to the known limited exercise endurance.

P158

Ebstein's anomaly

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There is very little information about exercise performance in older patients with Ebstein's anomaly, particularly those that have not undergone surgery. Between 1996 and 2000, we studied 10 patients with Ebstein's anomaly (8 males and 2 females) ranging in age from 22 to 55, mean age 28). All patients had at least 2–3+ tricuspid regurgitation and 2 patients underwent tricuspid valve repair. Five patients exercised according to the Bruce protocol and 5 by the Cornell protocol. Normal VO2 ranged from 53% to 100% of predicted (mean 76% of predicted). The actual VO2s ranged 14.5 to 39.4 ml/kg/min. The mean heart rate was 178 (96% of predicted). Five patients had patent foramen ovales with a mean saturation of 96% prior to exercise, decreasing to a mean of 86% during the exercise test. Adult patients with Ebstein's anomaly and tricuspid regurgitation have mild to moderate impaired exercise performance, but are able to achieve predicted peak heart rates. Some patients desaturate significantly with exercise, which limits their exercise performance.

P159

Primary discreet subaortic stenosis in the adult: a 17-year experience

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There is little information about the presentation of discreet subaortic stenosis (DSS) in adults. Between 1973 and 2000, 39 patients were identified (15 men and 24 women) at a mean age of 38.0 years (range 18 to 83). All patients underwent evaluation by echocardiographic analysis and/or cardiac catheterization. Five patients (12.8%) were also diagnosed with a VSD. Thirty-two

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

patients had normal LV function and 5 were mildly dysfunctional. Three patients had aortic arrhythmia. Nine patients had no aortic insufficiency, 10 trivial, 16 mild, 5 moderate and 1 severe. Seven patients had a dilated ascending aorta. The mean maximum pressure gradient across the LVOT for 23 patients was 74 ± 12 mm Hg. Thirty-two patients (82%) underwent surgery. There was 1 (2.6%) early death. Late follow-up (1 to 207 months) of 23 patients showed no late mortality. ES in adults presents with symptoms, normal LV function, aortic regurgitation (though not severe), and a significant LVOT gradient. Surgery is safe and effective.

P160

The Older Patient With Ebstein's Anomaly: Medical and Surgical Follow-Up.

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We reviewed 51 patients with Ebstein's anomaly (EBS), mean age 44 (age range 18–77 years). Fifty-nine percent of the patients were female. Twenty-seven patients (53%) had surgery and 24 (47%) were treated medically. Eighty-three percent of the medical patients (mp) and 93% of the post op surgical patients (sp) were in sinus rhythm. Seven patients (13.7%) had Wolf-Parkinson-White syndrome. Nineteen of the 27 sp (70.4%) underwent tricuspid valve replacement, and 5 also had right atrial resection or reduction. Seven patients (13.7%) died, 5 sp and 2 mp. However, only 1 of the surgical deaths was related to surgery (4% mortality). There were 4 sudden late deaths. Despite the incidence of late sudden death, only 23 of 51 patients (45%) had follow-up Holter monitoring, and only 16 of 51 (31.4%) underwent stress testing. Older patients with EBS can be successfully treated with tricuspid valve replacement with good surgical results. Late sudden death remains a problem.

P161

There are still adults to be elected surgically: the operative spectrum of congenital heart defects in adolescents and adults

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Interventional catheter treatment plays an increasing role in adult congenital heart disease (CHD). Therefore, we wanted to evaluate whether the disease spectrum of adolescent and adult patients admitted to our heart centre for corrective surgery had changed during the last decade. 430 patients (pt) with CHD and an age over 14 years were included. 254 P (59% were females, mean age was 29.6 years (range, 14–86 years). Patients were divided according to the time of operation: before and after October 1995 Table1 [see attach]. There was no significant difference in diagnoses between the two groups and septal defects were most frequent. There were 27 reoperations P (group I n = 11, group II n = 16) either for palliations, final correction after palliation or for defects. Corrective surgery was performed in 401 (46.4%) P. Hospital mortality was 4.3% (16 P). Complication rate was low (4%) mainly dysrhythmias, bleeding and effusion (no significant difference between groups). Conclusions: There has been no significant change of diagnoses in adolescents/adult P with CHD admitted to our center during the last decade. The treatment of CHD in adolescents and adults even with simple septal defects is still part of the spectrum in cardiac surgery.

P162

An exploration of information and services required by adults with congenital heart defects

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During the past 25 years there have been rapid advances in treatment for congenital heart defects. These developments have resulted in the survival of a new group of adult patients many of whom require life long follow up. The Canadian Consensus Conference on Adult Congenital Heart Disease held in 1996 has published recommendations to guide the development of service provision for this group. While 'These recommendations have been written with cardiologists in mind', (Connelly et al 1998) experienced nurses are considered an essential human resource. There is, however, poorly developed research evidence on which to base nursing care for these patients. This paper will describe the proposal for a qualitative study the purpose of which is to gain a better understanding of the needs of adults with congenital heart defects. This study aims to inform the development of nursing nursing services and plan future services required by adults with congenital heart defects. A complex surgical patient is presently defined predominantly by the physical components of their defect. Adults with congenital heart defects have unrecognised problems with which they cope without the use of existing services. They therefore, fulfil roles within society with a potential disadvantage and may be able to contribute in a more meaningful way both to society and themselves. An ethnographic study is the research approach that can elicit information to identify services and information required by this emerging group. Approximately ten patients from across the clinical spectrum will be interviewed. Identification of other factors impacting on the patients' ability to lead a 'normal' life will contribute to our understanding. Use of this research approach can obtain the patients' perspective, rather than depending on assumptions made from experience in providing care. Connelly MS et al (1998) Canadian Consensus Conference on Adult Congenital Heart Disease 1996. Canadian Journal of Cardiology Vol 14 (5) 705–717.

P163

Thirty years following repair of Tetralogy of Fallot: Right ventricular function as a major determinant of the outcome

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Long term outcome of corrective repair of tetralogy of Fallot (TOF) is determined by several factors including anatomy at operation, residual defects requiring reoperation, ventricular failure and arrhythmias. Although good results are now expected with early correction and improvement of surgical procedures, this is not the case for many patients who underwent operation decades ago. This study was designed for identifying variables that might be involved in the persistence of symptoms (NYHA class II, III or IV versus I) in patients followed for thirty years after repair of TOF. Operations were carried out between 1961 and 1970 and patients (N = 51), 26 female, no deaths included, were aged 7 months to 26 years (median 6 years) at time of treatment. At the end of follow-up, 15 patients were completely free of symptoms whereas the remaining ones had class II (N = 25), III (N = 14) or IV (N = 1) symptoms. Variables analyzed as to differentiate symptom-free from symptomatic subjects were age at operation, need for enlargement of pulmonary annulus and trunk, need for a second (2ndOP) or third operation, residual VSD, residual pulmonary regurgitation, peak gradient across RVOT, right ventricular enlargement or hypertrophy (RVH), cardiothoracic index (CTI), right and left ventricular ejection fraction (RVEF/LVEF), and arrhythmias. Bivariate analysis showed a close association of symptoms with 2ndOP ($p = 0.031$), increased CTI ($p = 0.0001$) and residual moderate to severe RVH ($p = 0.0002$). Multiple logistic regression showed a combination of 2ndOP ($p = 0.037$), RVH ($p = 0.0028$) and decreased RVEF ($p = 0.012$) in determining the persistence of symptoms. A RVLF of 0.23, 0.45 and 0.35 was associated with a probability of persistence of symptoms of 0.5, 0.88 and 0.99 respectively. We therefore conclude that: despite the surgical procedure employed and age at operation, right ventricular remodelling and performance are major determinants of the late outcome following repair of TOF.

P164

Right ventricular physiology after repair of tetralogy of Fallot: Mid-term effects of type of outflow tract repair.

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After repair of tetralogy of Fallot (TOF) either restrictive physiology or progressive dilatation of the right ventricle (RV) can occur. The factors associated with the diverging states of RV performance are not clear. Catheterization data and biplane angiocardiograms were quantitatively analyzed from 62 patients 1 – 30 years (mean 14 years) after surgical correction. RV ejection fraction varied from 31 – 63 % (mean 45 %). End-diastolic volumes ranged between 80 – 390 % (mean 210 %) of normal, with close correlation to pulmonary regurgitant volumes ($r = 0.8$) and QRS prolongation ($r = 0.64$). RV compliance was approximated by the difference of early and late diastolic pressure divided by stroke volume. Compliance was compared in two subgroups with uncomplicated follow-up (no complaints, no arrhythmias, no medications, RV EF > 40 %). In the group with a transannular patch (TAP) ($n = 20$) compliance increased linearly with age from 60 % to 150 % of normal ($r = 0.67$). In the group with non-TAP ($n = 18$) compliance averaged 55 % of normal without age dependence. Thus (1) restrictive RV physiology is common in children irrespective of the type of outflow tract repair, (2) after adolescence RV compliance generally increases with age in TAP extensively, transient persists after non-TAP repair, (3) the impact of elevated compliance accompanying free pulmonary regurgitation favors

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

progressive RV dilatation and QRS prolongation; (4) restrictive physiology may result from regional stiffness of the outflow tract scar or patch rather than from a generally disturbed myoarchitecture, since mechanisms of eccentric hypertrophy for compensation of massive volume overload are still preserved.

P165

The second homograft compared to first homograft in the right ventricular outflow tract in the same patient

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With experience of use of the homograft in the right ventricular outflow tract(RVOT) for repair of complex congenital heart disease since 1966, the study is aimed to evaluate the longevity of the first and second homograft in the same patient. Thirty-three patients with tetralogy of Fallot(14), complex pulmonary stenosis(CPA)(7), pulmonary autograft(7), and other(5), aged 11–66 years(median 37±13 years), male/female: 21/12, survived reoperation of RVOT with homograft and left hospital 1975–1998 following the first operation using homograft 1966–1991. Clinical data was reviewed chronologically and information on pedigree of homograft included. Follow-up time from the first and second homograft was 7.3–33 years(mean 16) and 2.5–20(mean 8.9), respectively. Four(12%) late deaths occurred unrelated to failure of second homograft. Freedom from death at 10,20,30 years after first homograft operation was 97,90, and 77% respectively. Freedom from needing a third homograft at 10, 15, 20 years after the second homograft was 96.75, and 61%, respectively(n=5, 1996). The second homograft lasted for shorter time(9.0±6.2 years) than the first(13.3±7.0 year, p=0.03). 28 second homografts(85%) still remained without failure. Freedom from right ventricular dysfunction(RVD) at 10,20,30 years after first homograft operation was 94,91 and 60%, respectively, however, was significantly less freedom in CPA(p<0.0001). Three of four significant pulmonary valvular disease(PVD) were associated with CPA with poor clinical outcome. Age at both operations, gender(recipient & donor), type of homograft(aortic or pulmonary), did not affect failure of homograft. Despite the obvious shorter follow-up for second homograft, it appeared to have a shorter life compare to the first in the same patient. CPA with regard to RVD and PVD is an important risk factor for long-term outcome of the homograft function. We remain concerned about unknown factors influence the homograft in RVOT.

P166

Fontan procedure. haemodynamic evaluation during exercise

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The cardiac catheterization during exercise on the Fontan patients was performed to evaluate haemodynamic changes in the Fontan circulation. Twelve patients without fenestration (atriopulmonary connection:10, Björk: 2) aged 13 – 29 years (mean 21), operated 1973 – 1990 (3 – 19 years after operation: mean 11) had bicycle ergometric exercise (0.75W/kg, 5 minutes) in supine position with catheter in the pulmonary artery from left subclavian vein. Haemodynamic data obtained is shown with significance indicated. Two had past history of atrial arrhythmia, and there mean PA pressure(PAP) was higher than the others at rest and during exercise(p<0.01). Subaortic stenosis(subAS) was seen in 2 with a significant rise of PA wedged pressure(PAWP) only on exercise(p<0.01). There was no obstruction recognized in the Fontan circuit. Ventricular morphology (RV/LV) did not affect haemodynamic performance. In conclusion, CI at rest was always low, but increased on exercise. Arrhythmia might be an early sign of the increased PAP, and the raised PAWP during exercise was related to subAS. The factors influenced on the elevated PAP and PAWP during exercise such as pulmonary vascular properties and diastolic function of the ventricle, therefore, might predict the long-term outcome of Fontan procedure.

P167

Cardiac output and intracerebral blood pressures at rest and during exercise up to maximal level in GUCH-patients with Fontan circulation at a long-term follow up

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The hemodynamic situation in patients with Fontan circulation is quite different to that in normal patients, as lung perfusion is dependent on the central venous pressure. Our knowledge of how Fontan patients regulate cardiac output (CO) during exercise is limited. MATERIAL. Fifteen out of the 20 surviving patients with Fontan circulation operated between 1980–1991 volunteered to this study. There were 9 men and 6 women with a mean age of 26,4 yrs (19.9–34.2). The mean follow-up time after operation was 14,2 yrs (8,3–19,3). In two patients the CO determinations failed due to technical reasons. METHODS. An arterial cannula was implanted in the brachial or radial artery and a venous one in the opposite arms cubital vein. The Dye-dilution method was used to achieve the cardiac output using Cardiogreen as the indicator substance. Heart rate, oxygen uptake and ventilation were continuously measured using an automatic device and intra-arterial blood pressure were also measured. Exercise was performed on a bicycle ergometer. Double determinations of CO were achieved at rest in the supine position and at two submaximal exercise loads in the sitting position before the maximal load was reached. RESULTS. The patients had a normal mechanical efficiency. The values for cardiac output and stroke volume were lower at all exercise loads compared to the expected values. However, they did keep the stroke volume constant during exercise. The increase in oxygen uptake was achieved by a marked increase in the AV-oxygen difference, which is limited by the Hemoglobin's oxygen carrying capacity. Their mean heart rate at maximal exercise was 142 beats/min. Systolic and mean blood pressure increased in the normal way during exercise. This indicate that their total peripheral resistance were higher than in normal persons.

P168

Clinical profile of adult patients after total repair of tetralogy of Fallot

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To describe the post operative problems of adult tetralogy of Fallot(TOF), a retrospective study was carried out on 28 patients who were registered(1994.5–2000.7) at Samsung Medical Center for TOF. Results: 1) Mean age at this study was 30.6±10.2(range:16–63)year-old, age at total correction was 15.8±2.3(2–49)year-old and the postoperative follow-up duration was 15.1±7.7(0–33)years. 2) The significant clinical problems during adult life were as follows, arrythmia(11), significant pulmonary regurgitation(8), left pulmonary artery stenosis(6), residual VSD(6), significant mitral regurgitation(5), significant tricuspid regurgitation(4), myocardial dysfunction(3), significant aortic regurgitation(2), infective endocarditis(1), and protein losing enteropathy(1). 3) The postoperative arrythmias observed were atrial fibrillation(3), atrial flutter(2), AV block(6), and frequent VPCs(2). Among the 3 patients of atrial fibrillation and 2 patients of atrial flutter, right side maze operation was performed in 3 patients(one patient, atrial fibrillation, the other two patients, atrial flutter), after the operation, atrial fibrillation and atrial flutter were disappeared. The frequency of arrythmia was lower in the group who underwent surgery at a younger age(p<0.05). 4) The incidence of cardiomegaly(cardiothoracic ratio>0.55) was higher in the arrythmia group(p<0.06). 5) Reoperations were performed at 8 patients, and the causes were residual VSD(6) with or without pulmonary regurgitation and peripheral pulmonary artery stenosis(1). 6) Syncope was occurred in 3 patients during postoperative follow-up, the causes were atrial fibrillation and ventricular dysfunction in one patient, and unknown in the other 2 patients. Conclusions: There were various residual abnormalities or problems in adult postoperative TOF patients, so careful lifelong follow-up is needed.

P169

Health status and quality of life of adult survivors of tetralogy of fallot: comparison to sibling controls

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Background: Adult survivors with tetralogy of Fallot (TOF) are at risk for important morbidity and mortality. Health status and quality of life have not been studied in a controlled manner. Methods: A single institution inception cohort of patients with TOF who would have been 18 years of age or older as of December, 1999, were identified. Attempts were made to trace and contact all assumed survivors to obtain data regarding health status, quality of life, medical care and clinical status. Patients were asked to identify a sibling to provide control information. Analysis was based on matched pairs. Results: To date, 62 adult patients (median age 34 years; range 19 to 52) and their siblings have completed the questionnaire. General health was felt to excellent or good in 49% of patients and 68% of siblings (p=0.47), although 20% of patients vs. 5% of siblings felt that their health was worse than one year ago

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

(p=0.006). Limitations regarding vigorous activities were indicated by 71% of patients and 33% of siblings (p≤0.001). Patients vs. siblings did not differ regarding physical limitations or emotional problems impacting on work or social activities. 13% of patients vs. 25% of siblings were occasional or daily cigarette smokers (p=0.27). There were no differences regarding satisfaction with social life (p=0.83). However, 34% of patients vs. 55% of siblings had children (p=0.0119), and 45% of patients vs. 55% of siblings were presently married or living with someone (p=0.23). Conclusions. Compared to their normal siblings adults with repaired TOF had similar self-perceived health but more limitations. Although they had similar life achievements, they were less likely to have had children. Factors impacting on these differences need further exploration.

P170

Pulmonary function in adults with congenital heart disease with and without prior surgical intervention

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Impaired exercise tolerance is frequently noted in adults with congenital heart disease. This has been attributed to multiple factors including: decreased ventricular function, residual defects and chronotropic incompetence. Abnormalities in pulmonary function (PF) have been reported in postoperative patients, but there is little information on PF in patients who have not had surgery. We reviewed the cardiopulmonary exercise tests performed on 63 patients aged 18 – 71 between May 1999 and August 2000. 13 patients had not had prior surgery: Ebstein's: 6, corrected transposition: 4, partial AV septal defect: 1, absent pulmonary valve: 1 and left atrial isomerism: 1). 124 surgical procedures were performed in 50 patients. Mustard's operation: 12, Tetralogy repair: 15, Fontan procedures: 9 and other lesions: 14. Vital capacity (FVC) and forced expiration in one minute (FEV1) were normal in the group without previous surgery and were significantly decreased in patients who had undergone surgery. Results (able: FEV1 and FVC were not affected by age at surgery, type of surgery or number of procedures. Restrictive pulmonary changes are common in adults with congenital heart disease who have undergone surgery and should be considered as a possible contributor to diminished exercise tolerance in these patients.

P171

A method for identifying the basic electrophysiologic substrate for monomorphic ventricular tachycardia after repair of tetralogy of Fallot

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Sudden death after ventriculotomy is largely associated with monomorphic ventricular tachycardia (VT) which requires a zone of slow conduction capable of sustaining reentry. Current methods of risk stratification such as QRS duration and QRS rate of change are indirect reflections of the basic electrophysiological (EP) substrate. The purpose of our study is to define the role of signal averaged electrocardiogram (SAECG) in identifying slow conduction capable of supporting a reentrant circuit after repair of tetralogy of Fallot. Of 169 patients with tetralogy of Fallot, 129 had an intracardiac repair via right ventriculotomy. A SAECG was recorded in 41/129 patients. The mean age of the patients was 40 years, mean time from repair was 27 years, mean QRS duration was 163 ms. We developed an effective method for recording and interpreting SAECG with 2/3 criteria required to establish the presence of late potentials in patients with QRS duration > 120 ms: 1) filtered QRS duration > 145 ms, 2) root-mean-square terminal 40 ms voltage of the filtered QRS < 17.5 microvolts, and 3) duration of low amplitude signals of the terminal filtered QRS > 50 ms. SAECG was positive for late potentials in 22/41 patients. Exercise stress testing and Holter monitoring were used to identify an activating trigger in the form of three or more consecutive monomorphic ventricular depolarizations in three patients. Seven patients underwent re operation with revision of the ventriculotomy scar. The SAECG was negative in 19/41 patients, none of whom had documented monomorphic VT or sudden death after mean follow up of 26 months. We believe that SAECG is a cost-effective measure for risk stratifying patients likely to have an electrophysiologic substrate potentially capable of sustaining reentrant monomorphic VT after repair of tetralogy of Fallot.

P172

Management of grown-up congenital heart disease patients (GUCH): identification of the causes of death and main cardiovascular complications in a series of 80 cases

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The knowledge of the main causes of death and complications is a valuable information for the team of cardiologists dealing with GUCH patients. Data from 80 necropsies were analyzed (age range: 11–59 years; mean=28.64 y; median=26.5; 48 male). The most common types of defects were: tetralogy of Fallot (TF): 15 cases; isolated ventricular septal defect: 10; aortic stenosis: 8; tricuspid stenosis: 7; isolated atrial septal defect and aortic coarctation: 6 cases each. Fifty five patients (68%) had been submitted to cardiac surgery during life (75 operations). Death occurred within the first post-operative month in 58.2%, within the first year in 9.1% and after one year of surgery in 32.7%. The longest post-operative survival times were: 30 and 32 years, in two patients submitted to total correction of TF. From those presenting palliative procedures, the longest survival times were: 29 and 14 years (patients with tricuspid atresia and double inlet left ventricle, respectively). Pulmonary embolism prevailed among the non-operated patients (12.7% against 3.6%) and was the main cause of death in this group. In the operated group the leading cause of death was chronic cardiac failure (30.9%); the second was septicemia (9.1%). Arrhythmia was responsible for death in 5.5% and 2.6% of non-operated and operated patients, respectively. Aortic rupture was detected in 2 patients with aortic coarctation, one of them operated. Pulmonary hypertension was the main complication in non-operated cases (6.9% against: 2.3%). Infectious endocarditis was observed in 24.0% and 10.9% of non-operated and operated groups, respectively. Interventional catheterization related complications occurred in one non-operated (4.0%) and in 3 operated patients with residual defects (5.4%). In conclusion, the recognition of the main fatal cardiovascular events may help clinicians in the care of this particular group of neocardiob.

P173

Anesthetic and perioperative outcome of teenagers and adults with congenital heart disease

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Purpose: Adult and teenaged patients with previously palliated or corrected congenital heart disease require surgical revision or complete obstructive procedures with increasing frequency. The older group of patients presents anesthetic and surgical problems which may differ from the usual population of infants and young children. We describe the perioperative outcome of patients aged 12 years and over undergoing surgery for congenital heart disease in a children's hospital by a dedicated congenital heart surgery and anesthesia team. Methods: Medical records of all patients over the age of 12 from October 1997 to July 2000 requiring cardiopulmonary bypass were reviewed. A control group of patients age 5 years and under was also reviewed, and matched to each older patient when possible by diagnosis, surgical procedure, and repeat sternotomy. Data are reported as mean ± SD. T test or chi square were used to compare older (study) patients with younger (control) patients, with p ≤ 0.05 significant (*). Results: 86 patients were studied in each group. See table. There were no intraoperative deaths. All perioperative deaths, neurologic complications, cardiopulmonary resuscitation episodes in the operating room, massive hemorrhages with sternotomy, femoral cannulations for bypass, and severe hypotension on induction of anesthesia occurred in older patients undergoing repeat sternotomy. Older patients had longer anesthetic, surgical, and bypass times, and experienced more dysrhythmias requiring treatment. Younger patients more often required blood transfusions, and had difficult vascular access. A large number in both groups required inotropic support. Conclusions: Mortality and major morbidity were low in both groups; however all major incidents occurred in older repeat sternotomy patients. Based on these data, preparation for these infrequent but potentially catastrophic occurrences seems warranted in this group of patients.

P174

The use of self-expanding stents in stenotic aortopulmonary shunts in adults with complex cyanotic heart disease

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We describe the use of self-expanding stents in treating long-segment stenosis

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

of multiply stenotic aortopulmonary shunts (APS). Four patients underwent cardiac catheterization due to clinical deterioration with a view to stent implantation. Their age ranged between (23–32) years. The underlying diagnosis was complex cyanotic heart disease in all, 3 had a systemic interposition graft, and 1 had a classical Blalock shunt. The pre procedural haemodynamics were: resting oxygen saturation (O2 sat) range between (69–82%), exercise tolerance (ET) between (50yards–5min) with exercise O2 sat range (42–60%), pressure distal to stenosis (PDS) range (11–13) mmHg, ankle index (ABI) range (3–4). The post procedural haemodynamics were: resting O2 sat range (84–85%), ET range (4.5–15) min, exercise O2 sat range (59–68%), PDS range (14–26) mmHg, ABI range (2–4). There was one technical failure due to migration of the stent distal to an atrial stenosis. The early term functional results (follow-up 1.6–3.5 years) in the other 3 patients have been excellent. We conclude that this technique may further palliate adult patients with complex congenital heart disease, however the long term patency of stents is unknown.

Transplantation

P175

EKG changes after pediatric heart transplantation and their association with pretransplant parameters

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Objective: The study was performed to report the ECG changes in pediatric heart transplantation and to examine pretransplant factors that could be associated with these abnormalities. Patients and methods: Seventeen children underwent to heart transplantation from October 92 to February 98. The patients ranged in age from 12 days to 6 years (mean: 2.4 years). The immunosuppression protocol was: cyclosporine, azathioprine and antilymphocyte induction therapy. Acute rejection episodes were diagnosed primarily using noninvasive parameters. Actuarial survival rate was 94% at 1 year and 88% at 5 years. The mean follow-up period was 2.5 years. All 82 head electrocardiogram obtained in the study patients at the last year after transplantation were reviewed. The pretransplant factors studied were: pulmonary vascular resistance index, ischemic and donor age, donor/recipient weight ratio and ischemic time. Results: The right ventricular hypertrophy (n=79) was the most common morphology observed. Right bundle branch block was finding occurring in 2 (11.8%), incomplete right bundle branch block developed in 1 (17.6%). Pretransplant factors were not related to right ventricular hypertrophy and right bundle branch block abnormalities. Arrhythmias were associated with rejection episodes in 84.6% of occasions and in 15.4% with abnormal metabolic and electrolyte balance. Only one patient required definite pacemaker due to complete heart block. Conclusion: Right ventricular hypertrophy and right bundle branch block were the most common ECG findings in pediatric transplanted recipients, but in this study there was no pretransplant factor associated with these abnormalities.

P176

Factors associated with infection in pediatric heart transplantation

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Background: Despite the current wide-spread success of heart transplantation as a therapy for end-stage heart disease, infection represents one of the leading causes of death in pediatric heart transplantation. This study was performed to report factors associated with infection in children after heart transplantation using double immunosuppression. Patients and methods: Seventeen children underwent to orthotopic heart transplantation from October 92 to February 98. Age ranged from 12 days to 6 years (mean: 2.4 years). The immunosuppression was accomplished with cyclosporine, azathioprine and antilymphocyte induction therapy. Acute rejection episodes were diagnosed primarily using noninvasive parameters. All infections treated with oral antibiotics were entered into the analysis. We studied the following factors: diagnosis of cardiopathy indicated for transplantation, donor/recipient weight ratio, donor age, ischemic time, the number of rejection episodes, pulmonary vascular resistance index. results: Of the 68 infections in 17 patients, bacterial infections were most frequent (70.6%), followed by viral (16.2%), protozoal (10.3%) and fungal (2.9%). CMV infection accounted in 8 (47%) patients, with peaked during the first two months. The most common sites of bacterial infection were lung (50%) and blood (12.7%). Bacterial infections were the most common type of infection in infants less than 6 months of age. The mean number of infection per patient was 3.9+/- 2.5 and median 3. Greater number of rejection episodes were associated with increased number of infection episodes (p=0.002). There was no death due to infection. Conclusions: Although infections episodes were frequent in pediatric heart transplanted recipients, they were successfully treated. Greater number of rejection episodes were associated with increased number of infection episodes.

P177

Is there any difference between induction therapy with horse-(atg) and rabbit (ratg) derived -antilymphocyte preparations in pediatric heart transplantation

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Background: Immunoprophylaxis remains a controversial aspect of immunosuppression regimens. This study was performed to compare the medium-term follow-up between induction therapy with ATG and RATG in children undergoing to heart transplantation. Patients and methods: Seventeen children underwent to heart transplantation from February 93 to February 98. The patients ranged in age from 28 days to 9 years. The survival rate was 93.7% at a mean follow-up period of 2.5 years. All patients had their panel-reactive antibodies rate < 10% except one who had 11%. All cross-matches were negative. The immunosuppression protocol was consisting of cyclosporine, azathioprine, Methylprednisolone was given every 12 hours for four days or on rejection treatment. The induction therapy was defined as an administration of antilymphocyte preparations immediately after transplantation or on the first rejection episode. ATG (Group A, n=8) was administered at mean dose of 17.5+/- 2.7 mg/kg/day during a mean period of 6.9 +/- 2.8 days and RATG (Group B, n=8) at a mean dose of 11+/-0.7 mg/kg/day, during a mean period of 7.0 +/- 2.1 days. A decrease in T-cell populations was expected with total T-cell (CD3) counts dropping to at least 150 cmm/l by flow cytometry. Rejection, infection episodes and requirements of antilymphocyte globulin to treat rejection episodes were compared between the two groups. Results: There was a trend towards worse rejection history with group A (p=0.061). Group A required more antilymphocyte preparations to control rejection episodes (p=0.001). There was no death due to acute rejection on this study. Conclusions: The use of induction therapy with RATG has shown a better rejection history with decrease in the number of treated episodes of rejection and the use of antilymphocyte preparations for treatment of recurrent rejection in pediatric transplanted recipients.

P178

Clinical outcome using noninvasive methods for the evaluation of acute rejection and double immunosuppression therapy in pediatric heart transplantation

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The noninvasive method of rejection diagnosis requires a synthesis of various clinical and laboratorial parameters. The purpose of this study was to evaluate the noninvasive methods for diagnosis of acute rejection with double immunosuppression and clinical outcome in pediatric heart transplantation. Patients and methods: Seventeen children have undergone to heart transplantation from October 92 to February 98. The patients ranged in age from 12 days to 6 years (mean: 2.4 years). The diagnosis of rejection was based on noninvasive methods. The noninvasive parameters used were: clinical findings, ECG changes, echocardiogram abnormalities. Endomyocardial biopsy was performed only if there were persistent signs of acute rejection despite initial treatment or for confusing clinical situations. There were 94 episodes of acute rejection and we studied the number of episodes of rejection per patient, the necessity of biopsy and clinical outcome. Results: The noninvasive surveillance detected acute rejection on 90.8% of the occurrence and the diagnostic endomyocardial biopsy could aid in the diagnosis was required on 9.2%. The episodes of rejection per patient ranged between one to eight (mean: 3.2+/-1.9 and median: 3.0). Actuarial survival rate was 94% at 1 year and 82% at 5 years. The mean follow-up period was 2.5 years. Only one patient died due to acute rejection (40 days after transplantation [on the second day after initial rejection treatment]). Conclusions: An evaluation of the medium-term follow-up of children undergoing heart transplantation indicates that excellent results can be obtained with a rejection management and surveillance protocol that emphasizes noninvasive techniques for diagnosing rejection.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P179

Growth and survival outcome of children less than seven years of age using double immunosuppression therapy after heart transplantation

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Background: The precise role of steroids in children is still being debated due to growth retardation. This study was performed to report the growth and survival outcome using double (cyclosporine and azathioprine) immunosuppression therapy. Patients and methods: Seventeen children underwent to heart transplantation from October 92 to February 98. The patients ranged in age from 12 days to 6 years (mean 2.4 years). The immunosuppression was accomplished with cyclosporine, azathioprine and antirhymocyte induction therapy. Acute rejection episodes were diagnosed primarily using noninvasive parameters. Cyclosporine target range (parent compound in a whole-blood matrix) were 250 to 300 ng/ml during the first year and 150 to 200 ng/ml thereafter. Azathioprine (2 mg/kg/day) was adjusted to maintain a white blood cell count > 3500/mm3 and after one year the dose was reduced to 1 mg/kg/day. The Methylprednisolone was not routinely used in any age group. We studied the Z-score values for weight and height for children at their last routine visit using epi Info. Results: There was an increase in all percentages of Z-scores values in children less and more than one year follow-up of transplantation. However, the Z score percentage increase for height of children less than one year of transplantation was smaller than children with more than one year of transplantation and with others Z scores. Actuarial survival rate was 94% at 1 year and 85% at 5 years. Mean follow-up period was 2.5 years. Six patients used chronic prednisone for a median of 63.5 days. Conclusions: Double immunosuppression regimen allowed development growth of children with less and more than one year of heart transplantation, with excellent survival outcome.

P180

Pediatric heart transplantation: risk factors associated with acute rejection

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Background: Acute rejection is one of the most important complications in the heart transplantation. The purpose of this study was to better understand factors that might predict a worse rejection history. Patients and methods: Seventeen children underwent to heart transplantation from October 92 to February 98. Actuarial survival rate was 94% at 1 year and 85% at 5 years. Mean follow-up period of 2.5 years. Age ranged from 12 days to 6 years (mean 2.4 years); 52.9% were male. The diagnosis of rejection was based on noninvasive methods. The immunosuppressive protocol was cyclosporine, azathioprine and induction therapy with polyclonal antilymphocyte serum. There were 54 episodes of acute rejection and we studied the distribution of rejection episodes, the number of episodes of rejection per patient. The factors evaluated were the following: donor/recipient sex match, blood type, age at transplantation and number of infections per patients. Results: The episodes of rejection per patient ranged between one to eight (mean 3.2 +/- 1.9 and median 3.0). The major incidence of rejection episodes occurred on the first three months after transplantation (72.2%), between 3 to 6 months was 5.6%, 6 to 12 months was 13% and more than 12 months was 9.3%. There was no correlation between donor/recipient mismatch for gender, blood type and age at transplantation with rejection episodes (p=0.335 / 0.000 / 0.305). Greater number of infections were associated with more frequent rejection episodes (p=0.002). Conclusions: An evaluation of the medium-term follow-up of children undergoing heart transplantation indicates that excellent results can be obtained with rejection management and surveillance protocol that emphasizes double immunosuppression regimen with induction therapy and noninvasive techniques for diagnosing rejection. The only factor associated with more frequent number of rejection was the greater number of infections.

P181

Survival after late intubation in pediatric heart transplant recipients

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BACKGROUND: Cardiac transplant has excellent outcomes and survival in children with severe heart disease. Long term predictors of poor outcomes in these patients are limited. This study evaluates the effect of late intubation (> 30 days post-transplant) on survival in these patients. METHODS: Data on 357 pediatric heart transplant recipients (0–18 yrs) were reviewed to evaluate outcomes with late intubation. RESULTS: Overall mortality of 30% (106/357) was noted. 53 patients (15%) required late intubation and mechanical ventilation. 35 of these patients (38%) suffered multiple episodes of respiratory failure requiring multiple intubations. The etiology of the respiratory failure was an infectious agent in 32 of the patients (60%). The need for late intubation was not affected by age at transplant. The 30 year actuarial survival of the late intubation patients was significantly lower than those not requiring such intubation (40% vs 69%, p=.03, see graph). The relative risk of mortality following late intubation was 1.59 (CI 1.11-2.27). The relative risk of death was exponentially higher in those patients requiring multiple intubations, 2.38 (CI 1.6,3.53). Infection as the cause of the respiratory failure did not significantly influence the relative risk of mortality, 1.02 (CI 0.54,1.92). There was no difference noted due to age at first intubation (p>0.05). CONCLUSION: Intubation and mechanical ventilation after 30 days following pediatric cardiac transplantation significantly increases the risk of mortality. This risk is further increased with multiple late intubations.

P182

The assessment of exercise tolerance, cardiac function and wall motion using stress echocardiography in children who have undergone cardiac transplantation

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Purpose: This study compared exercise tolerance, cardiac function and wall motion in 5 transplant patients (T) with 12 healthy controls (C) at rest and during progressive exercise using stress echocardiography. Methods: Subjects exercised on a semi-supine cycle ergometer to volitional fatigue. M-Mode 2-D echo and Doppler were performed before, during, and immediately after exercise. Results: Coronary angiography and biopsy results were normal for all T. The median interval between transplantation and testing was 10.9 years for T. While T were 2.4 years older than C, height, weight and BSA were similar. T did less work than C (960 vs 1219 joule/kg-1; p<0.05). LV mass and resting and exercise LV dimensions were similar. SF (in mm/s) rose from 36 to 58% in T and from 38 to 52% in C. Diastolic BP was higher in T, both at rest (90 vs 78 mmHg; p<0.01) and at peak exercise (160 vs 80 mmHg, p<0.005). Peak VTI was similar (29.9 vs 29.5 cm) for T and C. Resting CI (3.23 vs 3.29 l/min-1/m-2) and SVI (31.7 vs 41.7 ml/m-2) were similar for T and C. At peak exercise both CI (5.87 vs 9.26 l/min-1/m-2, p<0.002) and SVI (39.1 vs 49.1 ml/m-2, p<0.02) were lower in T. HR reached 83% and 91% of predicted maximum at peak exercise in T and C. Wall motion was normal in all subjects. Conclusions: T achieved near normal exercise tolerance, although they were able to do less work than C. T had lower CI and SVI, and higher diastolic BP at peak exercise than C. However, HR, systolic BP, LV contractility and wall motion response to exercise were normal in T.

P183

First Experience with Daclizumab in Pediatric Heart- and Heart-Lung-Transplantation

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Daclizumab, a new adjunct immunosuppressant, is a human IgG monoclonal antibody against the IL-2-receptor of activated T-cells (CD-25+). So far data after thoracic transplantation are only available in adult patients. Three selected patients with additional pretransplant or postoperative expected organ dysfunction were treated with Daclizumab after heart and heart-lung-transplantation. Patient one, age two years, had primary pulmonary hypertension, patients 2 and 3, age 12 and 18 years, had Eisenmenger-complex with severe cardiac failure. Daclizumab was administered pre and post transplantation (0.5 mg/kg) and on p.o. day 8, 22, 36, 50 (1 mg/kg) (pts 2 and 3), patient 1 had only pre operative dosage. In addition patients received cyclosporin A (pt. 2 and 3) or tacrolimus (pt. 1) combined with mycophenolate mofetil and prednisone. The aimed drug levels of cyclosporin A and tacrolimus were reduced by one third (200–250 ng/ml and 10–12 ng/ml, respectively). CD-25+ lymphocytes were measured every third day. Daclizumab was well tolerated, no adverse effects were noted. CD-25+ lymphocytes were completely eliminated in all cases. There was no acute rejection during follow-up. Patient 3 developed transient renal failure after an excessive CPB time, no other organ complications were observed. These preliminary results show an excellent suppressant effect of daclizumab on CD-25+ lymphocytes in pediatric thoracic transplantation. The dosage of other immunosuppressive agents was

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

reduced to protect from renal and other organ failure: no acute rejection was observed. Further investigations are necessary to assess the potential advantage of daclizumab as adjunct immunosuppressant in selected patients with potential organ complications caused by standard immunosuppression.

P184

Infection and Rejection in Pediatric Heart and Heart-Lung Transplantation: Comparison of Mycophenolate Mofetil and Azathioprine

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Currently either Mycophenolate Mofetil (MMF) or Azathioprine (AZA) are used as immunosuppressive adjuncts in pediatric heart- (HTX) and heart lung-transplantation (HLTX). We examined the difference between these agents regarding infection and rejection. Thirteen patients, age 6 months to 19 years treated with MMF (group 1: HTX n=9, HLTX n=4) were compared to a group of 10 children, age 2 to 15 years, treated with AZA (group 2: HTX n=4, HLTX n=6). All patients received Cyclosporine A, FK 506 and Prednisone in addition. The mean follow-up was 283 (group 1) and 186 patient months (group 2). All episodes of infection and rejection were registered. The number of simple infections without hospital admission was two times higher in the AZA-group (AZA 0.44 vs. MMF 0.21 simple infections/patient months). The number of severe infections necessitating hospital admission was less in group 2 (AZA 0.1 vs. MMF 0.17 severe infections/patient months). Acute rejections (AR) were only seen in the AZA-group in 5 of 10 patients with more than one episode per patient in 2 cases (AZA 0.13 rejection/patient months). No AR occurred in group 1. One case of chronic rejection was seen in the MMF-group. No patient died during the follow-up period. In pediatric patients after HTX and HLTX the overall number of infections was low in both groups. Simple infections were more frequent under AZA; in comparison, severe infections occurred more often under MMF. However, AR was not seen under MMF. In conclusion MMF seems to be an acceptable alternative to AZA in the pediatric population after HTX and HLTX. Further follow-up is necessary to compare the incidence of long-term graft survival in both groups.

P185

The mid-term outcome of living-donor lobar lung transplantation in 3 Japanese pediatric patients with primary pulmonary hypertension

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Lung transplantation has been practical choice of treatment for the patients with end-stage primary pulmonary hypertension (PPH) in US with encouraging early results. However, mid-term follow-up of cardiac and pulmonary function have not yet been fully documented. From 3/1995 to 10/1996, living-donor lobar lung transplantation (LDLT) was performed at Children's Hospital of Los Angeles for 3 Japanese pediatric patients with PPH in whom oral PGI2 were ineffective. Their ages at operation were 11, 12, and 14 y/o. The preop NYHA class were IV, III, and III, respectively. The average preop mean PAP was 71 mmHg, and mean pulmonary/systemic vascular resistances were 17.9/20.4 u·m2. Each recipient received a right lower lobe from recipient's father as a right lung and a left lower lobe from recipient's mother as a left lung. Mean follow-up period was 58 months. No one of the donors had any complication. Their initial immunosuppressive regimen consisted of cyclosporin, prednisolone, and azathioprine. No serious complications nor chronic rejection has been reported. Cardiac cath at 1yr after the LDLT revealed normal mPAP and mean pulmonary/systemic vascular resistances of 2.0/21.7 u·m2. NYHA class was I in all patients. TBLB revealed No – A1. Mean PFTs results were as follow. FEV1.0/FEV2% at 1yr, 78% at 2yrs, and 77% at 3 yrs. FVC were 100% at 1 yr, 109% at 2yrs, and 107% at 3yrs, respectively. Conclusions: Mid-term follow-up of 3 pediatric LDLT patients with PPH has demonstrated favorable cardiopulmonary function and quality of life. Consequently, LDLT appears to be one of alternate therapeutic options in pediatric patients with PPH who are rapidly deteriorating.

P186

Ascending Aortic Aneurysm after Pediatric Heart Transplantation Case report of an unusual complication.

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Pediatric orthotopic heart transplantation is a recognized and well stablished surgical procedure to treat some congenital heart defects or different acquired cardiomyopathies. The occurence of problems related to immunosupression are well known and the objective of that report is to present an uncommom and potencially lethal post-operative complication in a child after heart transplantation. In our experience with 30 children transplanted since November 1992, with ages ranging from 12 days to 12 years, a 28 month-old boy, weighting 11 kg, with diagnosis of severe dilated cardiomyopathy was transplanted on December 1995. The child had pre operative left ventricle ejection fraction of 14%. Orthotopic heart transplantation was performed on December 1995, no surgical problems or immediate post operative complications were observed. Immunosuppressive therapy consisted of cyclosporine and azathioprine as basic drugs. The child was discharged from the ICU on 12th PO day and from the hospital in the 38th PO day, after had created a respiratory infection. After 5 months of clinical asymptomatic follow up, in a routine ECHO, a dilatation in the ascending aorta was observed. An NMR confirmed the diagnosis, with observation of an ascending aortic aneurysm, with 3.8 cm in its transverse diameter. Surgery was performed by midline sternotomy and before opening the sternum heparinization (4mg/kg) was done and the right femoral artery was cannulated. The ascending aorta was exposed and CPB was initiated. After opening the aorta, a left anterior aortic aneurysm was confirmed, a bovine pericardium patch was sutured, reconstructing the aortic wall, excluding the aneurysm. Good heart recovery was obtained, hypertension was detected in ICU, well controlled with captopril. Immunosuppressive drugs were maintained during post operative period. The child was discharged from the hospital on 7th PO day; post operative ECHO and NMR showed absence of aortic aneurysm. After 48 months he is in a good clinical condition, with normal ECHO studies performed every 3 to 6 months, followed by outpatient clinical visits. We conclude that the surgical solution was safe, with no morbidity after 18 months of operation.

P187

Indicators for Pacemaker Dependency in Patients after orthotopic Heart Transplantation

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Introduction: Indications for peroneent pacemaker implantation (ppi) after heart transplantation (HTx) are uncertain, especially in the early period (four weeks) after transplantation. As well uncertain are parameters concerning pacemaker dependency in the early versus late period after HTx. Results: 1230 HTx have been performed between March 1985 and December 1996. In 96 patients (8.3%) ppi was necessary. 71 patients (74%) received a dual chamber (DDD-R) ; 25 (26%) a single chamber system (21 pts VVI-R, 3 pts. AAI-R and 1 pt VDD). Indications for VVI-system were symptomatic brady-arrhythmias during atrial fibrillation in 6 pts, intermittent total AV-bl in 12 pts and symptomatic SA-block in 3 pts. DDD were implanted in 31 pts with intermittent total AV-bl, in 21 pts with bradytachycardia in 7 pts with sick sinus node and in 3 pts with SA-block. The single lead system was implanted due to AV-bl and the AAI in 3 cases due to isolated bradycardia. Early implantation (< four weeks after HTx) was performed in 27 cases (28%) and late implantation (> four weeks) in 68 pts (72%). 24 pts (25%) have died in the following 0.6 to 73 months (mean 13) after ppm. 11 pts (46%) due to severe rejection, 9 pts (38%) due to sepsis and multiple organ failure and 2 pts (9%) due to myocardial infarction. In two other patients (9%) a sudden heart death was documented by holter monitoring. Recipient- or donor age, ischemic time, number of myocardial biopsies, heart rate before ppm and early or late implantation of the pm were not related to long term pacemaker dependency. In the early period 31% of the pts were pm dependent vs. 29% after a mean of 43 months follow up. The one year mortality rate in pm-pts (25%) after HTx was higher than in pts without pm (21%), but did not reach statistical significance. Conclusion: Most symptomatical bradyarrhythmias in the early period after HTx are emerging on the base of mild, moderate or severe rejections and can be sufficiently treated by modification of the immunosuppressive protocol in combination with positively chronotrope medication and temporarily epicardial pacing. In lack of reliable parameters predicting pm dependency after HTx early implantation should be avoided whenever possibly.

P188

Influence of mechanical circulatory support on the incidence of neoplasia following long-term survival of heart transplantation

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https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Introduction: Incidence and mortality of malignant neoplasia after long-term survival of cardiac transplantation (HTx) are higher than in non-transplanted people and depend to risk factors. Predominantly are skin-, bronchial-, urogenital- and lymphoproliferative neoplasia (NPL) in 3–15% of organ recipients. Methods and Results: Between March 1989 to May 2000 1060 HTx were performed analog to Lower and Shumway. 162 pts. (15%) were bridged to HTx with five different assist devices. The centrifugal pump-Biomedicus was implanted for mean 3 days (1 hour – 15 days), the Abiomed-system for mean 7 days (1–26). Thoratec for mean 55 days (3–288), Novacor for mean 150 days (80–194) and Heart Mate for mean 145 days (84–730). All pts. were successfully transplanted. 102 of 1060 pts (9.7%), m/w 94/8, developed under the influence of immuno-suppressive medication a malignoma. Cutane NPL n=24 (24%) m/f=23/1, bronchial-carcinoma n=21 (21%), m/f=22/1, gastrointestinal-NPL n=20 (20%): esophagus ca. n=3, m=3, gastric-ca n=5, m/f=4/1, colorectal n=6, m=6, pancreatic-ca n=3, m=3, liver/gall bladder-NPL n=2, m=2, urogenital n=19 (19%), Hodgkin and Non Hodgkin lymphoma (HL/NHL) n=16 (16%), m/f=14/2, gynecologic n=2 (2%), glandula thyreoidea n=1, f=1. 12 pts. suffered from two malignant NPL and 1 pt from three different NPL. 4/162 pts (2%) developed a neoplasia after MCS as bridging to HTx: Heart: Marr-gastric-carcinoma and prostata-ca., Novacor: mamma-ca. and Thoratec-bronchial-ca. 58/102 pts. (57%) died in the follow-up period of mean 43 months (9–120). pts. with gastrointestinal tumors had the highest mortality of 90%, followed by bronchial-NPL 71%, gynecologic NPL 67%, HL/NHL 63%, urologic-NPL 37% and cutane 21%. Discussion: This retrospective analysis of the incidence of different malignoma in pts who underwent cardiac transplantation (9.7%) correlates to reports of other investigators with a dispersion of 3–15%. In contrast to implanted hyperbaric mechanical circulatory support has an influence on the development of malignant neoplasia. Prospective studies comparing different parameters and their influence on NPL induction are necessary.

P189

Results of bicaval heart transplantation in children

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Purpose: Review our results with the bicaval anastomosis technique for heart transplantation in children. Methods: From 1985 through 2000, 55 consecutive pediatric patients underwent cardiac transplantation using a bicaval technique. Diagnoses were cardiomyopathy (18), complex congenital heart disease (6), failed Fontan (4), failed atrial baffle (3), and adriamycin toxicity (4). Age ranged from 1 month to 17 years, mean age was 6.2 ± 7.7 years. Results: There were 2 operative deaths (6% early mortality) at 1 and 6 weeks post transplant from fixed pulmonary hypertension and from a massive unrepairable abdominal lymphangioma. Donor ischemic time ranged from 71 to 238 minutes (mean 156 ± 92 minutes). No patient required a pacemaker or late tricuspid valve replacement. No patient had a postoperative tricuspid or inferior vena cava pressure gradient. Perioperative echocardiograms showed tricuspid regurgitation (TR) to be trivial (24), mild (7), and moderate (3) for a mean early TR score of 1.3 ± 0.5. Follow-up echocardiograms ranged from 0.5–4 years post-transplant (mean 1.9 years). TR was then assessed as trivial (14), mild (15), and moderate (3) for a mean late TR score of 1.5 ± 0.7, not statistically different from perioperative TR ($p = 0.5$). Conclusion: In children the bicaval technique of heart transplantation can be performed with acceptable donor ischemia times, has a low operative mortality, and is not associated with progressive tricuspid regurgitation. The bicaval technique greatly facilitates cardiac transplantation after failed atrial baffle procedures, failed Fontan, and after Glenn procedures. We recommend the bicaval technique as the procedure of choice for cardiac transplantation in children.

P190

Outcome of pediatric heart transplant recipients without induction of immunotherapy with polyclonal or monoclonal antibodies

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Background: The regular usage of polyclonal or monoclonal antibodies in the immediate postoperative period after pediatric heart transplantation is controversial. Alternatively the induction of immunosuppression could be started with Cyclosporine A, Azathioprine and Glucocorticoids subsequent to the reduction and adjustment of the dose. We report in this retrospective evaluation the outcome of 72 pediatric heart transplant recipients, who have not received induction immunotherapy using antibodies against cell-surface proteins. Patients: Between 1988 and 2000 cardiac transplantation were performed in 72 pediatric patients at an age of one week up to 16 years (mean 7.4) in our center. The induction immunotherapy was composed of Cyclosporine A, Azathioprine and Methylprednisolon. After the early stages of the posttransplant period, a Cyclosporine monotherapy was possible in 27 patients. Combination with Methylprednisolon was necessary in 15 patients. The acute rejections were detected non-invasively and treated primarily with steroid-pulse-therapy. Results: We observed 74 cases of acute rejection in 18 patients (25%). Nine patients were treated sufficiently in the above described manner. In 5 cases, where repeated rejection episodes occurred, we replaced Cyclosporine A with Tacrolimus. In 4 patients with severe and steroid-resistant acute rejection we administrated monoclonal antibody OKT3 as an antirejection agent. Two patients survived. The early and late mortality of 20 patients were 16.1% and 6.4% respectively. We observed 14 cases of renal insufficiency, one case of graftvasculopathy and one neoplastic disorder. Steroid-related diabetes was noticed in two Patients. Conclusion: The induction of immunotherapy with Cyclosporine A, Azathioprine and Glucocorticoids following the reduction and adjustment of the necessary dosage established a sufficient immunosuppressor standard in the early and late posttransplant period.

P191

Daclizumab induction therapy in pediatric cardiac transplant recipients

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Acute rejection is a major source of morbidity in the first twelve weeks following cardiac transplantation. Repeated or severe episodes of rejection may predispose to accelerated graft vasculopathy or chronic vascular rejection. Triple drug immunosuppression has been the mainstay in most centers, but the incidence of acute rejection remains relatively high. The addition of induction immunotherapy may increase the risk of opportunistic infections and lymphomas. Daclizumab, a molecularly engineered human IgG1 monoclonal antibody to the IL-2 receptor, has been shown to reduce acute rejection in adult cardiac and renal transplant recipients without an increase in the incidence of infections or lymphoproliferative disorders. The experience in pediatric transplant patients has been limited. We evaluated 8 consecutive pediatric cardiac transplant patients during the first 12 weeks following transplant. The first 5 recipients received standard triple immunosuppressive therapy only. The subsequent 3 recipients received daclizumab, 1.0 mg/kg IV at the time of reperfusion, at 8 days and then every two weeks for a total of five doses in addition to standard triple therapy. Neither group had any reactive T or B cell antibodies at the time of transplant. Each patient in the non-treatment group had at least one episode of ISHLT grade 3 rejection with no grade 3 episodes in the treatment group ($p=0.018$). The mean frequency of acute rejection episodes (ISHLT grade 2 or greater) was 0.25 per patient in the non-treatment group and 0 for the treatment group ($p=0.018$). There were no adverse reactions to daclizumab and no significant difference in the incidence of opportunistic infections. Daclizumab induction in pediatric cardiac transplant recipients was associated with a significant reduction in the frequency and severity of acute allograft rejection without significant adverse effects.

P192

Echocardiographic surveillance following paediatric cardiac transplantation: which parameters are reliable?

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Objective: The role of echocardiography to detect rejection following cardiac transplantation (CTx) remains controversial. Thirty-eight children have undergone CTx at Hopital Ste-Justine between 1988 and 2000. Rejection surveillance was accomplished preferentially by serial echocardiogram and by periodical endomyocardial biopsies. A total of 376 biopsies were performed during a mean follow-up period of 66 months (range 1–157 months). Rejection requiring pulse steroid therapy was identified in 29 biopsies (Group 1) and 347 biopsies showed no need for intervention (Group 2). To determine which echocardiographic parameters were useful to detect rejection, we analyzed echocardiographic data collected at the time of these biopsies measuring ventricular mass and systolic/diastolic function. Results: Group 1 and 2 were comparable with respect to mean body surface area (1.31±0.4 vs 1.26±0.4, $p = 0.21$). The left ventricular mass index (LVMI) was significantly increased in Group 1 (114.4±40 g/m2) compared to Group 2 (95.7±32 g/m2, $p = 0.005$). Systolic function was significantly decreased in the presence of rejection, as measured by shortening fraction (Group 1 = 0.35±0.07 vs Group 2 =

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

0.36±0.06, p = 0.03) and heart-rate corrected velocity of circumferential fibre shortening (Group 1 = 1.72±0.42 vs Group 2 = 1.99±0.55, p = 0.03). Parameters examining diastolic function were not significantly different in the presence or absence of rejection (isovolumetric relaxation time, p = 0.17, ratio of mitral E and A waves, p = 0.49; mitral E wave ascending pressure half-time, p = 0.34 and descending pressure half-time, p = 0.27). Conclusion: Serial echocardiographic surveillance is a reliable non-invasive method for identifying rejection following cardiac transplantation. An increase in LVMI and/or a decrease in systolic function is suggestive of rejection.

MAY 28 Time: 14:00–15:30

Session 2
School Health/Preventive Cardiology

P193

Physical fitness of urban US children

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There has been a growing concern in the US that physical fitness (PF) in children is decreasing. The objective of this study was to measure the level of PF in a large US urban population and compare it to the results of a widely used reference study from Canada. Bruce treadmill protocol was utilized to evaluate the endurance time (ET) of 525 children 4 to 18 year old (303 males and 222 females). The subjects (S) were from Chicago and they were referred for evaluation of chest pain, syncope, shortness of breath, innocent murmurs or suspected arrhythmias. All S had history, physical examination and electrocardiogram and when indicated chest x-ray, Holter monitor and/or echocardiogram. They were excluded in the study only if cardiopulmonary pathology was excluded. The mean maximal heart rate in males was 192 +- 15 and in females was 194 +- 14 beats/minute. ET increased with increasing age in males it increased up to 10–12 years of age in females and thereafter leveled off. Results were compared with those of the reference study that included 327 S from a small city population in Canada. Mean ET of our S were significantly lower (p<0.01). When the S were placed in percentile groups based on the reference study, 61% of males and 81% of females performed below the 25th percentile. There was a strong negative correlation between BMI and ET (p<0.0001), suggesting that obesity is a major contributor to decreased PF. For each unit rise in BMI the ET fell by 0.0695 minute. Compare to S in the reference study, children from a large US metropolitan area have markedly reduced exercise capacity suggesting poor PF. Inactivity and lack of fitness, if unchecked, are likely to lead to cardiovascular problems in adulthood. Efforts should be made to promote an active life style in childhood.

P194

Psychosocial inventory method for children/adolescents with heart disease

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The extensive development of paediatric cardiology has given result in a new group of surviving patients with complex congenital heart disease and a complex psychosocial situation. Along with medical development, there is a need for specific psychosocial instruments to measure and describe the psychosocial complexity. Such a method is now invented. STUDY GROUP: Ninety-seven patients, 47 boys and 50 girls were graded into three categories (I–III) with respect on complexity of congenital heart disease (CHD). Group I comprised 42 patients, group II 20 patients and group III 35 patients. METHOD: The inventory method is based on a life quality model divided into 3 spheres: personal, interpersonal and external. Each sphere is composed of a part of symptoms and a part of interventions. The personal sphere describes the child's individual psychological distress symptoms and interventions needed. The interpersonal sphere evaluates symptoms/problems in the family, school and corresponding interventions. The external sphere describes the need for and execution of medical, social and economic support related to the heart disease. The severity of the symptoms/ interventions is evaluated using a 4 point Lickert scale ranging from 0 to 3. RESULTS: The most frequent symptoms were somatic symptoms-personal (19/97), family symptoms-interpersonal (68/97) and health care related needs-external (71/97). The most frequent interventions were support of parents/psychotherapy- interpersonal (408/1526), health care contacts-external (307/1526) and social investigation/ information – external (229/1526). Family symptoms constituted the most severe separate interpersonal variable and support of parents/psychotherapy the most extensive intervention. As expected the group of patients with the most complex CHD showed the most common and severe needs of support (fig.1). CONCLUSION: This method unveil psychosocial symptoms and measures the need of psychosocial support in patients with complex congenital heart disease and should have implications in clinical work. A manual for the method is presented as an appendix.

P195

Dental health in children prior to cardiac surgery

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Children with congenital heart disease have an increased risk of infective endocarditis from bacteraemia induced by invasive dental procedures or poor dental hygiene. The aim of this study was to record the dental health behaviour and experiences of parents and children requiring open heart surgery. Factors associated with the presence of dental disease were also investigated. Children attending cardiac surgical clinic over 2 consecutive years participated in the study. A dentist (J.C.) used a structured interview with both parents and children. The family's experience of dental services and dental behaviour were also recorded. Treatment required prior to surgery was noted. Out of 89 children, 17 were found to have untreated dental decay severe enough to place them at risk from bacteraemia. 10 children had concurrent evidence of dental abscesses. 53 Children (60%) had previously seen a dentist and the dentists were aware of the underlying cardiac condition in 41 (77%). 62 children brushed their teeth regularly but only 50% brushed their teeth twice daily. Only 11 children (12%) had additional fluoride supplementation. 28 children (45%) used a high fluoride toothpaste. Variables included in logistic regression model were dental attendance, parental awareness of endocarditis risk, previous dental treatment, brushing teeth and fluoride supplements. None of the variables reached significant values (p<0.05). The study emphasises the need for better education of parents and children, regarding the importance of better hygiene and dental attendance. Integration of dental services into the care of children with cardiac defects and collaboration with the cardiac team is essential.

P196

Primary prevention of coronary artery disease in children of czech republic

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Total cholesterol level is higher than mmol/l in 26% of school children in Czech Republic. The individual approach in coronary artery disease (CAD) prevension begins in early childhood by identifying children at risk. Pediatricians in Czech Republic are obliged to measure lipoprotein profile in children at the age of 5 and 13 years. The 7 year experience of the population approach in children oriented on a decrease in lipid levels especially in children with familial hypercholesterolemia and also on a decrease of other risk factors of CAD (hypertension, obesity, physical inactivity and smoking will be discussed.

P197

Efficiacy, safety and compliance of bile acid sequestrants in children with familial hypercholesterolaemia

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Heterozygous familial hypercholesterolemia (HeFH) is a common disorder (1/500), associated with an early coronary artery disease. In the vast majority of patients low saturated fat and cholesterol diet have only a small effect on reduction of cholesterol levels. The age at which the drug therapy should be started is controversial. The US NCEP recommended drug therapy should be considered if after appropriate diet remains LDL-C level higher than 4.9 mmol/l in boys over 10 years of age with family history of premature CAD. 27 boys with HeFH aged 10–12 years were treated by Colestipol 2x5g/d or Cholestyramin 2x4g/d. TC and LDL-C levels decreased significantly (p<0.001) by 21% and 23% after 3 years. There were no significant changes in HDL-C, TG, BMI, hepatic transaminases, ALP, routine hematology and thyroid function. Conclusion: This study in boys with HeFH has shown the cholesterol reducing effect without any adverse effects and with good compliance of bile acid sequestrants.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P198

Autonomic function in adolescent patients with orthostatic dysregulation according to heart rate variability (HRV)

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Many adolescent patients with orthostatic dysregulation have been thought to have autonomic dysfunction in Japan. But there is no report to clarify the relationship their symptom and autonomic dysfunction in adolescence. We assessed autonomic function in such patients and investigated the etiology of their symptom using heart rate variability (HRV). Methods: Subjects were 16 healthy controls (Controls) and 16 patients with symptom of headache, nausea, abdominal pain and vertigo at standing (OD). We performed 24-hour electrocardiography and calculated the subjects' autonomic function using a heart rate variability method (Analyzed software: MemCalc/CHIRAM). We also performed a time-domain analysis (SDNN, rMSSD, pNN50) and a frequency-domain analysis (LF, HF, LF/HF) and figure analysis (Triangular index, Lorenz plot). Results: Controls, OD: SDNN(ms) 194+/-142, 165+/-26.7 rMSSD(ms) 45.1+/-14.2, 53.0+/-21.2 pNN50(%) 21.6+/-9.27, 29.8+/-10.1 LF(ms2) 1960+/-492, 971+/-62.6 HF(ms2) 522+/-302, 630+/-477. LF/HF 2.04+/-0.663, 2.36+/-1.70*(p<0.05). Triangular index 23.7+/-10.6, 35.6+/-13.8* Conclusion: Patients with orthostatic dysregulation showed significantly higher sympathetic nerve activity (LF/HF ratio) and total variability of heart rate (Triangular index) than controls. These data showed hyperactivity and easily variable autonomic functions in patients with orthostatic dysregulation.

P199

Which sign is conspicuous for detection of atrial septal defect in Japanese screening system for school children?

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Objectives: To accurately detect atrial septal defect (ASD) in Japanese screening system of cardiac disease for school children using phonocardiogram (PCG) and electrocardiogram (ECG), findings of screened students were analyzed. Method: Sixty students were screened as suspicious ASD because of systolic murmur on PCG or incomplete right bundle branch block on ECG from 16 districts of Tokyo between 1996 and 1999. As final diagnosis reported from cardiologists, thirty-two were diagnosed as ASD and twenty-eight were not. Several signs on PCG and ECG were researched and compared between ASD group and non-ASD group. Welch's test and chi square test were used for statistical analysis and differences were considered to be significant with p values less than 0.01. Result: Concerning to PCG, averaged Q-I time were 121.3 and 105.9msec, the variance of width were 1.70 and 3.81, in ASD group and non-ASD group respectively, and both differences were significant. Concerning to ECG, averaged height of R wave in V1 were 0.196mV and 0.094mV, higher R' than R was seen in 66.7% and 25.0%, notch phenomena in inferior leads were seen in 87.5% and 42.9%, and abnormal progressions of T wave in precordial leads were seen in 62.5% and 3.6%, respectively. All of these comparisons had significant difference. Conclusion: Signs on PCG were non-specific for detecting ASD except Q-I time and variance of width in second sound. ECG shows more signs than PCG, such as notch phenomenon with high sensitivity or abnormal T progression with high specificity. However, PCG has the possibility to disclose other cardiac disease than ASD. It is necessary to interpret both instruments for screening cardiac disease in school children.

P200

Echocardiographic evaluation of wheelchair-bound basketball players with paraplegia

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A marked detriment in cardiopulmonary function has been shown in sedentary men whose lower limbs has been immobilized for years. the cardiopulmonary function of paraplegics who regularly activate their upper limps and trunk has been suggested to be near normal in a few studies. The purpose of this study was to evaluate the left ventricular dimensions, left ventricular mass, systolic and diastolic functions in paraplegic basketball players by echocardiography. The study group consisted of 11 paraplegic male basketball players who regularly play basketball for at least 2 years (on a high school basketball team) and the control group consisted of 11 healthy male adolescents of similar age and weight. The study group were all paraplegic subjects. The median age of study and control group were 17 (15–20) and 17 (14–18) years respectively. Also, the median body weight of study and control group were 46 (40–65) and 55 (42–65) kg, respectively. There were no significant differences in left ventricular dimensions and wall thickness, left ventricular ejection fraction, shortening fraction, aortic root, left atrium diameters, left ventricular filling characteristics between study group and control group ($p>0.05$). Previous reported studies state a significant reduction in cardiopulmonary function in sedentary paraplegic individuals. So, with the combination of our results and previous studies, we can conclude that cardiac functions in paraplegics can be improved to the normal levels by activating upper limbs and trunk regularly like playing basketball.

P201

Autonomic function by Tilt test in patients with orthostatic dysregulation: usefulness of monitoring heart rate variability (HRV)

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In adolescence, there are so many students with symptom which deemed autonomic dysfunction. But mechanism of this autonomic dysfunction is not clear. In Japan, this condition is called as orthostatic dysregulation. Their symptom is occurred at standing position. So we tried to clarify the activity of autonomic function by use of heart rate variability method in Tilt test (passive standing). Methods: All population (16 healthy controls and 16 OD) underwent continuous electrocardiographic monitoring while Tilt test. They were in supine position for twenty minutes and head by Tilt table until eighty degree and continued their passive standing position for twenty minutes and were again in supine position for ten minutes. Holter tapes were analyzed with a MARS 8000 analyzer (Marquette, Milwaukee, USA). After arrhythmia analysis, we studied heart rate variability. We analyzed their autonomic function every five minutes in Tilt test. We also performed time-domain analysis (SDNN, rMSSD, pNN50) and frequency domain analysis (LF, HF, LF/HF). Results: Time-domain analysis: In two groups, there were no significant difference in SDNN and rMSSD and pNN50. Frequency-domain analysis: In two groups, there were no significant difference in LF and HF and LF/HF. But there was strong variability in OD group. Conclusion: There is strong variability in autonomic function in patients with orthostatic dysregulation. This instability of autonomic function is etiology of symptom at standing position in OD.

P202

EQUIPP your heart. The educational quest for understanding in promotion and prevention for your heart

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Hypercholesterolemia is a major risk factor for atherosclerosis. It is evident that the atherosclerotic process begins in the young and is related to known cardiovascular risk factors: a large epidemiologic study, The Bogalusa Heart Study, has shown fatty streaks and fibrous plaques in the coronary arteries and aorta of children. Early management and primary prevention of coronary heart disease in children is supported by the fact that a health related diet and lifestyle, along with behavioral patterns, do affect a child's lipoprotein levels. The Hospital For Sick Children, Toronto, manages a Familial Hypercholesterolemia clinic of more than 250 children. We emphasize that a heart healthy lifestyle is a family affair. An important aspect of the child/family management of care is a EQUIPP class which uses group dynamics and is an efficient and effective use of the dietitian and nurses' time. EQUIPP is a program of primary prevention, focusing on heart and cholesterol function, discussion about controllable risk factors, nutritional goals, dietary fats, the 'more often - less often foods, activity, smoking status. The patient/family are instructed in the use of the Clinic self-evaluation tools, i.e. Food Frequency Checklist, Activity Questionnaire and the Report Card of lipid profile results. EQUIPP classes promote family life style changes with age appropriate recognition of control and responsibility. It addresses issues of the prevalence of the sedentary life style and obesity.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Surgery – Research and New Techniques, Prosthetic Materials, Surgical Management and Results: Innovative/Experimental Surgery

P203

Corrosive behavior of amplatzer nitinol devices in vitro and in a biological environment

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Purpose: to evaluate the long term corrosive behavior of the alloy in vitro and in vivo in biologic environment Methods: The follow samples were subjected to electron microscopy from 50 to 5000 times magnification. Sixty-four devices that were randomly selected from one hundred ninety Amplatzer Atrial septal defect (ASD) devices, which were exposed to saline solution of 37C for thirteen months. Two muscular ventral septal defect occluder were explanted from canine hearts after fifteen months. An unopened dacron graft that was implanted into a dog was explanted and studied after two years. An ASD device from a patient who died of other causes was studied in the same manner after eighteen months of implantation. An ASD device from a heart transplantation patient who had received the device implantation for 18 months. Results gross examination and 500,5000,5500 times magnification were used In vitro study, no gross evidence of corrosion or wire fractures Magnification was also showed an intact titanium oxide layer indistinguishable from control. Animal and human Studies: gross pathologic examination reveal the grafts to be covered by a thin layer of neointima. Electron microscope examination after cleaning revealed a typical intact titanium layer on the surface of both devices without evidence of corrosion or wire fractures. However, at 5,000 times magnification

P204

Surgical treatment of anomalous origin of the left coronary artery from the pulmonary artery

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Anomalous origin of the left coronary artery from the pulmonary artery is a very rare congenital heart defect. From 1991 Aug – 2000 Aug, 9 cases procedure was performed in 7 patients, coronary artery bypass grafting in 1 patient A new surgical method was used in a 6-year-old boy Mitral valve incompetence was repaired in 2 patients 8 Patients were followed up, altogether follow-up 41 1 months, mean 46 8 ± 9.7 months All patients are alive and in NYHA class I One patient because the suture of aortapulmonary fistula leak, aorta-pulmonary artery leakage happened and he was re-operated successful. Mean LVMD (46 1 ± 3/mm) significant descent compared to pre-operation (64 1 ± 2 8); (P<0 01) EF (69 4 ± 2 3%) significant raise compared to pre-operation (59.6 ± 2 8%) (P<0 05). We conclude that a two-coronary system appears more physiological and concomitant pathology may be corrected simultaneously The new procedure has not been described before in the literature.

P205

Modified rastelli procedure for double outlet right ventricle with left-malposition of the great arteries: report of 10 cases

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Background A Double outlet right ventricle with l-malposition of the great arteries is a very rare type of double outlet right ventricles This article will report our experience Methods: Between September 1995 and May 1999, 10 patients with double outlet right ventricle underwent a modified Rastelli procedure. Nine of them are DORV (S, D, L), and one is DORV (I, L, D). All but one patient had stenosis of pulmonary valve or subpulmonary stenosis. The location of VSD is subpulmonary or remote from both great arteries. The modified Rastelli procedure was performed under cardiopulmonary bypass. Right ventriculotomy was made to repair the VSD with Teflon patch or artificial blood vessel patch. An intercrural tunnel was made between the left ventricle and the aorta The main pulmonary artery was divided and the proximal end was closed. Using allograft conduit an external valved conduit was reconstructed between the inlet of the right ventricle main pulmonary artery. Results: All patients survived and recovered uneventfully Echocardiography showed that the internal tunnels and valved and external conduits performed well. The results were excellent Conclusions The Modified Rastelli procedure is a satisfactory method for treatment of DORV with left malposition of the great arteries. It can completely correct the stenosis of pulmonary outflow tract and right to left shunt, and avoid injuring the right coronary artery

P206

Pulmonary blood distribution after total cavopulmonary connection (TCPC) of different types

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Objective To assess the feature of pulmonary blood distribution after TCPC of different types Methods 23 survival patients after TCPC of four different types underwent radionuclide lung perfusion According to the radionuclide counts in left and right lungs to analysis the blood distribution from superior venous cava (SVC) and inferior venous cava (IVC) and the whole pulmonary blood flow to both lungs. Results when the anastomosis of IVC shift to left, the flow ratio of the IVC to left lung was greater than that to the right, p<0.01, the flow ratios of the SVC to right lung was greater than that to the left, p<0 01, and the whole pulmonary blood flow go dominantly to left lung, p<0.05 When the anastomosis of IVC and SVC directly opposite each other on the right pulmonary artery (RPA), the flows from the SVC and IVC were mixed and turn toward both lungs evenly in half, the whole pulmonary blood flow go to both lungs, p>0.05 When the IVC anastomosis shift toward the RPA with widening anastomosis, the flow ratios of the SVC go to both lungs in half, p>0.05, and major part from IVC go to right, P<0.01, the whole blood flow go dominantly to right lung, p<0.05 For patients after TCPC with bilateral bi-directional cavopulmonary connection the flows from right SVC go to right lung by 100%, and that from left SVC go to left lung by 100%, the flows from IVC dominantly to left lung and little part to right lung Conclusion: Different designs of TCPA can result in different pulmonary blood distribution, the best flow distribution between the left and right lungs can be obtained for an offset of the IVC anastomosis toward the RPA with widening anastomosis for the patients

P207

Shallow sutures close to the posteroinferior rim of the perimembranous ventricular septal defect avoid damage to the right bundle branch and the his bundle

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Referring to the three portion theory of the right bundle branch (RBB), which had been proposed by Lev, we introduced a novel surgical technique into surgery of the perimembranous ventricular septal defect (VSD) and analyzed the prevalence of the postoperative conduction disturbance. Methods: Three autopsy specimens having experimentally created VSD were subjected to the study of the conduction system. From March 1996 through April 2000, 42 consecutive patients with VSD who underwent surgery using the novel suture technique, in which sutures were placed shallow and close (< 4 mm) to the rim (group 1). Fifty-eight consecutive patients, who had previously undergone surgery using the conventional technique with sutures being placed remote (3–5 mm) from the rim, were subjected to the comparative study (group 2). Postoperative electrocardiogram was reviewed to investigate whether the novel suture technique was superior to the conventional one in preserving the function of the conduction system. Results: The second portion of the RBB penetrated through the septum with the third portion beyond the ventricular septum 5 to 6 mm inferior to the rim. Suturing sites at the inferior rim of group 1 corresponded to the myocardium lying on top of the second portion and were free from danger; whereas those of group 2 neighbored the third portion and were in proximal danger of damaging RBB. Persistence of the complete right bundle branch block and left axis deviation were significantly less in group 1 as compared with that in group 2 (p<0.0001 and p<0.005, respectively). Atrioventricular block was noted in neither of the groups. Conclusion: The novel suture technique for surgery of the perimembranous VSD was superior to the conventional one in preserving function of RBB and the His bundle

P208

Avoiding homologous blood transfusion ameliorates postoperative lung oxygenation in pediatric open heart operation

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https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

OBJECTIVE: We examined the beneficial effects of avoiding transfusion in the lung function during the pediatric open heart operations using cardiopulmonary bypass (CPB). METHODS: Study 1: 46 patients who underwent ventricular septal defect closure were divided into (a) a control group (n=22) in whom homologous blood was transfused, and (b) a leukocyte removal (LR-1) group (n=24) in whom a leukocyte removal filter was used during and post operation. Study 2: 32 ventricular septal defect patients were divided into (a) a non-blood transfusion (NB) group (n=14) consisting of patients in whom homologous blood was not transfused, and (b) a LR-2 group (n=18) consisting of patients in whom it was used with the filter during and post operation. An arterial blood gas analysis was carried out several times and the respiratory index (RI) was calculated. RESULTS: Study 1: RI immediately after CPB was significantly lower in the LR-1 group than in the control group (2.23+-0.22 vs. 3.90+-0.68, p<0.05) but was not thereafter. Study 2: RI immediately after CPB did not differ between the NB and LR-2 groups, but RIs 3 and 6 hours after the operation were significantly lower in the NB than in the LR-2 group: (1.43+-0.09 vs. 1.92+-0.16 and 1.36+-0.08 vs. 1.91+-0.15, p<0.01). CONCLUSIONS: These results suggest that avoiding transfusion of whole homologous blood elements could work more effectively to prevent lung dysfunction after CPB.

P209

Availability of cavopulmonary bypass using a centrifugal pump without a membranous oxygenator in the right heart bypass operation.

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Although a temporal bypass between the caval vein and the atrium has been introduced in a right heart bypass operation to prevent disadvantages for the postoperative Fontan circulation due to cardiopulmonary bypass, an elevated pressure of the drained caval vein would be concerned to bring down congestive organ damages. We performed the right heart bypass operation under the support by a cavopulmonary bypass using a centrifugal pump without a membranous oxygenator for three patients who had a functional univentricular heart. The two patients underwent a bidirectional cavopulmonary shunt and the third patient underwent a total cavopulmonary connection with an extracardiac conduit. The mean pressure of the drained caval vein during the bypass with a flow of 80 – 100 ml/kg/min was maintained around less than 10 mmHg with stable hemodynamics and sufficient systemic oxygen saturation. As the pseudo-right heart bypass circulation had been established during surgery, postoperative hemodynamics were stable in all patients, who had weaned from mechanical ventilation without any deterioration of respiratory function or any signs representative of congestive organ damages early after surgery. In conclusion, a temporal cavopulmonary bypass which could be set up only by a minimal dissection for the procedure, using a centrifugal pump might be one of beneficial techniques at a right heart bypass operation without intracardiac repairs.

P210

Dorsal mini incision and q-clip extrapleural dissection for pda clip closure in premature neonates.

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To present a new minimally invasive surgical technique for extrapleural closure of the ductus arteriosus (PDA). From January/96 through November/2000, 30 consecutive premature neonates were operated on. Patients were positioned semi-prone with the left hemithorax raised 45 degrees upward by a soft roll. Through a 1.5 cm dorsal incision, the auriculoscapular triangle fascia and the intercostal space were entered, and PDA extrapleural dissection with q-tips was performed. Isolation of the ductus arteriosus end was completed with sharp scissors and the PDA was clipped. Thoracotomy closure was performed without pleural drainage. Gestational age (weeks), patients age (days) and weight (g), and operative time (min) were, respectively (mean ± standard deviation): 27 ± 4, 20 ± 10, 980 ± 212 and 36 ± 18. One pt. (3.3%) required reoperation for residual shunt early in the series. No other surgical complication occurred. Hospital mortality was 6.6% (enterocolitis - 1 pt., sepsis - 1 pt.). Q-tip PDA extrapleural dissection was easy to perform, resulting in no lung lacerations. As the PDA is approached at a right angle relative to its long axis and at the shortest possible distance through the dorsal skin incision, very good exposure is obtained, leading to expeditious and secure clip closure of the PDA.

P211

Simple one patch method for the surgical repair of atrioventricular septal defect

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Generally patch closure has been performed for atrial septation of atrioventricular septal defect. Recently we evolved a new surgical technique for atrioventricular septal defect to avoid the use of any atrial septal patch material. We report our experience with this technique. Methods: Seven patients (complete type: partial type2) underwent this technique. The diameters of atrial septal defect were measured by transesophagus echocardiography. The electrocardiograms before surgery were compared with those after surgery. Results: The diameters ranged from 3 to 9mm. There was no early death and one late death with severe pneumonia. The comparison of electrocardiograms before and after surgery showed no significant difference. Significant valve regurgitation and residual shunt were not detected by postoperative echocardiography. Conclusions: This single one patch method simplifies the repair of atrioventricular septal defect. In short-term results, this method does not cause arrhythmia nor valve regurgitation.

P212

Congenital tracheal stenosis and heart defects: one-stage repair

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We reviewed our experience with one-stage repair of congenital heart defects and long-segment tracheal stenosis. Four children, mean age 22 months (8 months – 3 years), mean weight 8.4 kg (5.3 – 12.0 kg), with preoperative mechanical ventilation up to 3 months, underwent one-stage cardiac and tracheal surgery. Diagnosis: dextrocardia, atrial septal defect, persistent left superior vena cava (PLSVC) (1), pulmonary artery sling (1), ventricular septal defect (VSD) (1), double outlet right ventricle with VSD, pulmonary atresia, patent ductus arteriosus and PLSVC in left auricular appendage (1). Tracheal stenosis with circular rings extended to 80% tracheal length in 2 children, 75% in one and 60% in one, internal tracheal diameter was 2 mm in one, 3 mm in two and 4 mm in one. Slide tracheoplasty and intracardiac repair in one case requiring a right ventricle to pulmonary artery conduit, was performed with a single period of cardiopulmonary bypass, mean duration of 365' (range 145–185'). There were no early or late deaths. Mean ICU stay was 6.5 days (range 4–11 days), with extubation after a mean period of 54 hours (range 48–72 hours). Mean hospital discharge was 16 days (range 15–18 days). Endoscopy showed 4.4 mm (3.0–5.0 mm) mean increase of internal tracheal diameter, equivalent to mean 160% of preoperative size (75–250%). Endoscopic excision of endoluminal tissue was required twice in one child and once in two. All children remain well, without medication and respiratory problems, with a mean follow up of 26 months (range 15–60 months). One-stage repair of congenital heart defects and slide tracheoplasty for long-segment tracheal stenosis is feasible and provides adequate treatment of both cardiac and tracheal problems.

P213

A new technique for the management of anomalous left coronary artery from the pulmonary artery

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Objectives: Several operative approaches are utilized for the management of anomalous origin of the left coronary artery from the pulmonary artery, each with some limitation. A new technique that facilitates direct and tension free implantation of the anomalous artery in the aorta is described. Methods: From 1/1/92–6/30/01 ten consecutive patients with anomalous left coronary artery underwent operation using this new technique. It consists of isolating an anterior and posterior transverse segment of pulmonary artery in continuity with the origin of the anomalous coronary artery. The two segments are folded with the orifice of the coronary at its fulcrum and the edges sutured together to form an extension cone of pulmonary tissue. This lengthens the coronary artery and allows direct aortic implantation (posterior to the pulmonary artery) without tension. The pulmonary artery is reconstructed with autologous pericardium. Results: Patient age ranged from 3 weeks to 3 years (mean 35 weeks), with 80% less than 11 weeks. Average weight was 7.7 kgm (3.7–21 kgm). The left ventricle was dilated with an end diastolic diameter z-value of 1.1 to +3, and the LV shortening fraction was markedly reduced to 16±6%.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

(7–28%), with 8/10 patients having a shortening fraction less than 20%. Mitral regurgitation was severe in 5, moderate in 2, and 3 patients were in CHF. Post repair, there were no hospital deaths. Inotropic support was needed in all patients, but none required mechanical assistance. At a follow up of 3.8±2.3 years (0.5–7 years) nine patients are asymptomatic and 1 has intermittent chest pain. All patients (10/10) have echocardiographic documented patency of the reimplanted coronary artery, as well as marked improvement in the LV shortening fraction (37±5%)* and decrease in the end diastolic diameter (z-value +1 to +1.5)*. Mitral regurgitation was absent in 4, mild in 4, and moderate in 2. Four pts. have mild supravalvar pulmonary stenosis (15–32 mmHg). Conclusion: This new technique allows a tension free direct aortic connection, has a low rate of occlusion and avoids pulmonary artery dissection and stenosis making it a viable alternative for this lesion. *p>0.05 vs group

P214

Selection of infants with tetralogy of Fallot for surgical repair: is it justified now?

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Is there enough evidence in favor of the selective carrie for surgical repair of tetralogy of Fallot (TF) in infants? We have subjected to the retrospective analysis records of 124 consecutive patients who were admitted to the Bakoulev Center over the period between 01.01.97– 01.07.00 aged 0.2 – 12 months (median 7.5). 12 pts required ventilation prior to surgery. All were dramatically limited in their physical activities due to severe hypoxia. 19 were on beta blockers for longer than 3 months. 64 pts (52%) had undergone angiography. 2 pts had coronary artery anomalies. 2 others had PA-valve agenesis syndrome; pts with true PA atresia were not included. 52 pts benefited from urgent Gore-Tex shunts indicated by the severity of their state. The same palliation was performed on 4 pts for marked hypoplasia of pulmonary branches (Nakata index < 120 mm2/m2). There was no mortality in the palliation subgroup. One hundred pts underwent repair of TF including 32 pts with previously performed shunts with no cases of procedure-caused PA distortion registered. In 96 pts we used transatrial-transpulmonary approach, traditional technique in was used in 4. One pt suffering from Di George syndrome died shortly after the surgery (mortality 1%). During 3–41 months follow up period no one died or required reoperation. Two-staged repair in infants with TF is still justified, more so in the communities where TF is diagnosed relatively late with ensuing severe arterial hypoxemia and deteriorating general state of the pts. Our experience suggests that selection of these pts for one or two-staged repair can yield excellent results.

P215

Atrioventricular groove patch plasty for anatomically corrected malposition of the great arteries

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Objective: In anatomically corrected malposition of the great arteries concordance-discordance i.e. dextroposition of the posterior pulmonary artery and levoposition of the anterior aorta, causes the leftward deviation of the proximal portion of right coronary artery away from the right atrioventricular groove, associated with levoposed anterior aorta. This anatomical feature allows a transannular subpulmonary incision of the right ventricular outflow tract into the right atrioventricular groove between the right coronary artery and the tricuspid anterior anulus for relief of subpulmonary stenosis without jeopardizing the right coronary artery. Method: This report describes the mid-term results of a new surgical technique atrioventricular groove patch plasty with a autopericardial transannular patch for subpulmonary stenosis in 3 patients with anatomically corrected malposition of the great arteries by utilizing the aforementioned morphological advantages of 'concordant criss cross', with a concomitant closure of ventricular septal defects. Results: There was no operative or late death. Preoperative catheterization revealed adequate relief of pulmonary stenosis with a pressure gradient of 8.0±3.5mmHg and with normalized right ventricular pressure (33±10 mmHg), contributing to excellent mid-term results with no late death and reoperation during postoperative follow-up period of 60±47 months. Conclusion: This technique provides a promising alternative to Rastelli type conduit repair for subpulmonary stenosis in anatomically corrected malposition of the great arteries.

P216

A new surgical technique for one-stage repair of interrupted aortic arch with valvular aortic stenosis

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We developed a new operative technique for reconstructing aortic arch of neonatal type B interruption of the aortic arch (IAA) without the use of autologous or prosthetic material. Case report: The diagnosis of the neonate was type B IAA, hypoplasia of AAo with normal great arterial relation, PDA, subarterial VSD, and valvular AS. The main PA and AAo were transected at the level of pulmonary bifurcation. PA bifurcation was translocated anterior to the AAo and main PA stump. The inferior half of the descending aorta (DAo) orifice was anastomosed to the posterior half of the main PA orifice. The posterior half of the distal AAo orifice was directly anastomosed on the superior half of the DAo orifice. The proximal AAo stump was anastomosed to the right posterolateral wall of the main PA. Aortic arch reconstruction was accomplished by a direct end-to-end anastomosis between the anterior aspect of distal AAo stump and the main PA stump. After intraventricular repairing through the ventriculotomy, continuity between the RV and PA bifurcation was established using an autologous pericardial roll equipped with tricuspid Gore-Tex valve. Postoperative three-dimensional helical CT demonstrated non-obstructed smooth aortic arch. Doppler echocardiography showed laminar flow through the distal DKOT routes and the aortic arch. Conclusion: A suitable combination of the great arterial stump provides a wide neo-aortic arch with laminar flow. This technique can be adapted to most cases of IAA.

P217

New surgical technique for creation of inter-atrial septal defect, an animal study

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Background: In some of the conginital heart diseases such as transposition of great arteries, mitral atresia and tricuspid atresia an adequate inter-atrial communication is essential for patient survival. Inter-atrial balloon septostomy is now frequently performed for these patients early in infancy. Blade septostomy versus surgical septectomy may be suggested for older children due to thickness of the septum. Because of lack of facilities for blade septostomy in many centers and difficulties with the present methods of surgical approach, we are now presenting a new experimental technique for surgical inter-atrial septostomy which we found is a simple and safe method with high success and low complication rate. Method: 10 mixbreed dogs were chosen for the study. They were properly anesthetized under blood pressure and ECG monitoring and right thoracotomy was performed. After brief dissection of inter-atrial groove a purse string was placed at the junction of both aorta and auricle prepared. Two small wholes were made on both sides of the inter-atrial septum (right and left atrial sides) inside the purse string. The 2 limbs of a specialized Metzenbaum scissors were placed in the two wholes and septostomy was performed by 2 cuts in 2 right angled directions while the purse controlled bleeding. Pre and post operative echocardiography and autopsy studies were performed. Results: No complication or mortality happened. Creation of appropriate size defect in the septum was confirmed both by echocardiographic evaluation and autopsy observation of the cases. Conclusion: Although in most of the cases inter-atrial septostomy can be safely done with current methods in selected cases this method of septostomy can be a very good alternative.

P218

Practical, technical improvements in the construction of the modified Blalock-Taussig anastomosis

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Authors describe technical aspects of modified Blalock-Taussig anastomosis and propose few practical improvements of it's performance. Proposed modifications are based on the personal surgical experience of 98 shunts constructed in 96 patients over the period from January 1992 through September 2000 and the results of the simulations on the transparent models as well as computer simulation of such an anastomoses. Authors present description of few technical improvements as well as analysis of results in comparison to control group.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P219

Correction of atrial communications with submammary minimal thoracotomy

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The purpose of this paper is to show the posibility of surgical correction, with extracorporeal circulation (EC), in atrial pathology leaving the scar concealed under the growing breast in young females with the submammary minimal thoracotomy technic (SMT). Fourty two female patients (p) between 18 months to 18 years (x 8 years) weithing 10–54 Kgs (x 12 kgs) were operated. A 3 to 6 cm semicircular incision is made over the 6th rib (1st and 2nd. intercostal) or under the submammary wolves in the sexu. The thorax is entered at the level of 4th rib. A third of the anterior portion of the rib was resected in 15 p (35.5%) and not in the other 27p (64.5%). The EC was established cannulating the femoral artery and both caves vein. The time of follow up was from 1 month up to 5.5 years. The corrected pathology was: secundum ASD in 36 p (85.7%), ostium Primum in 2 (4.7%) sinus venosus ASD with partial anomalous pulmonary venous return in 3p (7.1%) scimitar syndrome in 1 (2.3%) all defects were repaired with no mortality. No hypertrophic scars were late. In conclusion the SMT permits the correction of pathology in the atrial and caval levels. The results are similar to the median sternal approach and minimizes the anesthetic scars and its psychological effects since they are concealed under the breast.

P220

Pedicled pericardium patch for RVOT and pa reconstruction in TOF

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Pericardial patches have been widely used for repair of the RVOT. But after using such "ideal" material as autologous pericard RVOT aneurysms still form 2% to 25% complications of the follow-up period. Today, when many centers have adopted early repair of TOF one more problem appeared. Even autopericardial patches don't grow with somatic growth of patient. Because patch can't receive a natural neuro- and blood supply it means that RVOT obstruction will be the reason for reoperation also. Numerous techniques were suggested for construct of pedicled pericardium patch to avoid reoperation. We tried a new one. The main task was to prevent just the stitches throughout non-tissue part of patch. For that purpose pericardial flap was raised before cardiopulmonary bypass began. This flap had a wide lower part and more narrow top part which kept contact with mother" pericardial sac. Then with two inclined incisions were formed two leaflets which were sewed together behind of top part. After intracardiac stage of operation was completed pedicled pericardium patch of appropriate size was implanted within RVOT or/and PA incision. In that way due to atypical mushroom – shaped for pedicled pericardium patch we preserved its natural blood supply as much as possible. We consider that our method of RVOT and PA reconstruction is useful especially in early infancy.

P221

Right ventricular outflow tract reconstruction using a PTFE pulmonary monocusp valve

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Background: Pulmonary regurgitation after transanular patch reconstruction of right ventricular outflow tract following total correction of Tetralogy of Fallot(TOF) has been a problem in the long term. Monocusp valves prepared from various materials have been fabricated & amplanted ensuring varied degrees of competence in the short term. Objective: The objective of this paper is to describe our experience with fabrication and construction of a 'monocusp valve' from PTFE pericardial membrane and the short term outcome. Methods: 29 patients with Tetralogy of Fallot and needing a Transanular Patch were chosen as the subjects of this study out of a total of 200 patients of Tetralogy of Fallot operated in our institution from May 1998 to November 2000. All the patients underwent intracardiac repair using moderate hypothermia and cold blood card oplegic arrest. VSD was closed transatrially in all except 5 patients where a transventricular route was adopted. All the patients except 2, did well post operatively. The two mortalities were due to severe low cardiac output immediately post operatively out of which one was due to a residual VSD. The patients who did well had a mean ICU stay of 2 days. Post op echocardiography revealed No Pulmonary regurgitation in 2 patients, mild to moderate PR in 22, and free PR in 3 patients. At follow up all the hospital survivors were alive. 24 patients in whom the monocusp was functioning well in the early postoperative period were asymptomatic and were not on any medications. The 3 patients in whom the monocusp was incompetent, decongestive medications were necessary. Conclusion: A monocusp valve fashioned from PTFE pericardial membrane is an inexpensive, and easily reproducible technique that restores reasonable valvular competence at the pulmonary level. It ameliorates and hastens the post-operative recovery in patients needing a transanular patch. Short term follow-up has not revealed any deleterious effects of the PTFE membrane, though long term results remain.

P222

Tricuspid leaflet detachment in transatrial congenital heart repairs – revisited

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Background: Temporary Detachment of Tricuspid Leaflets to enable transatrial repair of VSD, Tetralogy of Fallot and DORV has been described earlier. However, its use has diminished fearing resultant tricuspid regurgitation (TR). Objective: It is the aim of this paper to enforce on our advantages of detachment of the various leaflets of the Tricuspid Valve in unison or sequentially to obtain better visualization, improve the accuracy and security of the repair of lesions such as VSD, Tetralogy of Fallot and Double Outlet Right Ventricle via the right atrium. Methods: In the period between May 1998 to October 2000, 1500 congenital cases were operated at our center. Of these 204 were VSD's, 15% were Tetralogy of Fallot, and 1 (%) was Double Outlet Right Ventricle, to give a total of 575 patients. Of these, 225 patients had transatrial repair with temporary detachment of tricuspid leaflets. They form the subject of this report. 75(%) of these had part of the ATL (Anterior Tricuspid leaflet) detached, 24% had part or whole of the STL (Septal Tricuspid leaflet) detached, and 1(%) had part or whole of the PTL (Posterior Tricuspid leaflet) detached. ATL detachment was predominantly done in patients with Tetralogy, DORV and perimembranous VSD with malalignment. STL detachment in patients with inlet VSD, and PTL detachment in patients with posterior muscular VSD. In all the patients the detached leaflets were reattached once the repair was completed, and the valve tested for competence. Results: No Tricuspid regurgitation or compromised valve motion was noted by echocardiography postoperatively in any of these patients. Conclusion: Temporary detachment of Tricuspid leaflets for transatrial repair of the lesions alluded to above is a safe technique without resultant TR. It improves the exposure of the angle between conal septum, atrial annulus and Ventriculo infundibular(VI) fold where sutures could be taken safely and securely, thus reducing the incidence of residual VSD in this area. This is particularly so in adults or those with extreme atrial dextroposition with VI fold hypertrophy. The extreme retraction of the Tricuspid valve leaflets needed is obviated thus protecting the subvalvar apparatus.

P223

Limited posterior thoracotomy in the repair of atrial septal defects

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Cosmetically appealing incisions and approaches are being increasingly used for closure of Atrial septal defects. Anterolateral thoracotomy has been described as an approach, but as a prepubertal girl the incision scar could fall on the ipsilateral breast as the developing breast line is ill defined. Hence, another approach – the posterior approach is being suggested, which circumvents the problem.

P224

The usefulness of sensory and motor evoked potential monitoring for operation of mild coarctation of the aorta to avoid the spinal cord ischemia

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Paraplegia induced from spinal cord ischemia demands one of the most serious consideration of aortic surgery. But there is no sure monitoring method for aortic operation to avoid the spinal cord ischemic injury, especially for children. We tried to monitor a sensory evoked potential (SEP) and motor evoked potential (MEP) through the subclavian flap coarctectomy operation in case of mild coarctation of the aorta without major collateral arteries, these cases were thought to be a high risk group of paraplegia. In the almost cases, except one, no remarkable changes of SEP and MEP were

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

detected through the operation. In one case, serious changes of SEP and MEP were recorded through the operation. The patient was 4-month-old boy who was diagnosed as coarctation of the aorta with ventricular septal defect. The pressure gradient of the aortic coarctation was 40 mmHg. The aortography showed poor collateral arteries. In this case, the MEP amplitude decreased after 10 minutes from aortic clamp, and SEP also disappeared right away. After 25 minutes from aortic clamp, aortic unclamp and reperfusion of descending aorta were performed. Soon after aortic unclamp, the MEP and SEP began to be detected. The MEP and SEP amplitude recovered to the initial levels after 12 minutes and 18 minutes from aortic unclamp. In this case, neurological deficiency was not detected after operation. Ischemic spinal cord injury occurs before we are aware of it under the operation in cases of mild coarctation of aorta. The evoked potential monitoring for children is useful to know the spinal cord function through the operation. In case of disappearance of evoked potential we have to reperfuse distal blood flow as soon as possible to avoide paraplegia after operation.

P225

Successful excision of a rare benign intracardiac teratoma – case report

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An 11-month-old boy with no previous medical problems was admitted to hospital in heart failure due to sudden sepsis (Staphylococcus aureus), endocarditis and ventricular arrhythmia, while a term uneventfully created. ECHO showed a huge mass in the right ventricular cavity indicating a cystic teratoma. Three weeks later the boy was submitted to surgery. A cystic tumor (6 x 4.5cm) was excised from the right ventricular cavity. The tumor originated from the interventricular septum but it was also enough adherent to the apex and parietal to the right ventricular free wall. The papillary muscles of the tricuspid valve were partially involved in the tumor mass. The child recovered uneventfully and pathologic examination revealed a mature cystic teratoma. The operating procedure, pre- and postoperative ECHO studies, pathologic gross and microscopic findings, four-year follow up and a review of the literature are presented.

P226

Limited posterior thoracotomy for open heart surgery

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There is a progressive enthusiasm recently on minimally invasive techniques for accessing the heart. We present our technique of correction of congenital heart defects employing the limited posterior thoracotomy approach. Since June 1997 to Nov 2000, 92 patients underwent correction for various intracardiac defects. Eighty-three patients were females and 9 were males. The median age was 7 years and the median weight was 20 kgs. There were 69 patients secundum defects with or without other associated anomalies. There were 12 sinus venosus defects with partial anomalous pulmonary venous connection. 3 patients had perimembranous ventricular septal defects while 5 patients had partial atrioventricular defects. In 2 other patients, pulmonary stenosis was repaired, using pulmonary valvotomy in 1 patient, whereas the other patient required sharp transannular patch, one patient had mitral valve replacement. All the patients were extubated within 12 hours following surgery and the median ICU stay was 24 hours. The mean chest drainage was 82 cc at 24 hours and 7 patients required blood transfusions in the ICU for significant blood loss. None of the patients had phrenic nerve palsy. No patient required additional analgesics other than routine analgesia. Short and mid term follow up revealed no functional or physical disability of the thoracic wall, the right arm and the right breast. All patients who underwent surgery with this approach were happy with the limited visibility of their scars. Limited posterior thoracotomy offers a viable alternative for midsternotomy and submammary thoracotomy. It has the advantage of a scar in the back that does not impede the future growth of the breast tissue and the pectoralis major. Our approach does not need any new instruments. Our short and mid term results are good with better cosmesis.

P227

Closure of muscular VSD by a sandwiching method via a preexisting larger VSD or an interatrial septostomy

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Results of closure of muscular VSD (mVSD) by a sandwiching method well in 9 patients are summarized and technical details are demonstrated on video. Patient's age ranged from 0.3–9 (mean 4.6) years. Muscular VSDs were closed along with the repair of other complex cardiac anomalies in 5 patients and perimembranous VSD in 3. Operative technique: A right-angled forceps are introduced through the pre-existing perimembranous(5) or muscular outlet(1) VSD or through an interatrial septostomy(3) into the left ventricle (LV), and the interventricular septum is pressed gently from both left and right ventricular (RV) sides to locate the openings in the septum. Once the opening is located, guiding rubber catheter is passed through the mVSD. An oversized stiff Dacron felt mounted on a 3–0 Neupolen suture is connected to the rubber and pulled into the LV, gently pulling the suture length through the mVSD toward the RV side. The suture ends are then passed through a similar Dacron felt on the RV side of the septum. The Neupolen suture is then tied firmly, thereby sandwiching the septum between the 2 stiff Dacron felt patches. Results: Mean sizes of mVSD, Dacron felt patch on LV side and that of RV side of 12 mVSDs were closed by this technique were 5.3+/-0.6, 14.8+/-0.5 and 11.7+/-0.4 mm, respectively. All of the patients survived operation well and no significant left to right shunt was noticed on postoperative evaluation but for 1 patient who had a swiss-cheese type mVSD. In this patient, a residual left to right shunt of Qp/Qs 2.3 at 1 month decreased to Qp/Qs 1.4 at 16 months after operation. Conclusion: 1) This technique is simple and less invasive without the need of elaborative procedures or left ventriculotomy. 2) In case, minor residual shunt remained in the immediate postoperative period, it is expected to disappear in long-term.

P228

Pulmonary artery line placement via the right atrium: a safe approach in congenital heart operations

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Purpose: Pulmonary artery (PA) lines can provide important monitoring information following congenital heart operations. PA lines are typically placed in the operating room, through the right ventricular free wall, and are removed in the intensive care unit. However, ventricular bleeding and tamponade can complicate the removal of such lines. The aim of this study is to evaluate the safety and effectiveness of an alternative approach, which avoids ventricular puncture: placement via the right atrium. Methods: From January 1999–October 2000, 23 patients undergoing congenital heart operations had PA lines placed via the right atrium. PA lines were placed in patients in whom postoperative PA hypertension was anticipated. Lines were placed in the operating room, across the right atrial free wall and tricuspid valve. The results of this approach were retrospectively reviewed. Results: Median patient age was 3.5 months (range 20 days–9.7 years), median weight was 4.2 kg (range 2.0–19.2 kg). Operations were: TAPVC repair (4), VSD closure (4), DORV repair (3), heart transplant (3), ALCAPA repair (2), PA unifocalization (2), and other (5). All lines functioned well for determination of PA pressures, PA oxygen saturations, and subpulmonary ventricular outflow tract gradients (measured during line withdrawal). PA lines were removed in the intensive care unit after a median postoperative duration of 3.8 days (range 1–7 days). No patient had bleeding requiring transfusion, or tamponade at line removal. There were no cases of line entrapment. Conclusion: PA line placement via the right atrium avoids ventricular puncture with its attendant bleeding risk. This is a safe and effective approach to PA line placement in patients undergoing congenital heart operations.

P229

Less invasive aesthetic approach for atrial septal defects surgery: the right posterolateral thoracotomy

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All incision or antero-lateral approaches for open heart surgery carry some aesthetic prejudice or uncertainty (specially in young female). We evaluate here a particulary aesthetic, safe less invasive approach, through a short right postero-lateral thoracotomy that could be extended on individual basis for other defects. Since July 1995, 38 patients underwent ASD closure through this approach. Mean age was 9 years (0.5 to 52, median = 6, SD = 13). The third (n=5), the fourth (n=30) or the fifth intercostal space (n=3) was entered after a 7 to 12 cm skin incision posterior to the mid axillary line. CPB was established with ascending aorta cannulation (n=37, femoral cannulation in 1) and two angled venous cannulae. Aortic crossclamping, anterograde cardioplegia and de-airing were conducted as usual. Diagnosis were 47 ASD secundum

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

(4 low ASDs), 1 sinus venosus, 6 Ostium primum(1reoperation), 2 prematurity-borne VSDs, 1 right coronary-RV fistula and 1 mitral replacement. There was no major intraoperative problem. Mean aortic cross-clamping time was 26 mn (14 to 52, SD=11). There was no post-operative mortality, but one hemothorax needing reentry. Mean post-operative hospital stay was 7.6 days (SD=2,6). Follow up is uneventful. As long as great care is given to achieve good exposure, with a short technical learning, the procedure appears safe and reproducible. Cosmetic results are excellent, with no scar on the anterior aspect of the chest. When percutaneous closure is not feasible, surgery through this very aesthetic posterior approach appears as a good option.

P230

Bloodless open heart surgery in neonates and infants

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Since November 1989 four Jehovah's Witness children less than 1 year old, weighing 4 - 8kg, had corrective heart surgery without the use of blood products in the operating room or during their post-operative course. Operations included Norwood Stage 1 procedure (1), repair of complete atrio-ventricular canal (1) and bi-directional Glenn procedure (2). Successful bloodless heart surgery requires a coordinated strategy for the pre-operative, intra-operative, and post-operative care. A key element has been our minimalization of the cardiopulmonary bypass circuit to a priming volume of 180cc. Using a hemodilute prime this microcircuit yields post dilutional hematocrits within an acceptable range even for newborns undergoing complex heart surgery. Other strategies include the intra operative use of aprotinin and a cell-saver as well as the pre- and post-operative use of erythropoietin.

P231

Ministernotomy for the closure of subarterial ventricular septal defect associated with mild aortic regurgitation

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OBJECTIVE: To minimize cosmetic or surgically invasive problems, ministernotomy has been introduced to pediatric cardiac surgery. Recently we have applied ministernotomy for the closure of subarterial ventricular septal defect (VSD) associated with mild aortic regurgitation (AR). METHODS: Between July 1998 and November 2000, seven alone 14 patients with supracristal VSD with mild AR in age between 6 months and 15 years and in body weight (BW) between 7 and 38 Kg, underwent VSD patch closure through a lower sternal split incision using a 5 to 9 cm skin incision. In patients above 22 Kg of BW, a reversed J-shape incision at the left third intercostal space was added. Cardiopulmonary bypass was established by aortic and bicaval cannulation. Antegrade cardioplegic arrest was achieved. The pulmonary trunk was opened horizontally and VSD was closed through the incision using an oval Hemashield patch. RESULTS: There was no mortality. Extracorporeal and aortic cross-clamping times were 102+/-32 and 63+/-24 minutes, respectively. All but two infants were extubated in the operating room and were not transfused. Although AR was not repaired, AR disappeared in 11 patients and decreased to trivial in 2 patients after VSD closure. Pulmonary regurgitation or stenosis were not detected. All school age patients went to school or kindergarden within 2 weeks after surgery. CONCLUSION: Ministernotomy for the closure of subarterial VSD associated with mild AR is technically feasible and may provide better outcome with respect to cosmetics and less surgical invasion than full sternotomy.

P232

Patch augmentation and chorda reconstruction of left atrioventricular valve in complete atrioventricular septal defect (cAVSD) - A case report

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Scarcity or deficiency of leaflet tissue may compromise satisfactory repair of the atrioventricular valve (AVV) in a small number of patients with cAVSD, especially in patients with normal karyotype. In such a situation, the insertion of a valve prosthesis is an unsuitable solution for infants. We experienced an early infant with deficiency of valve tissue of left AVV and this was successfully repair by the augmentation of leaflet and reconstruction of chorda using single bovine pericardium. A one-month-old boy with normal karyotype and congestive heart failure continued since the neonatal period, was diagnosed to have cAVSD. AVV regurgitation was noted after birth. Echocardiography showed severe AVV regurgitation, Rastelli type A anatomy, and small VSD. Operation was performed using standard cardiopulmonary bypass with systemic hypothermia. VSD was located under superior bridging leaflet and was closed directly. Left side valve leaflets, particularly superior bridging and lateral leaflets, were hypoplastic. After closing ASD with autologous pericardium, the defect of the cleft

P233

Adjustable pulmonary artery banding: alternatives for rapid pulmonary ventricle prepare

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Objective: Two models of adjustable devices for pulmonary artery (PA) banding were applied in young goats, in order to assess and compare each method of pulmonary ventricle hypertrophy induced by a progressive systolic pressure load training program. Methods: Three groups of seven animals each were used, as follows. Group I, PA banding was achieved by a balloon catheter. Group II, an extravascular hydraulic cuff banding device was applied, and Group III was the control group for septum, left and right ventricles weights. All the goats from group I and II were submitted to progressive systolic pressure load imposed by banding device adjustments at 24-hour interval, during a 96 hour period. The behavior of the right ventricle (RV) muscle mass was assessed by echocardiogram and morphologically. Results: Right ventricle to pulmonary artery pressure gradient, RV to LV ratio, and RV systolic pressure were significantly higher in group II ($p<0.05$). A significant increase in the RV wall thickness was observed in groups I and II. RV dry weight was higher in groups I and II ($p<0.05$), as compared to control group. Myocyte perimeter and cross-area showed a significant increase after the 96-hour training period. Conclusions: Progressive systolic pressure load training program was able to induce a significant degree of pulmonary ventricle hypertrophy in a 96-hour period, regardless the PA banding device used. Such a program may eventually be a useful tool for RV preparation to the Jatene operation beyond the neonatal period, and even for the failed atrial baffle procedures in patients with transposition of the great arteries.

P234

Transsiphenid approach without sternotomy for the repair of ostium secundum atrial septal defects

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Objective: Analyze the results achieved by repair of ostium secundum atrial septal defects through the transsyphenid approach without opening the sternum. Method: It was a longitudinal and prospective study of 35 patients operated from July 96 to July 99. Ages ranged from 6 months to 34 years, with a median age of 5.1 ± 3.1 years. By a longitudinal median skin incision of 5 cm at level of the xiphoid, total resection of the appendix was performed, and an especially retractor was positioned in order to expose the right atrium. A normothermic cardiopulmonary bypass was performed by left femoral artery and bicaval cannulation. Aorta was unclamped and crystalloid cardioplegic solution was administered anterogradely. Atrial septal defect was closed through the right atrium by running suture or with a pericardium patch. After closure of the right atrium, air is exhaustively removed from the left cavities and bypass is interrupted. Results: Average time of bypass was of 33.8 ± 11.3 minutes and clamping 20.7 ± 9.6 minutes. There was no intra-operative complications. Reversal to a median sternotomy was required in one case for repair of an anomalous drainage of pulmonary veins. In 22 (62.9%) patients extubation was performed in the operating room. Average time of stay at the ICU and hospital stay was of 19.9 ± 15 hours and 3.8 ± 1.89 days respectively. One patient presented pericardium effusion and superficial infection of the scar in two. In a median follow-up of 21 ± 10 months, all patients are in functional class I (NYHA), without medication or residual defect. Two (5.7%) patients presented less than 20% left femoral arterial stenosis by Doppler. Conclusion: This technique is feasible, safe, with excellent cosmetic results.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P235

Biocompatibility of microdomain structural extracorporeal circuits in infants

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<Purpose> Cardiopulmonary bypass (CPB) induces nonspecific systemic inflammatory reactions in infants. The purpose of this study was to examine the biocompatibility of microdomain structural extracorporeal circuits in infants. <Material> Twenty-one VSD patients who underwent elective cardiac surgery were randomly divided into three groups. group C, conventional circuit and oxygenator group (n=7), group M; microdomain structural circuit and heparin bonded oxygenator group (n=7), and group H, heparin bonded circuit and heparin bonded oxygenator group (n=7). <Methods> Measurements of blood cell count, fibrinogen, AT-III, D-dimer, bradykinin and complement system were made before CPB, after 5 min of CPB, just after CPB, 2 hrs after CPB and 24 hrs after CPB. <Results> There were significant differences for group M in platelet reduction ($p<0.05$) and AT-III reduction ($p<0.05$) between group M and group C or H. But there were no significant differences in fibrinogen reduction and D-dimer production. There was tended to be less bradykinin generation in group M ($p=0.06$ vs group C and $p=0.06$ vs group H). There was tended to be less C3a activation in group M ($p=0.08$ vs group C and $p=0.08$ vs group H). <Conclusions> microdomain structural circuit could reduce early systemic inflammatory reaction compared with heparin bonded circuits and non-bonded circuits.

P236

Our experience of surgical atrial septal defect closure without cardiopulmonary bypass

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As usually the correction of atrial septal defect (ASD) is performed under conditions of cardiopulmonary bypass by medial sternotomy. This approach is traumatic and leads to more complications than others. The purpose of the study is to review our experience of surgical correction of ASD by minimal invasive technique without cardiopulmonary bypass. We performed surgery in 79 patients in age from 3 to 48 years, 28 were male and 45 were female. Secondary ASD was in 66, partial A-V canal in 1 and ASD with partial anomalous right pulmonary venous connection in 6 patients. The protection of the body was made by general hypothermia with decreasing of rectal temperature till 29–27 C. General hypothermia was made with covering the body with the small pieces of ice, after giving anesthesia. The active hypothermia was stopped after reaching rectal temperature 32 C, then continuing hypothermia of the head to obtain the required temperature. The approach to the heart was performed by anterior minithoracotomy in 4th right intercostal space. We performed longitudinal pericardiotomy anterior to the diafragmal nerve. The stepwise occlusion of both vena cava and aorta ascendens was carried out. In 43 cases the ASD was plasticed by autopericardium and 32 ones by sewing. Air embolism was prevented by aorta ascendens puncture and defining the blood flow outside. The occlusion time of the magistral vessels during sewing ASD was from 3 to 13 minits with temperature ranges 29 C and during the plasty was from 10 to 25 min. with temperature lower 28 C. Defibrillation was use in 38 cases. The was observed hypoxic encephalopathy in 4 patients that pressure for 3 days. Artificial ventilation was during over 4 to 20 hours. Other complications were not observed. Patients stayed in the hospital for 4 to 10 days. Conclusions: Generally hypothermia protection of the body complete with minitoracotomy from right side without CPB elaborated by us release to perform closure of ASD without dangerous and it have all positive kinds of the minimasive technology in the cardiosurgery (economical, commercial).

P237

Pedicled autologous pericardial flap method of right ventricular outflow tract reconstruction using new sutureless technique

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Background, Extracardiac conduit stenosis should be one of major obstacles for long term freedom from reoperation after Rastelli type operation. We have repeated pedicled autologous pericardial flap method of Right ventricular outflow tract reconstruction (RVOTR) for it and its efficacy by preliminary animal experiment which revealed living tissue layer on its lumen and possible growth of flapped area in the late phase. We have recently had two successful cases who underwent corrective surgery with this method using new sutureless technique. Method, As Rastelli type operation, extracardiac conduit was created with ePTFE monocusp valve attached to distal graft for posterior wall side and pedicled autologous pericardial flap for anterior side, which were fastened at distal end of its conduit. Flap base remained intact just at the distal end of conduit so that blood supply was preserved. We applied this technique for two clinical cases which were 2 years girl of VSD with PA and 6 years boy of congenital AS. Concomitantly, we performed experimental study which simulated this technique on anterior wall side of pulmonary trunk in mongrel dogs to examine morphological and histological change. Results: In our experimental study, autologous pericardial flap method using sutureless technique suggested of Lectin positive endothelial layer which was examined with immunohistochemistry as living tissue in its lumen and growth in the flapped area. There was no clinical events in postoperative period (6–24 months). No deformity or stenosis of RVOTR revealed. The mean pressure ratio of RV to LV measured 0.38 at one year after operation in 2 patients. Thus method could be effective procedure for Rastelli type operation in terms of growth and freedom from reoperation, and may improve prognosis.

P238

The hemodynamic change of RV-PA shunt in modified Norwood procedure for HLHS and its equivalent hearts

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The blood flow pattern and the influence of the anastomotic stenosis after the modified Norwood procedure with RV-PA shunt was analyzed using the 2DE and pulse Doppler echocardiography. Seven neonates and infants (HLHS 6, DILV+AS 1), weighing 1.7 to 3.7Kg, were followed up since January 1998. The size of the PTFE graft was 4mm in 2 and 5mm in the remaining 5. SaO2 was analyzed concomitantly. The investigation was conducted at 2 or 4 weeks(immediate), 1–5 to 2 months(midterm) and 3 to 6 months(prior to the second stage bidirectional Glenn anastomosis) after the operation respectively. Graft flow pattern consisted of the systolic forward flow(FF) and the diastolic reverse flow(RF). The ratio of the velocity-time integral(VTI) of RF and FF(RFV/FFV) decreased along the time course in all cases. The stenosis developed after the anastomosis at the distal anastomosis. SaO2 concomitantly decreased after 1.5 to 2 months. (1)Both the changes of RFV/FFV and SaO2 were inversely correlated with the change of the pressure gradient. (2)The rate of the decrease in RFV/FFV was consistent with the increase in the peak gradient. (3)RFV/FFV ratio shifted to below 0.30 after 90 days of age irrespective of the severity of the stenosis, suggesting the predominant influence of the reduction of the pulmonary resistance. (4)The change of the gradient had the stronger influence than RFV/FFV on the decrease in SaO2. Both the reduction of the pulmonary vascular resistance and the valve-like mechanism of the distal stenosis played a role in the change in RFV/FFV ratio with the more strong influence of the former after 90 days of age. As a conclusion, this hemodynamic study implicated the rationale of the second stage BDG after 90 days of age with or without the development of the stenosis.

P239

The effects of pretreatment with FK506 on the neuropathological changes in the brain of neonatal piglets undergoing cardiopulmonary bypass (CPB) with deep hypothermic circulatory arrest (DHCA)

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Introduction. Brain injury and altered psychomotor development still occur following corrective cardiac surgery in early life. The immunosupressive drug FK506 (Tacrolimus) involves inhibition of calcineurin in T-lymphocytes by a complex of FK506 and an FK506 binding protein. Recent studies have suggested a protective effect of FK506 against ischemia in neuronal cells. We evaluated the effect of pretreatment with FK506 on neuronal cell injury in neonatal piglets undergoing CPB with a prolonged DHCA period. Methods: 15 neonatal piglets (age < 10 days, weight: 2.1 +/- 0.5 kg BW) were included in this study. Ten animals without pharmacological intervention were served as control group. Five animals were pretreated i.v. with FK506 (0.2mg/kg BW) five hours preoperatively. All animals were anaesthetized, intubated and

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

mechanically ventilated. After median sternotomy the animals were connected to CPB by cannulation of the aorta and right atrium. Full flow CPB (200ml/kg/min) was initiated for homogeneous systemic cooling. Circulatory arrest for 120 min was induced when rectal temperature of 14 °C was achieved. After rewarmed reperfusion the animals were weaned from CPB and monitored for 6–8 hours. Then the animals were sacrificed and the brain was immediately removed, cut in standardized sections and fixated for further histological studies. Neuronal cells were counted in sector CA 1 – CA 4 and dentate gyrus of hippocampus formation in respect to apoptosis and hypoxic necrosis. Results: The main preliminary findings in this brain ischemic model were the quantitative evaluated differences in necrotic and apoptotic neuronal cell injury according to pretreatment with FK506. A reduction of necrotic neuronal cell changes in hippocampus sector CA1 – CA4 was found in the group of FK 506 treated animals. In the dentate gyrus the mode of neuronal cell injury changed from necrosis to apoptosis. Conclusion: The application of FK506 seems to have protective effects on the neuronal cell necrosis but not on the apoptosis in the hippocampus formations. The reduction of neuronal cell necrosis and the increase of apoptotic cell events may suggest possible protective actions. Further studies however, are necessary to evaluate the role of apoptosis in the brain after deep hypothermic ischemia.

P240

Anatomical repair of complete complete atrioventricular septal defect

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Background: A better understanding of the morphology of complete atrioventricular septal defects (CAVSD) has impacted surgical techniques greatly. Competency of the left atrioventricular valve play an important role in the outcome of repair of these defects. On some occasions the leaflet tissue is deficient and repair becomes difficult. We present a modified technique for the repair of this defect. Material and Method: The repair is performed on standard cardiopulmonary bypass with cardioplegic arrest. After evaluation of the anatomy, both superior and inferior leaflets are divided coronarily to completely expose the VSD. The 0.4 mm thin Gores VSD patch is sutured to the right side of the ventricular septum leaving 3 mm of patch above the annular line to augment the divided leaflet tissue of the left A-V valve. The autologous pericardial atrial septal patch is attached 3 mm from this suture line, thereby allowing 3 mm of the VSD patch to augment the left AV valve. These 3 mm allow better coaptation of the leaflets thereby improving the competency of the valve. Results: This technique was performed on 14 infants with CAVSD. Mean age was 7 months. There were no deaths among these infants. There was no significant postoperative left AV valve regurgitation or residual VSD. By echocardiography performed postoperatively, the contribution of patch augmented left AV valve to competency is clearly seen. The right AV valve is apically offset as in normal subjects. Conclusion: The use of this modified technique yields good anatomical repair and helps in cases with deficient left AV valve tissue.

P241

Neuromonitoring and CPB strategies for avoiding Neurological Injury after Hypothermic Circulatory Arrest in a Survival Piglet Model

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Background: Widely different protocols have been used to achieve hypothermic circulatory arrest (HCA) with considerable ongoing controversy regarding optimal pH strategy, optimal hematocrit and optimal temperature. Near-infrared spectroscopy (NIRS) is a relatively new technique for assessment of cerebral oxygenation. We studied the interaction of pH strategy, hematocrit, temperature and duration of HCA and their combined impact on cerebral oxygenation and neurological outcome in a survival piglet model including monitoring by NIRS. Methods: Seventy-two piglets (9.29 ± 0.14kg) underwent HCA under varying conditions with continuous monitoring by NIRS (hematocrit 20 or 30%, temperature 15 or 25°C, pH-stat or alpha-stat strategy; HCA time 60, 90 or 100 minutes). Neurological recovery (NR) was evaluated daily by a veterinarian and the brain was fixed in situ on POD 4 to be examined by histological score (HS) in a blinded fashion. Results: Cerebral oxygenation indicated by Tissue Oxygenation Index was associated with difference of pH strategy ($p < 0.001$), temperature ($p = 0.03$) and hematocrit ($p = 0.06$) at the end of cooling. Oxygenated hemoglobin signal declined to a plateau (nadir) during circulatory arrest. Time to nadir was significantly shorter with lower hematocrit, higher temperature and alpha-stat strategy, $p < 0.008$. Duration from reaching nadir until reperfusion

P242

Two-staged balloon dilatable pulmonary artery banding

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A dilatable pulmonary artery banding (DPAB) may be useful for temporary palliation in neonates or for long palliation in infants with complex lesions. In lab work a banding tied with a 5/0 prolene mattress stitch and a second 5/0 prolene stitch was banded with different sizes of balloons and pressures. In swine a DPAB was performed and 3 months later it was dilated with balloons 1,5 to 2,1 times larger than the bandings and pressures of 4 to 8 atm. Three children aged 7 days to 5 months old, with muscular VSD, aortic coarctation or interrupted aortic arch were operated and a DPAB was performed. An average of 18 months later the DPAB were dilated with 10 or 12 mm balloons with pressures of 3,5 to 12 atm. The 5/0 prolene stitch was bursted but never 5/0 stitch in all instances. In all animals and thereafter in children dilation was safe and effective. The gradient across the banding decreased from 83 to 31 mmHg in average. In 1 case the banding was left open and the child does not need further procedures; the other 2 wait for a definitive repair. This simple technique permits a programmed dilation for a definitive opening or a staged enlargement of the banding.

P243

Evaluation of Amplatzer devices by cardioscopic assessment

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Purpose: To assess dynamic and anatomic function compatibility of implanted cardiac devices using a new ex vivo technique with an endoscope (cardioscope). Methods: Seven out of forty one animals who underwent testing of Amplatzer cardiac devices had a final ex vivo cardioscopic evaluation 2–38 months after device placement. Of the seven animals four had a ventricular septal defect closure (3 perimembranous, 1 muscular) and three had patent foramen ovale closure. Ultrasound and angiography evaluation suggested atrioventricular valve insufficiency and aneurysmal expansion in three animals. The animals were anesthetized and ECG, blood pressure, and blood gases were obtained. The heart was explored and infused with cardioplegic solution. The heart was explanted. Cannulas and tubing were attached for circulating a clear oxygenated perfusion solution. Using the Langendorff technique, hemodynamics and ECG were adjusted to simulate in vivo values. The cardioscope was introduced into various cardiac chambers for real time imaging. After cardioscopy the hearts were submitted for pathologic evaluation. Results: In vivo and ex vivo ECG and pressures were similar ($p = ns$). The heart beat spontaneously in a Langendorff mode until the rhythm stabilized and became self-sustaining. With our cardioscope one could obtain clear detailed 360 degree visualization of the interior of the beating heart for functional dynamic evaluation of valves, chordae tendineae, cardiac chambers, and implanted devices. The etiologies for valve insufficiency and vegetations were clearly demonstrated. Conclusions: This new technique provides an accurate clear look at the interior of a beating heart and functional and anatomic assessments of implanted cardiac devices. Cardioscopy Ex vivo, is a valuable assessment procedure for the study of and optimization of implanted cardiac devices.

P244

The role of oxidative stress in the development of pulmonary arteriovenous malformations following cavopulmonary anastomosis

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OBJECTIVE: Cavopulmonary anastomosis (CPA) is often used for palliation of cyanotic heart disease. Clinically significant pulmonary arteriovenous malformations (PAVM) can occur in up to 29% of patients following surgery. CPA causes several modifications to the pulmonary circulation that may contribute to PAVM development. Our objective was to examine the role of one such alteration, reduced pulmonary blood flow (PBF), in PAVM formation by studying angiogenic and stress-related gene expression following pulmonary artery banding (PAB) and CPA. METHODS: Lambs aged 35 to 45 days were placed into three groups: CPA (n=6), PAB (n=4), and sham

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

controls (n=6). In our model, PAVMs are detectable by bubble-contrast echocardiography 8 weeks following CPA. To examine genes involved in PAVM development, tissue was harvested at 2 and 5 weeks after surgery. Expression of angiogenic and stress-related genes was determined by Western blot and scanning densitometry. RESULTS: CPA and PAB both increased angiogenic gene expression, but only CPA influenced the expression of endothelial stress-related genes. Vascular endothelial growth factor (VEGF) was upregulated 2.5 fold following both CPA (p=0.002) and PAB (p=0.007). However, CPA alone upregulated two markers of oxidative stress, heme oxygenase-1 and glucose transporter 1, 2 fold (p=0.004) and 5 fold (p=0.003), respectively. PAB failed to induce expression of either protein. Expression of CD62, a marker of endothelial activation, was also unchanged following PAB, but increased 4 fold (p=0.008) following CPA. CONCLUSIONS: Reduced PBF induces a pulmonary angiogenic response, but not an endothelial stress response. These results suggest that oxidative stress is more relevant to PAVM formation than angiogenic signaling, as PA banding does not result in PAVMs. The chronic oxidative stress of the pulmonary endothelium resulting from cavopulmonary anastomosis may predispose the affected vasculature to arteriovenous shunting.

P245

Bovine-albumin-glutaraldehyde surgical adhesive impairs growth and causes strictures at aortic anastomoses in neonatal piglets

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Purpose: Bleeding complications remain a significant cause of mortality and morbidity in patients undergoing congenital heart surgery. A new surgical adhesive composed of bovine albumin and glutaraldehyde is currently under clinical investigation in adults as an adjunct for securing hemostasis at cardiovascular anastomoses. Interference with vessel growth, however, would preclude its use during congenital heart surgery. The purpose of this study was to determine if reinforcement of aortic anastomoses with bovine-albumin-glutaraldehyde surgical adhesive (BAG-SA) impairs vessel growth and causes stricture using a neonatal piglet model. Methods: Ten 4-week old piglets (8.0 ± 1.4 kg) underwent primary aorto-aortic anastomoses with interrupted polypropylene sutures after baseline aortic measurements were obtained. Following aortography, 5 piglets were randomly assigned to anastomotic reinforcement with BAG-SA. After a 7-week growth period, aortography was repeated and the aortas were excised for morphometric analysis and histopathology. Results: After 7 weeks, mean weight gains were similar in BAG-SA animals (24.5 ± 5.1 kg) and control animals (22.0 ± 4.0 kg, p = 0.41). In BAG-SA animals, however, aortic circumference increased only 1.5 ± 1.1 mm (vs. 3.2 ± 1.4 mm in controls, p = 0.035) and aortic luminal diameter increased only 0.9 ± 0.9 mm (vs. 2.5 ± 0.6 mm in controls; p = 0.004). Strictures producing stenosis exceeding 25% developed in 4/5 BAG-SA animals (80%) vs. 0/5 control animals (p = 0.047). Aortic histopathology revealed adventitial changes - macrophages, micropyogranulomas, giant cells, and moderately increased connective tissue - in all 5 BAG-SA animals vs. none of the control animals (p = 0.007). Conclusions: Reinforcement with BAG-SA impairs vascular growth and causes stricture when applied circumferentially around an aorto-aortic anastomosis. This adhesive should not be used on cardiovascular anastomoses in pediatric patients.

P246

Bovine-albumin-glutaraldehyde surgical adhesive causes acute phrenic nerve injury and paralysis of the diaphragm in young pigs

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Purpose: A new surgical adhesive composed of bovine albumin and glutaraldehyde is currently under clinical investigation as an aquitet for securing hemostasis at cardiovascular anastomoses. The use of this adhesive to reinforce anastomoses of the aorta, vena cavae, and pulmonary arteries places the nearby phrenic nerves at risk for injury. Phrenic nerve injury with diaphragmatic paralysis complicating congenital heart operations is associated with increased morbidity and mortality, especially in neonates. The purpose of this study was to determine if bovine-albumin-glutaraldehyde surgical adhesive (BAG-SA) causes acute phrenic nerve injury. Methods: Via median sternotomy in 12 young domestic pigs (age 10–15 wks, weight 32 ± 4.8 kg), baseline diaphragmatic excursion was measured using cinefluoroscopy during direct phrenic nerve stimulation. Diaphragmatic excursion was remeasured 5 and 30 min after exposing the nerve to BAG-SA (n = 6) bovine albumin (negative control, n = 3), or glutaraldehyde (positive control, n = 3). Results: All animals exposed to glutaraldehyde had complete diaphragmatic paralysis at 5 min; diaphragmatic paralysis did not occur in any of the albumin exposed animals. The mean diaphragmatic excursion in the BAG-SA group was lower than in the albumin group both 5 min (1.7 ± 3.6 mm vs. 18.7 ± 11.1 mm respectively, p = 0.0068) and 30 min after exposure (0.33 ± 0.8 mm vs. 18.7 ± 10.1 mm, respectively, p = 0.002). Five of 6 animals (83%) exposed to BAG-SA had complete diaphragmatic paralysis by 30 min (p = 0.047 vs. albumin). Conclusions: Bovine-albumin-glutaraldehyde surgical adhesive causes acute phrenic nerve injury with diaphragmatic paralysis. When using this adhesive, contact with nerves must be avoided. Further study is needed to delineate the degree and duration of impaired nerve function and to evaluate possible neuroprotective strategies.

P247

Topical bovine-albumin-glutaraldehyde surgical adhesive causes sinoatrial nodal degeneration and persistent bradycardia in young pigs

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Objective: Bleeding complications remain an important cause of mortality and morbidity during congenital heart surgery. A new surgical adhesive composed of 10% glutaraldehyde and 25% bovine albumin is currently being used to reinforce suture lines and improve hemostasis. The superficial location of the cardiac conduction system makes it vulnerable to acute injury, particularly during congenital heart surgery. The purpose of this study was to determine if bovine-albumin-glutaraldehyde surgical adhesive (BAG-SA) penetrates through the epicardium and causes injury to the underlying sinoatrial node. Methods: Eleven young domestic pigs (age 10–15 wks, weight 35.2 ± 5.6 kg) underwent median sternotomy. After obtaining baseline electrocardiograms, BAG-SA was applied to a 2 x 2 cm area at the coronary junction overlying the sinoatrial node. Electrocardiograms were obtained at 15-minute intervals for 1 hour. The heart was then excised for histopathological examination. Results: Histopathology revealed coagulation necrosis extending through the entire epicardium and into the myocardium in all 11 animals; ubiquitous hocytes were nuclear pyknosis, cytoplasmic eosinophilia, and contraction band changes. Two animals (18%) also had focal degeneration involving the sinoatrial node. In contrast to the other 9 animals, both pigs with nodal degeneration developed bradycardia after application of the adhesive, the bradycardia persisted for the entire 60 minutes (see figure). Conclusions: When applied to the surface of the heart, BAG-SA uniformly causes severe coagulation necrosis that extends into the myocardium and can involve underlying conduction tissue. Application near the sinoatrial node can cause nodal degeneration and persistent bradycardia. When used during cardiac surgery, this adhesive should not be applied near the cardiac conduction system. Further studies are required to delineate the duration of injury and evaluate potential protective strategies.

P248

Bilateral Superior Vena Cavae are not a Risk Factor for Single Ventricle Palliation

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Background: Concern has been raised regarding the presence of bilateral superior vena cavae as a risk factor for single ventricle palliation. We have not observed this in our clinical practice. Methods: 84 patients have undergone bilateral CPA between 1/92 and 11/00. Median age at operation was 7.6 months (range 1 mo – 22 yr). Median weight at operation was 6.6 kg (range 3.5 – 100 kg). The most common diagnoses were heterotaxy and tricuspid atresia. Cavopulmonary shunts were often performed without cardiopulmonary bypass (CPB). CPB was utilized in patients who required additional surgical procedures. Cannulation of the superior vena cavae was avoided whenever possible. Anti platelet therapy was utilized in all cases. Results: Takedown of bilateral CPA was required in one patient (1.3%). This patient died early (n=1, 1.9%) due to sepsis after takedown and common AV valve replacement. There were 6 late deaths (11%): 2 due to bronchopneumonia, 1 due to pulmonary arteriovenous malformations, 1 due to pulmonary venous obstruction and 1 after Fontan operation. Thrombosis in the cavopulmonary circulation was not seen in any case. 20 patients have had subsequent Fontan operation.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

One patient had one and one-half ventricle repair. Conclusions: Bilateral CPA can be performed in patients with minimal morbidity and mortality. Bilateral CPA is not associated with thrombus formation in our experience. The presence of bilateral superior vena cavae does not signify increased risk for single ventricle palliation.

P249

Evaluation of pulmonary blood flow distribution after corrective surgery for Tetralogy of Fallot

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Abnormal pulmonary blood flow (PBF) distribution was sometimes seen before and after surgery in patients with Tetralogy of Fallot(TOF). The purpose of this study is to evaluate the PBF distribution and its clinical implications after corrective surgery for TOF. Quantitative lung perfusion scintigraphies (LPS) with 99mTc MAA were performed in 51 patients. Abnormal PBF distribution was defined as left lung receiving less than 30 % or more than 60 % of the total PBF. The patients were divided into two groups. Group A included 23 patients who had abnormal PBF distribution. Group B included 28 patients who had normal PBF distribution. Eight patients in group A and 6 patients in group B had pulmonary atresia (NS). Previous shunts were performed in 19 patients in group A and 14 in group B ($p<0.05$). Pulmonary angioplasty was performed at the time of corrective surgery in 16 patients in group A and 11 in group B ($p<0.05$). Peripheral pulmonary artery stenosis was morphologically showed after surgery in 14 patients in group A and 2 in group B ($p<0.01$). Postoperative catheterization was performed in 18 in group A and 6 in group B. Systolic main PA pressure was 35.2 +/- 11.1 mmHg (mean +/- SD) in group A and 33.3 +/- 11.1 mmHg in group B (NS). Five patients in group A and 2 patients in group B had the systolic main PA pressure of more than 40 mmHg. Postoperative balloon angioplasty was performed in 10 patients in group A and 1 in group B. Eleven patients in group A had natural improvement of PBF distribution on the second postoperative LPS. It is concluded that: the patients with abnormal PBF distribution after corrective surgery had high incidence of morphological peripheral pulmonary artery stenosis. However, systolic main PA pressure was not elevated and the abnormal PBF distribution would not have deleterious influence.

P250

Intrapulmonary reconstructions of the pulmonary arteries in a dog model.

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Severe congenital or acquired malformations of the intrapericardial pulmonary arteries remain a serious risk for correction of congenital heart defects. In some of these patients the intraparenchymatous pulmonary arteries are reasonably developed. The objective of the experimental work was to elaborate a method of a reconstruction of the pulmonary arteries by means of a conduit – neopulmonary artery – connected with intrapulmonary branches of the pulmonary arteries. In 11 dogs (age 1 – 4 years, weight 10 – 45 kg) a total of 15 operations was performed. The intrapulmonary arteries were approached through postero-lateral thoracotomy and interlobar fissure. The conduits were created from different materials: native pericardium, xenopericardium (porcine pericardium – Polystan) or vascular prosthesis (expanded polytetrafluoroethylene – GoreTex). The distal portion of the conduit was sutured end to side to the lower lobe pulmonary artery, the proximal one end to side to the pulmonary trunk. The relevant intrapericardial branch of the pulmonary artery was ligated and pressure in the conduit was measured. Unilateral pulmonary reconstruction was created in seven and bilateral in 4 animals. Two dogs with unilateral reconstructions died 11 and 13 days after surgery. The patency of the conduits was confirmed by autopsy. Angiographic examination performed in 9 animals surviving two weeks after operation revealed patency of all conduits with regular pulmonary distribution. Postoperative pressures in the conduits remained unchanged compared with preoperative state (< 18 mm Hg) in all 4 animals with bilateral reconstructions. In conclusion, intrapericardial pulmonary arteries can be fully replaced by conduits connected to the intrapulmonary arteries. It is possible to create the conduit from the native pericardium, porcine pericardium or vascular prosthesis (GoreTex).

P251

An experience of a patient with HLHS undergoing a bidirectional cavopulmonary shunt after the modified Van Praagh operation

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A 11-day-old boy with the diagnosis of hypoplastic left heart syndrome without hypoplastic aortic arch or coarctation of the aorta, which is a very rare combination, underwent the modified Van Praagh operation which consisted of bypass grafting between the main pulmonary artery and the brachiocephalic artery associated with bilateral pulmonary bandings, to avoid disturbances for pulmonary circulation and reoperation following cardiopulmonary bypass. A balloon atrioseptostomy was performed 8 days later. The dimensions of the ascending aorta, aortic arch and left coronary artery by postoperative echocardiography or angiocardiography developed from 5.2 mm, 4.2 mm and 1.2 mm to 8.9 mm, 5.8 mm and 2.9 mm, respectively, 5 months after the operation. At the age of 6 months, the patient underwent a bidirectional cavopulmonary shunt with the Darnia-Kay-Stansel anastomosis and the DeVega annuloplasty for tricuspid regurgitation. His postoperative course was uncomplicated. Postoperative management after the modified Van Praagh operation was easier compared to that after the Norwood operation. The ascending aorta and aortic arch developed sufficiently. We conclude that the modified Van Praagh operation, which has a possibility of development of the hypoplastic aortic segment and coronary system before arch reconstruction, might be the treatment of choice as an alternative stage-I palliation for hypoplastic left heart syndrome.

P252

A vad in the right circulation of patients with univentricular hearts : an animal model.

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Background : In the long-term, patients with univentricular hearts and high pulmonary artery pressures are doomed to either death or heart-lung transplantation. The same fate is reserved to patients with failing total cavopulmonary connection. Implantable assist devices offer a new hope for these patients. Methods : Five piglets weighting a mean of 33 ± 12 kgs were placed under cardio-pulmonary bypass. Univentricular hearts were obtained by ligating the pulmonary artery and resecting 2 leaflets of the tricuspid valve and the muscular septum. A MEDOS HIA assist device was interposed between both vena cavae and the main pulmonary artery. Results : The animals were maintained under right Ventricular Assist Device for a median of 4 hours (range : 3–6 hours). Conclusion : Definitive implantation of a Ventricular Assist Device in the right circulation of patients with univentricular hearts might become an alternative to heart-lung transplantation in those with elevated pulmonary artery pressures or with failing cavo-pulmonary connections. An animal model has been developed to study this possibility.

P253

Redirection of hepatic venous drainage resolves pulmonary arteriovenous malformations in patients after total cavopulmonary anastomosis

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Background. The development of pulmonary arteriovenous malformations (PAVMs) is a known complication of Total Cavopulmonary Anastomoses (TCCA) wherein hepatic veins are excluded from the pulmonary circulation. They are considered analogous to those associated with liver disease, which are known to resolve after liver transplantation. Patients and Method: Five patients with TCA (Kawashima procedure) underwent re-direction of hepatic veins to the pulmonary circulation. All were profoundly desaturated (60–73% 73) and had clinical and angiographic evidence of pulmonary arterio-venous malformations. The TCA was performed between 7 and 57 months (27.6) of age. Cardiac catheterisation prior to the redirection of hepatic veins showed mean PA pressure of 15.6+/- 1.52. Age at Kawashima procedure was 7–57 months (27.6+/- 13.2). The age at redirection of the hepatic veins to the pulmonary circulation was 29–144 months (97.8+/- 32.66) and the mean interval between Kawashima and completion of Fontan was 70.2+/-26.15 months. A Gore-Tex tube was interposed between the hepatic veins and the pulmonary artery under cardiopulmonary bypass using a 16–18 mm tube conduit. Patients were followed up for a period of 12–26

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

months (18.8 +/- 4.24) The arterial saturation steadily improved to >95% in all but one patient (75%) who had the hepatic effluent preferentially going to the right lung. Conclusion: Redirection of hepatic venous drainage to the pulmonary circulation resolves pulmonary arterio-venous malformations in those patients palliated with Kawashima operation

P254
Congenital heart surgery with small submammary incision in girls
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Interest in minimally invasive procedures has recently increased. Especially in simple congenital heart disease, there is less surgical trauma, decreased patient discomfort, and better cosmetic appearance that are important. Based on these facts, we have been using the small submammary incision and partial sternotomy approach for correction of simple congenital heart defects in girls. From December 1997 to June 2000, small submammary incision within the bilateral mammary lines and partial sternotomy approach was performed in thirteen girls with ventricular septal defect or atrial septal defect. The average age of the patients was 6.5 years (range, 1 to 16 years) and the average weight was 25.0 kg (range, 7.8 to 57kg). In all but one patient, extracorporeal circulation was carried out by means of cannulation of the aorta and bicaval veins. The average aortic cross clamp time was 48.3 minutes (range, 35 to 77 minutes). Two patients who had atrial septal defect had ventricular fibrillation electrically induced. The average length of the hospital stay was 6.8 days (range, 5 to 12). No patients had blood transfusions and there were no operative or late deaths. There were two postoperative complications of subcutaneous fluid retention. The advantages of this modification include excellent cosmetic results in girls and smaller invasion of the mammary glands than the conventional submammary approach and concomitant security and full sternotomy when required.

P255
Nitrous oxide delivery during spontaneous breathing
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Objectives: The role of nitrous oxide (NO) in the treatment of pulmonary hypertension following surgical repair of congenital heart defects is well recognized and up to date has been delivered in our institution during mechanical ventilation. However there are patients such as post Glenn or Fontan procedure that could benefit from NO administration but are hemodynamically better on spontaneous breathing. A home made system has been recently developed to deliver NO through mask and used in two patients. Method: The system is composed of a 400 PPM tank connected to a double stage manometer (CONCOA, Virginia Beach, VA) coupled to an electronic flowmeter. The NO is measured by electrochemistry analyzer with a 0.1ppm precision (SensorMedics Critical Care Corporation, Yorba Linda CA). NO is inhaled through a ventimask with a reservoir without rebreathing (Hudson Respiratory Care Inc., Tecoma, CA) using continuous air/oxygen flow. A minimum required gas flow to maintain the reservoir inflated and to avoid an important NO2 formation is 10 liters/min. As an extubation system the child is placed under an oxygen tent connected to vacuum tubes to prevent contamination of the room air. Results: NO was administered in this way to two patients. First was 11 month old child with severe Ebstein anomaly 24 hours after a Glenn procedure and closure of tricuspid valve. The second was a newborn who underwent complete repair of a truncus arteriosus and presented recurrent pulmonary hypertension after extubation. None required reintubation, oxygenation improved and no complications were noted. Conclusion: NO can be safely administered to extubated patients through a ventamask and this mode of administration increases its therapeutic.

P256
Cardiopulmonary bypass reduces bronchial blood flow: a potential mechanism for injury of the lung during extracorporeal circulation
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Background: During total cardiopulmonary bypass (CPB), blood flow to the lung is limited to flow through the bronchial arteries. We tested the hypothesis that bronchial blood flow during CPB does not prevent ischemia of the lung and that perfusion of the pulmonary arteries with oxygenated blood during CPB reduces subsequent lung injury. Methods and Results: Of 24 piglets (5.2±0.5kg), 18 were subjected to 120min of normothermic total CPB without aortic cross-clamping, followed by 60min of post-bypass perfusion. Nine of them received continuous pulmonary perfusion with oxygenated blood during CPB. Six piglets served as control and were ventilated for 180min only. We quantitated bronchial arterial blood flow, tissue lactate content, alveolar septal thickness and alveolar surface area and obtained bronchoalveolar lavage fluids (BALF). With the beginning of CPB bronchial arterial blood flow was decreased to 15% of baseline values (42.1±10.4 to 5.6±1.0ml/min), remained decreased until the end of CPB and returned to starting levels 60min after CPB. The decrease in bronchial blood flow was associated with a 3-fold increase in lactate content of lung tissue. At the end of reperfusion there was a 2-fold increase in alveolar septal thickness and a significant accumulation of albumin, lactate dehydrogenase, neutrophils and elastase in the BALF vs. control. Controlled pulmonary perfusion significantly ameliorated all the observed changes. Conclusions: 1.) CPB causes a reduction in bronchial arterial blood flow which is associated with injury of the lung. 2.) The inflammatory response, as evidenced by BALF, may be caused by ischemia. 3.) Controlled pulmonary perfusion reduces injury to the lung during CPB.

P257
Small diameter transvenous permanent pacing leads in children
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The smaller venous caperitance in children may hamper transvenous pacemaker lead implantation. The purpose of this study was to review the intraoperative performance of smaller diameter active-fixation transvenous pacing leads in children. Methods: All transvenous pacemaker implant procedures at a single pediatric institution during a 3-year period from November 1997 to November 2000 were retrospectively reviewed. Modern small diameter active fixation bipolar leads include the Tendril SDX 1488 (St. Jude Medical), Thinline 4388-13 (Intermedics), and CapsureFix Novus 5076 (Medtronic). Measured outcomes included successful venous passage and anatomic location, electrical implant characteristics, procedural variables and complications. Results: A total of 355 leads in 224 patients (age 2 year – adult) were implanted during the period. Tendril SDX leads were utilized 77 times in 53 patients, Thinline fixed-helix leads 9 times in 6 patients, and CapsureFix Novus leads 7 times in 5 patients. Therefore, small diameter leads were chosen 25% overall but account for 75% of lead implants in 2000. All Tendril SDX and CapsureFix Novus leads were located without complication. There were no differences in capture, sensing, or impedance characteristics compared with analogous 5-9 French bipolar active-fixation leads (i.e. Tendril 1388, CapsureFix). Two of 8 [25%] sutureless leads failed due to acute bends in the conductor. At short-term (10±7 months) follow-up 92/93 (99%) implanted small diameter leads remain functional, comparable to 1 year standard diameter transvenous lead survival. Conclusions: The study demonstrates the feasibility of using smaller diameter transvenous pacing leads in children. The extendable retractable steroid-eluting active fixation leads had 100% acutely-successful implantation without complications, whereas difficulty was occasionally encountered with the fixed screw lead. Further long-term follow-up will be necessary to evaluate a potential value of improved preservation of venous patency, lead longevity, and lead extraction feasibility.

Surgical Management and Results: Valves/Conduits

P258
New technique of preparing and preserving acellular heart valve homografts
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The basic purpose of the research was the development of a technique of preparing heart valve homografts. The technique consists of preservation of the natural leaflet matrix carrying out the decellularization process. One hundred eighty seven homografts were dissected within 24 hours of death (mean 17.11.25) from hearts removed from cadaver donors. The age of donors was at the range from 1 to 48 years (mean 32.45.7). In 28 donors the following viruses were found: Cytomegalovirus 7; Hepatitis B virus 11; Hepatitis C virus 2; Human herpes virus 8. These homografts were discarded. A cellular lysis step was initiated after dissection of the homografts and proceeded up on the moment of operation. Homografts were incubated

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

in a combined solution including the hemeno-peptid complex with osmolality of 550-600 mmol/kg (Russia, Patent 2083105, August 27,1994) Results: 67 preparations were investigated by standard electron microscopy After exposure to the decellularization protocol, leaflet's endothelial cells could not be detected in all cases. The stromal cells were severely damaged. The leaflet matrices and conduit linearly arrayed collagen fiber structure after decellularization retained the trilaminar structure of native tissue Thirty four were successfully implanted with the same heart valve homografts placed in the right ventricular outflow tract. We have no reoperations in this group for narrowing leaflets during 6.7 + 1.2 mean patient follow up In summary we offer a technique of preparing homografts which effectively keeps matrices and carries out decellularization. The valves were fully competent, with no evidence of insufficiency or prolapse after implantation

P259
Early experience with the shelugin no-react pulmonary heterograft
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The performance of a porcine pericardial conduit with anticalcification treatment was evaluated as a possible alternative to homografts in newborns and infants requiring RV-PA connection Method From March 1998, 22 Shelugh No-React Heterografts (SNRB) were implanted in 21 patients for palliation or repair of complex malformations. Mean age and weight at surgery were 13.3 mos (range 0.3–156) and 6.3 kg (range 2.6–30). Eleven patients were younger than three mos. Results Three pts died at surgery Two deaths were unrelated to the conduit: One death occurred after multiple procedures due to Aspergillus endocarditis. One patient required two conduit replacements due to Candida infection, at last receiving a homograft In another 3.8 kg infant, a size 12 SNRB could not be fitted in the chest and was acutely replaced by a size 17 homograft, the pt died 2 months later of progressive heart failure. Four pts required 3 conduit replacements 2 to 20 mos after implantation (mean 9 mos): homografts were finally implanted in all. Histology of explanted conduits did not show calcification nor inflammatory reaction in the conduit wall and valve Actuarial patency and reoperation-free survival were 74% and 46% at mean follow-up interval of 13 mos (range 3–33). Conclusion. The durability of SNRB approached that of homografts of comparable size. This prevents preemptive infection to homograft in case of pacemaker surgery and, possibly, in resistance to infections, as suggested by two cases of endocarditis Early calcification did not occur

P260
Collagen synthesis and collagenase activity of cryopreserved heart valve
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Background Durability of the valve is assumed to be dependent on the remodeling ability of the valve Valvular remodeling seemed to be controlled by the collagen synthesis and collagenolytic activity of the valvular fibroblasts and endothelial cells However, the balance of the collagen synthesis and the collagenolysis of the cryopreserved valve have not yet been clearly revealed. We assessed the collagen synthesis and collagenolysis of the cryopreserved valve Methods: Twelve valves were devided in two groups, freshly harvested valves (FRES group, n=6) and cryopreserved valves (CRYO group, n=6) We assessed the collagen content by the Sirius red, known as a selective dye to the collagen Collagen synthesis itself was evaluated by 3H proline incorporation method Non collagenase-digestive counts (NDC), represent protein synthesis, and collagenase- digestive counts (CDC), collagen synthesis were estimated Collagenase activity of the valves were assessed by the gelatin zymography Results: Collagen content of the CRYO group was not diminished. NDC of the CRYO group decreased to 23Å|42% of the FRES group, CDC of the CRYO group was maintained about 78Å|35%. Collagenase activity (MMP-2 gelatinase-A, MMP-9 gelatinase-B, and MMP-1: interstitial collagenase) was same level in the supernates of the both valves Intensity of the collagenolytic activity was same in both groups. Conclusions. Although collagen content of cryopreserved valves was maintained, ability of collagen synthesis was slightly diminished Activities of MMP-2, MMP-9 and MMP-1 were maintained even after the cryopreservation Cryopreservation procedure itself may lead the collagen metabolism to the degradative side

P261
Midterm results of total aortic root replacement with pulmonary autograft (Ross operation)
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Midterm Results of Total Aortic Root Replacement with Pulmonary Autograft (Ross Operation) Between February 1995 and January 2000, 18 patients underwent Ross operation Age at the operation ranged from 2 to 31 years Diagnosis includes congenital aortic stenosis and/or regurgitation in 15, and adult aortic regurgitation in 3. In all cases autograft was implanted by the method of total aortic root replacement, associated with annuloplasty for the dilated aortic annulus in 2 and anterior mitral valvotomy by the Konno procedure in 3 (Ross-Konno) Right ventricular outflow tract was reconstructed by a pulmonary homograft in 12, a xenopericardial conduit in3, or the other reconstructive procedures with autologous tissue and outflow patch in 3. There was no operative and late death. Reoperation was needed in 1 patient due to stenosis of pericardial conduit 4 years after the initial operation Pressure gradient across implanted autograft was negligible (4.8± 0.5mmHg), and echocardiography revealed no aortic regurgitation in 12 cases and trivial to mild in 6, over a mean follow-up period of 23±18 months (range 2 to 60 months), signifying excellent durability of implanted autograft. Right ventricular outflow tract reconstruction with the homograft resulted in excellent mid-term performance as showing pressure gradient of 9.0± 4 mmHg and no regurgitation in 11 of 12 cases, whereas pressure gradient was17.9±13 (mmHg in the patients underwent the other reconstructive procedures We conclude that Ross procedure associated with the concomitant procedures to adjust the size discrepancy between the native aortic annulus and autograft has provided good midterm results with excellent autograft durability And the procedure was thought to be a preferable method for children as well as young adults with congenital aortic stenosis KEY WORDS Ross procedure, pulmonary autograft, homograft, aortic stenosis, aortic root replacement

P262
Percutaneous transseptal mitral commissurotomy in children during acute rheumatic fever
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Although closed mitral valvotomy has been done during acute rheumatic fever (ARF) whether percutaneous transseptal mitral commissurotomy (PTMC) can be performed during ARF is not known We performed PTMC in 4 children(age 10.3±9.2 years, 3 females) with severe symptomatic mitral stenosis during ARF.All patients had severe mitral stenosis and class IV symptoms. Mean height was 135.1±14.8 cm and weight was 23.7±9.1 Kg. Rheumatic fever was diagnosed on the basis of Jones criteria. All patients were receiving steroids. The mitral valve area increased from 0.8±0.16 to 1.7± 0.6 cm2, the mean diastolic gradient and pulmonary wedge pressure decreased from 21.9±3.3 to 8.2±1.5 mmHg and 30 ±1.4 to 17.5±5.4 mmHg respectively. The mean pulmonary artery pressure decreased from 56.9±19.3 to 38.1±16.4 mmHg. The cardiac index increased from 2.56±.66 to 3.29±.57 l/m2/min. One patient developed moderate mitral regurgitation who was well tolerated. Marked symptomatic improvement occurred in all the patients. On a follow-up of (3.4±1.1 months), no atrial thrombosis was seen. Hence PTMC may be considered in selected patients with severe mitral stenosis even during ARF if required

P263
A 22-year experience of surgical management of congenital heart defects with conduites
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The application of different types of conduits became routine for surgical restoration of RV to pulmonary artery continue over past decades. It is still uncertain what kind of conduit should be used for good outcomes We reviewed 112 patients with different CHD which undergone operation at one institution from 1978 to 2000. Patients ages ranged from 6 months to 22 years (mean 12.5 ± 5.8 years) We used conduits for repair TOF in 23 cases, CTGA (30), TGA (24), PA (36), hemi TA (3), TA (4), DORV (7), DOLV (3). Autopericardial conduits were implanted in 36 (32%) patients All these grafts had an autologous pericardial monocusp. Allografts were used in 27 (24%)

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

parents. In 7 of them it was combined with xenovalve. 14 autologous pericardial monocusp, 6 without valve. Xenografts were used in 5 (8%) patients. Aortic and pulmonary antibiotically treated homografts were used for 28(19%) and 19 (13%) patients correspondingly. Hospital mortality was 17% (19/112). Since 1995 mortality has been 13% despite younger age at repair. Early deaths were caused severe bi-ventricular insufficiency. Median follow-up was 18, 2 years (range 1 to 22 years), and was complete for 90,2% patients. 68,1% of patients had a systolic pressure gradint between RV and conduit 15,7 ± 4,5 mm Hg and were in I NYHA class. 29% had gradient 30,1 ± 3,6 mm Hg and II NYHA class. 2.9% had 72,4 ± 4,7 mm Hg and III NYHA class. Majority of patients with low gradient had a pulmonary homograft. There were 3 (2.5%) cases of late death for reason 1-conduit obstruction, 1- sudden and 1- cardiac cachexia. We conclude that pulmonary autografts usually are maximum calcified, have small gradient and more facile the repair.

P264

The left atrioventricular valve in ostium primum atrial septal defect: management strategy and surgical outcome

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We tested the hypothesis that in patients with a partial atrioventricular septal defect (PAVSD), i.e. ostium primum atrial septal defect, and a competent left atrioventricular valve (LAVV), sutures should be placed at now the line of apposition of the superior and inferior bridging leaflets (septal commissure, SC) to prevent the development of regurgitation. Outcome of surgery and risk factors for the need for reoperation were also evaluated. 152 children with PAVSD underwent surgical repair between January 1979 and December 1999. The median age and weight at repair were 4 years and 15 Kg. The interatrial communication was closed using a pericardial patch in 62% and with synthetic material in the remainder. In 84% sutures were placed across the SC partially to close the so called, but incorrectly named, mitral valve cleft. Hospital mortality was 2.6% (95% CI 0.7, 6.7%) which did not differ statistically over 21 years. No risk factors for early death were identified. 21 patients (13.8%, 95% CI 8.8, 20.3%) required reoperation: 19 for LAVV regurgitation, 1 for RAVVR and 1 for subaortic stenosis. Univariate analysis of risk factors for LAVV reoperation were young age, low weight, the severity of preexisting LAVVR, small size LAVV and the presence of a small interventricular (IV) communication. The hazard ratio for the need for reoperation when preoperative LAVVR was moderate to severe was 4.7 times higher than with no to mild regurgitation and 6.5 times higher in patients with a small IV communication. 19(15%) of 127 patients in whom the SC was sutured required LAVV reoperation, but none of the 25 in whom the commissure was left alone ($P=0.04$). The hypothesis that in the absence of preoperative LAVVR it is necessary to place sutures in the SC has not been proven.

P265

Spiral pericardial tube conduit for extracardiac fontan procedure

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Extracardiac fontan procedure gained increased acceptance recently due to better blood flow dynamics, preservation of ventricular and pulmonary function and avoidance of dysrhythmias. PTFE tube grafts are the conduit of choice. We would like to present an alternative conduit substitute for extracardiac Fontan procedure. Between January 1998 to October 2000, 8 extracardiac Fontan procedure were performed by using spiral pericardial tube conduit. Five patients were male. Median age was 7.5 years (range 4 to 16 years). Preoperative diagnosis were tricuspid atresia in 2 patients and double inlet univentricular heart and pulmonary stenosis in 6 patients. Operations were performed under mild hypothermia and cardiopulmonary bypass without cross clamp. Pulmonary arteries were mobilized extensively. Superior vena cava was anastomosed to right pulmonary artery in an end to side fashion. Main pulmonary artery was transected and inferior vena cava was connected to main pulmonary artery by using spiral pericardial tube graft. This was created from a large piece of autologous pericardium. Reversed fresh pericardium was wrapped around a 20 mm Hegar cabinet in a spiral fashion and was sutured to create a tubular shape. Then it was immersed 10 minutes in 0,6 % glutaraldehyde solution to obtain appropriate shaped conduit. There was no mortality and major morbidity. Mean intensive care unit and discharge time were 2,4 and 11 days respectively. Follow up was complete for all patients. Recurrent pleural effusion was detected in 3 patients. Routine echocardiographic examinations for all patients and magnetic resonance angiography in 3 patients revealed no problem in conduits and fontan circuit. Spiral pericardial tube conduit may be a useful alternative for extracardiac Fontan procedure. We think that its inherent low thrombogenicity may be attractive; excellent handling and hemostatic properties are other advantages.

P266

Experience with the stentless bioprosthesis at an extracardiac conduit in complex congenital heart disease

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Extracardiac conduits invariably need to be replaced due to growth of the patient or conduit failure. Homografts may be the best choice but results of stentless bioprosthesis at the same position is not clear. From January 1998 to October 2000, 32 patients were operated. The median age and weight were 6.1 ± 3.2 years (range, 2,5 to 15 years) and 17,4 ± 7.6 kg. (range, 9.5 to 40 kg.) respectively. Preoperative diagnosis was; TOF with pulmonary atresia or absent pulmonary valve syndrome in 18 of the patients, TGA,VSD and PS was present in 9 patients, Corrected TGA, PS and/or VSD in 7, DORV and PS in 2; Previous arterial switch operation and PS in 2; DIOLV,VSD and PS in 1 patient. We used Baxter – Edwards Prima bioprosthesis in 13 patients, Medtronic Free-Style in 13, Cryo-Life Bois in 9; and Medtronic Contegra bovine jugular vein conduit in 1 patient. Hospital mortality was 6,2 % (n=2 patients). The mean postoperative RV/LV was 0.56. The postoperative complications were reexploration for bleeding (n=2), bacterial endocarditis (n=1) and complete heart block (n=1). Cardiac compression occurred in 1 patient. Late sternal closure at fifth postoperative day was employed. The mean intensive care unit and hospital discharge time were 3.9 ± 1.9 and 12.5 ± 6.7 days respectively. All patients were evaluated echocardiographically before hospital discharge and normal pulmonary valve function was detected in all patients. The median follow-up was 19 ± 11,2 months (range, 1 to 36 months). Three patients died at follow-up period. Two of them died of low cardiac output soon after reoperation. Reoperation was performed in 6 of the patients (22 % %) due to tricuspid regurgitation in 2, residual VSD in 1 and LVOT obstruction in 1 patient. Conduit stenosis developed in 1 patient at 6th postoperative month who had 21 mm Baxter – Edwards Prima bioprosthesis. Unrestricted life - style was possible for 82,7 % of survivors. Stentless bioprostheses may be an alternative for right side reconstruction of complex congenital cardiac anomalies. Pulmonic porcine and bovine jugular vein conduits look promising.

P267

Mid-term follow-up after pulmonary autograft replacement of the bicuspid aortic valve in the young

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A dysfunctioning congenitally bicuspid aortic valve may require surgical treatment within the fourth decade of life. This retrospective study was undertaken to evaluate the mid-term results achieved with the Ross procedure in adolescents and young adults with a bicuspid aortic valve. Between July 1994 and October 2000, 51 patients, 44 males, with a mean age of 27±10 years (range, 7 to 48), underwent replacement of a diseased bicuspid aortic valve (stenosis 6; 12%; insufficiency 32; 63%; combined 13; 25%) with a pulmonary autograft (PA). Mean NYHA FC was 1.6. Four patients (8%) had healed endocarditis and 5 (10%) had previous cardiac surgery. The PA was inserted as a subcoronary implant in 1 case (2%), as a root in 39 (76%) and as a cylinder in 11 (22%). The right ventricular outflow tract was reconstructed with a cryopreserved pulmonary homograft in all cases. Mean cardiopulmonary bypass and aortic crossclamp times were 207±55 min and 161±38 min respectively. No early or late deaths had occurred at a mean follow-up of 29±17 months (range, 1 to 78). Two patients (4%) were reexplored for bleeding. 2-D Echo evaluation of neo-aortic valve competence at 6 months revealed no evidence of aortic valve regurgitation in 43 (88%), trivial regurgitation in 7 (14%), and mild-to-moderate in 2 (4%). The latter patients (subcoronary implant) required reoperation. At six months, the degree of regression of left ventricular mass compared to pre-operative data, was 37±14% ($p<0.05$). Two patients (4%) showed mild dilatation of the neo-aortic root after 2 years of follow up. All patients are asymptomatic, in NYHA FC I, and enjoy normal social interaction. In conclusion, the Ross procedure can be offered as a low-risk alternative to adolescents and young adults with a bicuspid aortic valve. Continued patient evaluation with regard to evidence of neo-aortic valve degeneration, root dilatation and homograft dysfunction in the long term is warranted.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P268

Long-term outcome of conduits in the right ventricular outflow tract for repair of truncus arteriosus in early infancy

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Objective: Right ventricle to pulmonary artery (RV-PA) conduits are needed for the repair of many congenital heart defects. Homografts and heterografts are options, yet the optimal choice remains unclear. To evaluate the potential differences, we performed a retrospective cohort study on patients with truncus arteriosus, a group with relative uniformity in anatomy and repair technique. Methods: Between March 1991 and April 1998, 49 patients less than 3 months of age underwent repair with an RV-PA conduit. Patients received either a heterograft (9) or homograft (40). Major outcomes were time to conduit replacement and patient survival. Results: The mean +/- SD age at the time of repair was 21 +/- 20 days, with a mean weight of 3.1 +/- 0.6 kg. Mean weight of patients undergoing homograft versus heterograft placement was 3.0 kg versus 3.4 kg, respectively ($p=0.1$). Homograft size ranged from 8–15 mm and heterografts were 12 mm. Mean length of follow-up was 56 months (range 0–112 months). Data are inclusive of all patients, including early deaths. Time to replacement (in years) for: 50% of the conduits was: all conduits, 2.8; homografts, 2.8; and heterografts 1.9. By 5 years, 100% of heterografts and 69% of homografts required replacement ($p=0.27$). For technical reasons, homografts were placed preferentially in smaller infants (wt<2.7 kg, $p=.05$). Adjustment for weight or age at the time of surgery, aortic versus pulmonary homograft, or conduit/weight ratio did not affect the time to conduit replacement. There was no difference in survival. Conclusion: Advantages to homograft placement include smaller size availability and technical ease of placement. In addition, no significant difference in overall survival or time to conduit replacement between homografts and heterografts was demonstrated in this study, however our results may be limited by the sample size.

P269

Application of Stentless Bioprosthesis (Freestyle Aortic Valve) as a pulmonary homograft substitute for RVOT reconstruction for the patients with severe pulmonary regurgitation associated with severe RV dysfunction

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Although pulmonary homograft (P-homograft) is ideal for RVOTR in the patients(pts) with severe PR associated with severe RV dysfunction late after RVOTR in various congenital heart disease. However, severe donor shortage of p-homograft had been forced us to use other p homograft substitute such as monocusp xenograft, RVOT patch, valved pericardial roll or stented xenograft valve. But RVOTR in the pts with severe RV dysfunction associated with sever PR or sever PH might require competent PA valve to ensure RV function, postoperatively. Better midterm results of recently introduced stentless aortic bioprosthesis for AVR, expecting larger effective valve orifice and new anticalcification treatment encouraged us to apply the new stentless valve to the pulmonary position for 3 pts expecting less pressure gradient and longer competent valve function. Case 1, 10 years old girl, 4 years after monocusp patch RVOTR for VSD+PA. Case 2, 37 years old male, 20 years after total correction of TF with RVOT patch. Case 3, 41 years old male, 25 years after repair of TF/PA with Conduit repair. All 3 pts had severe PR and TR with dilated RV and severe RV dysfunction(RVEF 23% to 37%). For these 3 pts, PVR with Freestyle Bioprosthesis(23mm, 27mm, 25mm) were performed successfully. TAP(Case 1) and TVR with 31 mm Hemopark II(Case2) were also done. After operation, all 3 patients had dramatic improvement of RV function and reduction of RV volume, and great improvement of clinical symptoms were also obtained. Conclusions: Although there must have long term follow up for valve function and its durability, midterm results indicate that this new approach using stentless bioprosthesis seems to be a better alternative to p-homograft PVR for RVOTR in the pts with severe RV dysfunction.

P270

The Metras modification of the REV operation: a prosthesis-free procedure for reconstructing the right ventricular outflow tract in children

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A prosthesis-free procedure to reconstruct the right ventricular outflow tract (RVOT) in children is described. Limitations to the use of prostheses and homografts are those of cost, availability, and the need for later replacement. Our experience with a modification of LeCompte's réparation à l'étage ventriculaire (REV) operation, initially described by Metras, forms the basis of the present report. During a two-year period ending in October 2000, 12 consecutive children with d-TGA, VSD, and PS underwent the Metras modification of the REV operation. The mean age at operation was 12 ± 8 months and the mean weight was 10 ± 4 Kg. Concomitant cardiac defects were common and included an ASD in 7 patients, a complete AV canal in 1, a restrictive VSD in 4, a PDA in 4, and coronary artery abnormalities in 6. The procedure entails harvesting a portion of proximal ascending aorta and storing it in saline. Following closure of the VSD and division of the main pulmonary artery, the aortic autograft is interposed between the distal divided end of the pulmonary artery and the right ventriculotomy. The ventricular anastomosis is augmented with a patch of glutaraldehyde-treated pericardium to complete the operation. There were 2 hospital deaths (25 percent), all of which occurred early in our experience, and all due to right-sided heart failure progressing to biventricular failure. The remaining 9 patients are in NYHA FC I or II, and 6 patients are medication free. Postoperative echocardiography revealed normal biventricular function with unobstructed RVOTs in all patients, and mild to moderate pulmonary insufficiency with no stenosis in 3 patients. These findings demonstrate that the Metras modification of the REV operation is an acceptable prosthesis-free alternative in reconstructing the RVOT in children. The procedure is associated with an acceptable incidence of postoperative pulmonary insufficiency and after an initial learning curve, with excellent early postoperative survival. Careful follow-up is needed to assess the functional outcome and long-term durability of this new procedure.

P271

RVOT – Reconstruction with bovine valved jugular veins as an alternative to homografts and porcine xenografts

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Background: Paediatric RVOT-Reconstruction with homo- or porcine xenografts is problematic for limited availability and lack of material for reconstruction. Frequently, these grafts show early degeneration and tissue ingrowth. Venpro offers a bovine jugular vein graft to overcome these problems. Patients and Methods: Within an FDA-controlled study, we implanted 54 Contegra® Pulmonary Valved Conduits (PVC) from May 1999 to November 2000 in 54 infants (m/f 23/31) aged 2 days – 17.4 years, median 1.6 years. 20 were primary repairs, 20 had previous graft implantation, 14 other repairs. Preoperative diagnoses: TAC (17 patients), TOF (22), DORV (12), TGA+PS+VSD (2), and rare malformation. Echocardiography is performed at 1 and 3 months, then every 3 months postoperatively. Total follow-up: 9.5 years. Survival, freedom from explantation and from redo/explantation were compared to our 52 homograft- and 20 xenograft recipients. Results: Having enough tissue on both sides of the valve, the PVC enables the surgeon to perform all anastomoses without additional material. The PVC tissue is very apt for suturing. There were four deaths (3 early, 1 late). PVC insufficiencies are common, but without clinical significance. Redos: 3 peripheral pulmonary arteries (pPA) stenoses, 1 bronchus compression by enlarged pPA, 2 unplanned VSD-closures. There were no device related adverse events. Transvalvular peak gradients remained constantly below 25 mmHg. Distal PA-gradients developed in TAC, less in DORV and nearly not in TOF. PVC grafts were advantageous to homografts concerning survival and freedom from explantation (without reaching statistical significance). Freedom from explantation/redo was equal for the latter two groups. Xenografts were significantly inferior. Conclusion: The Venpro Pulmonary Valved Conduit offers unique stitching and suturing options for primary and redo RVOT reconstruction. Its durability seems at least equivalent to homografts and is superior to porcine xenografts.

P272

Repair of Common Arterial Truncus (CAT) using pulmonary homografts achieves improved results compared to other valved conduits

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Study of progress and management of early primary correction of CAT. 16 consecutive pts undergoing primary correction of CAT from 1993 to Feb 2000 were reviewed. Median age was 29 days (range 11–125), 5 pts had

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

anatomical type I, 4 pt's had type I-II, 6 pt's had type II and 1 pt had type III. The RVOT was reconstructed with aortic homograft (7 pt's), pulmonary homograft (8 pt's) and valved xenograft (1 pt). There was no perioperative or late mortality at median follow-up of 51 months (9 to 88). One pt, who had severe tricuspid stenosis and regurgitation, subsequently required replacement of both the aortic root and the homograft conduit. Obstruction occurred proximal or at the homograft in 2 and 3 pt's respectively requiring replacement (1 pt), balloon dilatation (2 pt's) and stenting (2 pt's). 4 pt's had pulmonary artery branch stenosis which were treated by balloon dilatation in 3 pt's and stenting in the fourth. 57% of pt's with aortic homografts developed stenosis as compared to 10% repaired with pulmonary homografts and this difference is statistically significant (p<0.001). All pts are asymptomatic, only one pt has significant tricuspid regurgitation and it is enlarged. We believe that early surgical repair, preferably using a pulmonary homograft, yields good medium term results. Subsequent therapeutic catheterization delays inevitable conduit replacement.

P273

Posterior Annular Plications: A Simple and Effective Operative Technique in Ebstein's Anomaly

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Background: Ebstein's anomaly (EA) is a rare congenital anomaly which is treated operatively in both pediatric and adult patients. Many complex operative techniques have been described for repair of the aberrant tricuspid valve and the atrialized segment of the right ventricle which characterize EA. We have recently adopted a simple operative strategy for the treatment of patients presenting with symptomatic EA. Methods: Our operative approach to EA is a posterior plication of the anteroposterior commissure to the septoposterior commissure with closure of the valvular to the fungiform. This creates a functionally bicuspid valve with an aperature determined by the amount of plicated leaflet. After discontinuation of cardiopulmonary bypass mean central venous pressures are measured and compared to the mean pulmonary artery pressure to determine the need for a bidirectional Glenn's shunt (BDG). A retrospective review of our patients was conducted (n=6). Results: The mean age of our patients was 41+15 (range 15-56). Preoperatively all patients had 4+ tricuspid regurgitation and were functional NYHA Class III. Five of the patients underwent a posterior annular plication and one (the first patient) had a prophylactic BDG established at the time of the operation. The sixth case had a relatively stenotic tricuspid orifice and did not require reductive annuloplasty. No patient (apart from the first one in which the BDG was constructed prophylactically) has required a BDG shunt despite aggressive reduction in the tricuspid orifice. The post operative length of stay was on average 5.3+1.2 days (range 4-6) with no post operative complications. Mean follow up was 5.2+4 months with all patients symptomatically improved to NYHA Class I. Transthoracic echocardiography at follow up reveals 1+- 2+ tricuspid regurgitation in the cohort. Conclusion: Plication of the posterior annulus without specific intervention for the atrialized right ventricle or aneurysm to make the tricuspid valve appears to be a reasonable operative approach to Ebstein's anomaly. Prophylactic BDG is not routinely necessary and should be reserved for those few patients in whom the post repair central venous pressure is greater than the mean pulmonary artery pressure.

P274

Right ventricular outflow reconstruction in the Ross procedure : combinedautologous aortic and bicuspid Gore-Tex valve

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Concomitantly with the Ross procedure, the right ventricular outflow tract (RVOT) was reconstructed with the aortic autograft with noncoronary cusp (NCC) and a pericardial patch bearing a bicuspid valve made of a 0.1-mm-thick Gore-Tex membrane. Early and midterm results of this technique were assessed. Patients and methods: Seven patients 8 to 17 years underwent Ross or Ross-Konno procedure. The diagnoses were aortic stenosis with regurgitation (n=4) and aortic regurgitation (n=3). At the operation, the noncoronary cusp was harvested along with the adjacent aortic wall. The right side of the aortic fibrous ring of NCC was preserved to avoid injury to the atrioventricular conducting tissue beneath the annulus. After reconstruction of the aortic root with the pulmonary autograft, NCC was anastomosed directly to the septal aspect of the RVOT opening and the adjacent autologous aortic wall was anastomosed on the pulmonary bifurcation. A prosthetic patch bearing bicuspid Gore-Tex valve was anastomosed to the anterior aspect of RVOT. Results: Neo pulmonary annular diameter ranged from 119 to 148 % of normal value. There was no operative and late death. Follow-up ranged from 2 weeks to 48 months (median 24.3 months). Postoperative echocardiography showed mild pulmonary regurgitation in five and moderate in two. Both NCC and Gore-Tex valve motion were maintained well in all patients. Postoperative cardiac catheterization was performed in five patients. By the catheterization, pressure gradient across the neopulmonary annulus ranged from 13 to 25 mmHg (median 18.4 mmHg). Conclusion: Combined autologous aortic and bicuspid Gore-Tex valve is an excellent alternative at modest cost for RVOTR concomitant with the Ross procedure. This technique can be used in almost all patients with aortic incompetence and/or aortic stenosis who have a mobile NCC.

P275

Aortic valve replacement in pediatric patients

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The ideal choice of aortic valve prosthesis in children remains controversial. We report a retrospective review of the computerized records of 175 consecutive children undergoing 249 aortic valve replacements between April 1968 and September 2000. The mean time to follow-up is 13.4 years. The median age at first operation was 13.1 years with a range of ages from one day to 19.8 years. The type of valvular dysfunction included aortic insufficiency (n=87), aortic stenosis (n=70), and a combination of both aortic stenosis and insufficiency (n=58). The etiology of the aortic valve pathology included congenital (n=131), rheumatic (n=13), Marfan's (n=10), VSD with aortic insufficiency (n=9), previous infection (n=8), ascending aortic aneurysm (n=3), and other (n=1). One hundred of these children had a previous intervention and in 73 patients it was for the aortic valve disease. Concomitant surgical procedures were required in 105 patients. Thirty-two percent of patients (n=56) underwent between one to five reoperations. Freedom from reoperation at the mean follow-up time of 13 years is better for mechanical versus either autograft/allograft or porcine/pericardial aortic valves (75% vs. 38% vs. 10%; p<0.001). There was a 6.8% rate (n=12) of 30 day perioperative mortality. The Kaplan Meier survival for the entire cohort (n=175) at 11 years from surgery is 65%+/-4%. There is no significant difference in Kaplan Meier survival among mechanical, porcine/pericardial, and allograft/autograft aortic valves (p=0.55). Kaplan Meier survival is less favorable for infants aged less than one year of age versus those one year old or greater (45% vs. 70% at mean follow-up of 13 years). This study demonstrates the importance of valve type and age at initial surgery as contributing to long term survival and morbidity. The pediatric patient with a mechanical aortic valve has a lower incidence of reoperation as compared to tissue valves although all groups have similar survival.

Surgical Management and Results: Univentricular Heart/Hypoplastic Ventriculoarterial Discordance

P276

Respiratory effect on pulmonary blood flow after total cavopulmonary anastomosis

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Objective To document whether the action of breathing influence the pulmonary blood flow in patients after total cavopulmonary anastomoses (TCPA). Methods Doppler echocardiography was used on 15 patients after TCPA for the treatment of complex heart malformation in assessment of superior vena cava (SVC) inferior vena cava (IVC) and pulmonary artery blood flow at rest and during deep respiration. Results: the cavovenous and pulmonary flow patterns were affected by respiration obviously: During inspiration, the flow was forward and increased during deep inspiration the basic waves were not changed. for nonvalved extracardiac conduit TCPC, there was distinct reversal flow in IVC the valved conduit can reduce or prevent the reversal flow. Conclusion: The acting of breathing provides an additional energy supply to pulmonary circulation after TCPS. It can augment forward flow for pulmonary circulation. The valved conduit can prevent retrograde flow in IVC which is benefit to patients for their recovery.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P277

Application of the extracardiac conduit total cavopulmonary anastomosis

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Objective To evaluate the value of application of extracardiac conduit total cavopulmonary anastomosis (TCPA) in complex heart disease. Methods From June 1998 to August 2000, 18 patients with functional univentricular complex heart diseases. There were 13 males, 5 females with a mean age of 9.15.6 years (from 2.3 to 21 years), mean weight 25.1 ± 14.25kg (from 11–45kg). Associated heart abnormalities were TGA PS or ASD. All surgical procedures were performed under cardiopulmonary bypass with general anesthesia and hypothermia. Results No operative and postoperative deaths. All patients were followed up from 6 to 19 months. All patients are clinically asymptomatic. There were no evidence of systemic venous pressure high level. EKG revealed no arrythmias. The Sao2 are approximately 96%, the cardiac function are in NYHA class I. Conclusion: The extracardiac conduit TCPA is a simple procedure, it is easy to perform. Compared with other type, it has more advantages.

P278

Ddd-pacemaker implantation after fontan-type operations

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Purpose. Bradyarrhythmias developing after fontan-type operations may severely impair the function of the univentricular heart, leading to symptoms such as fatigue, headaches, ascites, and even protein-losing enteropathy (PLE). Because of the inaccessibility by a transvenous route, pacemaker implantation requires epicardial access, which may account for a certain reserve to perform such operations in this setting. Material & Methods: Between 1997 and 8/2000, 24 (of 67=36%) patients with fontan-type operations received DDD-pacemaker systems with atrial steroid-eluting screw-in-on electrodes (mean threshold 1.9V at 0.5ms, range 0.4–3.5V) and ventricular screw-in electrodes (mean threshold 1.7V at 0.5ms, range 0.1–3V). Three were implanted at the time of a conversion from atrio- to cavo-pulmonary connections (3–56 months), mean 18) in 13 patients. Mean age was 9.5 years (range 6 months – 19 years). A right ventricular anatomy was present in the pacemaker patients in 54% (13/24), whereas in the overall Fontan group 39% (26/67) had right ventricles. Results: 23/24 pacemakers are currently functioning in DDD mode. Average length of stay in the 18 patients requiring repeat sternotomy was 5 days without procedure-related complications. In 3 children repeat sternotomy could be avoided by implantation of atrial electrodes during the Fontan operation. All patients improved clinically, including resolution of near PLE in 4 instances. Conclusions: Development of symptomatic bradyarrhythmias may lead to significant morbidity after fontan-type operations. Electrophysiologic evaluation is advised for follow-up. The indications for implantation of a (DDD-) pacemaker system should be handled liberally. Placement of atrial electrodes at the time of the fontan operation, especially in right ventricular anatomy, helps to avoid repeat sternotomy.

P279

High frequency of arrhythmias after fontan operation indicates earlier anticoagulant therapy

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Background: As patients could survive for longer periods following modified Fontan operations (conventional atrio-pulmonary connection), the late morbidity after this procedure became increasingly apparent. The purpose of the present study was to evaluate late sequelae of modified Fontan operations in long-term survivors (n=114) at our institute. Method and Results: The subject consisted of patients who underwent a modified Fontan operation between 1981 and 1990. Thus, all patients were examined at least 10 years postoperatively in this study. Early mortality, within 30 days of the operation, was 17.6% (3 of 17 patients died due to low output syndrome). Excluding these early deaths, the cumulative survival rate at 5 and 10 years was 100 and 79%, respectively. Arrhythmias including atrial fibrillation or flutter were the main late causes of morbidity. The arrhythmia-free rate at 5 and 10 years was 77% and 50%, respectively. Although the quality of life was considered good because all patients (n=111) who survived for 10 years or more were in class I or II according to the New York Heart Association classification, most of them in fact suffered from potentially life-threatening arrhythmias. Conclusions: Meticulous attention and utilization of established treatment strategies for arrhythmias including anti-arrhythmics, anticoagulants, catheter ablation of re-operation concerning the circulation to the total cavopulmonary connection must be considered in long-term survivors following the modified Fontan operation. The fact that no one knows when the thrombogenic arrhythmias occur suggests anticoagulants should be initiated in the early postoperative period.

P280

Arterial switch operations in which neo-aortic anastomosis is performed prior to coronary anastomosis

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Arterial switch operation is universally accepted standard repair of transposition of great arteries. Different methods are proposed for coronary translocation, because coronary kink is the most important cause of the mortality in our clinic, in 40 arterial switch operations which were made between years 1997–2000 we performed neo-aortic anastomosis preceding coronary anastomosis. The coronary anastomosis are made on the swollen neo-aorta. According to Leiden classification, there were 22 cases with usual coronary pattern (A1), 13 cases with AB1, 4 cases with single coronary ostium, and 1 case with intramural course of left coronary artery. In cases with AB1, and single coronary ostium, after neo-aortic anastomosis, we applied pericardial and pulmonary hood technique at the medial side of the coronary anastomosis. Among these 40 cases, the patients with intramural course of the coronary artery is lost. Also one case died because of prolonged intubation period, pneumonia and sepsis. After arterial switch operations, the most important cause of death is the coronary kink. The cases with different position of aorta and pulmonary arteries (A-P, side by side), the coronary implantation positions can be different. And also, coronary implantations sites can be different if there is size difference between aorta and pulmonary artery. The formation and working of neo-aorta facilitate the determination of the exact coronary implantation points, and this minimizes the risk of coronary kink.

P281

Early results of bidirectional glenn anastomosis performed with external shunt in patients with single ventricle physiology

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In single ventricle physiology patients due to its satisfactory effects on left ventricle functions and good results in long term palliation the bidirectional cava-pulmonary anastomosis has been an acceptable procedure. In our department 15 patients ages ranging from 6 to 18 months old have undergone bidirectional cavapulmonary anastomosis. The operation was executed by applying a transient shunt between the vena cava superior and right atrium. Bilateral cavapulmonary anastomosis was applied to 4 patients who had bilateral vena cava superior. Systemic shunt was performed on 7 cases in their neonatal period. Two cases were lost because of low cardiac output in early postoperative period. All cases were extubated within 2 to 12 hours in the postoperative period. In children possessing single ventricle physiology, bidirectional cavopulmonary anastomosis can be obtained by transient shunt. It will allow for good palliation in the early period.

P282

Extracardiac total cavopulmonary connection (ETCPC) as the method of choice for the final palliation in children with functionally single ventricle

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Between 1988 and 2000 109 children with various forms of functionally single ventricle underwent different surgical techniques of cavopulmonary connections (29 bidirectional Glenn anast., 15 Doty, 38 DeLeval, 27 ETCPC). In ETCPC group there were 9 (33.3%) direct IVC-PA anastomosis and 18 graft interposition – 66.7% (8 Gore-Tex, Dacron and 10 ascending aorta homograft). Mean age at surgery was 7.44 (95%CI 6.69 – 8.18). The hospital mortality in the whole cohort was 11% (3 bid Glenn 13.3%, Doty 13.3%, DeLeval 13.5% ETCPC 11%). Late complications occurred in 39% of patients, 9.1% in ETCPC group. Cardiac index late postoperatively was highest and mean PA pressure lowest in ETCPC group. Late mortality (5/97 – 5.1%) occurred only in DeLeval group of patients 5/29 – 14.7%. Probability

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

of survival after 12 years in the whole group of patients is 83%, the highest – 88,9% in E-TCPC group (p = 0,0078). Conclusions. Extracardiac TCPC applies to any type of anatomy in children with functionally SV. Direct IVC – PA connection was possible in 33,3% of patients without mortality. The best late hemodynamic outcomes and the lowest rate of late complications were observed in ETCPC group. Extracardiac TCPC is our method of choice as the final palliation in children with functionally single ventricle.

P283

Extracardiac Fontana performed without cardiopulmonary bypass

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Compared to intracardiac tunnel technique, extracardiac TCPC has advantages regarding, arrhythmias, atrial thrombus formation, and no use of cardiopulmonary bypass. In our department extracardiac TCPC without cardiopulmonary bypass was performed on 5 patients ages between 2 to 10 years old with diagnosis of single ventricle physiology. During the operation after median sternotomy incision, extensive dissection was performed to prepare the pulmonary artery, vena cava superior, vena cava inferior, innominate vein, left atrium, and the pulmonary veins. Bidirectional caval pulmonary anastomosis was performed using the transient innominate vein to right atrial shunt. Next vena cava inferior was thoroughly prepared down to the diaphragm and the transient arterial shunt was placed in the vena cava inferior. After achieving a transient shunt between the vena cava inferior and right atrium, vena cava inferior was dissected at the inferior atrial junction. End to end anastomosis was performed between size 22mm gore-tex tube graft and vena cava inferior. Upper end of the graft was anastomosed to the inferior side of the right pulmonary artery. The patients were extubated in early peoperative period. Excessive inotropics were not in need, and patients were discharged on the seventh and eighth day. Extracardiac fontane due to advantages regarding early extubation, and less inotropic usage, can be performed without cardiopulmonary bypass. The answer, in which patients we must use cardiopulmonary bypass is one questions.

P284

The application of pericardial and pulmonary hood techniques in arterial switch operations to provide the exact coronary geometry

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In arterial switch operations, coronary flow disturbance is the most important cause of mortality. Varied methods are proposed to provide the exact coronary geometry. We used pericardial and pulmonary hood techniques in 14 cases for the achievement on the medial side of the coronary button anastomosis. In 12 of 14 cases, innominate coronary artery was taken as origin from right coronary artery. There was single coronary ostium in 3 of 14 cases. After performing neo-aortic anastomosis, we decided the exact coronary implantation points, and appropriate angle of the coronary button. The piece which is removed from the neo-pulmonary artery is anastomosed on the medial side as pulmonary hood. We didn't find low cardiac output due to coronary ischemia in patients whom we used pericardial and pulmonary hood techniques. In arterial switch operations, pericardial or pulmonary hood techniques are important methods to provide exact coronary geometry.

P285

Do the patients grow somatically well after the fontan procedure?

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Objective. To determine whether somatic growth is normal in patients with the Fontan circulation. Patients. Consecutive 100 patients were investigated who have been followed up for 3 years or more after the Fontan type procedure. Age at operation was 6±4 (1–18) years old. The Fontan circulation was established by atriopulmonary connection in 66 and total cavopulmonary connection in 34. Construction of a systemic-to-pulmonary shunt had been previously employed in 53, banding of the pulmonary trunk in 12, the Glenn procedure in 5. Results. The Z values for body weight and height calculated in comparison with the anticipated standards in the normal children did not change significantly, from preoperative -1.3±0.7 and -1.0±1.0 to postoperative -1.0±0.8 and -1.0±1.0 respectively. The operative procedures chosen for the Fontan circulation did not affect these values. The preoperative and the postoperative Z values were smaller in patients previously undergoing palliative procedures than in those undergoing a primary Fontan procedure ($p<0.01$ and $p<0.05$, respectively). Prolonged administration of diuretics ($p=0.006$), the presence of a dominant morphologically RV ($p=0.03$), and smaller postoperative Cardiac Indexes ($p=0.04$), were other factors deleteriously affecting their somatic growth. Younger age at the Fontan procedure did not promote somatic recordation. Conclusions. Somatic growth may be slightly impaired in patients undergoing the Fontan procedure. In terms of better growth, an earlier Fontan procedure can be performed if preparative procedures are to be avoided.

P286

Reoperation for tricuspid regurgitation after correction of tetralogy of Fallot

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Objective. The aim of this study is to review outcomes in reoperation for tricuspid regurgitation after tetralogy of Fallot. Materials and Methods: Twelve (2.4%, 9 male and 3 female) of 507 patients who had corrective surgery for tetralogy of Fallot at the Sapporo Medical University between 1955 and 2000 required reoperation for severe tricuspid regurgitation. The mean age at the time of reoperation was 17 years (range 1–39 years). The mean interval between the initial correction and the reoperation was 7.2 years (range 10 days to 19 years). The functional class was New York Heart Association (NYHA) class II in 2 patients and class III or IV in 10. Five patients had associated residual ventricular septal defect, 2 had residual right ventricular outflow tract obstruction, 2 had pulmonary insufficiency, 2 had atrial septal defect, and 2 had left ventricle-right atrium communication. Six patients underwent tricuspid valve repair, and the other underwent tricuspid valve replacement. Results: Hospital mortality rate was 16.7% (2/12). Three patients (30%, 3/10) required a second reoperation at 1.6, 9.2 and 15.6 years after the most recent reoperation with no deaths. The reasons for second reoperation was failure of the tricuspid repair in two and thrombosis in valve in one. There were two late deaths. Conclusion: Tricuspid regurgitation after corrective surgery for tetralogy of Fallot must be promptly diagnosed and cured, as tolerance is poor because of postoperative right ventricular insufficiency.

P287

Results of one-stage and two-stage fontan in patients with single ventricle

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Purpose. To compare the results of one-stage and two-stage Fontan procedures performed in a large group of patients in a single center and determine which perioperative factors may influence mortality and morbidity. Methods. We analyzed 164 consecutive patients with single ventricle operated on between 1980 and November 1999. One-stage Fontan procedure was performed in 41 (25 %) children (group I). In the remaining 123 (75 %) patients, two-stage fontan procedure with baffle fenestration was recommended since 1989. From these group 81 (65.8 %) children are after completion fontan procedure (II group). We analyzed about 30 factors that may influence mortality and morbidity. We compared early, late and total mortality rates between two groups, the incidence of postoperative effusions and arrhythmia and analyzed other factors such as age at operation, type of single ventricle, atrio-ventricular and ventriculo-arterial connection, association of mortality and concomitant cardiovascular malformations (anomalous pulmonary venous connection, obstructions to atrial outflow), type of palliation before fontan-procedure, quality of life. Results: Early ($p=0.001$), late ($p=0.042$) and total ($p=0.013$) mortality was significantly higher in group I. No significant differences were demonstrated in the incidence of pleural and pericardial effusions and arrhythmia depending on the employed surgical method ($p>0.05$). Among the remaining variables, the time between the creation of the systemic-pulmonary shunt and Fontan procedure was a factor that significantly increased the risk of early mortality ($p=0.0002$). Assessing the quality of life we found that a significantly greater number of children after two-stage procedure are in NYHA class I ($p=0.0001$). Conclusion: The use of two stage technique in Fontan procedure results in a significant decrease in early, late and total mortality and ensured a better quality of life in the late postoperative period.

P288

Cardiorespiratory response to exercise in children after modified fontan operation

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https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Purpose: Examination of exercise function of Fontan patients after two-stage operation and comparison with healthy control subjects. Methods: Fifteen patients (7 male, 8 female, age: 9.7–17 years, mean: 9.1) after two-stage Fontan repair as NYHA class I with rest O2sat>95% requiring no cardiovascular medications performed graded exercises on treadmill using a modified Bruce protocol 0.5–3.2 years postoperatively (mean: 1.8). During the tests the heart and respiratory rate, blood pressure, oxygen uptake, carbon dioxide production, minute ventilation, tidal volume and O2sat were recorded. Spirometry was performed before and during exercise. Results: The peak VO2max in Fontan patients was significantly reduced compared with normal values (14.4±6.1 vs. 30.5±7.6 ml.kg-1.min-1, p=0.0001) and constituted 52.8–94.2 % of the mean control group VO2max. Other parameters: anaerobic threshold (0.26±0.14 vs. 0.72±0.19 l.min-1; p<0.00001), pulsO2 (2.57±1.23 vs. 5.14±2.25 ml beat-1, p=0.00005), peak minute ventilation (25.8±10.6 vs. 45.5±12.6 l.min-1; p=0.0014); physiological dead space to tidal volume ratio at peak exercise (0.34±0.09 vs. 0.18±0.05; p=0.0004), maximal work rate (80.0±45.7 vs. 236.4±63.5 W, p=0.00008); exercise time (502.2±240.3 vs. 1048.9±175.7 s, p=0.00003) were significantly reduced in univentricular patients. The heart rate at peak exercise was lower in the patients (142.2±24.8 vs. 183.4±23.6 beats.min-1; p=0.0003) and O2sat dropped significantly (92.6±4.1 vs. 87.1±4.3 %; p=0.005). Conclusion: The aerobic capacity in Fontan patients is markedly reduced compared with controls. Subnormal VO2max and Hrmax were demonstrated. The total work and exercise time was reduced. The anaerobic threshold was significantly lower in univentricular patients. The ventilatory response to exercise showed a decreased breathing reserve and an increased dead space/tidal volume ratio. The decreased O2sat at peak exercise suggested intrapulmonary shunting. Exercise spirometry may be useful in evaluating exercise tolerance in children after congenital heart defect surgery.

P289

Radiofrequency ablation of patients with wpw-syndrome, tricuspid atresia and fontan-like circulation

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Introduction: Caused by dilation and scarring of the right atrium patients with Fontan-like circulation suffer from atrial arrhythmia. We report on three patients with a coincidence of tricuspid atresia and WPW syncrome, where atrial arrhythmia triggered atrio-ventricular reentry tachycardia. Methods and results: Three patients (1 female, 13, 14, and 31 years old) had a tricuspid atresia type 1b and concomitant WPW-syndrome. They were palliated by a Fontan-Björk procedure with the age of 2, 4 and 15 years, respectively. All of them had frequent orthodromic AV reciprocating tachycardia and at least one syncope during tachycardia. Medical treatment with up to 3 drugs was unsatisfactil. During electrophysiologic studies only anterograde conducting accessory pathways could be found in all three patients in a septal position at the floor of the right atrium. Anterograde accessory path-way effective refractory period was 300 ms, 355 ms, and 310 ms, respectively. All were successfully treated with radiofrequency ablation. Conclusion: In patients with Fontan-like circulation suffering from arrhythmia, accessory pathways must be considered and treated by radiofrequency ablation. Furthermore, in patients planned for extra- or intracardiac caval conduits a accessory pathway must be excluded carefully or removed by ablation.

P290

Is the capillary distribution in hypoplastic left heart syndrome adequate? A morphometric analysis of the ventricular capillarisation

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In order to assess the quantitative differences between the distribution of capillaries in the ventricles of hypoplastic left heart syndrome and normal hearts, we examined the myocardium of 15 hearts and compared them with 5 aged matched controls. The capillaries were demonstrated using immunohistochemistry, staining the endothelium with von Willebrands Factor (Factor VIII). Using the technique of the 'closest unbiased method of analysis' we analysed the distribution of the myocardial capillaries. The hearts with hypoplastic left heart syndrome demonstrated that the mean and maximal capillary diffusion distance, from any arbitrary point in the tissue to a capillary, is significantly more than compared to normal hearts in both the left and right ventricle. This finding suggests that ventricular perfusion is inherently abnormal in hearts with hypoplastic left heart syndrome. In theory, unless formation of new capillaries accompanies growth, myocardial oxygenation can become a problem.

P291

The ventricular myoarchitecture in hypoplastic left heart syndrome

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To demonstrate the three dimensional architecture of the myofibres in hearts with hypoplastic left heart syndrome. 8 hearts representing the four morphological subtypes, namely: mitral atresia or stenosis combined with aortic atresia or stenosis were selected. Using fine dissection and serial photography the fibers were dissected from the epicardium inwards. The myoarchitecture of the left ventricle followed the same principles of distribution as normal hearts, however, the right ventricle showed the presence of a distinct middle layer, arranged in a circumferential pattern and sandwiched between the superficial and deep layers. The mechanism of development of this middle layer is not known but may be hemodynamically acquired or represent an inherent abnormality in the right ventricle of hearts with hypoplastic left heart syndrome. Either way, this may have significant implications for ventricular function following surgical reconstructive procedures.

P292

Unsuccessful hepatic conversion for pulmonary arteriovenous malformations after total cavopulmonary shunt placement

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Pulmonary arteriovenous malformations (PAVMs) is a well-known complication following total cavopulmonary shunt (TCPS) procedures. Some reports suggested that a resolution of PAVMs occurred following a completion Fontan procedure. We report a child who developed severe PAVMs after TCPS and underwent hepatic conversion, which resulted in no resolution of PAVMs. There was no report of hepatic conversion for such severe PAVMs. A girl who had polysplenia, interrupted inferior vena cava with azygos continuation, bilateral superior vena cava, and univentricular heart underwent pulmonary artery banding at 2 weeks and total cavopulmonary shunt (TCPS) placement at 10 months. Four months after the TCPS she developed cyanosis with extremely low arterial saturation of 23.7% under room air. Cardiac catheterization and lung perfusion scan confirmed diffuse PAVMs. She therefore underwent a completion Fontan procedure to redirect the hepatic vein by interposing a 14 mm tube between the hepatic venous orifice and right pulmonary artery. She was successfully weaned from cardiopulmonary bypass and extubated 7 days after surgery. Her arterial saturation was transiently elevated to almost 80% under 50% oxygen, but subsequently decreased to 60.8% under 50% oxygen 3 months after the surgery. Lung perfusion scan revealed almost unchanged PAVMs. Cardiac echo revealed an absence of flow in the tube, however, abdominal scan demonstrated an absence of dilatation of the hepatic vein. The existence of a well-developed collateral between the portal vein and the systemic azygos vein was suspected. In conclusion, a failure of hepatic conversion for severe PAVMs following TCPS can occur. The presence of collaterals from the portal vein to the azygos vein needs to be considered.

P293

Increased fibrosis in hearts with tricuspid atresia: a possible basis for ventricular dysfunction following fontan

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An abnormal ventricular function is considered one of the factors for the worsening aspects seen in the late follow-up of the Fontan circulation. It has been postulated that the myocardium in hearts with tricuspid atresia might be intrinsically abnormal. Methods: We examined microscopically 29 hearts with tricuspid atresia and 24 normal hearts (mean age: 9.6 months). For analysis, we considered two age groups: 0–2 months (13 hearts) and 2–30 months (16 hearts). Interstitial fibrosis was assessed by computer assisted morphometry (Quantimet Leyca) in the endocardial and epicardial halves of the myocardium at the tube, apex and outlet of left ventricle. Perivascular fibrosis was analysed separately and normalized to the vessel diameter. Results: The mean total interstitial collagen area fraction was greater in the malformed hearts than in controls in both age groups (p< 0.001) and increased with age (r= 0.53, p= 0.02). Above 24 months of age the total content of fibrosis was 7-fold greater than in controls. The tube and apex samples were more fibrotic than the outlet ones in hearts with tricuspid atresia (p=0.004), but not in the controls. Fibrosis was greater in the subendocardial compared with the

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

subepicardial zone in both age groups. Perivascular total collagen content was greater in the intes of the malformed hearts ($p<0.001$). Conclusions: Hearts with normal stress were more fibrosis than normal controls, even at younger ages. A disturbance of oxygen supply and demand with chronic tissue injury could result in increased fibrous content in the subendocardial zone. Regional differences seen in our study could be related to those seen in clinical studies. Such myocardial changes could explain, at least in part, the impaired ventricular performance seen in some patients following a Fontan-type procedure.

P294

Atrial volume and pulsatility after modified Fontan procedure in the long term period

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To evaluate the right atrial pulsatility after modified Fontan procedure, postoperative right atrial maximal volume (RAVmax) and right atrial ejection fraction (RAEF) were measured in 15 patients with biplane cineangiogram in the long term period (10.6 ± 4.2 years, average ± SD). Oblique parallel method with direct right atrial-pulmonary artery (RA-PA) anastomosis was performed in 8 patients (oblique group). Atrial septal defect (ASD) closure with direct RA-PA anastomosis was performed in the other 7 patients with tricuspid atresia (ASD group). RAVmax in the long term period was 61 ± 14% of normal in the oblique group, 116 ± 26% of normal in the ASD group. RAEF in the long term period was 24 ± 7% in the oblique group, 34 ± 6% in the ASD group. All 15 patients showed normal sinus rhythm. Another data acquired in the same time period regarding RAVmax and RAEF in the early period (about 5 weeks after operation) kept almost the same values of these RAVmax and RAEF in the long term period. Forward flow synchronized P wave was detected at the pulmonary artery by echocardiogram in the both groups. These data suggest that direct RA-PA anastomosis generates good RA pulsatility and better hemodynamics after operation in the long term period.

P295

Double inlet and double outlet right ventricle: anatomic variants and systematics

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Double inlet and double outlet right ventricle: anatomic variants and systematics. The following anatomic features are pathognomonic for double-inlet right ventricle (DIRV): (1) both AV fibrous rings open entirely (or mainly) in the cavity of morphologically right ventricle (RV); (2) there is a rudimentary left ventricle (LV) in the ventricular segment of the heart. We describe the results of angiocardiographic and echocardiographic studies of 31 patients. Age ranged from 5 months to 11.5 years. 5 – male. There was vascular atrioventricular discordance in all cases. With the anatomy of the heart segments as a guide we identified the following anatomic types (Table 1). The spectrum of associated anomalies of the patients with DIRV was the following: PA stenosis (n=7), anomaly of AV valves (n=7), ASD (n=8), PDA (n=3), multiple VSD (n=1). Conclusion: We have 6 anatomic variants of DI-DO RV. Most frequently the pathology associates with pulmonary stenosis (64%) and malformations of AV valves (58%).

P296

Hemodynamic and angiographic evaluation of patients with bidirectional Glenn anastomosis for a possible Fontan procedure

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Background: Bidirectional Glenn Shunt is often performed as a bridge to the Fontan procedure in patients with functional single ventricle who are considered high risk for the primary Fontan surgery. However it may be the ultimate form of palliation in certain patients who remain at high risk for Fontan procedure. Methods: We analysed the hemodynamic and angiographic data in 44 patients who had undergone bidirectional Glenn shunt between 1993 and 1999 at our center. Their age at the time of surgery ranged from 6 months to 25 years with a mean of 45.9 +/- 59.8 months. Risk factors for primary Fontan procedure were : age below two years (n=24), small size and/or abnormal anatomy of pulmonary arteries (n=13), pulmonary arterial hypertension (n=7), ventricular dysfunction (n=10), atrioventricular valve regurgitation (n=4) and need for associated procedures like repair of total anomalous pulmonary venous drainage, pulmonary arteroplasty (n=10). Two or more of these adverse factors were present in 15 cases. Results: Postoperative cardiac catheterization and angiography performed 5 months to 5 years (mean 27 +/- 13 months) after the Glenn shunt revealed favourable data for completion of Fontan procedure in 31 of these 44 cases. Young age was the only reason for doing the Glenn shunt initially in 18 of these 31 patients. However 13 cases were not considered suitable for Fontan procedure, either due to small size and/or abnormal anatomy of pulmonary arteries (n=8) or significant ventricular dysfunction (n=5). Twelve of these 13 patients were in functional class I or II. Conclusion: Majority of cases who had undergone a bidirectional Glenn Shunt are suitable for a Fontan procedure subsequently. However, Glenn shunt may be the final form of palliation in a small proportion, who were considered high risk for Fontan procedure due to factors other than a young age.

P297

Long-term outcome after Senning operation

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The purpose of this study was to determine the incidence of sinus node dysfunction (SND), atrial arrythmias, right ventricular dysfunction (RVD) and late sudden death in the long-term follow-up of a group of patients operate on with Senning procedure for complete transposition of the great arteries (TGA). Patients and methods: From november 1978 to november 1987 73 consecutive patients underwent the Senning operation for TGA. The 70 survivors have an average follow-up of 16 years (11–20 years). A standard 12-leads ECG an echocardiographic study and a 24-hours Holter monitoring were performed once or twice a year. Results: 1) cardiac rhythm: with the time there have been a progressive fall in the stable sinus rythm with appearance of junctional rhythm and supraventricular tachyarrythmias. At 5 years of follow-up 86% of patients had sinus rhythm, this percentage decreases on 80% at 10 years, 55% at 15 years and 40% after 20 years. The active arrythmias requesting therapy were present in 3% at 10 years, in 5% at 15 years and in 10% at 20 years. 2) Right ventricular function: 20% of the patients had reduced ejection fraction of the right ventricle (< o > 45%) by echocardiography. 3) late mortality: no late death until 1991 (mean follow-up 9 years); in the last 9 years 2 sudden deaths (2.8%) occurred. 4) Functional status: 80% was in NYHA class I, 17% in class II and 3% in class III. Conclusions: Our results confirm that the Senning patients have a progressive loss of sinus rhythm, increase in active arrythmias and other important problems as late sudden death and decrease of right ventricular function, however most of these patients are alive (93% in our series) and in good functional status.

P298

Is pulsatility of benefit after bidirectional Glenn shunt?

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In patients with univentricular physiology, bidirectional Glenn (BDG) is an important stage before TCPC. In developing countries, it is the final surgery in many patients who are unable to afford another surgery. In the premise that leaving pulsatile flow allows for better saturation (SaO2), thus reducing the need for conversion to TCPC, many patients undergo pulsatile BDG (PBDG). The aim of this study is to compare outcome after PBDG Vs non (N)PBDG. We reviewed records of 330 patients with functionally univentricular hearts operated between 1988–98. 125 patients underwent BDG. Among them, 94 had BDG as the first stage. BDG alone was performed in 13.9%, 51.6% underwent BDG + atrial septectomy and 9.8% underwent PA reconstruction + BDG. Rest of the patients underwent either a BT shunt (17.2%) or PA banding (5.7%) prior to BDG. 1.6% underwent Kawashima repair. Results: Group I consisted of 88 pts with PBDG and 37 patients who underwent NPBDG formed Group II. In group I, 88% had PS and 12.9% PAH. In group II, 40% had PS, 40% PAtresia and 11.4% PAH. In the early postoperative period, Group I patients had higher incidence of pleural effusion (6 Vs 4), arrhythmias (5 Vs 1). The in hospital mortality was higher in group II (11.3% Vs 8.1%). On late follow up, group I pts. had higher SaO2 ($p<0.05$), higher incidence of venovenous collaterals (6 Vs 4) and arteriovenous malformations (PAVMS (2 Vs 0). Conclusions: We conclude that, though patients with PBDG had better SaO2, they have a higher incidence of pleural effusions, venovenous collaterals and PAVMs.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P299

Phenoxybenzamine prevents hemodynamic deterioration at high arterial saturation in neonates after the Norwood procedure for HLHS

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Mortality for HLHS remains high because of impaired myocardial function and inefficient parallel circulation even after palliation by the Norwood procedure (NP). Traditional management has attempted to limit pulmonary overcirculation, presumably detected by high arterial saturation, by induction of hypercapnia-hypoxia pulmonary vasoconstriction. This strategy has obvious limitations in that arterial saturation is a major determinant of venous saturation. We have previously reported higher venous saturation, lower systemic vascular resistance, lower Qp/Qs, and improved survival with the perioperative use of phenoxybenzamine (PBZ) and continuous venous oximetry. In this report, we provide evidence that PBZ prevents runaway pulmonary overcirculation, thereby eliminating the need for limitation of arterial saturation. Neonates undergoing the NP received PBZ 0.25 mg/kg or placebo on CPB in a randomized prospective study design. Perioperative hemodynamic management targeted SvO2>50%. Prospectively acquired hemodynamic data was used to assess the effect of PBZ on the relationship between arterial and venous saturation, arterio-venous oxygen content difference (Da-vO2), and Qp/Qs during postoperative hours 1-48. Data was analyzed across intervals of SaO2 from 60% to 95% and between groups using repeated-measures ANOVA with Tukey's WSD post-hoc comparisons, with $p<0.05$ considered significant. Data from 71 consecutive patients and 2826 patient hours was available for analysis. 61 patients received PBZ, 10 who did not served as controls. In control patients, SvO2 peaked at an SaO2 of 90%, with reduced SvO2 at SaO2>90% and SaO2<70% ($p<0.01$), while Da-vO2 increased with SaO2>80% ($p<0.0001$). In patients receiving PBZ, the SvO2 increased linearly with SaO2>65% ($p<0.0001$), and Da-vO2 was constant at all SaO2 ($p=ns$). The SvO2 was higher, and the Da-vO2 lower, across the whole SaO2 range with PBZ ($p<0.0001$). These data show that PBZ eliminates the critical range of SaO2 for optimizing systemic oxygen delivery, specifically by eliminating the systemic hypoperfusion associated with high SaO2. This effect makes higher SaO2 a useful target for postoperative management in patients receiving PBZ for NP.

P300

Univentricular heart: medium to long term survival in a group of 155 patients

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The objective of our study was to evaluate the medium to long term survival of 155 patients (81 males, 74 females), affected by either univentricular heart or tricuspid atresia (TA), observed in our institution over a time period extending from 1974 to 1997. A hundred-forty pts. (90.3%) had situs solitus (94 with double inlet left ventricle and 46 with TA), thirteen pts.(8.4%) situs ambiguus, 6 (3.8%) right isomerism and 7 (4.6%) left isomerism and two pts (1.3%) situs inversus. Left ventricular morphology was present in 123 pts (79.2%), 77 (49%) with DILV and 46 (29.6%) with TA. A right ventricular morphology was present in 29 pts (18.7%) and 3 (2%) had an undefined morphology. Forty-nine patients (31.6%) had a discordant ventriculo-arterial connection. All patients had associated cardiac anomalies: 58 (37.4%) pulmonary stenosis, 33 (21.3%) pulmonary atresia, 25 (16%) aortic or subaortic stenosis, 11 (7%) anomalies of pulmonary venous drainage. Of the 155 pts., 138 (89%) underwent surgery at least once: 77 pts. (group A) underwent a Fontan operation or a total cavopulmonary anastomosis, while 147 pts.(group B) received merely palliative surgery, including systemic to pulmonary shunting in 109(80.9%), pulmonary artery banding in 27(19.5%), stent-angioplasty in 25(18.8%); Group C comprised seventeen pts. (10.9%) who were not treated surgically. Patients were followed-up for 8±9.7 years. Six of the 155 pts. were lost at follow-up. In conclusion, 72 pts. (46.4%) died, 3 (1.9%) were judged inoperable or consent was not given; 30 pts. (19.3%) died in the postoperative period (0-30 days), 39 pts. (25%) in the successive period. By the end of the follow-up study 77 pts. (49.7%) were still alive, median age was 14±11 years (2-59 years); Of those, 59 (76.6%) received definitive treatment (group A), while 13 (16.9%) received palliative treatment(group B) and 5 (6.5%) received no surgical treatment (group C). Survival was significantly different ($p<0.01$) between groups A+B (62% and 47% respectively at 5 and 20 year follow up) versus group C (46% and 25% respectively at 5 and 20 year follow-up). There was a significant difference in 5, 10 and 20 year survival between groups A and B (81% versus 30%, 74% versus 24%, 40% versus 21% respectively) $p<0.0001$. Conclusions: Patients with a univentricular heart for whom no operative approach was taken had very poor survival at 5 to 10 years follow up. Palliative surgical treatment while improving short term prognosis, did not alter the long term prognosis. Fontan operation or total cavopulmonary bypass provided the greatest survival rate (>70% at 15 years follow-up). However, the improvement in survival observed in group A rapidly declines 20 years post operatively, thus a strict clinical follow-up is necessary.

P301

Clinical and echocardiographic results with the extracardiac pedicled pericardial Fontan operation.

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Objective: Despite improved results after standard Fontan operations, morbidity remains high. We report midterm results using a simple viable pericardial extracardiac tunnel (PET) technique. Methods: Since 1996, 23 patients (14M/9F) with univentricular physiology underwent the PET Fontan using cardiopulmonary bypass and mild hypothermia (34 degrees C) and no myocardial arrest (96 +/- 24 min). Results: There were no deaths. Serious perioperative complications occurred in 4 patients (17.4%) including, bleeding (n=3) and laryngeal edema (n=1). Pleural effusions (> 14 days) occur in 5 patients. Hospitalization was 11.0 3.4 days and ICU stay was 3.6 +/- 1.5 days. Follow up was 100% (n=23) mos and showed all patients in NYHA class I with no evidence of delayed TE events, effusions or critically affected exercise tolerance. One patient had stenosis at the IVC anastomotic site (20 mo) and 1 patient required subxiphoid intervention (18 mo) for LVOT obstruction. Serial EKG and Holter evaluation revealed no arrhythmias and echo-cardiographic follow up disclosed atrial/ventricular function, AV valve function and evidence of growth of the tunnel. Conclusion: The extracardiac tunnel cavopulmonary connection using viable pericardium is a safe and simple operation that results in low perioperative morbidity. Furthermore, growth of the tunnel may decrease delayed morbidity of conventional Fontan procedures.

P302

Evolutionary trends in univentricular repair over one decade

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Five hundred and eighteen patients with a functional single ventricle (194 with tricuspid atresia and 324 non tricuspid atresia) underwent univentricular repair since January 1980. A nearly equal distribution between atriopulmonary connection and total cavopulmonary connection upto 1992 gradually changed to a policy of TCPC as the only used method for univentricular repair from Jan 1994. Since this time a policy of routine fenestration of the intra atrial baffle also was followed. That the total experience includes 405 TCPC, of which 296 have received a fenestration. Lately, the extracardiac (synthetic, n=15) and pedicled pericardial (n=3) TCPC has been added to the armamentarium. Overall Fontan failure rate has been 14 % and effusions occurred in 27%. Fenestration of the baffle appears to decrease both Fontan failure ($P=0.002$) and effusions ($p<0.001$). There have been 12 late deaths in a mean follow up period of 47 +/- 17 months (range 6 to 154 months). The extracardiac TCPC has done well over the short term with respect to Fontan failure and effusions. However, larger numbers and longer follow up is required before it can be recommended for widespread use.

P303

Pulmonary stenosis after arterial switch operation for TGA – efficacy of the nontreated autologous pericardium

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One of the major sequelae after arterial switch operation for transposition of great vessels is pulmonary stenosis. That is to evaluate the efficacy of non-treated autologous pericardial patch in the reconstruction of the neopulmonary artery. We operated 70 consecutive babies with transposition of great vessels with intact ventricular septum from April 1991 to October 2000 at our center. Among them 66 cases were neonates and 3 cases were older than 2 months of age. All patients were repaired by using non-treated autologous single pericardial patch. There were 3 hospital deaths and 2 late deaths. All survival cases were followed up without loss with a mean 43.7+/-30.7 months (range,

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

from 2 to 111 months). We indicated significant pulmonary stenosis as more than 2m/sec velocity by Doppler echocardiography at the time of discharge. There were 6 cases (9.1%) of pulmonary stenosis according to the criteria. Among them 5 cases showed spontaneous regression of pressure gradient, and only 1 case with 90mmHg pressure gradient confirmed by cardiac catheterization underwent operative repair 5 years postoperatively. In our experience, untreated autologous pericardial patch for the reconstruction of unipulmonary artery in the repair of simple transposition showed reliable and durable repair in midterm follow up. Video demonstration will be also included.

P304

Bidirectional cavopulmonary anastomosis reduces pulmonary artery banding induced systemic ventricular hypertrophy in Fontan candidates

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Ventricular hypertrophy has been suggested as one of the risk factors in Fontan procedure. Severe hypertrophy of the systemic ventricle develops in patients who have had pulmonary artery banding (PAB) before Fontan procedure. We sought to identify the ventricular response of patients with previous PAB and the effects of bidirectional cavopulmonary anastomosis (BCPA) on ventricular response in these patients. Since 1990 thirty-five patients underwent Fontan procedure in our institute (median age: 5 years). In 14 patients who had previous PAB, 9 patients underwent Fontan procedure in one stage (PAB-1 group) and 5 patients underwent BCPA prior to Fontan procedure (PAB-2 group). In 21 patients who underwent cavopulmonary shunt or had native pulmonary stenosis, 7 patients had Fontan procedure in one stage (NPAB-1 group) and 14 patients had staged Fontan procedure (NPAB-2 group). The following hemodynamic parameters at cardiac catheterization prior to the Fontan procedure were analyzed retrospectively in each group: ventricular end-diastolic volume index (EDVI), ventricular mass index (VMI), mass-volume ratio (VM/EDV) and end-diastolic pressure (EDP). There was no significant difference in EDVI and EDP among the groups. In PAB-1 group, VMI (152+/-33 g/m2) and VM/EDV (0.93+/-0.13) were significantly higher than those in NPAB-1 group (94+/-26 g/m2, 0.66+/-0.11) and NPAB-2 group (82+/-30 g/m2, 0.73+/-0.14). However, PAB-2 group showed significantly lower VMI (101+/-19 g/m2) and VM/EDV (0.78+/-0.10) compared to PAB-1 group, where these values were equivalent to those in NPAB-1 and NPAB-2 group. In conclusion, PAB did induce ventricular hypertrophy and BCPA can reduce the ventricular hypertrophy induced by PAB in Fontan candidates.

P305

Suitable shunt size for regulation of pulmonary blood flow in a canine model of the univentricular heart

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Objective: We examined the influence of shunt size on regulation of the pulmonary blood flow using a canine model of a univentricular heart, because the specific guidelines of suitable shunt size in Norwood operation remained to be determined. Methods: Female Beagle dogs (n=8), 3–7 months old and weighing 3.3–5.0 kg, were used. Atrial septectomy and patch closure of the tricuspid valve were performed and a systemic-to-pulmonary arterial shunt was created by interposing a 3.5 or 4.0 mm polytetrafluoroethylene (Gore-tex) graft between the origin of the right subclavian artery and main pulmonary artery. Hemodynamic variables, including pulmonary blood flow and systemic blood flow, were measured consecutively and systemic arterial blood gas analysis was performed simultaneously while voluntarily changing the respiratory conditions. The shunt size (mm)/body weight (kg) ratio ranged from 0.8 to 1.3 (mean 1.1). Results: Negative correlation between the pulmonary/systemic flow ratio and arterial carbon dioxide tension was found when the shunt size/ body weight ratio was smaller than 1.1, but not found when the ratio was larger than 1.1. As inspired oxygen tension became higher, the pulmonary/systemic flow ratio increased significantly when the shunt size/ body weight ratio is smaller than 1.1, but no fixed relationship was found when the ratio was larger than 1.1. Conclusion: It is concluded that when the shunt size/ body weight ratio is smaller than 1.1, the pulmonary/systemic flow ratio is controllable by respiratory manipulation. However, the larger shunt makes pulmonary flow excessive and uncontrollable in the parallel circulation after Norwood operation.

P306

Staged Fontan operation for Single Ventricle With Subaortic Stenosis – Damus-Kaye-Stansel/ Norwood operation or Pulmonary Artery Banding as First Palliation

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Single Ventricle (SV) with Subaortic stenosis (SAS) is difficult subset of patients to achieve successful Fontan operation. Between June 1993 and November 2000, fifteen patients with SV/SAS were enrolled Fontan protocol. 9 patients underwent PA banding +- CoA repair (Group P) and 6 patients underwent Damus-Kaye-Stansel (DKS)/Norwood operation as first palliation (Group D). Indication for DKS/ Norwood operation is diameter of Ascending Ao or LVOT less than (Body weight +Thirty and/or subaortic ratio potentially cause SAS rapidly. LVOT/AoV/Asc Ao diameter (mm) were 3.8+-0.5/5.3+-0.5/ 6.0+-1.0/ in Group D and 7.6+-3.2/7.9+-1.8/9.3+-5.3/ in Group P respectively. Age at first palliation were 24+-11 days in Group D, 92+-8 days in Group P. In Group D, 1 patient with Coarctation died due to progressive PVO. 4 patients proceeded with bidirectional Glenn (BDG) operation at age 6 PA 13.1 months, and 1 patient is waiting for BDG. Fontan operation was performed in 2 patients and another 2 patients are waiting for Fontan as good candidate. In Group P, there was no early and late mortality. 8 patients underwent DKS/BDG operation as second palliation and 1 patient underwent DKS/Fontan without second stage. Mortality rate for entire protocol was 6.6%. Conclusion: Excellent result could be achieved in Fontan protocol for SV with SAS utilizing DKS/Norwood or PA banding as first palliation and early conversion to DKS/BDG before developing SAS to the patients after PA banding.

P307

Operative results of Fontan operation for complex hearts: Importance of staged strategy for isomerism heart

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Introduction: Many patients with complex heart (IH) have atrioventricular valve regurgitation (AVVR) and/or pulmonary vein stenosis (PVS) which may prevent these patients from reaching to Fontan operation. We have introduced repair of AVVR and PVS, and other strategies to Fontan operation for IH. Operative results of Fontan operation for IH were reported and compared with those for non-IH. Patients: Among consecutive 30 patients who underwent Fontan operation (age 1.5 to 9.1 years), nine patients have IH with single ventricle and common atrioventricular valve (CAVV). Operation for AVVR: Valvular separation of CAVV into two valves was effective to reduce AVVR in 3 patients. Other types of valvuloplasty were done in 5. These operations were done prior to Fontan operation in 2 patients and done concomitantly with Fontan in 4. Operation for PVS: Cuts of pulmonary veins often show PVS by hypertrophic atrial wall itself in IH. Resection of atrial wall covering ostia of the veins was done prior to Fontan operation to 2 patients and done with Fontan in 3. Other Strategy in IH: Bidirectional Glenn procedure was applied to 8 patients prior to Fontan. Extracardiac conduit was used in 8 patients. Operative Results: There were 1 operative death (high pulmonary vascular resistance) and 1 late death (sudden) in IH group. Central venous pressure during the early postoperative period was almost same between IH and non-IH (12±2mmHg vs. 11±3mmHg). Conclusion: It is very important to improve AVVR and PVS in IH to achieve Fontan circulation. These procedures enable reduction of ventricular overload and pulmonary vascular resistance, which are essential factors for Fontan circulation. Staged strategy for Fontan operation for IH showed good operative results compared with the results for non-IH.

P308

Distribution of pulmonary blood flow in the presence of bilateral superior caval vein after total cavopulmonary anastomosis – potential clinical implication

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The pulmonary blood flow distribution after total cavopulmonary anastomosis(TCPA) may be influenced by surgical modifications and associated anomalies, such as persistent left superior vena cava(SVC), but adequate evidence for the appropriateness of the conventional surgical method is lacking. To assess the appropriateness of the conventional surgical method and to evaluate the influence of bilateral SVC on the distribution of pulmonary blood flow after TCPA, we investigated the pulmonary arterial growth, hemo-

dynamic data and the pulmonary blood flow distribution in 41 patients(M:F=23:18, age:54.9±45.6 months) who were followed-up by cardiac catheterization, angiography and lung perfusion scan 35.1±40.0 months after the operation. The cross-sectional area index of the pulmonary artery in regard to the variability of body surface area was reduced after the operation(298±128 vs 195±77, p<0.05). A larger amount of pulmonary blood flow was distributed to the ipsilateral side of IVC than the contralateral side, with an ipsilateral to contralateral perfusion ratio(i/cPR) of 1.05±0.58. Comparing the subgroups by the type of superior vena caval inflow, the unilateral superior cavopulmonary anastomosis(SCPA) group showed significantly higher i/cPR(1.57±0.79) than the bilateral SCPA group(0.96±0.52, p<0.05). We concluded that the bilaterality of superior vena cava may have a major influence on the distribution of the pulmonary blood flow and the details of surgical methods should be evaluated case by case in respect to the associated anomalies in order to achieve adequate postoperative pulmonary blood flow distribution.

P309

One and a half ventricle repair for complex congenital anomalies

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Objective:one and a half ventricle repair is a surgical option for congenital cardiac anomalies characterized by right ventricle (RV) hypoplasia and/or dysplasia. Methods: from March 1994 to November 2000, 7 patients (mean age 10.4 years, range 7 months–35 years) with hypoplastic and/or dysplastic RV underwent correction of their intracardiac anomalies in association to bi-directional cavo-pulmonary shunt (BCPS). Diagnosis included Ebstein anomaly (2 pts), PA with IVS (1 pt), CAVC+TOF (1 pt), heterotaxy sindrome with VSD and systemic venous return anomalies (1 pt), truncus arteriosus (1 pt), VSD with straddling tricuspid valve TV(1 pt). Previous surgery included pulmonary valvotomy, central shunt and RVOT reconstruction. PA banding and PA separation with Blalock-Taussig shunt (MBTS). Cardiac procedures associated to BCPS included DV reconstruction (3 pts),VSD closure (3 pts), ASD closure (2 pts). CAVC+TOF repair (1 pt) supra-hepatic vein-right atrium tunnel (1 pt), RVOT reconstruction with heterograft (1 pt), MBTS takedown and PA plasty (1 pt). BCPS alone was performed in 1 patient. Results:there were no hospital death. All patients were discharged home asymptomatic and well. At a mean follow up of 29 months (range 1 month–6.0 years), all patients are in good hemodynamic conditions. There were no late deaths or reoperations. Pulmonary artery balloon dilation was performed 24 months after CAVC+TOF repair in one patient. Conclusions. BCPS in association to repair of complex malformations with hypoplastic/dysplastic RV is a low risk procedure which allows separation of pulmonary and systemic circulation, maintaining a pulsatile flow in pulmonary arteries and a low IVC pressure. Venous hypertension in SVC is well tolerated and no complications are reported in the midterm. Long term follow up is needed to evaluate the effect of pulsatile flow in pulmonary arteries together with a continuous systemic flow.

P310

Aortic growth in patients with aortic stenosis or atresia following Norwood operation

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Aim To assess ascending aortic growth after Norwood palliation in patients with aortic atresia (AA) versus aortic stenosis (AS). Patients and methods: Fifteen patients with hypoplastic left heart syndrome with AA (n=12) or AS (n=3) had serial measurements of ratios of aortic (ascending and descending aorta (AAO, DAO)) and pulmonary dimensions (PA) at the time of cardiac catheter. Mean age at first assessment prior to cavopulmonary shunt was 0.2 years (0.15 – 0.42), and 3.6 years (3.0 – 4.7) at second assessment prior to Fontan operation. Ratios of AAO/PA, AAO/DAO, PA/DAO were calculated to elevate age and size independent variables. Statistical analysis was performed using student t-test. Results: Mean ratios of AAO/PA in aortic atresia patients were 0.32 (SD 0.06) initially and decreased to 0.26 (0.04) [p<0.02]. AAO/PA was higher in aortic stenosis patients at first (0.42 (0.09) [p<0.05]) and second assessment (0.34 (0.02) [p<0.06]). There was no significant difference of PA/DAO ratios amongst the two groups and the time of assessment [first: 2.02 (0.28) vs 2.06 (0.35), second: 1.85 (0.65) vs 1.66 (0.06)]. There was no significant difference of AAO/DAO ratios between the two groups at first assessment (0.60 (0.14) vs 0.76 (0.13)). However, there was a significant difference of AAO/DAO ratios between the two groups at second assessment (0.51 (0.11) vs. 0.80 (0.02), [p<0.005]). Discussion: In patients with aortic stenosis the growth of the ascending aorta is maintained whereas in patients with aortic atresia its growth falls further behind, as judged against the growth of the PA and DAO. This may have implications for coronary perfusion and thus long-term right ventricular performance.

P311

Right ventricle-pulmonary artery conduit as a pulmonary blood supply for neonates with hemodynamically single right ventricle

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Maintenance of high diastolic systemic blood pressure is thought to be necessary to keep adequate coronary blood flow in particular for neonates. Instead of systemic pulmonary shunt, we performed right ventricle-pulmonary artery (RV-PA) conduit procedure as a pulmonary blood supply for neonates with hemodynamically single right ventricle. We evaluated the effects of the conduit operation on the early postoperative hemodynamic condition and the outcome. Between 1991 and 2000/11, RV-PA conduit was placed in 10 neonates with HLHS [age: 3 to 19 days] and in 4 infants with single right ventricle and pulmonary atresia [asplenia, age: 20 to 38 days]. A handmade semipericardial roll and an ePTFE graft were used as the conduit in 8 and 6 cases, respectively. The diameter of the conduit was from 4 to 6.5 mm with the median of 5 mm. The most concomitant operative procedures were aortic arch reconstruction in 10 (HLHS and TAPVC repair in 3 asplenia patients. There was no operative death. All cases maintained stable hemodynamic condition with high diastolic systemic blood pressure during the early postoperative period. Conduit stenosis occurred in all except 2 cases 1 to 5 months after the operation followed by conduit replacement in 2, additional BT shunt in 2, Glenn procedure in 6, and transcatheter balloon dilatation in 4 cases. Of 7 presently alive patients (4 HLHS, 3 asplenia), 3 are candidates for Fontan operation and another 2 are waiting for Glenn procedure. Even though early conduit stenosis occurred, RV-PA conduit procedure brought stable hemodynamic condition early after the operation in all patients and may be beneficial as a short term pulmonary blood supply for neonates with single right ventricle.

P312

Borderline hypoplastic left heart malformations: Norwood palliation or two-ventricle repair ?

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Hypoplastic left heart syndrome (HLHS) and associated malformations are treated with Norwood staged palliation. Some patients, however, are potential candidates for a two-ventricle repair. We report our experience with both surgical strategies. Since 8/99, 12 patients presented with HLHS physiology. Eight patients with typical HLHS and 1 with DILV, (S,L,L) and arch obstruction underwent Norwood/Fontan palliation (group I). Anatomy in the other 3 pts (group II) was: pt 1: aortic atresia, 2mm ascending aorta, coarctation, two normal sized ventricles and AV-valves, unrestrictive VSD; pt.2: Shone complex, aortic valve 4.5mm, Z-score -2, ascending aorta 4.5mm, hypoplastic arch (2–3mm), coarctation, mildly hypoplastic LV, MV 7.5mm, Z-score -2; pt.3: aortic stenosis, bicuspid valve (5mm, Z-score -1.5), hypoplastic arch (3mm), coarctation, MV 7mm, Z-score -1.5, mildly hypoplastic LV, tiny VSD. Age at surgery was up to 3 weeks. Pts. in group I underwent (Norwood/Fontan pathway. Two patients died due to RV-dysfunction with severe TR (14d p.o.) and sepsis (40d p.o.). Five pts. underwent Glenn procedure successfully. Surgical procedures in group II were: pt 1: Norwood reconstruction of aortic arch, Rastelli with VSD-patch-closure connecting LV to neoaorta, RV to PA homograft; pt. 2: aortic arch reconstruction with homograft patch, ASD-closure; pt.3: aortic arch reconstruction with homograft patch, aortic valve commissurotomy, ASD-closure. Postoperative course was uneventful, except elevated LAP (<20 mmHg) in pts 2 and 3 for 24 h. Follow-up echocardiography showed an LVOT-gradient of 20 mmHg in pt 3, no other pathological findings. The Norwood concept has proven its validity in the treatment of hypoplastic left heart malformations. However, some anatomical subsets with borderline MV and small LV may undergo two-ventricle repair despite severe LVOTO. Mortality and morbidity seem to be lower. Further experience is necessary to define selection criteria for a two-ventricle repair.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P313

Morphologic and Clinical Spectrum of the Rare Anatomic Subgroup of {S,D,L} Segmental Complexes

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The anatomic subtypes of hearts with the segmental subsets of {S,D,L} ie solitus–atrial situs solitus, D-looped ventricles, and L-malposed great arteries, comprise of a rare but heterogeneous group and present important diagnostic and surgical issues. In this study we analyzed 14 such patients evaluated at our institute between January 1999 and October 2000. 11 patients belonged to {S,D,L} and 3 to {A,D,L} subset. There were 10 male and 4 female patients. Mean age was 27 months (range 2–144months). Saturations ranged from <75% in 8(57%), 75–92% in 3(21%) and >92% in 3(21%). 42% cases had associated cardiac malposition. A VSD was present in all cases: conoventricular in 43%, conseptal in 29% and single ventricle (large) in 21% of the cases. Pulmonary stenosis was present in 93% of the patients. Ventricular abnormalities were common, including double outlet right ventricle in 36%(5), hypoplasia of the right ventricle in 36%(5), supero-inferior ventricles in 36%(5) and criss-cross AV connections in 14%(2). Other common associated findings include atrial septal defects in 57%(8), juxtaposed atrial appendages in 29%(4), atrioventricular valve (AVV) straddle in 14%(2), AVV atresia and a common AVV in 1(7%) patient each. Normal systemic venous return (71%) and pulmonary venous return (86%) was found in most cases. 4/14 patients) underwent surgical intervention, 2 had biventricular repair (arterial switch and Rastelli procedure), 1 each had a modified BT shunt and bidirectional glenn shunt. One patient with physiologically corrected (normal) circulation had a spontaneously closing VSD. There was one death due to low cardiac output. In conclusion, the segmental subset of {S,D,L} hearts comprise of a heterogeneous group of patients with varied presentation. Proper delineation of their anatomy is essential for understanding their varied physiology and planning management.

P314

Circulatory arrest versus antegrade cerebral perfusion in the Norwood procedure – a retrospective comparative study

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Many working groups demonstrated, that even short periods of deep hypothermic circulatory arrest (DHCA) lead to measurable neurologic injury. Reducing DHCA should not complicate the procedure or prolong bypass time. Since 1997 33 unselected consecutive patients underwent a Norwood procedure. Group 1: in 17 consecutive patients (14 HLHS, 3 single ventricle) DHCA was used for aortic arch reconstruction. Group 2: in 16 consecutive patients (14 HLHS, 2 single ventricle) antegrade cerebral perfusion via a mod. BT-shunt was used for aortic arch reconstruction and DHCA was restricted to excision of the atrial septum. DHCA time was 51.8+/-11,1min in group 1 versus 3,2+/-1,5 min in group 2. Examined parameters: survival, total bypass time, serum lactate levels, ICU stay, total hospital stay, neurologic events. Survival: group 1: 10 pts (59%), group 2: 12 pts (75%); mean bypass time: group 1: 202+/-30min, group 2: 197+/-15min; no serum lactate levels (6.8 +/-4.2mmol/l in group 1 versus 5.9 +/-2.3mmol/l in group 2: p = 0.56). No sign. difference resulted in length of hospital stay and ICU stay. No sign. residual anatomic lesion was seen in any group. There was 1 stroke in group 1, no neurologic event in group 2. Replacing circulatory arrest by antegrade cerebral perfusion via the mod. BT-shunt does not complicate or prolong the Norwood procedure, has no negative influence on the anatomic result and on the length of stay. Survival might be significantly improved in a larger patient population.

P315

Plasma hormonal and renal water–electrolyte excretion responses to water loading in patients with one ventricle heart: influence of type of surgery

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Background: A major long term complication in patients with one ventricle heart is salt and water retention which eventually becomes resistant to diuretics and hence contributes to morbidity. Methods: Plasma hormonal and renal water-electrolyte excretion response to water loading were studied in 19 patients with one ventricle heart, 10 aged 30±9 years with Fontan type repair and 9 aged 37±10 years with aorta to pulmonary shunts. All patients received an oral water load of 10 ml/kg after an overnight fast. Blood samples and all urine passed were collected before (T0), immediately (T2), one hour (T3) and two hours (T4) after water loading. Results: Free water clearance was impaired and hormonal levels was raised and neither changed with water loading. On the other hand there a water-electrolyte excretion response only in GI patients. Conclusion: Although the right ventricle is absent in Fontan repair for one ventricle heart the circulatory response to water loading is better than non-Fontan.

P316

Early and intermediate-term results of Norwood stage 1 operation

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To assess early and intermediate-term outcomes of Norwood stage I operation, a retrospective study of 42 consecutive patients who underwent a Norwood stage I palliation between Oct. 1996 and Nov. 2000 was conducted. Postoperative survival at 1 month was 88%. Hospital survival was 72%. Between the survivors and non-survivors, there were no significant differences in parameters including age at operation, body weight, size of the ascending aorta, circulatory arrest time, and aortic cross-clamp time. However preoperative hemodynamic instability manifested as metabolic acidosis was associated with higher hospital mortality. Restrictive ASD resulted in adverse early mortality. During a median follow-up period of 20 months (range 1–49), 21 patients underwent bidirectional cavopulmonary anastomosis with 95% of survival. 5 Fontan with 100 % of survival.

P317

Growth velocity of infants with HLHS. A comparison of enteral feeding strategies

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To examine postoperative nutritional support patterns and their impact on growth velocity (GV) in infants with HLHS, a retrospective chart review of 28 consecutive pts undergoing the Norwood procedure (NW) and surviving to bidirectional Glenn (BDG) palliation was performed. Feeding strategies and growth measurements from birth to BDG were recorded. Pts were classified as entirely oral fed (OF, n=15), combination oral and tube fed (OTF, n=8), and entirely tube fed (TF, n=5) based on feeding method at discharge following NW. Groups were statistically similar at birth weight (wt), sex, age, size, CPB, and circulatory arrest time at NW. Caloric intake (70–132 kcal/kg/d; mean 109), fluid volume (95–166 cc/kg/d, mean 135), and calorie density (20–30 kcal/oz, mean 24) at discharge were similar between groups. Hospital length of stay (LOS) was significantly longer for TF pts compared to OF (62.2±29.7 vs 24.8±7d, p<0.001). Twelve of 28 pts failed to regain birth wt prior to discharge; this was most likely in OF pts (7/15; p<0.01). Wt gain in grams/d during hospitalization was also lowest in OF (-2.6 vs 6.6, p=0.03), but OF demonstrated the best wt gain following discharge (20.9 vs 15.0, p<0.03) and had the greatest overall GV compared to the OTF and TF groups (p<0.001). No significant differences were found between the OTF and TF groups. We conclude that achievement of full oral feedings is associated with shorter LOS and greater growth velocity in infants with HLHS following NW. Although OF infants struggled initially on growth ability on feed orally appears to be an important indicator of wellness in this population. We speculate that necessity for tube feeding after NW identifies a population at risk who may benefit from earlier BDG rather than prolonged feeding supplementation.

P318

Hemodynamic response to esophageal pacing in patients with fenestrated fontan, sick sinus syndrome, junctional rhythm, and brisk retrograde AV conduction

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INTRODUCTION: The lack of atrio-ventricular synchrony (AVS) and normal heart rate are known to affect the hemodynamics of Fontan patients more than patients with normal structural heart. However, difficult venous access, increased risks from thoracotomy for epicardial leads, bleeding from anti-coagulation, extracardiac anomalies, and arrhythmia complicate the decision for pacemaker implantation. Predictive factors for the hemodynamic effect of atrial pacing for junctional rhythm (JR) in fontan patients are unclear. METHOD: Esophageal pacing (EsP) was performed in 5 fenestrated Fontan patients with JR and brisk retrograde P waves, to establish AVS and normal heart rate. Intracardiac or central venous pressures, and oxygen saturations

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

were obtained before and after EsP. Results: Oxygen saturations increased by 7.9% +/- 1.9%(SD) with EsP in this selective group of patients(paired t test, p< 05). Hemodynamic response to EsP in patient A is summarized in the table below. Figure 1 shows a striking phasic waveform in the pulmonary arterial wedge pressure tracing. The upstroke of this waveform occurs in systole, immediately after each retrograde P wave in JR. Immediately after EsP, the abnormal waveform and other hemodynamic parameters significantly improved. Conclusion: These data suggest that an atrial contraction against a closed AV valve can result in a retrograde pumping force in systole in Fontan patients. Nearly simultaneous atrial and ventricular contraction can adversely affect cardiac output. The existence of a P wave via fast retrograde pathway in JR on ECG may predict increased hemodynamic improvement with atrial pacing. The striking hemodynamic improvement with EsP underscores the potential importance of this observation.

P319

Limb threatening ischemia in a newborn after Cardiac Surgery: successful surgical and medical treatment

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Thrombosis and ischemia are rare but limb threatening complications after line insertion into the groin in newborn patients undergoing cardiac surgery. A premature newborn, 31st week of gestation, with TGA, underwent arterial switch operation at age 5 weeks, weight 2.3 kg. Arterial and venous lines were inserted into the right groin. Surgery was performed under CBP (150 min) with uneventful weaning. Postoperatively, the right leg was congested and dark. Both lines were removed immediately and systemic heparinization (500 IE/kg/d) begun. Venous congestion improved, but the leg stayed completely ischemic without doppler signals distal to the groin, the foot and ankle were black. Local thrombectomy was performed by a minimal incision of the femoral artery. A 4cm (distal) and a 0.8cm (proximal) thrombus were removed with a 2F coronary embolectomy catheter (Baxter, Deerfield, Ill., USA). There was weak orthograde flow and good backflow. Two 22G silastic catheters (Medexmedical, Ratingen, USA) were inserted distally and proximally to the incision. Local rTPA lysis was commenced 24h after surgery of TGA (0.25mg/kg increased to 0.5mg/kg, every 4-8h, 2/3 via proximal and 1/3 via distal catheter). Clinically the leg improved slowly. On day 5 pericardial tamponade had to be drained through a subxiphoidal incision. rTPA therapy was continued for 8 days. Doppler signals were available in the groin (day 3), the popliteal (day 4) and the foot (day 5). The leg recovered completely. Fragmin (s.c. 520 IE/d) and Aspirin (3mg/kg/d) were administered for mid-term therapy. Limb threatening leg ischemia can occur, if venous and arterial lines are inserted into the same groin. Immediate surgical embolectomy and local lysis can be performed even early postoperatively after complex cardiac surgery for leg salvage without life threatening bleeding complications.

P320

Outcome, morbidity risk factors and mid-term follow-up of surgically treated ventricular septal defects

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Objectives: The study reviews the morbidity, complications, and residual findings in patients (pts) after surgical correction of VSD's in our institution from 12/97 to 11/03. Methods: Ninety pts, 49 males, 41 females were treated. Median age was 1.9 years (yrs) (range, 0.3 -53.3 yrs), 26 pts were infants (28.8%). VSD type: 79 conoventricular (87.8%), 4 conal septum type (supracristal) (6.7%), 4 atrioventricular canal type (4.4%) and 3 multiple muscular (3.3%). Thirty-four pts (37.8%) had significant associated defects consisting of aortic and pulmonary valve anomalies, right and left ventricular outflow tract stenosis. Fifty-five pts (61.1%) had symptoms and 50 (55.6%) were on medication. Dacron patch was used in 89 pts (98.9%), and direct suture closure in 1 pt (1.1%). Data analysis were analyzed with t-test. Results: Median ICU stay was 2 days (ds) (range 1-28), infants' was 6.5 ds and non-infants' 1.5 ds (p< 0.005). Median hospital stay was 7 ds (range 4-40): infants 14 ds, non infants' 6 ds (p<0.005). Pericardial effusions developed in 25 pts (27.8%), 18 (20%) needing medical treatment and 7 (7.8%) pericardiocentesis. Fourteen pts (15.5%) received antibiotics for infections, 2 (2.2%) pts needed antiarrhythmics, and 1 (1.1%) chest tube placement for pneumothorax. One pt needed ECMO postoperatively and subsequently died (1.1%). There were no reoperations. Median follow-up (f/u) was 11 weeks (range 1-140). Three pts (3.3%) had small residual VSDs of no hemodynamic significance. Incomplete right bundle branch block (RBBB) developed in 24(23.3%) while complete in 34 (37.8%). Conclusions: Surgical VSD closure has low mortality and morbidity. Infants have higher ICU and hospital stay. Pericardial effusion, infections and arrhythmias are the commonest complications usually treated medically. Postoperative RBBB is common and requires further f/u to decide its significance.

P321

Primary cardiac neoplasms in children. Early and mid-term results of surgical treatment

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BACKGROUND AND OBJECTIVE: Primary cardiac neoplasms (PCN) are rare lesions and include both benign and malignant histological types. The aim was to study the early and mid-term outcome in a series of PCN consecutive children. MATERIALS AND METHODS: Between 1987-2003, 21 consecutive children underwent PCN resection. There were 17(81%)males, mean age 6.2±3 years. The clinical presentations included: congestive heart failure-2(9.5%), palpitations-4(19.0%), neurologic symptoms-2(9.5%), dyspnea-4(19%), chest pain-2(9.5%). 9(43%)patients were asymptomatic. Associated anomalies were: interventricular defect-2(9.5%) supravalvular aortic stenosis-1(4.8%), interatrial septal defect-1(4.8%) and bicuspid aortic valve-2(9.5%) patients. Inflow obstruction of the left or right ventricle and left ventricular outflow tract obstruction were present in 10(47.6%) and 4(19%)patients respectively. RESULTS: All patients underwent PCN resection and associated congenital anomalies correction. Hospital mortality resulted to be 2(9.5%) patients. One of them presented multifocal PCN. Histology revealed rhabdomyoma. The hystologic examination demonstrated benign PCN in all patients: myxoma-15(71.4%), papillary fibroelastoma-2(9.5%), fibroma-1(4.8%), rhabdomyoma-2(9.5%) and lipoma-1(4.8%)patient. 2(10%) and 5(23.8%)patients presented right and left atrial myxoma respectively. 1(4.8%) and 2(9.5%) other patients presented atrial myxoma and mitral valve myxoma respectively. In one patient we found papillary fibroelastoma in concomitance to the mitral valve myxoma. Total PCN resection was performed in 18(85.7%)patients. 3(14.6%) patients presented multifocal PCN: mean age 9.4 ±4 months (significantly younger than other patients, p<0.001). Mean follow-up was 4.5±1.7 years (range 6 month to 13 years). One patient, with bicuspid aortic valve, underwent reoperation due to severe aortic stenosis at 3.2 years after operation. There was no recurrence, significantly lower than the tumor recurrence in a series of 77 consecutive survived adult patients with benign tumors operated during the same period(p<0.017). CONCLUSION: Benign PCN in childhood have an excellent prognosis when completely excised and seems to have acceptable early and mid-term outcome even when excision is incomplete; prompt surgical resection should be anticipated in all children. Tumor recurrence incidence seems to be lower than the adult with PCN.

P322

Early and long term outcome of the arterial switch operation for transposition of the great arteries. Our experience

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OBJECTIVE: The aims of this study were 1)to review the early and late postoperative outcome in patients with transposition of the great arteries(TGA) undergoing arterial switch operation(ASO), 2)to identify the risk factors in this pool of patients. METHODS: Between 1992 and 2000, 126 patients underwent ASO, 83(66%)males, mean age: 41±32days. They were divided in:Group I(n=78)-simple TGA;Group II(n=32)-TGA with ventricular septal defect;Group III(n=7)-corrected TGA;Group IV(n=9)-TGA with ventricular septal defect and aortic coarctation. Other anomalies were: preoperative coronary anomalies(n=35(27.8%)), interatrial septal defect(n=37(29.4%)), left ventricular outflow obstruction(n=10(8%)), hypoplastic aortic arch(n=6(4.8%)), straddling mitral valve(n=4(3.2%)), thromboembolism of left ventricle(n=3(2.4%)). 86(68.2%) patients in Group II presented Taussig-Bing anomaly. All Group III patients underwent double switch operation (Senning technique and ASO). The aortic coarctation was repaired simultaneously to ASO in Group IV patients. Rashkind procedure was employed preoperatively in 51(40.5%)patients. RESULTS: The overall hospital mortality was 20(15.7%) patients. The mortality in Group II and Group IV resulted to be significantly higher than Group I, p=0.022 and p=0.046 respectively. Low cardiac output was identified in 8(6.4%) patients, acute renal failure necessitating ultrafiltration in 5(4%) patients

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

and hemi-diaphragmatic paralysis in 5(7%) patients. The univariate analysis using only preoperative and intraoperative variables revealed the complex anatomy (p=0.008), coronary anomalies (p=0.011), low weight birth (p=0.001) and prolonged bypass time (p<0.001) resulted as risk factors for early mortality. The multivariate logistic regression model revealed the low weight birth and complex anatomy as independent risk factors. There were 3(6.25%) hospital death in a subgroup of 48 patients operated during the last 3 years, resulting in a significantly lower hospital mortality than in other patients operated in the previous years(p=0.029). Mean follow-up time was 5.6±2.3 years (ranged 4 months-11.8 years). The overall survival in 136 survived patients resulted to be 97%, 95%, and 91% at 1, 3 and 5 years respectively. Freedom from reoperation was 100%, 98% and 91% at 1, 3 and 5 years respectively. CONCLUSIONS: ASO can be performed with acceptable post-operative mortality and morbidity. The associated anatomic malformations, anomalous coronary arteries and low weight birth influence significantly the early and late mortality. Growing experience with ASO of the surgical and intensive care teams is a necessary prerequisite for improvement of the post-operative outcome.

P323

Surgical correction of the coronary arteries with anomalous origin from the pulmonary artery

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BACKGROUND AND OBJECTIVE: Anomalous origin of the coronary arteries (AOCA) from the pulmonary artery (PA) carries a poor prognosis,most patients die early in life from myocardial infarction and congestive heart failure. The aim of this study is to review our experience in the AOCA from PA treatment and to evaluate its anatomic presentations. METHODS: Between 1991–2000, 17 patients with AOCA from the PA, 7(41%) males, mean age 30.6±23 months (range 6 days to 10 years), LVEF = 30±11%. They were divided in Group I(n=11, AOCA with associated anomalies) and Group II(n=6, simple AOCA). The AOCA anatomic findings included: left main coronary artery(CA)-n=7(41%), left anterior descending artery-n=3(18.7%), circumflex artery-n=1(6%), right CA-n=5(29%) and both CA-n=1(6%). The associated congenital anomalies in Group I included: transposition of the great arteries[n=4(23.5%)], atrial defect[n=2(12%)], ventricular septal defect[n=3(17.6%)], right aortic arch[n=1(6%)], subaortic membrane[n=2(12%)], severe mitral valve regurgitation[n=1(6%)] and tetralogy of Fallot[n=3(17.6%)]. One patient (4.5 years old) from Group II presented homozygote hypercholesterolemia and coronary artery disease. Another patient from the same group presented anomalous origin of both CA: the left CA from the right PA and right CA from the main PA. RESULTS: The overall hospital mortality was 3(17.6%)patients. No death resulted in Group II patients (p=NS). The reimplantation of the coronary ostia was performed in 8(47%)patients. Takeuchi operation in 2(12%)patients and coronary artery bypass grafting employing left internal mammary artery in 4(23.5%)patients. Rethoracotomy for bleeding and low cardiac output were identified in 1(6%) and 2(12%)patients respectively. Mediastinitis was identified in the patient with homozygote hypercholesterolemia. At 3.8±1.7years follow-up, 7(87.5%) and 5(83%)patients from Group I and II respectively survived(p=NS). In 12 survivors the mean LVEF increased significantly 47±13%(p<0.001). CONCLUSIONS: Patients with AOCA from PA may undergo surgical correction with acceptable early and mid-term mortality and morbidity. Various surgical procedures seems to be valuable alternatives for the AOCA corrections.

P324

Reconstruction of the Right Ventricular Outflow Tract (RVOT): A 26-Year experience with Valved and Nonvalved Conduits

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Objective: RVOT in congenital heart disease often requires the implantation of a valved or nonvalved extracardiac conduit (EC). Early results of reconstruction of the RVOT with EC are excellent. However, late follow-up demonstrates failure of these EC due to stenosis and valve insufficiency. We compared the long-term durability of all conduits in the RVOT over a 26-year period. METHODS: Between 2/1974 and 7/2000, 287 patients (mean age 12.1 years, mean weight 32.6 kg) with congenital malformations received a conduit (243 valved and 44 nonvalved). The EC size ranged between 8 and 33mm (mean size, 20.1mm). RESULTS: There was 10% early mortality (28/278). Long-term follow-up data were available on 253 (88%) patients. Seventy-two EC (28.5%) required replacement, 16% for nonvalved EC (7/44) and 27% for valved EC (65/243). The interval between first and second surgeries was between 3 months and 16 years (mean 4.4 years). Stenosis was the main mode of failure (66/72, 88%). The mean reoperation-free intervals were 3.5 and 6.2 years (P<0.04) for patients with non-valved and valved EC respectively. Comparing both groups, we found no difference in patient survival probability (p=0.7), but there were significant differences between xenografts (n=168) and homografts (n=216) in valved EC group (P<0.02) with respect to obstruction. At 10 years, the freedom from reoperation for EC obstruction was 84% for nonvalved EC and 73% for valved EC (P=0.1). CONCLUSION: EC give good results in spite our long-term results are disappointing. In the past 10 years pulmonary homografts use in reconstructing RVOT has been the conduits of choice for patients with RVOT pathology. Recently more potentially durable RV-PA conduits have been introduced. Further follow up is needed to test their durability.

P325

Repair of Tetralogy of Fallot associated with Total Atrioventricular septal defect

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Between February 1984 and November 2000, 22 patients with Tetralogy of Fallot (TOF) and atrioventricular septal defect (AVSD) were submitted to total correction in our institution. 17 patients were males (81.5%), age ranged from 1 to 15 years (median 5.0 yrs). 21 had Down Syndrome (95.5%). 20 patients were Rastelli Type 'C' (85%)and 8 had a previous modified Blalock-Taussig shunt (24.2%). Associated lesions were: PDA (3), pulmonary atresia (2), anomalous origin of LPA from Aorta (2), Total anomalous pulmonary venous connection (2), pulmonary valve agenesis (1), anomalous origin of LAD from RCA (1) and Situs Inversus (1). The surgical technique starts with a right atrial approach to the AVSD, dividing the AV valve into a left and a right components and partially closing the VSD with a patch. After a right ventriculotomy,the RVOT stenosis was relieved and the subaortic portion of the VSD was closed with another patch. 28 patients needed a transannular patch (85%), with a monocuspid valve. Mean postoperative RV/LV systolic pressure ratio was 0.65 (0.55 to 0.9). Two patients died postoperatively (6%): one of cardiogenic schock and one of multiple organ failure. The mean follow up time was 46 months (4 to 160 mo). There were 2 left AV valve replacements in the early postoperative phase (1 death). Mild left AV valve regurgitation was found in all patients except 3 (moderate). Among survivors, 15 are in functional class I (88.2%) and 2 in Class II (11.7%). CONCLUSION: TOF associated with AVSD can be successfully treated with a low mortality rate, low morbidity and satisfactory late hemodynamic results.

P326

Midterm results of arterial switch operation in transposition of the great arteries

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Objective: Since introduction of the arterial switch operation (ASO) in the 1980's it has been widely accepted and became the treatment of choice for transposition of the great arteries (TGA). We present our midterm results after ASO. Patients: Since 1994 thirty-five patients with TGA were corrected by the ASO in our institution. Thirty-nine of them were operated in a one-stage, or in a two-stage approach. The age ranged from 1 day to 2.5 years (median age 6 days). The weight at operation ranged from 2145g to 11.6 kg (median weight 3520 g) and the lenght ranged from 46 cm to 90 cm (median lenght 51cm). 11 patients had the diagnosis of simple transposition, 12 had additional anomalies. In 21 patients a Rashkind manoeuvre had to be performed pre-operatively. Methods: All infants were operated on normothermic cardiopulmonary bypass. The bypass time ranged from 94 minutes to 215 minutes (median 137 minutes), the aortic cross-clamp time ranged from 42 minutes to 106 minutes (median 74 minutes). All infants underwent postoperative catheterisation after 3 months. Results: There were two early deaths, no late deaths. In 11 patients interventional dilatation because of pulmonary stenosis or coarctation became necessary. In 3 patients coronary stenosis could be dilated successfully. All patients are in good clinical condition with good ventricular function. 9 infants show mild semilunar valve regurgitation. Two patients underwent re-operation due to supravalvular pulmonary obstruction. Conclusion: Arterial Switch Operation shows low early and late mortality. Midterm functional results are excellent. Routinely performed postoperative

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

catheterization allows early diagnosis and intervention of coronary, pulmonary and airway stenosis. In the presented patient group except two children all these stenoses could be successfully treated interventionally without the need of reoperation.

P327

Delayed Sternal Wound Closure in the Pediatric Population

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We retrospectively reviewed our entire population of delayed sternal wound closures (DSWC) from March 1997 to November 2000. DSWC was utilized in 6.5% of our total pediatric cardiothoracic surgical cases (69 of 1069) due to actual or predictable hemodynamic compromise related to chest wall edema or pulmonary congestion. Methods and Results: Mean patient age was 89 days and chest wounds remained open for a mean of 2.2 days. Temporary chest wound closure was accomplished with a Gore-tex patch in 61 (88%) of patients, silastic membrane in 3 (4%) and skin closure only in 5 (8%) of patients. All chest wounds were covered with betadine ointment and occlusive sterile dressing; chest was not intact until the time of definitive sternal closure. Sternal wound closure commenced when hemodynamics stabilized or chest wall edema resolved. Chests were prepped with multiple layers of betadine scrub, alcohol and betadine solution. The Gore-tex patch or membrane was removed and the wound irrigated with Vancomycin solution, followed by sized wire sternal closure and vicryl suture fascial closure. Delayed sternal wound closure was performed in the pediatric intensive care unit in 61 (88%), operating room in 7 (10%), and the neonatal intensive care unit in 1 (2%) patients. No delayed wound infections occurred. Conclusion: Delayed sternal wound closure is a safe and effective method of postoperative management in infants and neonates undergoing complex open heart surgery and can be performed in the intensive care unit, improving efficiency and saving operating room costs.

P328

Non-neonatal Transatrial-transpulmonary repair of Tetralogy of Fallot

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152 patients underwent transatrial-transpulmonary repair for Tetralogy of Fallot (TOF), between June 1995 and October 2000. TOF with pulmonary atresia was excluded. Associated major abnormalities were complete atrioventricular canal in 8 and absent pulmonary valve syndrome in 3 patients. The median age was 12.1 months (range 4.1 to 843), with a median weight of 7.5 kg. Palliative surgery (systemic-to-pulmonary shunting) was performed in 31. At repair, 52.8% of cases required a right ventricular outflow tract (RVOT) trans-transannular patch. Coronary artery anomalies were seen in 14 patients, of whom 9 required a trans-transannular patch. Median ICU stay was 2 days (range 1–19) and the median hospital stay 6 days (range 3–46). There was one mortality (0.7%) from a severe transfusion reaction. The mean follow-up was 14.0 months (range 1 to 100). There were 10 reoperations (one patient required 3), including revision of the RVOT in 7 and repair of residual VSD in 3. The reoperation rate was 5.3%. Two RV to pulmonary artery conduits were inserted at this time. In conclusion, non-anomalous, transatrial-transpulmonary repair of TOF can be performed with a very low mortality and short hospital stay. Coronary artery anomalies do not preclude a transatrial- transpulmonary repair.

P329

Respiratory syncytial viral infection increases postoperative morbidity and mortality among infants with correction of AV canal defects

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Background: Respiratory Syncytial Virus (RSV) is the most common cause of lower respiratory tract illness in infants. We sought to quantify whether the presence of RSV increases morbidity, mortality, and hospital costs in infants with correction of atrioventricular septal defect, a patient cohort with otherwise similar operative and postoperative risk factors. Methods and Results: Forty-one patients were identified from 1997 through the year 2000. Three patients tested positive for RSV by immunoassay; one death occurred in the RSV group. All three contracted RSV during the winter months (November-April). Intensive care length of stay (8.1 vs 15.7 days p= 0.00021), total length of hospital stay (7.4 vs 33.7 days p=0.0001), and inflation corrected hospital cost ($53,631 vs $122,202 p=0.0001) were significantly increased. All other risk factors including age, preoperative comorbity, operative time, and technique were similar. Additionally, no other significant difference was identified between patients with operation during the summer months (May–October, n=16) and winter months (November-April, n=25) except the presence of RSV. Conclusions: RSV increases icu and total hospital length of stay, and inflation corrected costs among infants undergoing corrective surgery for congenital heart disease. Infants should be extensively screened both clinically and by laboratory analysis for RSV prior to surgical consideration.

P330

Hypertrophic obstructive cardiomyopathy (HOCM) in pediatric patients: results of surgical treatment

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Between May 1977 and May 2000, 39 pediatric patients on one hospital service underwent extended left ventricular septal myectomy for HOCM. Ages ranged from 2 months to 18 years (median, 14 years). Twenty-five patients (64%) had moderate or severe mitral valve insufficiency. Medical therapy failed in all patients and 8 had undergone dual-chamber pacemaker implantation without improvement. Preoperative resting left ventricular outflow tract (LVOT) gradients ranged from 27 to 150 mmHg (median, 91). Fifteen patients (38%) had one or more concomitant procedures including division of abnormal papillary muscle attachments in 5, aortic valve repair in 4, mitral valve repair in 4, ASD closure in 2, resection subaortic membrane in 2, and division coronary artery muscle bridging in 1. Intraoperative post-myectomy LVOT gradients ranged from 0 to 41 mmHg (median, 6 mmHg). Postmyectomy mitral insufficiency was moderate in only 5 patients and severe in none. There were no operative deaths. Complications included temporary heart block in 1. Follow-up ranged from 6 months to 22 years (median, 7 years). There were 2 late deaths, neither of which was sudden. Echocardiography in 24 patients demonstrated a median LVOT gradient of 4 mmHg. Electrocardiography in 22 patients showed sinus rhythm in all. Five patients had late reoperation: re-myectomy in 2, and aortic valve replacement, Ross procedure, and heart transplant in 1 each. One of these patients required subsequent mitral valve replacement 19 years after initial myectomy. NYHA functional class at follow-up was I in 31 patients (84%) and II in 6 (16%). Extended septal myectomy relieves rapid symptoms and LVOT obstruction safely and effectively in pediatric patients with severe HOCM. Late survivorship compares very favorably with the natural history of the disease.

P331

Tetralogy of fallot with multiple ventricular septal defects

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Tetralogy of Fallot (TOF) with multiple ventricular septal defects (VSD's) has been considered a high risk group due to their higher incidence of residual VSD's and hence were grouped as incremental risk factors for adverse events following surgical repair. We present our experience with this subset of patients. From January 1997 to November 2000, 762 patients were operated for TOF with pulmonary stenosis. Eight patients were found to have multiple VSD's in addition to a sub aortic VSD. Patients with pulmonary atresia were excluded from this retrospective study. The age group of these patients were from 1.9 years to 11 years (mean 4.25 years). The sex distribution was equal. Echocardiogram detected the additional VSD's in seven of the cases, one was detected intra operatively. A cardiac catheterization was performed in seven cases where the additional VSD's were confirmed. The additional VSD's were found to be mid muscular in six, apical muscular in one and inlet with apical muscular in one. In addition to the VSD closures, infundibular resection done was required in three, while the remaining five required right ventricular outflow tract (RVOT) reconstruction using trans annular patch in three, a sub annular patch in one and a right ventricle to pulmonary artery homograft conduit in one. All patients were in sinus rhythm post operatively, no pacing was required in any of them. We have had one mortality in this group due to right ventricular failure. The mean duration of ventilation required was 49.6 hours (range 10 to 168 hours) and the mean ICU stay was 3.87 days (range 2 to 8 days). Renal failure was seen in one patient and right ventricular failure in two patients. One patient developed right bundle branch block on follow up. All of them are asymptomatic and no residual VSD was detected. TOF with multiple VSD's can be repaired satisfactorily with no added risk to the patient.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P332
Results of multiple patches technique for Double Outlet Right Ventricle (DORV) with noncommunicated ventricular septal defect (VSD)
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We report our experience with biventricular repair of DORV with non communicated VSD, using multiple patches technique. From April 1987 to November 2000, 22 patients underwent biventricular repair using this technique to construct a tunnel, connecting the left ventricle to aorta. Age ranged from 2 months to 13 years (m=4.3 yrs) and one patients had previous palliative operation. Under moderated hypothermy the right atrium is opened and the VSD anatomy and the distance between the VSD and aorta are evaluated. If VSD is restrictive, an incision is made on its anteroseptal margin, in order to enlarge it. The first patch is tailored and sutured with interrupted stitches on the inferior margin of the VSD. Through a right ventriculotomy, the subaortic conus and a portion of the infundibular septum are resected. A second patch is placed around the aorta and subaortic conus. Finally a third patch is sized according to the distance and spacial disposition between the others two. Pulmonary stenosis is treated with pulmonary valvotomy and/or infundibular resection. A transannular patch with monocusped valve or an extracardiac conduit is placed if necessary. Early mortality was 4.5%. Survival patients were followed for a mean period of 50 months. There were 4 late deaths. Survivals patients are in functional class I (14 patients) or II. Residual lesions are moderate left outflow stenosis (2), moderate pulmonary stenosis (2), severe pulmonary regurgitation (2) and minimal VSD (4). In conclusion the use of multiple patches technique for this anomaly simplifies the biventricular repair.

P333
Management of complete ectopia cordis
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Although ectopia cordis is a rare defect representing less than 0.1% of congenital heart defects, surgical and postoperative management can be challenging. Classification of this defect can include the partial and complete forms. Of the complete forms these have been divided into various classifications including the cervical, thoracic and abdominal types. A fourth type for consideration is the thoraco abdominal type also called the Pentalogy of Cantrell. Between 1984 to 1999 five cases of complete or true ectopia cordis have been encountered by the author. Three were diagnosed early with ultrasound. Pregnancy was terminated in one because of multiple other anomalies. Our infants died immediately after birth because of multiple anomalies and three were surgically repaired. Surgical correction includes 1.) coverage of the naked heart; 2.) palliation or complete repair of major intracardiac defects; 3.) placement of the heart into the thoracic cavity, and 4.) sternal or thoracic reconstruction. Only the first two steps are essential. We have performed surgery on three infants with acute ectopia cordis in which the heart was returned within the thoracic cavity. Two infants were survivors of this type of surgical repair while the third died of infection within eight weeks of repair. Postoperative management of respiratory mechanics are extremely important because of the absent anterior chest wall. Finally because of the overall outcome of this condition, ethical questions of management should be considered as soon as the diagnosis is determined.

P334
Concepts and results in surgical management of complex double outlet right ventricle
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Objective: The surgical management of double outlet right ventricle (DORV) is determined upon 1. the position and size of the VSD, 2. the ventriculoarterial connection and 3. the presence of associated lesions. We retrospectively analyzed the results after definitive surgical treatment of DORV with subpulmonary, remote and doubly committed VSD (complex DORV) and excluded 158 pts. with TOF- type of DORV. Methods and Results: Between 3/1982–10/2000, 123 pts. (mean age 5.7±5.5 years) underwent repair of complex DORV (44% of all DORV repairs, 1.1% of all cardiosurgical pts.). Follow up time is 4.7±4.9 years. Table 1 summarizes early and late results. The overall, early survival rate increased from 74% (1982–1995) to 84% (1996–2000). The surgical procedure did not influence early survival. The re-operation rate was 24% (29/94) during follow up (n=19 for recurrent outflow tract obstruction). Conclusion: The repair of complex DORV may require a variety of surgical procedures. Early and late results after repair of complex DORV in the current area are improving in all subsets. The high re-operation rate is related predominately to recurrence of outflow tract obstructions.

P335
Intermediate results after complete repair of tetralogy of Fallot in infants
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Objective: Timing and operative method for complete repair of tetralogy of Fallot is still under discussion. We present our intermediate follow-up results. Patients: From 1994 until November 2000, 51 infants underwent corrective surgery for tetralogy of Fallot at our institution. The age ranged from 2 days to 16 years, median age of 4 months. Fourteen infants had a diagnosis of tetralogy of Fallot with pulmonary stenosis (group I) and 5 infants with pulmonary atresia (group II). Nine infants had a two stage correction, 7 from group I and 2 from group II. In the first group 5 patients received a transannular patch and 2 a BT-shunt. In the second group one infant was primarily palliated with an aortopulmonary shunt and the other with a transannular patch. All patients with diagnosis of tetralogy of Fallot were corrected by resection of the pulmonary valve, subvalvular resection of hypertrophic muscular bundles, transannular patching and VSD patch closure. There were two early deaths. No late deaths occurred. In the median term follow-up no reoperation was necessary. In 8 patients pulmonary stenoses were treated interventionally by balloon dilatation and in 4 cases by stent implantation. All children are in good clinical condition. None of these children suffers from arrhythmia or right ventricular dilatation. Transannular patch technique for corrective surgery of tetralogy of Fallot shows good midterm results. No reoperation was necessary so far. To protect the right ventricle from late failure pulmonary stenoses are treated early and aggressively by interventional methods. Close follow up is needed but to miss the time of indication for a homograft implantation. Implantation of homografts in older aged children avoids several reoperations caused by the growth of the patients.

P336
Staged palliation of patients with single ventricle pathology and arch obstruction without circulatory arrest – focus on the timing of Damus-Kaye-Stansel (DKS) connection
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Aims: Retrospective evaluation of repair strategies for neonates with double inlet left ventricle and L-tga, and similar lesions. These often present with arch obstruction and duct-dependent circulation necessitating early repair. Methods: 'Primary' repair utilising a Norwood type approach may be performed. Our group has preferred to repair the arch and limit pulmonary blood flow, initially deferring the DKS to a point when cavo-pulmonary connection can also be performed. Potential narrowing of the bidirectional cavo-pulmonary connection (BVF) may warrant early unbanding of the systemic ventricle. Incompetence of the pulmonary valve remains a concern. Results: Our strategy was examined in the 19 patients who underwent DKS since 1990. Arch repair and PA banding was performed by thoracotomy with a second stage DKS and cavopulmonary connection at a median of 14 months after the first operation. At the second stage, 2 patients required repair of the pulmonary arteries because of the pulmonary band. To date the Fontan circulation has been achieved in 11 of 19 patients at a median 48 months later. A subset of children where BVF narrowing was not initially apparent (4) underwent completion Fontan but subsequently required DKS. This was performed by resection of the pulmonary valve (3) and xenograft (1). There was one unrelated death 9 months post operatively. The remainder are well with good physical capacity, with no chronic neurological injuries, no instances of tricuspid valve dysfunction, no progressive ventricular impairment nor ventricular dysfunction. Conclusion: Good results have been achieved with delayed DKS utilising a strategy that avoids circulatory arrest and extensive surgery in a sick neonate. Small numbers of patients representing with late BVF narrowing suggest that early DKS connection should be performed in patients with susceptible pathologies as part of the repair strategy.

P337
Factors associated with palliation for Tetralogy of Fallot (TOF)
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https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

To explore factors associated with initial palliation or repair for TOF, we analyzed discharge abstract data from 5 states (CA, IL, MA, PA, WA) in 1996. Using ICD-9-CM codes, we identified infants with TOF <1y who underwent complete repair or shunt placement. Patients with codes indicating complex anatomy or additional surgical procedures were excluded. We used multivariate models to determine independent risk factors for palliation. Risk of death, length of stay, and hospital charges were compared for each type of procedure. Of 379 cases, 286 were infants <1y (67 shunts, 219 repair) and were analyzed further. Among 37 institutions, 22 performed >=5 total cases, with 0-56.2% of cases as palliative. Premature infants (p = 02), neonates <=30days (p<.0001, those transferred from another facility (p= 001), admitted urgently (p< 001), or at institutions performing <20 cases (p< 001) were more likely to undergo palliation. Gender, race, insurance, and major chromosomal or structural anomaly/syndrome were not associated with palliation. By multivariate analysis, neonatal status, urgent admission, and surgery at an institution performing <20 cases (OR 6.8, p<.001) remained significant. Infants undergoing palliation had a higher risk of dying (11.9% vs. 4.3%, p=.03), and longer length of stay (12 vs 8 days, p=.006), but similar hospital charges ($63842 vs $59063). Length of stay was similar after controlling for clinical factors. However, in adjusted analyses mortality differences were similar for infants receiving complete repairs or shunts <=30 days, and were higher for shunts performed in older infants (OR 6.7, p=.009). Conclusion: In risk-adjusted analyses, complete repair of TOF had a similar mortality to shunt placement in neonates, and lower mortality in older infants. Adjusting for other factors, shunts were more common at centers performing fewer cases.

P338

Leaving the sternum open in small children after complex cardiopulmonary bypass procedures is not a complication: it is a benefit!

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Delayed sternal closure after cardio-pulmonary bypass (CPBP) in children has been considered as a complication or at a risk factor for increased post-operative morbidity. Hence, the technique is usually applied whenever there is a risk for haemodynamic or respiratory instability on sternal closure. From July 1998 to November 2000, we performed 51 complex CPBP procedures in small children aged from 2 to 120 days (median, 9.5 days), with weights between 2.0 and 5.5 kg (median 3.8 kg). There were corrective surgeries for transposition of the great arteries (TGA) (18, 35.2%), TGA with ventricular septal defect (VSD) (13; 25.4%), hypoplastic left heart syndrome (8, 15.7%), total anomalous pulmonary venous connections (4, 7.8%), truncus arteriosus (4, 7.8%), interrupted aortic arch with VSD (2; 3.9%), and TGA with VSD and aortic coarctation (2, 3.9%). All the patients were ventilated and returned from theatre with open sternum and closed skin. Fourty-nine patients (96.1%) required <10 mcg/kg/minute of inotropic drugs. Adrenaline was used in 9 (17.6%) patients. Three (5.8%) patients required $NaHCO_3$. Sternotomies were closed within 5 to 48 hours (median, 24 hours), and withdrawal of assisted ventilation occured between 8 and 240 hours (median, 40 hours). Mean ICU stay was 3 days. There was 1 sternal wound infection and no mediastinitis. Four patients (7.8%) died (3 Norwood, 1 TGA with VSD). Timed sternal closure was not correlated neither with death, nor with modification of haemodynamic or ventilatory parameters. Routine delayed sternal closure after complex CPBP procedures in small children ensures a clinical course with haemodynamic stability and low requirement for inotropic drugs, short ICU stay, and very low incidence of complications. It seems to be a benefit rather than a complication or a detrimental factor.

P339

Ebstein's anomaly and related conditions – complete repair in the severely symptomatic neonate

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Ebstein's anomaly in the severely symptomatic neonate is usually fatal. Until recently, successful repair had not been reported and various palliative operations were associated with prohibitive mortality. Single ventricle palliation (Starnes operation) has been somewhat more successful. We present our experience with complete biventricular repair in the symptomatic neonate, with emphasis on the evolution of our surgical technique and the medium-term follow-up of the patients. Since 1994, 7 severely symptomatic neonates underwent repair by one surgeon in Oklahoma city. Five had Ebstein's anomaly and 2 had gross cardiomegaly, tricuspid valve dysplasia and pulmonary atresia (n=2). One Ebstein patient had undergone a Starnes operation elsewhere and was transferred to our facility in a critical condition. Weight at operation ranged from 2.1-4.6 kg (mean 2.7kg). Five had either anatomical (n=3) or functional (n=2) pulmonary atresia. Severe (4/4) tricuspid regurgitation was present in all except 1 (poor Starnes operation), and cardiothoracic ratio exceeded 0.85 in all patients. Great Ormond Street risk scores were >15 in six (grade 4/4) and 13 in one (grade 3/4). Repair consisted of (i) tricuspid valve repair, (ii) reduction atrioplasty, (iii) relief of RVOTO, (iv) partial closure of ASD. One patient died in hospital – a 2.1kg pt with tricuspid dysplasia, anatomical pulmonary atresia and hypoplastic pulmonary arteries. The other six patients are in functional class I. Five take no medications, and all are in sinus rhythm. Although two patients had symptomatic SVT preoperatively, no child has experienced SVT after discharge. Three patients are now almost six years old, one is one year old, and the remaining 2 patients are six months old. At most recent follow-up, tricuspid regurgitation was considered mild (n=4) or moderate (n=1). Primary repair of the severely symptomatic neonate with Ebstein's anomaly or related pathology is feasible and safe. The surgical repair appears durable and associated with good medium-term outcome.

P340

Unbalanced atrioventricular canal and coarctation of the aorta: discrepancy in severity of atrioventricular valve unbalance and ventricular volume predicts operative failure using biventricular repair

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Selection of appropriate surgical strategy (biventricular repair (BVR) versus single ventricle palliation (SVP)) in patients with unbalanced atrioventricular canal defects and coarctation of the aorta (UAVC/CoA) is difficult, especially when the degree of unbalance is not extreme. We hypothesized that pre-operative measures of left-sided structures, affected by unbalance, may predict outcome. Retrospective review of the surgical database at our institution from 6/84 to 9/98 identified 43 patients with UAVC/CoA who underwent surgical intervention. Those with heterotaxy syndrome or malposition of the great arteries were excluded. Data on all surgical interventions were collected, in addition to survival status as of 6/00. Available preoperative echocardiograms were reviewed for anatomy, left ventricular end-diastolic volume (LVEDV) and severity of atrioventricular valve (AVV) unbalance. Results: Of the 43 patients with UAVC/CoA, 28 had echocardiograms suitable for review. No difference in baseline characteristics was noted at either study availability. Median age at time of initial surgery was 15 days (Range: 2 days – 11 months). BVR was the primary strategy in 12 patients (42.9%), one was subsequently converted to SVP. Six months after surgery, 6/16 SVP patients remained alive (37.5%, 95% CI: 15.2–64.6), compared with 7/12 BVR patients (58.3%, 95% CI: 27.7–84.8). Neither LVEDV nor degree of AVV unbalance predicted mortality after SVP. However, survival at 6 months was significantly worse for the subset of BVR patients in whom AVV unbalance was mild, yet LVEDV was small (See Graph, log rank, p=0.03). Conclusion: While survival with either surgical approach remains low, attention to the relationship between AVV unbalance and LVEDV may result in better candidate selection for BVR. Mild AVV unbalance may lead to the false impression of LV adequacy.

P341

Outcomes following total cavopulmonary connections in children with single ventricle and atrial isomerism/heterotaxy syndromes

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Background: From January 1984 to April 2000, 29 patients had total cavopulmonary connection for single ventricle and isomerism/heterotaxy syndrome. Patients and Methods: Right (n=18) or left (n=11) atrial isomerism/heterotaxy was associated with DORV in 16, a common atrioventricular valve in 21, pulmonary atresia or stenosis in 21, anomalous pulmonary venous drainage (APVD) in 17, interrupted IVC in 12 (azygous continuation n=9, hemiazygous n=3), and absent coronary sinus in 2 patients. A previous systemic to PA shunt was performed in 21 patients and 3 had previous PA banding. The Fontan procedure was staged in all but three children using a bidirectional cavopulmonary anastomosis (BDCPA, n=9), Kawashima anastomosis (n=12), bilateral BDCPA (n=3) and a hemi-Fontan (n=1) at a median age of 12 months (range=3–114 months). Prior to Fontan, atrial arrhythmia was present in 5 (17%, junctional=4, pacer-dependent=1, wandering atrial pace-

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

maker=1). Results: An extracardiac conduit was constructed in 22 patients (median=20mm, range=16–27mm) and a lateral tunnel in 4. Associated procedures included PA augmentation (n=16), repair of APVD (n=1), and repair of a regurgitant atrioventricular valve (n=2). The mean CPB time was 134+/-57 minutes. Cardioplegia cardiac arrest was used in 12 patients for a mean duration of 62+/-28 minutes. There were 4 hospital deaths (13%). 2 of which occurred in children having concomitant repair of APVD (previously undetected in 1). Twelve (41%) children developed early postoperative atrial arrhythmias, 4 of which required temporary pacing. There was one late death due to circuit thrombosis. Follow-up (3 mo-1.6 years) was available on 19 patients. All but 2 were in sinus rhythm. Conclusions: Early and mid-term outcomes following the Fontan procedure in this patient group are good and may be improved by detecting and repairing associated lesions (APVD) at the time of BDCPA.

P342

Is associated intracardiac anomaly a risk factor for repair of complete atrioventricular septal defect?

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<Background> The combination of complete atrioventricular septal defect (CAVSD) and major intracardiac anomaly has been difficult for total repair. This study is to evaluate retrospectively whether associated intracardiac anomaly is a risk factor for repair of CAVSD or not. <Methods> 35 consecutive patients underwent repair of CAVSD between January 1992 and April 2000. There were 12 patients (34.3%) associated with other intracardiac anomalies (7 tetralogy of Fallot including 2 hypoplastic RV, 2 double outlet right ventricle, 1 pulmonary atresia, 1 hypoplastic LV, 1 unbalanced common atrioventricular valve). Operative results of patients associated with other intracardiac anomalies (complex group) were compared to those without major intracardiac anomalies (isolated group). <Results> Age, gender, the incidence of trisomy 21, follow up period were not significantly different between two groups, however body weight in complex group was statistically bigger than isolated group. There were no operative death in both groups. The incidence of reoperation for mitral regurgitation was 16.7% (2 cases) in complex group, 5.7% (2 cases) in isolated group (p=0.48). There have been 1 non-cardiac late death in isolated group (Reye syndrome). All patients had no medication except early postoperative period in both groups. <Conclusion> Operative results of combination of CAVSD and major intracardiac anomaly were excellent. Associated intracardiac anomaly was not a risk factor for repair of CAVSD.

P343

Surgical repair of pulmonary stenosis in children and adults

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Surgical experiences of repair of pulmonary stenosis (PS) in children and adults, including hypoplastic right ventricle, is reviewed. 15 patients underwent operation, not balloon valvotomy because of concomitant infundibular stenosis. Preoperative pressure gradient was 96.6 mmHg in average. ASD was associated in 9 patients, 4 of whom showed marked cyanosis. Severe hypoplasia of right ventricle was recognized in two patients. Cardiopulmonary bypass (CPB) was used in all patients but one, who underwent infundibular resection under the shunt between SVC, IVC and pulmonary artery. One patient needed transannular patch but others was repaired through trans-atrial, trans-pulmonary approach. Two patients with hypoplastic right ventricle underwent one-and-one-half ventricular repair. All patients survived with postoperative pressure gradient of less than 25 mmHg. In conclusion, some patients with relatively favourable anatomy can be repaired without CPB. One-and-one-half ventricular repair can be an option for repair of PS with hypoplastic right ventricle.

P344

Postcardiotomy left ventricular assistance with centrifugal pump in neonates, infants, and children

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Background: Left ventricular assist devices have been useful for postcardiotomy myocardial failure and as a bridge to transplantation. However, our experience in children is limited. In isolated left ventricular failure after cardiac surgery in children, centrifugal pump assistance can be used, thus avoiding ECMO. Methods: Between February 1992 and October 2000, a retrospective review was performed in 28 patients who required postcardiotomy left ventricular assistance. The support was achieved with a centrifugal pump circuit (Bio-Medicus Inc., Eden Prairie, MN). The diagnoses were HLHS in 7 patients, other Single Ventricle physiology in 4, ALCAPA in 3, TGA in 3, Severe Mitral Regurgitation in 3, Senning Takedown with Arterial Switch in 2, Double Switch in one, and others in 5. There were 12 neonates, 7 infants and 9 children. The age of the patients ranged between 2 days and 11 years (median 29 months). The weight ranged between 1.9 kg and 35 kg (median 6.6 kg). Results: The mean duration of the left ventricular assist was 5 days (SD. = 1.95). Twelve patients (42.8%) died while on support (Inability to be weaned from LVAD in 6 patients, brain damage in 3, sepsis in 2, liver failure in 1). Sixteen patients were successfully weaned off (57.2%). Three (10.7%) of these patients died while still in hospital (1 from cardiac, 1 from respiratory, and 1 from neurological cause). Three patients (10.7%) died late with a follow-up duration between 13 and 40 months (all cardiac related). At the present, mean follow-up of 22 months, in one of the ten survivors, the left ventricular function was significantly improved to normal. One survivor (14.3%) was lost to follow-up. Conclusions: The percutaneously left ventricular assist device is hemodynamically effective in neonates, infants, and children. This support can allow otherwise unsalvageable patients to survive with good long-term outcome. Our best results were obtained in the patients with diagnosis of ALCAPA.

P345

Recovery of myocardial function following aortic implantation of anomalous left coronary artery arising from the pulmonary artery

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Background: Patients with anomalous origin of the left coronary artery (LCA) from the pulmonary artery (PA) often present during early childhood with severe myocardial ischemia and left ventricular dysfunction. The degree of improvement in ventricular performance following implantation of the anomalous LCA into the aorta was evaluated. Methods: The clinical and echocardiographic records of 12 consecutive patients who underwent transfer of the anomalous LCA from the PA to the aorta between January 1992 and October 2000 were reviewed. Results: The median age at surgical repair was 10 months and the median time to follow-up was 35 months. There were no early or late deaths in the group. Ischemic mitral insufficiency was present in 5 patients (42%). Preoperative echocardiography revealed major wall motion abnormalities and left ventricular ejection fraction (LVEF) <20% in 8 patients (67%). Of those patients, postoperative left ventricular assist device placement was required in 3 patients (38%). Circulatory support was weaned off by the third postoperative day in these patients. Echocardiography in the immediate postoperative period revealed significant recovery in ventricular performance in 2 patients (25%). At follow-up, all patients had echocardiographic evidence of markedly improved myocardial function. LVEF was greater than 50% in all patients. Resolution of wall motion abnormalities occurred in 7 of 8 patients (88%). Six patients (75%) demonstrated normalization of left ventricular dimension. Conclusion: Reversal of severe left ventricular dysfunction can be achieved following aortic implantation of the anomalous LCA. The establishment of a two-coronary system may promote remodeling of the left ventricle and regression of ischemic structural changes.

P346

Primary Repair of Critical Congenital Heart Defects in Neonates

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<Objective> Surgical treatment of congenital heart defects (CHD) has been moved to total repair to neonates and small infants. During these 10 years, there has been several modification in cardiopulmonary bypass, such as hemofiltration of the priming blood, high flow bypass, ultrafiltration, cerebral perfusion, avoidance of circulatory arrest and perioperative echocardiogram. We reviewed our 10 years experience in primary repair of critical CHD in neonates. <Methods> Since 1995, 263 neonates were operated on at Okayama University Hospital. There were 148 open heart surgeries and 115 palliative surgeries. Out of 148, 101 were total repair and 47 were open palliation. In 101 neonates, there were 39 TGA, 23 TAPVD, 14 CoA, 13 IAA, and other anomalies. Since 1995–1996, our protocol of perioperative management has changed and retrospectively we compared our results in 2 periods. <Result> There were 7 hospital death (14%) from our initial series and this

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

decreased to 1 hospital death(2%) in subsequent 51 cases. <Conclusion> The results of primary repair of critical CHD in neonates have been improved by these modifications. Surgeon's experience was a significant risk factor for operative mortality in this

P347

26 years' experience of anomalous left coronary artery from pulmonary artery (ALCAPA).

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Objective: To review an institutional experience of the treatment of ALCAPA and see how this management has influenced outcome over the years. Patients: Since 1972, 27 children with ALCAPA have been operated. Median age was 6 months (range 57 days to 5 years). From 1972-1986, 2 patients had Takeuchi procedure, 1 had ALCAPA ligation, 1 head a saphenous vein graft, and 4 had coronary transfer. From 1987–2000, 10 patients had coronary transfer and 1 had ligation. Results: 9 patients (33%) died prior to early postoperatively (1 vein graft, 3 Takeuchi, 6 transfer), 6 out of 8 (16%) in the early period and 3 out of 19 (1.6%) in the recent period. Since 1992, mechanical support was employed in 4 patients and they all survived. More recently, tissue Doppler echocardiography was used to assess myocardial viability (hibernating myocardium). Conclusion: Results of ALCAPA have improved over the years. This has coincided with improved results of neonatal and infant cardiac surgery, specifically with the introduction of postoperative mechanical assistance. Further improvement in treatment may be guided by the tissue Doppler echocardiography.

P348

Total anomalous pulmonary venous drainage: analysis of outcome and risk

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Objective: To evaluate results and identify risk factors associated with surgical correction of total anomalous pulmonary venous drainage (TAPVD). Methods: Between January 1987 and July 2000, 93 neonates, infants, and children underwent repair of TAPVD. Their ages ranged from 2 days to 15 years with a mean of 8 months. There were 33 females and 60 males. Mean weight was 4 kg (range 1.8 – 34 kg). Excluded were patients with Scimitar Syndrome and TAPVD associated with complex congenital defects like single ventricle physiology. There were 36 supracardiac type, 32 intracardiac, 14 infracardiac and 9 mixed types. One patient had dextrocardias and 3 had associated ventricular septal defect. Preoperatively 16 patients had obstruction, 26 were mechanically ventilated and 14 were on prostaglandin PGE. Twenty-five had documented sepsis preoperatively. Diagnosis was made by echocardiography. Cardiac catheterization was utilized early in the experience but very occasionally after 1990. Surgical technique was standardized. The confluence in supra and infra-cardiac types was widely anastomosed to the back of the left atrium with Polydioxalone (PDS) suture. A patch was used in the repair of intra-cardiac types and the mixed types were created according to anatomy but along the same surgical lines. Results: Overall hospital mortality was 11/93 (12%). Ten of the deaths were in patients less than 6 months of age. 6/11 weighed less than 3 kg. 9/11 had severe uncontrolled pulmonary hypertension. 6/11 had obstruction and 7/11 were emergency operations while 9/11 had sepsis. Among the 49 patients operated since 1995, only 2 deaths occurred and since 1996 no deaths occurred. We believe the reduction in hospital mortality is related to (1) better understanding and management of pulmonary hypertension including use of nitric oxide and (2) aggressive therapy of sepsis with early use of wide antibiotic coverage of both gram positive and gram negative organisms till results of cultures become available. Conclusion: Repair of TAPVD, though curative, remains a surgical challenge. Significant risk factors of adverse outcome include pulmonary hypertension, sepsis, pulmonary venous obstruction and emergency operation. Control of these factors together with experience seems to improve results.

P349

Growth of the pulmonary valve ring after total correction of tetralogy of Fallot

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Background: From 1998 we have performed total correction of tetralogy of Fallot (TF) without incising the pulmonary valve(PV) ring to prevent pulmonary regurgitation if the PV ring diameter was within a suitable range. In the present study, the growth of the PV ring after total correction of TF was evaluated. Patients and Method: 22 patients(pts) who underwent total correction of TF and both preoperative and postoperative cardiac catheterization were reviewed. The period between surgery and postoperative examination was 4.7+/-3.2 yrs. Their age at surgery was 2.5+/-2.4 yrs and 17 pts were male. PV was biscuspid in 15 and tricuspid in 7. Relief of right ventricular outflow obstruction was done by commissurotomy in 17 and infundibulectomy in 19 pts. In 15 pts, the pulmonary trunk was enlarged with a pericardial patch. Results: The postoperative PV ring was significantly larger than the preoperative one (16.7 +/- 5.7 vs 10.0 +/- 2.2 mm, p<0.05); and that of 14 of 22 pts was over 90% of normal PV ring. Postoperative systolic RV pressure was 40.6+/-10.8 mmHg. Pulmonary regurgitation was 3 degree in 5 and less than 2 in 17 by echocardiography. Conclusion: The PV ring may naturally grow after total correction of TF even without incising the PV ring.

P350

Pulmonary vascular disease in atrial septal defect and indications for surgery determined by lung biopsy diagnosis

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Pulmonary vascular disease in atrial septal defect and Indications for surgery determined by lung biopsy diagnosis. Indications for surgery were determined by lung biopsy diagnosis in 85 patients of atrial septal defect (ASD) with pulmonary hypertension. Eight of these 85 patients associated with primary pulmonary hypertension were eliminated from this study. Lung biopsy was performed if the patients had systolic pulmonary arterial pressure more than 70 mmHg and/or pulmonary vascular resistance of more than 8 units.m2. They ranged in age from 45 days to 71 years. Results: Pulmonary vascular disease (PVD) in ASD was classified into 4 types. 1) Plexogenic pulmonary arteriopathy. Surgery is indicated for an index of PVD 2.2 or less. Surgery was performed in 23 of the 30 patients. The remaining 7 patients for whom surgery was not indicated are under follow-up observation. No cases of death have occurred among any of the 30 patients. 2) Muscularization consisting of longitudinal muscle bundles and elastic fibers. Surgery is indicated no matter how severely the peripheral small pulmonary arteries are occluded. Surgery was performed in all of the 19 patients and the postoperative course was uneventful. 3) Mixed type of plexogenic pulmonary arteriopathy and muscularization. Surgery is indicated if the collateral is not observed or is incomplete. Surgery was performed in 14 of the 24 patients. The remaining 10 patients for whom surgery was not indicated are under follow-up observation. 4) Thromboembolism of small pulmonary arteries. Surgery is indicated for all such cases. Surgery was indicated in all of the 4 patients. Conclusion: No death has occurred at this time among the 77 patients who underwent lung biopsy diagnosis.

P351

Repair of complex aortic coarctation using extended aortic arch anastomosis

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Extended end-to-end aortic arch anastomosis was used to correct both the ischemic stenosis and hypoplasia of the transverse arch. Between May 1991 and July 1996, 28 consecutive neonates and children underwent repair of aortic coarctation and tubular hypoplasia using the extended arch repair technique. Median age 8.9 days (range 1 day to 15 yrs). Antenatal diagnosis was made in 11 patients. Preoperatively in 16 patients (57.1%) the mean systolic upper to lower extremity pressure gradient was > 20, in 10 (36%) < 20 mmHg and in 2 (7%) was not recorded. Additional procedures performed at the time of repair included, ligation of a patent arterial duct, closure of ventricular septal defect, atrial septostomy, pulmonary artery banding, arterial switch. Median duration of stay in intensive care was 8 days (range 1 to 67 days). There were 2 perioperative deaths: 2 sick neonates, who had emergency surgery died on the same day of operation. Early transient postoperative problems included, lung collapse, acute renal failure, hypertension, ischaemic hepatitis, seizure, intropenia, heart block. There was 1 early death (4.9%). The follow-up period varied between 3.30 -10.3 years, median 5.6 years. All 25 (89.9%) operative survivors were free of recurrent coarctation at latest follow-up. The extended arch technique is the procedure of choice for patients with coarctation and hypoplasia of the arch. Recurrent aortic coarctation was not

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

identified so far. Further follow-up is necessary to see the long term impact on systemic blood pressure and ventricular mass regression.

P352

The scimitar syndrome, follow up and outcome

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The Scimitar syndrome consists of changes of the lungs including an extra-pulmonary collateral and an abnormal pulmonary venous drainage. The syndrome is very variable in its expression. This rare disorder (about 3–5% of all partial anomalous pulmonary drainage) was first described 1836. There is still no general therapy concept. We followed 14 patients (5 male, 9 female) with a scimitar syndrome, the age of diagnosis was 7,8 years. Of those 5 were just followed and 9 needed an operation. The operation were aimed at the main presenting symptom. If the child was thought to be compromised by the shunt-volume (Qp:Qs>1.6), lower airway infection or by other cardiac defects. Three techniques were used: I.) lobe-excision (n=3); II.) tunnel from the inferior vena cava to the left atrium (n=2); scimitar vein re-implanted in the right atrium and a tunnel from there to the left atrium (n=6). The post-operative follow up was 3,6 years (2,5 – 13 years). The patients with operation technique I and III fared well with a general improvement and no complication were observed. Patients with operation technique II suffered a venous thrombosis in the tunnel and there was no improvement, the pulmonary hypertension even persisted. In one case. Of the other 5 patients 3 fared well, one died due to a complex cardiac lesion including a hypoplastic left heart and the other declined any surgery though he had a marked volume-load of his right ventricle and becomes increasingly breathless. 2 patients had malignant supraventricular tachycardia, one of them died. We would recommend to re-implant the scimitar vein into the right atrium with a tunnel to the left atrium or in infancy probably also a vein.

P353

Anatomical correction of anomalous pulmonary vein drainage

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We have reviewed our results and experience with 37 children (15 females, 22 males; age range: 9 months to 15 years, mean value 6,9 months) operated on various forms of partial (right-sided, left or mixed) anomalous pulmonary vein return. Qp:Qs ratio ranged from 1.5:1 to 5:1 (mean value 3.38 ± SD 1.14). Ten patients (27%) had additional vascular problems e.g. patent ductus arteriosus, left superior vena cava (LSVC), pulmonary stenosis, aortic arch hypoplasia. We paid special attention to the accuracy of preoperative diagnoses based on 2 D echo and Doppler measurements (number of pulmonary veins connected to a wrong cardiac chamber or vessel, atrial septum and SVC anatomy), intraoperative epicardial echocardiographic assessment and post-operative follow-up evaluation. The repair employed bicaval cannulation, moderate hypothermia 30–32 deg C, transatrial approach and the technique of autologous pericardial baffle (tunnel) diverting pulmonary blood into the left atrium. In one case of anomalous left pulmonary vein return (to the coronary sinus and through vertical vein to brachiocephalic vein) we used left thoracotomy (no CPB) to perform direct anastomosis left pulmonary vein – left atrial appendage. Postoperative echocardiography showed unrestricted pulmonary blood flow without turbulence and no SVC narrowing; we have noted no early or late post-op cardiac disturbances.

P354

Sinus venosus syndrome: surgical technique and results

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The most frequent complications of surgical treatment of Sinus Venosus Syndrome are arrhythmias, due to sinus node dysfunction and pulmonary veins or SVC obstruction. Between July 1992 and April 2000 we operated on 20 patients affected with sinus venosus ASD and PAPVR: in SVC: 2 veins in 8 patients, 1 vein in 8 patients, 3 veins in 4 patients. The age ranged between 4 months and 15 years and weight 6/30 Kgs. All patients had cardiac arrest through median sternotomy, CPB, 5% Thomas cardioplegic solution. The PAPVR was reduced to LA by a tunnel of Dacron Sauvage by a vertical incision from the RA through caval-atrial junction to the SVC, which was almost always enlarged with a bovine pericardial patch. Only 1 patient showed atrial arrhythmia on early postoperatory watch converted spontaneously to sinus rhythm in ICU. At mean follow-up 4 years (range 6 months – 8 years) all patients are on sinus rhythm and a 24 hours Holter didn't show any arrhythmias. A transthoracic echocardiography didn't show any atrial residual shunt nor signs of obstruction of pulmonary veins or SVC. Conclusion: patients with Sinus Venosus Syndrome present arrhythmias (30%) and obstruction of pulmonary veins or SVC and many techniques have been suggested to avoid these complications. With our approach all patients are free from arrhythmias and obstruction. The bovine pericardial and Dacron Sauvage are good material for this procedure.

P355

Scimitar syndrome associated with coarctation of aorta: a case report

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The scimitar syndrome is a rare congenital cardiopulmonary malformation, characterized by anomalous right pulmonary venous drainage in to inferior vena cava below the diaphragm. It is visible roentgenographically as crescentic shadow of vascular density at the right border of cardiac silhouette called “scimitar sign”. Its association with coarctation of aorta is extremely rare. Herein, we report a 29 day-old male infant with scimitar syndrome who also had coarctation of aorta. He had symptoms of heart failure on admission. Chest X-ray demonstrated hypoplastic right lung and dextroposition of the heart. 2 D echocardiography and colour Doppler showed secundum type atrial septal defect and coarctation of aorta. Cardiac catheterization revealed pulmonary hypertension and mild left to right shunt. Angiocardiography disclosed abnormal drainage of right pulmonary vein in to the inferior vena cava below diaphragm and also a severe coarctation of aorta. Surgical correction of coarctation of aorta was attempted. In spite of successful repair of the coarctation (end to end anastomosis), the patient died on the 10th post operative day due to respiratory insufficiency. We thought that the degree of hypoplasia of lung and severity of associated cardio-vascular anomalies are important in predicting prognosis of patients with scimitar syndrome.

P356

Venoatrial connections in patients with left isomerism with reference to surgical repair

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Objective. To determine variations in venoatrial connection, and their surgical implications, in patients with left isomerism. Methods. Venoatrial connections were identified in 62 patients undergoing definitive repair and in another 56 postmortem specimens, all having left isomerism. Anatomic repair was carried out in 45 of the clinical series, and would have been feasible in 35 autopsied hearts. The Fontan procedure was, or would have been, chosen in the other 17 and 21, respectively. Results. SVC was bilaterally present in 68 (58%). IVC was interrupted in 58 (71%), with drainage via either the right-sided (in 47) or left-sided (in 42) azygous vein. The right and the left hepatic veins independently drained in 40%. In 5.9%, the right pulmonary veins drained to the right-sided atrium with the left ones to the left-sided atrium. The coronary sinus was absent in 89%. To establish anatomic repair, a complicated intracardiac baffling was, or would have been, needed in 27 (60%) patients and 28 (80%) specimens, because of their tangled venoatrial connections or their discordant ventricular topology. The presence of dual inferior vena, seen in 21 of 38 unsuitable for biventricular repair, prominent, as would have required, surgical devices when establishing cardiopulmonary bypass and removing these veins to the pulmonary arteries. In the clinical series, postoperative venous obstruction occurred in one in whom SVC occluded at the intraatrial baffle after biventricular repair. Conclusion. Abnormal venoatrial connections were common in this setting. With precise recognition of patterns of venous drainage, surgical repair can be efficiently achieved.

P357

Long-term follow-up in repaired partial anomalous pulmonary venous connection (papvc)

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Commonly, PAPVC is operated on after diagnosis. The purpose of the study was to evaluate the risks and possible benefits of routine surgical correction of PAPVC and to compare the results of the postoperative and the natural history of the pts until repair. 262 pts (123 m, 139 f) aged 1 to 68 (mean 23) yrs were

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

operated on consecutively between 1987 and 1993. Preoperative freedom from AF was 92 ± 5 % for pts under 50 yrs, from tricuspid regurgitation (TR) 93 ± 4 %, and from pulmonary hypertension (PH) 68 ± 6 %. 5 pts (2 of PH, 3 of unknown cause) died late (2 to 24 yrs postop). 74 pts aged 1 to 63 yrs were clinically reinvestigated. (Follow-up: 1 to 34 (mean 13) yrs). Either dilation of the right ventricle in 34, and mild TR in 16 cases. ECG: 64 pts were in sinus rhythm, 4 had AF, and 4 atrial flutter, and 2 a permanent pacemaker. Conclusions: Surgical correction can be done safely, postop morbidity is low but cannot be disregarded. According to the Kaplan-Meier curve for AF, TR and PH there is no urgent indication for surgery in uncomplicated cases.

P358

Partial anomalous pulmonary venous connection with intact atrial septum : clinical presentation and surgical results

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Partial anomalous pulmonary venous connection (PAPVC) with intact atrial septum (IAS) is a rare entity and so far there are no clinical series except few case reports. We report here our experience with 8 patients with this anomaly operated at our institute. The median age was 15 years (range 2yr – 38 yr). Three children had history of repeated chest infection. Four adults presented with exertional dyspnoea and atypical chest pain and one child was diagnosed while investigating for an asymptomatic murmur. The right pulmonary veins were draining to the RA – SVC junction in six patients, body of RA in one patient and RA – IVC junction in one patient. Additionally in one patient the left upper pulmonary vein was draining to the innominate vein through a vertical vein. In 2 patients the diagnosis was established by echocardiography while other six underwent cardiac catheterisation to confirm diagnosis. All patients were operated under cardiopulmonary bypass. A defect was created in the atrial septum and the anomalous pulmonary veins were baffled to the left atrium with a pericardial patch. Six patients underwent augmentation of RA-SVC junction additionally. In the child with additional anomalous left pulmonary vein the vertical vein was transected and was anastomosed to the left atrial appendage. There was no early or late mortality. The mean hospital stay was 7 days. At median follow up 7 months (range 1 month – 5 yrs) all were in sinus rhythm and postoperative echocardiography revealed no baffle leak, and no systemic or venous obstruction. Mostly the patient presents with chest infection or exertional dyspnoea in adolescents period. Angio cardiography is required to confirm the diagnosis in most cases and surgical correction of this rare anomaly can be done with very gratifying results.

P359

The effect of surgery on cardiac rhythm in patients with total abnormal pulmonary venous drainage

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After surgery in which the atrium is incised, (atrial) arrhythmias are frequently seen. To determine which incisions for total abnormal pulmonary venous drainage (TAPVD) affects cardiac rhythm, 29 patients (20 males, 9 females), median age 1.6 months (range 1 day – 19,5 months) who underwent operative correction for TAPVD between April 1986 and March 1999 were studied retrospectively. Different incisions were used depending on the anatomy. Median follow up was 5.8 year (0.25–10.7). Nine patients died, five of whom with in 30 days. EKG monitoring was performed in all. 24-h ambulatory EKG (Holter) monitoring in all but one patients. All patients were asymptomatic. On EKG monitoring the postoperative rhythm was sinus in 14 (58%) and ectopic in 10 (42%) patients. In 5 of these latter 10 patients sinus rhythm changed to atrial rhythm during follow up. Early in follow up one patient had junctional ectopic tachycardia followed by frequent ventricular ectopic beats. Two patients had supraventricular ectopic beats and one patient had supraventricular tachycardia. On Holter monitoring we found significant arrhythmias in 14 out of 20 patients, including intermittent atrial rhythm (8), sick sinus syndrome (2), supraventricular tachycardia (3), ventricular tachycardia (1) and multiform supraventricular or ventricular ectopic beats (10). There was no correlation between the presence of these arrhythmias and type of atrial incision, cross clamp time or cannulation technique. Nonetheless, absence of respiratory arrest, bradycardia, type and multifocality were risk factors for these arrhythmias. Thus, postoperative arrhythmias, though usually asymptomatic are frequently seen following correction of TAPVD. The most specific way to detect these arrhythmias is by Holter monitoring and may be of importance for future follow up. The meaning and implication of the above mentioned remarkable risk factors are under investigation.

P360

Cor triatriatum: report of two cases

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Cor triatriatum is an extremely rare congenital heart disease. It is characterised with a membranous diaphragm which divides the left atrium into two chambers, the proximal chamber accepts the pulmonary veins and the distal one communicates with left ventricle via mitral valve. The size of the orifice between distal and proximal chambers is the main determinant of physiologic abnormalities and clinical symptoms. The patients with severe obstruction between chambers develop symptoms early in infancy. When there is a nonobstructive cor triatriatum there are no symptoms. Herein, we report two cases of cor triatriatum. The first case was a seven year-old boy who admitted to our hospital with failure to thrive and palpitation. 2-D echocardiography and colour Doppler showed an obstructive type of cor triatriatum. Cardiac catheterization revealed elevated pulmonary artery and pulmonary capillary wedge pressures (mean pressures were 30 mmHg and 29 mmHg respectively), and angiocardiographs revealed double chamber left atrium. The membrane removed surgically and the patient is well after operation. The second case was a one year old boy who admitted to our hospital with symptoms of heart failure. 2-D echocardiography and colour Doppler showed 5 mm perimembranous VSD and nonobstructive membranous diaphragm which divided left atrium into two chambers. Cardiac catheterization revealed moderated left to right shunt (Qp/Qs: 1.6/1) and pulmonary hypertension with mean pressure of 28 mmHg. The patient underwent surgical procedure for VSD closure and even though it was not an obstructive one, the membrane of cor triatriatum removed. The patient is well after operation. The report shows wide symptomatology of cor triatriatum. The size of the orifice between chambers and additional cardiac lesions are the determinant of severity and severity of hemodynamic abnormalities and clinical manifestations.

P361

Clinical Spectrum and Outcome of Partial Anomalous Pulmonary Venous Connections in Children

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Partial anomalous pulmonary venous connections (PAPVC) involving one or more pulmonary veins may be found in the setting of other congenital heart defects (CHD) or rarely occur in isolation. This retrospective study includes 52 pediatric patients (<14 years) diagnosed to have PAPVC from January 1986 to March 2000. The ages ranged from 16 days to 14 years. Isolated PAPVC was the accounting for only 7.4%(4) cases. Atrial septal defects were the commonest associated defect, being present in 80%(46) cases. Right pulmonary vein alone were involved in 58% cases, left pulmonary vein alone in 6% and mixed drainage was found in 6% cases. The commonest site of drainage of the right veins was to the SVC-RA junction in 31(62%), SVC in 9(18%), RA in 7 (14%) and IVC in 3(6%) cases. All anomalously draining left veins connected to the innominate vein and one patient had dual connection to both left atrium and the innominate vein. Recurrent respiratory tract infection and failure to thrive was the commonest presenting feature in 82% cases, while 3 cases(5%) of isolated PAPVC presented for evaluation of chest pain. Echocardiography correctly identified all the lesions with 2 false positives. 37 patients (67.9%) underwent surgery and rerouting of pulmonary veins with good results. 4 patients had mild SVC obstruction with one death in the setting of complex CHD. In conclusion, current echocardiographic techniques allow for the accurate diagnosis and delineation of the various subgroups of PAPVC, though a high index of suspicion is required. Surgical results and outcome are excellent.

P362

Intermediate Outcomes in Total Anomalous Pulmonary Venous Drainage

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Early postoperative mortality for TAPVD ranges in recent reports between 9–14%, late mortality is 2–3%, and reoperation is as high as 11%. The purpose of this study was to determine outcomes of TAPVD repair in the current era. Patients with TAPVD as a primary lesion who underwent repair between 1990 and 2000 were included in study. Clinical course including morbidity, mortality, reoperation, and intermediate functional status was reviewed. Study population was 46 patients; mean age at repair was 45 days (range 1 day to 21

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

enendu). TAPVD was supracardiac in 24 patients (50%), infradiaphragmatic in 11 (23%), coronary sinus in 9 pts (19%), including 1 patient with unilateral lung aplasia, eight atresia in 2 (4%), and mixed in 2 (4%). Pulmonary venous obstruction was significant in 22/48(46%). Extracardiac repair was performed for supracardiac and infradiaphragmatic TAPVD. Drainage to the coronary sinus or the right atrium was managed by unroofing the coronary sinus or septation of the right atrium to enlarge the left atrium. Nitric oxide was utilized pre- or post-operatively in 7 patients; ECMO support was necessary in one patient, who is recovering. One patient with severe pulmonary venous obstruction died intraoperatively. Three patients required reoperation, 2 had successful relief of obstruction, 1 patient died 5 weeks post-operatively due to cardiopulmonary venous stenosis. At latest follow-up only one patient has evidence of mild pulmonary venous obstruction, the remaining are asymptomatic with no evidence of pulmonary arterial hypertension and normal growth and development. In conclusion, 1) surgical outcome is excellent for infants with TAPVD, regardless of site of entry and degree of obstruction 2) reoperation was necessary in 6% of patients and 3) the vast majority of repaired patients with TAPVD have excellent intermediate term status

P363

Modified Fontan procedure in the presence of supracardiac total anomalous of pulmonary venous connection

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The purpose of this report is to outline technical maneuvers dictated by supracardiac total anomalous of pulmonary venous connection (TAPVC) in the performance of Fontan procedure. Between 1979 and 2000, 5 patients underwent supracardiac TAPVC repair concomitantly with a modified Fontan procedure at the Heart Institute of Japan. Mean age was 5.2 years. Main diagnosis was single right ventricle in 2 patients and complete atrioventricular canal (type C) in 2. Right side isomerism was identified in 4 and pulmonary stenosis or pulmonary atresia was in 5 (all). Mean PA pressure was 16.6. The connection site of TAPVC was innominate vein in 2, SVC in 2, both in 1. Previous palliative operation had been performed in 1 patient. There was no remarkable preoperative pulmonary venous obstruction. Surgical repair was accomplished by bottom ligation or division of superior vena cava (SVC) for correction of TAPVC (5) and or by PV-LA direct anastomosis through superior approach for TAPVC (1a). In the presence of obstruction at TAPVC connection site in TAPVC (1b), PV-atria direct anastomosis was performed after isolation and cut back. In the case of TAPVC (1a+ 1b), PV-LA anastomosis, SVC isolation and vertical vein – PA direct anastomosis were performed. Fontan circulation was obtained communicably by atrial septation or intra-atrial conduit and by atrial appendage - PA anastomosis. There were 1 early death and 4 survivals and there was no pulmonary venous obstruction. We conclude that the outcome of the modified Fontan procedure with supracardiac TAPVC repair in patients with asplenia syndrome or SRV was satisfactory

P364

Aortic coarctectomy in children: long-term (30 years) single center observation

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Long-term single center results after aortic coarctectomy in children were reviewed. From 1990 to 1999 104 patients, 30 neonates (<1month), 29 infants (1month-1year) and 45 children (1–8years) underwent aortic coarctectomy. Aortic coarctation was isolated in 31 children, with associated lesions in 73: PDA (26), bicuspid aortic valve (21), VSD (18), hypoplastic aortic arch (14), LVOTO (14), mitral stenosis (6), ASD (5), TGA (2), aortic valve regurgitation (1), mitral regurgitation (1), univentricular heart (1). Surgical technique was end-to-end anastomosis (56), patch aortoplasty (28), subclavian flap (16), pilooplasty type (11), conduit (3). Associated procedures: PDA closure (36), PA banding (6), aortic valvotomy (2), VSD closure (1) and Senning (1). Thirteen children underwent another surgical procedure because of associated heart defects. There were no hospital deaths. Actuarial survival 97.1% at 10 years, remained unchanged at 20 and 30 years. All 3 late deaths were related to associated lesions. Freedom from re-operation because of re-coarctation was 99% at 1 year, 93% at 10 years, 89% at 20 and 30 years. Eight patients (6/8 operated on before 1981) required re-operation because of re-coarctation : four neonates (3 subclavian flap, 1 pilooplasty type), one infant (end-to-end anastomosis) and three children (all patch aortoplasty). Incidence of reoperation was 13.3% in neonates (4/30), 3.4% in infants (1/29) and 6.6% in children (3/45)(NS at Fisher's exact test), with regard to the surgical technique was 18.7% (3/16) for subclavian flap, 15.0% (3/20) for patch aortoplasty, 9.1% (1/11) for pilooplasty type and 1.3% (1/76) for end-to-end anastomosis (P<0.05 at Fisher's exact test). 77/104 survivors have an ultimate mean peak gradient <20mmHg at cuff and Doppler measurement. In our center resection and end-to-end anastomosis provided the best long-term results for repair of Aortic Coarctation.

P365

Aorto-ventriculoplasty after a prior aortic valve replacement and posterior root enlargement

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A determining finding is the need for reoperation, over a variable period of time in children who had been operated for aortic stenosis. This is more likely to be the case, patients with a narrow annular anulus. We describe 3 cases with left ventricular outflow tract obstruction who had an aortic valve replacement and posterior root enlargement. Several years later, due to residual aortic stenosis an anterior annular enlargement by the Konno procedure and an aortic valve replacement was successfully performed in all cases. Good relief of aortic and subaortic stenosis, along with absence of significant gradient across the left ventricular outflow tract, lead us to believe that, in a situation as encountered by us, successful and very effective relief can be obtained by adding a Konno type enlargement to a previously performed aortic valve replacement and posterior enlargement

P366

Evolving perioperative management for hypoplastic left heart syndrome during eight years gives evidence for improved outcome

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Background: Many modifications have been proposed since Norwood reported nearly 20 years ago on the first successful surgical treatment for hypoplastic left heart syndrome (HLHS). Timing of the initial and subsequent operations, surgical techniques and perioperative medical treatment vary among the institutions. The aim of the study was to look at these issues over the past eight years to pin down the causes for the improved outcome. Methods: We retrospectively analysed the pre- and post-operative management as well as the surgical techniques of the Norwood I procedure in all patients with hypoplastic left heart syndrome over the last eight years. Results: Since Aug. 1992, 29 Norwood operations were performed for HLHS in two four year time periods. The total mortality was 34%. The mortality rate in the first four years (Aug. 1992 to Jul. 1996; 13 pts.) was 61.5% and in the later four years (Aug. 1996 to Aug. 2000; 16 pts.) 12.5% respectively. Perioperative management is currently run as follows: Preoperative: Balancing of Qp/Qs on intensive care unit, avoidance of ventilation and urgent transfer for surgery in case of severe obstructive PH-O, otherwise timing of the operation in the second week of life. Surgical technique: Aortic arch augmentation distal to the isthmus using the curvature of a cryopreserved pulmonary artery bifurcation homograft, insertion of unrestricted coronary blood flow, smallest possible shunt size, reduction of cross clamping and circulatory arrest time, low dose inotropes. Postoperative: Balance of Qp/Qs primarily by ph-management, afterload reduction, early weaning from ventilator. Conclusion: Clinical outcome of the Norwood procedure for hypoplastic left heart syndrome has improved tremendously and can today be performed with an acceptable risk. Multiple factors during the perioperative period presumably account for the superior results, which are now based on a precisely defined regimen.

P367

Accessory mitral valve tissue. Three case reports and current literature review

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BACKGROUND: Accessory mitral valve (AMV) tissue is a rare congenital malformation causing left ventricular outflow tract obstruction(LVOTO). MATERIALS AND METHOD: We found this anomaly in 3 adult patients. The first patient, a 60-years old man, presented AMV leaflet arising from the antero-aortic commissure, accessory papillary muscles and mild LVOTO(dp=38mmHg) and coronary artery disease. He undergone successful coronary revascularization and AMV leaflet resection. The other

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

patient, a 27-years old man, presented atrial septal defect and LVOTO(dp=72mmHg) due to AMV leaflet. He undergone successful septal defect closure and AMV leaflet resection. The third patient, a 42-years old man, undergoing left anterior descending artery angioplasty, presented an AMV lesion not causing LVOTO. RESULTS: The data analysis of various reports revealed 89 patients presenting this anomaly. The age range from same hour old newborns to 72 years old, mean 9.2±6.7. The interventricular septal anomalies were found in almost 98% of cases. We classified this anomaly as follows: Type I-FIXED TYPE (A-nodular, B-Membranous) Type II-Mobile type (A-Pedunculated, B-leaflet like). The type IIB is divided in 1)rudimentary chordae and 2)developed chordae. Important LVOTO was present in most of the cases (mean 71.17±18mmHg), even though, patients with mild obstruction are reported. 67 (75%) patients undergone cardiac surgery with a postoperative mortality 5(7.5%)patients. Postoperatively residual mild-to moderate LVOTO was identified in 9(13.4%) patients preoperatively 9(13.4%) patients necessitated reoperation. Mild-to-moderate mitral or aortic valve regurgitation were found in 6(8.9%) and 5(7.5%) patients respectively. DISCUSSION: The removal of the AMV tissue should in no way compromise the mitral valve function and its excision should be made under careful evaluation of the surrounding structures. The availability of Doppler echocardiography has leaded to an appropriate identification of the AMV tissue before the development of symptoms or important LVOTO. CONCLUSION: Patients with AMV tissue causing LVOTO can undergo safe cardiac surgery and mass excision with acceptable mortality and postoperative morbidity.

P368

Aortic aneurysm after patch aortoplasty for coarctation: is it real? A study of aortic wall growth

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Background: Aortic aneurysm (AA) has frequently been reported after patch aortoplasty for aortic coarctation, leading many to abandon this method of repair. However, diagnosis usually based only on localized dilatation (>150% descending aorta). This study analyzes the size of the patch and adjoining native aortic wall to determine the incidence of true AA formation following coarctation repair. Method: Electron beam tomography (ebe CT) was done on 19 asymptomatic patients 3 months-17.5 years (mean 9.6 years) post gortex patch angioplasty for aortic coarctation to examine the repair site. The image with largest cross-sectional area was used to measure patch segment: (Cp), native aortic wall segment (Cw), and total circumference (Ct). Measurements were normalized to circumference of distal descending aorta (Cda), as Cp/Cda, Cw/Cda, Ct/Cda. The patch/native aortic segment ratio (Cp/Cw) was also determined. Result: In all patients aortic images showed smooth convexity of native aortic wall without outpouching, i.e.without true AA. Total circumference (Ct/Cda, mean 160±48%) showed excellent linear correlation (r=0.92) with patch size (Cp/Cda). Ct/Cda of >150% was associated with Cp/Cda >90%. However, total circumference (Ct/Cda) vs native aortic wall (Cw/Cda) showed poor correlation (r=0.55), likewise, patch (Cp/Cda) vs native aorta (Cw/Cda) showed no correlation (r=0.18). Based on size of total circumference (Ct/Cda), patients were categorized into group A (n=12, Ct/Cda >155%), and group B (n=7, Ct/Cda <153%). Total circumference (Ct/Cda) and patch size (Cp/Cda, Cp/Cw) were significantly higher (P<0.0001) in group A than group B, however, the native aortic wall segment (Cw/Cda) did not differ between groups, varying from 51% to 122%. This was higher than the coarctation/descending aortic diameter ratio of 26±4% on preoperative angiograms, indicating normal aortic wall growth post surgery. Conclusion: Localized dilatation of the aorta following patch aortoplasty does not necessarily represent a true aortic aneurysm. Enlargement is primarily due to the presence of a large synthetic patch. Native aortic wall growth does occur post surgery. Patch enlargement remains a safe method for repair of coarctation.

P369

Single stage repair for intra cardiac anomalies associated with arch obstructions without circulatory arrest

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Intracardiac defects associated with arch obstructions is a complex congenital anomaly. Conventional surgical treatment for this anomaly is two stage procedure. Recently aggressive approach has been adopted to treat the anomaly with one stage. We report our experience with single stage repair without circulatory arrest. >From June '99 to Nov 2003,9 patients were treated with single stage correction for various intra cardiac defects associated with aortic arch obstructions. The age ranged from 2 months to 3 yrs (mean 1.2 years). The clinical spectrum included: VSD-3, DORV,VSD-3, d-TGA,VSD-2, subaortic obstruction-1. Seven patients had coarction and two patients had significant hypoplasia of aortic arch with coarctation. All patients were operated under C.P.B. During cooling the arch and neck vessels and thoracic aorta distal to the coarction segment were dissected to a sufficient length. The count segment was arrested between the two clamps and end to end anastomosis was done. In arch hypoplasia the cerebral perfusion was maintained through innominate artery while rest of the neck vessels were dissed. Under cardioplegic arrest intracardiac repairs such as VSD closure, arterial switch operation and subaortic resection were performed. Tissue to tissue anastomosis was achieved in all the patients. There was no hospital mortality and no neurological deficit. Postoperative echocardiogram showed no residual coarction or intracardiac defects. There were no reoperations and late deaths. It reduces the number of operations, hospitalisation and cost. Most of the repairs could be done without circulatory arrest. Early and mid term results are excellent.

P370

Aortic Valve Stenosis: A Thirty Year Experience

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A thirty-year retrospective review of all patients undergoing therapeutic interventions for aortic valve stenosis at Children's Hospital of Wisconsin since September 1969 was performed. The included 238 procedures in 173 patients, including 54 infants (31.2%) with critical aortic stenosis and 37 patients (21.4%) with significant associated cardiac malformations. Initial operative approach included balloon aortic valvuloplasty in 30 (17.3%), open commissurotomy on cardiopulmonary bypass in 118 (68.6%), valvulotomy under inflow occlusion technique in 24 (13.2%), and mechanical valve replacement or Ross procedure in 5 (2.9%). Clinically significant operative outcomes were defined as: good (residual gradient <50 mmHg, regurg <grade 2), fair (grad <50 mm Hg, regurg 2-3) or poor (early reintervention, grad >50 mmHg, regurg >3). Results: The early operative outcomes in those infants with critical aortic stenosis (54 pts) are presented in table 1. The early results of all 173 patients are presented in table 2. Follow-up on 155 initial survivors was 5.7 yrs (range 0 to 25.7 yrs). Ten of 29 survivors (34.5%) having undergone initial balloon aortic valvuloplasty underwent reoperation at a mean of 3.8 + 3.04 yrs. Twenty-two of 106 pts (20.8%) who underwent open commissurotomy at an initial approach required reoperation at a mean of 7.3 + 5.30 yrs (Kaplan-Meier, p<0.01). Conclusion: The palliative nature of aortic valve surgery is again apparent. Inflow occlusion is of historical interest. Open commissurotomy and balloon valvuloplasty provide equally effective immediate results in the patient with critical aortic stenosis. Overall, open commissurotomy provides a more effective and longer lasting operative result.

P371

Use of the valve-sparing Konno procedure for complex left ventricular outflow tract obstruction

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Complex left ventricular outflow tract obstruction (LVOTO) with normal aortic valve function requires aggressive resection in the subaortic region and preservation of the aortic valve. The valve-sparing Konno procedure (vsKP) allows generous exposure of the LVOT from the left ventricular apex to the interleaflet trigones of the aortic valve. Widespread use of this procedure has been limited by concern over injury to the aortic valve, the conduction system, and residual VSD. The vsKP was used in eleven patients (age 1 to 31) for LVOTO associated with previous subaortic membrane resection (n=8), diffuse subaortic hypertrophy (n=2), and persistent closure of ventricular septal defect (n=1). Ten of the patients had undergone previous LVOT resection. There were no perioperative deaths. LVOT peak gradient by echocardiogram were 77+/-22 mmHg (preoperative) and 19+/-18 mmHg at most recent follow-up (p<0.001 vs preop). Aortic insufficiency was mild or less in all cases. There were no cases of permanent heart block. Small residual VSD's were present in 4 patients (36%). Median follow-up is 3.4 years. The modified Konno procedure can effectively relieve complex LVOTO and preserve aortic valve function. Small residual VSD's were common but clinically insignificant. Extension of this procedure for use in the initial presentation of LVOTO may be appropriate in cases at increased risk of recurrent LVOTO.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P372

Determination of the severity of aortic stenosis using cardiopulmonary stress testing

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Children with aortic stenosis (AS) may have impaired aerobic capacity. Studies however have demonstrated no relation between oxygen consumption (VO2) and the degree of stenosis. We sought to determine the relationship between the degree of AS and other measures of aerobic capacity using cardiopulmonary stress testing. Sixteen patients with an age of 14.4+/-4.1 years and weight of 59.4+/-21.9 kg were studied. Exercise variables were expressed as a percent of predicted (ppm) based on a healthy age-gender-race matched control group. Resting supine echocardiography performed prior to exercise demonstrated peak and mean gradients (Pkrest & Xrest) using blind CW-Doppler from the suprasternal notch of 48.6+/-22.1 and 25.5+/-14.2 mmHg respectively. Peak and mean gradients obtained with the patient upright on the cycle-ergometer at peak exercise (Pkex & Xex) were 97.5+/-40.4 and 44.7+/-19.6 mmHg respectively. There was no correlation between VO2 and echocardiographic gradients. Patients with higher PKex and PKrest had a depressed rise in oxygen pulse (r=-.76 p=.0006 and -.73, p=.001 respectively) while only PKex was correlated with an increased change in [D]-[D]ETPCO2/[D]work (r=.54, p=.03). Increased [D]VE/[D]VCO2 (r=.51 p=.04) and reduced [D]VO2/[D]work (r=-.57, p=.02). The degree of aortic stenosis did not correlate with the presence of ST changes or BP response. Patients requiring aortic balloon angioplasty had higher PKrest and Xrest (67 vs 38 mmHg p=.006 and 38 vs 18 mmHg, p=.02), higher PKex and Xex (135 mmHg vs 75 mmHg p=.001 and 59 mmHg vs 36 mmHg, p=.02), lower ppm [D]oxygen pulse/[D]work (49 vs 79%, p=.005), greater ppm [D]VE/[D]VCO2 (115 vs 96%, p=.05) and steeper [D]HR/[D]work (1.7 vs 3.6, p=.04). Cardiopulmonary stress testing is of greater utility than cardiac stress testing alone for identifying patients with significant aortic stenosis. Exercise echocardiographic gradients correlate better with measures of aerobic performance.

P373

Strategy and long-term results of the Fontan procedure with systemic ventricular outflow obstruction

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To achieve unobstructed blood flow from the systemic ventricle to the aorta is important at the Fontan procedure for complex cyanotic congenital heart disease when there is systemic ventricular outflow obstruction (SVOO). Twenty-three patients underwent the Fontan procedure with SVOO. Sixteen patients had had prior pulmonary artery banding and SVOO had been progressed in 6 patients after the Fontan procedure. Main diagnosis was single ventricle in 11 (SLV 8, SRV 3), TA in 4, d TGA in 2, L-TGA in 2, DORV in 2 and AV canal in 2. The average age was 6.6 years and the SVOO gradient ranged from 0-100 (average 30.4) mmHg. The Damus-Kaye-Stansel (DKS) procedure was performed in 16 (Lamberti's modification in 13, end to side anastomosis in 5), VSD or hulboventricular foramen (BVF) enlargement was performed in 9, and muscle resection was performed in 6. Lamberti's modification (double-barrel method) is our first choice if pulmonary valve is intact since 1994. There was no early death and one late death of SRV (4.3%). Follow-up ranges from 5 months to 14 years (average 4.7 years). Nineteen of the 23 patients have undergone recatheterization. There are 0-20 (average 1.8) mmHg gradients in the SVO area, and CVP ranges from 9-20 (average 14) mmHg. In all patients who underwent VSD or BVF enlargement, regular sinus rhythm is maintained. Regarding the DKS procedure, there is minimal progression of semilunar valve insufficiency except 1 patient who underwent the end to side anastomosis with moderate pulmonary regurgitation preoperatively. Long-term results of the Fontan procedure with SVOO are satisfactory. SVOO could be progressed after the Fontan procedure if there was morphological obstruction. Therefore the appropriate strategy to achieve obstruction to systemic blood flow should be performed concomitantly at the Fontan procedure.

P374

Subaortic stenosis – does anatomy predict clinical profile?

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Between 1982 and 1997, all patients diagnosed with subaortic stenosis (SAS) were identified from echocardiography log books and operative notes. 178 patients were divided into 6 anatomic groups. 1 isolated SAS (n=69), 2 ventricular septal defect (VSD) and SAS (n=27), 3 right ventricular (RV) muscle bundles with 2 (n=11), 4 coarctation (CoA) with SAS (n=11), 5 CoA with VSD and SAS (n=9), 6 major mitral valve abnormality associated with SAS (n=11). Isolated SAS had a significantly later age at diagnosis compared to other groups, by independent t tests (mean 5.65yr vs 3.53, p<0.0001). Intergroup analysis was done using ANOVA with Bonferroni post hoc testing. The gradient at diagnosis was significantly higher in Groups 1 and 6, however, age-corrected gradient (gradient divided by age) was significantly higher in Group 6. Group 1 was operated at a significantly older age than Group 6 (p<0.01) with a higher preoperative gradient. Group 6 had a higher age-corrected preoperative gradient than the other groups. Resection was done in 53, myectomy in 41, Konno-type operation in 2 and details unavailable in 1, with 4 deaths (1 in Group 1 and 3 in Group 6). 13 underwent reoperations (7 in Group 1 with 1 death, 3 in Group 4 and 6 with 2 deaths). There were 9 recurrences (gradient > 40mm Hg at followup), with no differences between various groups. 65 patients had serial followup echocardiograms with mean followup duration of 4.5 years. 23 showed significant progression (gradient > 50 mm Hg at end point) compared to 25 with no progression (gradient < 25 mm Hg). This did not vary significantly between various anatomical groups. In conclusion, anatomical type determines age of onset and severity of subaortic stenosis but do not predict progression.

P375

Mid to long term result of intraventricular repair for Taussig-Bing Anomaly – in the aspect of left ventricular outflow tract obstruction

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Current surgical approaches for the Taussig-Bing anomaly include arterial switch operation (ASO) with ventricular septal defect closure and Kawashima intraventricular repair (IVR). Left ventricular outflow tract obstruction (LVOTO) is a concern after IVR. Since 1969, 19 children underwent surgical repair of Taussig-Bing anomaly. Among these patients, IVR was performed in 5 patients and ASO was performed in 4 patients. Age at operation ranged from 8 months to 8 years. In IVR group, 4 patients had side by side great arteries and one had oblique relationship. In ASO group, two patients had anteroposterior great arteries and the other had oblique great arteries. After IVR, there was one operative death due to septicemia. In ASO group there were 2 operative deaths due to low cardiac output. In IVR group, the narrowest distance from the tricuspid ring or chordae to the pulmonary valve ring (T-P distance) was ranged from 8 to 18mm. These values were 79 to 126% of normal aortic diameters. Postoperative systolic gradient across left ventricular outflow was ranged from 0 to 20 mmHg. Two patients had no LVOTO after 13 and 31 years after repair. Other 2 patients revealed to have significant LVOTO after 9 and 14 years after repair. One patient underwent revision of the intraventricular baffle. At the revision, calcified thick gel on the baffle was thought to be the major cause of LVOTO. T-P distance was 16mm. (100% of normal aortic diameter), although it was 8mm. (79%) at the initial repair. The other is waiting for the revision. In conclusion, Kawashima intraventricular repair yields excellent early and late results despite of late LVOTO. LVOTO was safely treated by the revision of the baffle with the grown T-P distance.

P376

Subaortic stenosis : do associated anomalies predict presentation, progression and recurrence?

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Subaortic stenosis (SAS) was diagnosed in 178 patients between 1982 – 1997, and grouped according to associated abnormalities: gr 1 isolated SAS n=69, gr 2 with ventricular septal defect (VSD) n=27, gr3 with VSD and RV muscle bundles n=11, gr 4 with coarctation (CoA) n=11, gr5 with VSD and CoA n=9, gr 6 with LV inflow obstruction and CoA n=11. Patients with valvar aortic stenosis and AV septal defect were excluded. Isolated SAS was diagnosed later than other groups (6.67+3.92 v 3.53+3.87, p<0.0001). The gradient at diagnosis was higher in grps 1 and 6, however, age-corrected gradient (gradient/age) was highest in grp alone. 65 pts with mild SAS (gradient < 25mm Hg) had serial prospective (mean followup 4.5yr, progression of obstruction (gradient > 50mm Hg) occurred in 23, but this did not

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

vary between groups. Surgery for SAS was performed in 97 pts (resection 54, myectomy 41, Konno 2) with 4 deaths (1 gr 3, 3 gr 6). Age at surgery for SAS was younger for gr 6 pts compared to gr 1 (3.24±2.98 v 8.58±4.59, p<0.01). Reoperation for recurrence was required in 33 (7 gr1 with 1 death, 3 in gr 4 and 6, 2 deaths). Recurrence (gradient >40 mm Hg) occurred in 9 other patients with no difference between groups. Septal left heart obstructions was associated with more severe SAS at younger age, required earlier surgery and had high mortality. Associated abnormalities did not otherwise render progression of mild SAS preoperatively, nor post operative recurrence in survivors.

P377

Primary repair of coarctation or interruption complex using isolated cerebral and myocardial perfusion

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To prevent possible neurologic injury after hypothermic circulatory arrest, we have repaired aortic arch obstruction (AAO) with cardiac defects in our department using isolated cerebral and myocardial perfusion (ICMP). For the last 5 years, 28 infants with simple AAO (CoA: 11/VSD=8, IAA: 9/VSD=10) and 10 with complex AAO (CoA with critical AS=3, Taussig-Bing: DORV=4, VSD+PAPVC=1, IAA with AP window=1, VSD+SAS=1) underwent primary total correction. An arterial cannula was inserted either into the ascending aorta or into a polytetrafluoroethylene graft which was anastomosed to the innominate artery. A cross-clamp was placed between the innominate and left carotid arteries, and an end-to-end arch anastomosis was performed as much as possible with brain perfusion and heart beating. In 13 patients, the innominate artery proximal to the graft was then secured down and the arch anastomosis was extended to the distal ascending aorta providing isolated cerebral perfusion. After arch reconstruction, the clamp was moved onto the ascending aorta, and intracardiac defects were repaired with cardioplegic arrest. The mean ICMP time for all patients was 15+/-5 min (range, 3–25 min). Descending aortic clamp time was longer than 30 min in only 3 complex AAO patients. There were no early and 1 late death (96% survival) in simple AAO group, and 2 early and 2 late deaths (60% survival) in complex AAO group. Primary repair of AAO and cardiac defects can be performed without use of total circulatory arrest. The ICMP technique may offer substantial brain and myocardial protection during aortic arch reconstruction.

P378

The changing fate and survival of newborns with pulmonary atresia or critical stenosis and intact ventricular septum

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The hypothesis that new protocols may favourably affect the prognosis of individuals with PA or critical PS was tested. Methods: in surgical era 1, (1982–1987), a systemic to pulmonary shunt was performed in all neonates, followed by early (<1 mo) RV decompression or later Fontan repair. Subsequently (1988–1996), palliative procedures were adapted to RV morphology/size: Fontan repair was staged by intermediate BCPA +/- retrograde RV decompression. In the contemporary era, RV overbanding and one-and-a-half ventricle repair were increasingly employed. Results: one-hundred twenty-five newborns were treated. Hospital mortality at palliation was 47%, 15%, 12% in the 3 eras (1 vs 2 p=0.004, 2 vs 3 p= NS). Among 81 survivors, 53 were suitable for biventricular repair, 25 (47%) reached the final stage, 5(9%) died and 23 (43%) await repair. Eleven pts were considered for Fontan repair in period 1 (41%), 16 (25%) in period 2 (3 ip=0.21). In period 1, 7/11 candidates for Fontan died (mortality 64%); in period 2 and 3, mortality was 25% (1 vs 2+3 p=0.11). Biventricular repair was achieved with 12% hospital mortality in period 1, 14% in period 2 (p=0.67). Partial biventricular repair was elected for 11 pts (all in eras 2 and 3) and obtained by 3 pts so far. Actuarial survival proportion at mean follow-up interval (84 mos, range 0.5–219) was 61%. Conclusion: neonatal mortality was significantly reduced by relaxing initial palliation to RV morphology/size. Two trends may become significant in the future: 1) the reduction in mortality towards Fontan repair by intermediate procedures; 2) the increasing proportion of pts who reach complete or partial biventricular repair.

P379

Coarctation of the pulmonary artery: diagnosis and surgical treatment

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Coarctation of the pulmonary artery (CoaPA) is a spontaneous acquisition of proximal pulmonary artery branch stenosis often found in patients with pulmonary atresia and ventricular septal defect (VSD). From 1994 to October 2000, we reviewed all patients with critical pulmonary obstruction and normal confluent pulmonary arteries(CPA), survived to neonatal Blalock-Taussig (B-T) shunt. Eight of 71 patients (10.2%) were found to have left CoaPA 4.5 to 22 months after surgery. In addition to pulmonary stenosis, 8 patients had:a) tricuspid atresia(4), b)tetralogy of Fallot (TOF) (3); c) transposition of great arteries and VSD (1). All patients had left-sided aortic arch and single ductal arteriosus ematerale to CoaPA. Diagnosis was found , in all patients, by echo and confirmed by angiocardiography. One patient was lost to follow-up, nine underwent additional procedures of left B-T shunt: (6 cases) and TDI with CoaPA repair (1 case). Hospital mortality was 12.5% (1 case). To a mean follow-up of 30 months, clinical data were excellent in 5 patients, while differentiation data demonstrated growth of discontinuated pulmonary artery diameter to near normal values. CoaPA is a tedious complication, whose missing early detection, prolong surgical plan and compromise expected mortality and morbidity. CoaPA is to be considered an indipendent risk factor of death and surgical neonatal approach of the lesion might keep more chances of success.

P380

Natural history of the intraparenchymal vasculature in complex pulmonary atresia

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Objective: Progressive extrahilar obstruction of major aortopulmonary collateral arteries (MAPCAs) in complex pulmonary atresia (CPA) is well documented. We studied the natural history of intraparenchymal vasculature to observe difference whether it was connected to MAPCAs or native pulmonary arteries (PAs). Tracheal diameter was identified as an independent, age-related factor to calculate a ratio for vessels' diameter change with time. Patients and Methods: 48 patients with CPA were studied in whom the pulmonary circulation depended entirely or predominantly on MAPCAs. There were confluent PAs in 95%, absent arterial duct in 85%. Age at study-entry was 1.5–11months, (mean: 3.3 months), study-period spanned 0.5–15.4years (mean: 7.5years). Only systemic-pulmonary shunt cases were included. Contrast enhanced angiograms were available in 96% to assess the size at hilum, and branching pattern of MAPCAs (3.4/patient) and the PAs. Lower third cerebral diameter was measured to express vessel/trachea ratios. Results: Over the study period there was a decrease in R.PA+L.PA/trachea: 0.62–0.57 (NS), MAPCAs/trachea: 0.94–0.67 (p=0.01), (in the first year-of-life: 0.94–0.71 (p<0.001)), hilum(PA+MAPCA)/trachea: 1.20–1.17 (p=0.05). Ebbing of MAPCAs at hilum as intraparenchymal vasculature was parallel to, albeit, less pronounced than elongation rate of the extrapulmonary vessels. Dist/trachea: 1.21–1.29 (NS). Conclusions: MAPCAs fade were than native PAs. Attrition rate is faster at a younger age. Accompanying extrapulmonary obstruction with time the intrapulmonary vascular bed becomes less amenable for encouraging the whole cardiac output. Systemopulmonary shunts do not enhance PA growth. An early, aggressive surgical strategy is advocated. Intrapulmonary excavation rather than extrapulmonary unifocalisation, may be needed to create hila. Interventions should focus on maximizing the number of bronchopulmonary segments connected to hila. Plotting the tracheal diameter is ideal for the follow-up the change of the pulmonary vasculature as it is readily available, independent from the morbidity.

P381

Complete unifocalization in infancy for tetralogy of fallot with pulmonary stenosis and major aorto-pulmonary collaterals via median sternotomy: early experience

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Background: Tetralogy of Fallot with pulmonary stenosis and major aorto-pulmonary collaterals (TOF/PA/MAPCAs) is characterized by a heterogeneous pulmonary blood supply. The vasculature is at its healthiest state in infancy. Distal stenoses, hypoplasia or pulmonary vascular disease develop almost always. Attrition is highest in infancy. Results of staged unifocalization have not been convincing. Some investigators have accomplished promising early results of complete unifocalization via median sternotomy. This has led us to start correcting TOF/PA/MAPCAs by complete unifocalization in infancy. Methods: Since January 2000 four infants underwent complete unifocalization via median sternotomy. Our purpose was to obtain antegrade

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

blood flow to every single collateral via a right ventricle (RV) to pulmonary artery (PA) continuity after reconstruction of the central PAs using collateral to collateral anastomosis. Results: All MAPCAs regardless of size and position were recruited to create central hybrid pulmonary arteries using only native tissue with the help of autologous pericardium. RV to PA continuity was accomplished using valved cryopreserved homografts. The number of MAPCAs per patient ranged from two to five. One patient had no native central pulmonary artery, his VSD was left open due to multiple peripheral stenoses of the MAPCAs. After balloon dilation of the MAPCAs three months later, he currently awaits VSD closure. All infants survived the procedure with no complications and were discharged home. Conclusion: Our initial experience of repair of TOF/PA/MAPC shows that complete unifocalization of all MAPCAs via median sternotomy in infancy can be performed with low risk. It provides normal physiology early in life and potentially avoids a series of palliative procedures and prosthetic material. Long term follow-up is mandatory to determine whether subsequent operations are limited to homograft replacements only.

P382

Late results after right ventricular outflow tract reconstruction using pedicled autologous pericardio

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Background: Early removal of cyanosis and ventricular load has several potential physiological advantages. The potential for growth of a new pulmonary arterial trunk (PAT) using viable autologous tissue may reduce the need of reoperation to replace the PAT with an extracardiac conduit after the child grows up. The aim of this study is to assess the late results after PAT reconstruction using pedicled autologous pericardium (PAP). Methods: Between June 1993 and February 1999, when 7 patients including 4 infants with tetralogy of Fallot with pulmonary atresia, and 1 neonate with truncus arteriosus underwent complete repairs, and 1 girl with congenital aortic stenosis underwent Ross procedure at 15±17 months of age (range, 12 days-43 months). PAT reconstruction using PAP was performed. The PAT was reversed without undue tension to direct its anterior phase toward the lumen of the PAT. In the former 3 patients, the left atrial appendage and PAP were interposed longitudinally to create more viable posterior wall of PAT. Consecutively 6 patients underwent PAT reconstruction using PAP only. Results: There were no early or late deaths, and no re-operations. All patients have nearly normal arterial oxygen saturation (97%±1%). Three patients required balloon pulmonary angioplasty for branch pulmonary artery stenoses, and one of them showed an aneurysmal dilation of PAT. The other 6 patients remain free of results as a mean of 40±26 months (range, 12–85 months), and growth of the PAT was observed. Conclusions: The experience indicates that PAT reconstruction using PAP is associated with good late outcomes, and it might be applied to most infants to remove cyanosis and vent

P383

Repair of pulmonary atresia with ventricular septal defect without the use of a conduit

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Objective: Generally a conduit is needed to establish the continuity between the right ventricle(RV) and the pulmonary artery(PA) in case of repair of pulmonary atresia with ventricular septal defect (PA/VSD). However, there are many drawbacks for the use of conduits and there is always a need for reoperation. Therefore there are advantages in accomplishing a repair without conduits whenever feasible. Methods: Between Oct 1988 – July 2000, 29 patients diagnosed with PA/VSD underwent total repair without the use of conduit. Surgical technique: the pulmonary arteries are widely mobilized, systemic to pulmonary artery shunts, a PDA or a ligamentum are divided and not just ligated. The PA is then pulled to the edge of the RV ventriculotomy and a direct anastomosis established posteriorly. Anteriorly a transannular patch is used. The VSD is closed. Age ranged from 7 month to 7 years, median 2 years. Female 17, Male 12. All had previous palliation by either one or two shunt procedures. Three patients had hypoplastic central PAs that needed augmentation. Results: There was no mortality. There was one mediastinitis and one wound infection and two neurological complications. Mean RV/LV systolic pressure ratio was 0.6. Patients were followed for a mean of 4.5 years. All are in NYHA-FC I-II though 20/29 are on cardiac medications. One patient needed reoperation for implantation of a pulmonary homograft in the pulmonary position because of progressive RV dilation with dysfunction. Conclusion: This technique simplifies to some extent the surgical repair of PA/VSD and it can be applied for many patients considered for the repair. Long-term course of these patients may be at least similar to the long-term results of Tetralogy of Fallot repair.

P384

Management of infants with PA,VSD, very diminutive central PA and MAPCAs

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To assess the validity of treatment policy of central shunt creation with concomitant ligation of duct supply MAPCAs as staging procedure for infants with pulmonary atresia plus VSD (PA VSD) and very diminutive central pulmonary artery (CPA), surgical results of 9 infants in the last 7 years are reviewed. Patient's age ranged from 1–12(mean 4) months, weight 2.4–8.1(4.8) Kg, size of CPA 1.5–3.5(2.4) mm, number of MAPCAs 2–6 (3.4), and PA index 15–81(58). At the initial catheterization, minute distribution of MAPCAs and their relation to CPA are visualized using repeated segmental aortographies to distinguish whether they are a dual supply or a single source of blood supply to the affected area (arborization anomaly). As operation, a median sternotomy was used in 7 of 9 patients, and a thoracotomy in 2. A short 4 mm Gelaski graft was anastomosed end-to-end to the main PA and end-to-side or side-to-side to the aorta using continuous 7–0 or 6–0 polypropylene. Number of duct supply MAPCAs ligated concomitantly ranged from 2–4(2.6). During a mean follow-up of 47+/-11 (range 2–89) months, additional staging procedures such as Blalock-Taussig shunt, unifocalization, Ligation of MAPCA, and PA plasty were carried out in 6 patients on 7 occasions. All patients are alive and 4 have proceeded to biventricular repair, and 4 each waiting for definitive repair and postshunt investigation. Mean PA index of 6 patients who had reinvestigation at a mean interval of 37+/-4 months, increased significantly from 53+/-11 to 284+/-64 (range 102–495) (p<0.05). In conclusion, our treatment protocol has proven satisfactory in attaining symmetric PA growth and increases the chance of reaching to corrective operation in patients with very diminutive central pulmonary arteries.

P385

No difference in right ventricular myocardial fibrosis at correction of PA, VSD with or without SPCA's

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We investigated whether after staged correction of PA,VSD with SPCA's the right ventricular (RV) myocardium showed more fibrosis than at correction of originally duct-dependent PA,VSD. In RV biopsies at the time of correction, the expression of collagen and fibronectin as well as mRNA levels of collagen Ia and III and fibroblast were studied in 8 pts with PA,VSD,SPCA's and in 6 pts with originally duct-dependent PA,VSD (age at correction: 3.2 +/- 0.4 vs 1.7 +/- 0.2 yrs, p<0.05). Video image analysis did not show a significant difference in total collagen (10.4 +/- 2.3 vs 7.6 +/- 1.3%) or interstitial collagen (3.2 +/- 2.5 vs 7.2 +/- 1.3%) and total fibrosis (7.2 +/- 1.5 vs 7.5 +/- 1.2%), or interstitial fibronectin (6.9 +/- 1.6 vs 7.5 +/- 1.8%). Perivascular collagen and fibronectin was not significantly different in both groups. There were also no differences in mRNA levels of collagen Ia, collagen III and fibronectin. We conclude that the cuts necessary for staged unifocalization in PA,VSD,SPCA's does not result in more RV fibrosis compared to correction of originally duct-dependent PA,VSD at younger age.

P386

Rehabilitation of the pulmonary arteries (PAs): a logical approach in pulmonary atresia, VSD, Extreme hypoplasia of PAs

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Among 63 patients with pulmonary atresia an IVSD, 10 patients with extreme hypoplasia of the PAs (mean Nakata index 20 mm2/m2) and MAPCAS, have been submitted to a rehabilitation" of the PAs with several steps: 1) connection between RV and PAs, 2) interventional catheterization, 3) complete correction with or without unifocalisation. We report here the results of this approach. The RV-PA connection was direct (5) or with an homograft conduit (1), done under normothermic CPB in patients aged 4.9 months (range 0.1–19 months). Subsequently, 6 underwent interventional catheterizations (dilation and stents in the PAs, MAPCAS occlusion by coils)

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Complete correction was done in 7 patients (mean age = 29 months, range 6 – 51). One patient was waiting correction. One patient died after the first step. All patients having had the third step had a satisfactory development of the PAs, had a complete closure of the VSD and a satisfactory reconstruction of the PA bifurcation. All patients have been followed by catheterization and / or echocardiograms. With a mean follow-up of 45 months, all patients are improved. 4 have no cardiac medications, none has residual shunt, RV/LV pressure ratio is 0.6 (range 0.3 – 1). The strategy of "rehabilitation" of PAs allowing: 1) antegrade flow in the PAs, 2) interventional catheterizations, 3) growth of the PAs with possible angioplasties, 4) complete correction, is a logical approach to be undertaken in the young patient. The therapeutic sequences depend upon the individual anatomy."

P387

The outcome following definitive repair of pulmonary atresia with intact ventricular septum

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OBJECTIVE To evaluate the outcome following definitive repair of Pulmonary Atresia with intact ventricular septum (PA-IVS). METHODS Between 1989 and 2000, 34 consecutive patients underwent surgical treatment of PA–IVS in our unit. Nine of these patients had only palliative procedures(4 died early, 2 are not suitable for definitive repair and 3 await a Fontan procedure). The remaining 25 patients (mean age 3.4 +/- 3.3 years,range 1 day – 15.2 years) had a definitive repair and are the subjects of this study. A biventricular repair was performed in 12 (group I)(RVOT reconstruction in 9,pulmonary valvectomy in 3 and homograft PVR in 1), an univentricular repair (Fontan) in 12 (group II) and one and a half ventricle repair in 1 patient. Eighteen patients had previous palliative operations. Follow up was complete (mean 5.6 +/- 5.5 years,range 1.3 – 21.3 years). RESULTS: There were 4 early deaths, all among patients with a biventricular repair (3 from cardiac causes and 1 from suppurative pericarditis). Six patients (3 in group I and 3 in group II) required re operations and/or catheter re-interventions. Ten year freedom from re-operation/re-intervention(+/- SEM) was 82.1 +/- 8%(SE) 9 +/- 8.6 %Vs 69.4 +/- 15.5 % for group I Vs group II)(p=0.8). There were no late deaths. Ten-year survival was 84 +/- 7.1 % (100% Vs 66.7%)(p=0.03). At the latest evaluation,21 patients were in NYHA class I and 4 patients were in class II. CONCLUSION These data suggest that biventricular repair of PA-IVS may have a higher operative risk than univentricular repair. The prospects of late survival are excellent irrespective of the type of definitive repair.

P388

Pulmonary atresia, intact ventricular septum, hypoplastic right ventricle and right ventricle dependant coronary circulation = a difficult subgroup for Fontan

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Patients with Pulmonary Atresia (PA) and Intact Ventricular Septum (IVS) with Right Ventricle Dependent Coronary Circulation (RVDCC) are a difficult subgroup for surgical correction. RVDCC is known to be a one of the major risk factors for poor outcome in patients with PA and IVS. Although the ideal end result for these patients would be a completely separated in series two-ventricle repair this is rarely achieved. Right ventricular decompression is required early in life to enable it to grow but in the presence of RVDCC this is contraindicated as decompression may lead to myocardial ischaemia or infarction and subsequent left ventricular dysfunction. Operations on these patients requiring cardiopulmonary bypass (CPB) puts them at risk of ischaemic myocardial injury due to decompression of the right ventricle on CPB. Several techniques have been described to maintain adequate coronary perfusion intraoperatively. Since 1994, eight patients of PA-IVS with RVDCC and hypoplastic right ventricle were treated in this institution. Four patients have undergone completion of Fontan circulation. Two have had bidirectional Glenn shunt and await completion of the Fontan circulation and two have had only initial palliation with left modified BT shunt and await further procedures. There have been no deaths in this group of patients. One patient has developed regional left ventricular dysfunction after the initial palliation with systemic to pulmonary artery shunt and atrial septectomy and is judged to be unsuitable for a right heart bypass operation. The paper discusses our experience in the staged surgical treatment of this subgroup of patients with particular emphasis on the technique of preservation of myocardial perfusion during surgical procedures done on cardiopulmonary bypass.

P389

Surgical implications of the pulmonary arterial anatomy in patients with right isomerism

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Objective. To determine precise anatomic feature of the pulmonary arteries(PA), and its surgical implication, in patients with right isomerism. Methods: Since 1978, 174 patients with this particular feature of atrial arrangement have undergone any surgical procedure. Patterns of PA were identified in these patients. Results: The pulmonary pathway initially had no obstruction, presenting pulmonary hypertension(PH), in 13 patients, while pulmonary stenosis(PS) was present in 95 and atresia in 66. There was no significant difference noted between these 3 groups in terms of associated malformations. Eventually, 27% of patients with pulmonary atresia, 59% of those with PS, and 58% of those with PH, underwent the Fontan procedure. Of these, number of palliative procedures previously employed was 2.1 per patient in the pulmonary atresia group, 1.9 in the PS group, and 1.6 in the PH group (p<0.0001). The included one patient (2%) with pulmonary atresia, and 15 (19%) with PS undergoing no previous procedures. Biventricular repair was attempted in 5% of with pulmonary atresia, 7% with PS, and 37% with PH. Of the with pulmonary atresia, the pulmonary trunk was present in 21%. In another 50%, the pulmonary trunk was lacking, with the central PA detoured around the posterior of the arterial duct. This posed difficulties in maintaining balanced pulmonary perfusion and surgical intervention onto PA. The incongruence was even worse in 14% with non-confluent PA. In the remaining 15%, arborization abnormality was found, and PA reconstruction as a preparation aiming toward the Fontan procedure was almost hopeless. Conclusion: The presence of pulmonary atresia in the setting of right isomerism was regarded as one of the unfavorable factors militating against successful establishment of definitive repair.

P390

Multiple stage approach in pulmonary atresia with ventricular septal defect and multiple aorto-pulmonary collaterals

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Multiple stage approach is an alternative treatment for patients with pulmonary atresia, ventricular septal defect (VSD) and multiple aorto-pulmonary collaterals (MAPCA's). From January 1997 to March 2000, 5 patients (5–15 years old) underwent 9 unifocalization (U) procedures: 4 had final repair. Angiographic identification of pulmonary blood supply and MAPCA's was mandatory in all. Through separate thoracotomies, collaterals were ligated and interlobal arteries were U and anastomosed to a 16-mm Gore-Tex tube which was tacked to the side of the ascending aorta and a modified 6mm Gore-Tex BT shunt was placed from the subclavian artery to the ipsi-pulmonary artery. Between thoracotomies, a magnetic resonance was done to confirm the permeability of the previous U and prior to final repair all patients had cardiac catheterization. Repair consisted of VSD closure and placement of a valved conduit with shunt ligation. Prior complete correction, all patients were on warfarin aiming an INR of 1.4–1.9 to prevent clot formation in the 16 mm Gore-Tex graft exposed to a low flow and an average of 8 months (4–13 months) recovery was allowed between procedures. One patient required an emergency repair due to Gore-Tex thrombosis following viral infection despite adequate anticoagulation. Her recovery was uneventful. All unifocalized patients were candidates for repair with no but pulmonary segments. There was no mortality and on last follow up (2–28 months) all are in NYHA class I. Based on our experience, the final results with multiple stage approach are worth the associated morbidity (trauma, reperfusion injury, extensive peribronchial dissection and denervation, Gore-Tex oozing, atelectasis) and should be offered to patients presenting later during childhood despite the previous non-intervention approach. Delay between procedures should be as short as possible to prevent tamponade.

P391

Biventricular repair of pulmonary atresia or stenosis with intact ventricular septum

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Since 1991 we have performed a multistage palliative approach to biventricular repair of pulmonary atresia (PA) or critical pulmonary stenosis (PS) with intact ventricular septum (IVS) in infants with a right ventricular

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

infundibulum. A total of 25 patients (19 PA and 6 PS) underwent initial palliation consisting of a transatrial pulmonary valvotomy and a polytetrafluoroethylene shunt between the left subclavian artery and pulmonary trunk. Among the 23 survivors, 15 underwent balloon valvotomy after a mean interval of 9 months. Seven patients later required additional palliative surgery ('RV-overhaul') that consisted of repeat pulmonary valvotomy, adjustment of an atrial communication, and resection of the hypertrophied muscles in the right ventricle. Actuarial survival of the 25 patients was 92% at 12 months with no further deaths over 9 years of follow-up. Among the 7 patients who required 'RV overhaul', 4 patients underwent bi-ventricular repair, one Fontan procedure, and the remaining 2 are awaiting evaluation. The other 16 patients (10 PA and 6 PS) subsequently underwent biventricular repair. In all of 21 patients coming forward for definitive operation, 20 (95%) underwent biventricular repair. In the 20 patients right ventricular end-diastolic volume significantly increased but tricuspid diameter did not change. The multistage palliation procedure to promote right ventricular growth makes a definitive biventricular repair of PA or critical PS with IVS possible in the majority of infants with a patent infundibulum.

P392

The clinical and echocardiographic evaluation of children with reoperated sever congenital heart defects (CHD)

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The aim of our study is the long-term evaluation of re-operated children with severe CHD. Methods: with retrospective study were analyzed the clinical and echocardiographic findings of our patients- children followed on long term period -20 years (81–2000). The diagnostic of CHD was made in our center and cardiac corrective surgical interventions and reinterventions were performed on foreign cardiac surgical centers (London,Sofia) The patients were followed with regular clinical ,ECG-standard and 24 h – Holter monitoring, 2d-Doppler echocardiography, x-ray examinations. Results: 15 children (11 female, 2 male) had more than one surgical cardiac intervention. The follow-up was made between 3–25 years (X-14) from the first operation. Reoperation of Tetralogy of Fallot was made in 2 children, Transposition of GA 2, Single ventricle 3, Double outlet right ventricle- 1, Tricuspid valve stenosis- 1, Common arterial trunc – 1, AV-canal – 3. 6 children had 3 operation and 9 had 2. Protetic mitral valve was implanted in 2 children and pacemaker in 2. 2 Children developed the postoperative infective endocarditis. The ECG, x-ray, Echo- Cardiograms of repeated children and longitudinal clinical evaluation showed good condition with 9 patients, had condition with 3 and lethal ending with 1 child. Conclusion: the cardiac surgical reinterventions of sever CHD may provide long term survival and good life quality in majority of children.

P393

Ductus-associated proximal pulmonary artery stenosis in patients with right heart obstructive lesions: does the magnitude of the problem justify a change in management approach?

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Objectives: Proximal pulmonary artery stenosis is a common acquired lesion in infants treated for congenital heart disease. These patients exhibit a wide range of presentation, from asymptomatic to severely hypoxemic. Decreased blood flow to the affected artery may cause late complications if not recognized early. We hypothesized that a large number of stenoses develop at the site of ductal insertion, that this occurs more frequently in patients with right ventricular outflow tract obstructions (RHO), and that these patients are at risk for developing hypoplasia of the ipsilateral pulmonary arterial bed. Methods: We reviewed the records of all infants under one year of age diagnosed with proximal pulmonary artery stenosis by catheterization from 1988–2000 at our institution. To determine the incidence of stenosis in patients with RHO we also reviewed records of all patients undergoing catheterization or surgery in the first year of life for pulmonary atresia (PA) or tetralogy of Fallot (TOF). Catheterization data and medical records were examined for site of stenosis and ductus insertion, associated diagnoses and treatments. Measurement of the pulmonary arteries was made at the first branching and a diagnosis of hypoplasia was made if the diameter of the affected vessel was ≤80% the diameter contralateral vessel. Results: 102 infants had proximal pulmonary artery stenosis. Stenosis occurred at a surgical site in 32 infants and was associated with a genetic syndrome in 9. Of the remainder, 33/61 infants had stenosis at the site of ductal insertion. Diagnoses in this group included right heart obstructive lesions in 29 (PA 18, pulmonary stenosis 6, TOF 5). Incidence of ductus-associated stenosis in patients with PA was 29% (18/62) and in TOF 2% (5/303). Fourteen of the 29 patients with RHO and ductus-associated stenoses were asymptomatic at the time of catheterization. Pulmonary arterial hypoplasia was present in 15 (7 asymptomatic) of 22 with interpretable angiograms. Echocardiography reports, available for 32 patients, identified a stenotic lesion in only 5. Intervention was undertaken in 17 (55%) of the infants with ductus-associated stenosis. Conclusion: Over half of patients with non-iatrogenic proximal pulmonary artery stenosis had a lesion associated with the site of ductal insertion, predominantly those with pulmonary atresia. Though half of the patients with ductal stenosis were clinically asymptomatic, there was a high (60%) incidence of distal pulmonary bed hypoplasia. Echocardiography was frequently unhelpful, therefore additional diagnostic modalities, such as quantitative lung perfusion scanning, magnetic resonance imaging or earlier catheterization should be used in patients with pulmonary stenosis to define proximal pulmonary artery stenosis. Early re-intervention may reduce secondary morbidity.

MAY 29 Time: 11:00–12:30

Session 3
General Pediatric Cardiology, Prognosis/Natural History

P394

Comparison of formulae for heart rate correction of qt interval in exercise electrocardiograms of healthy children

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Background: We set out to investigate the differences in 4 different formulae for heart rate correction of the QT interval in serial electrocardiograms (ECG) recorded in healthy children subjected to graded exercise. Methods: Fifty three healthy children, median age 9.9 years (range 5–14 years) were subjected to graded physical exercise (on a bicycle ergometer or treadmill) until heart rate reached > 85% predicted maximum for age. ECG were recorded at baseline, maximum exercise, and 1, 2, 4 and 6 minutes post exercise. For each phase, a 12 lead ECG was obtained and the digitally analyzed medians printed for each lead (Marquette MacVU). In each ECG, heart rate, R-R and QT intervals were measured (lead 2). Bazett, Hodges, Fridericia and Framingham formulae were used to obtain QTc interval values for each ECG. A paired t test was used for comparison of QTc at rest and peak exercise for each formula and analysis of variance for the QTc of all the stages. Results: The Bazett and Hodges formulae led to significant prolongation of QTc intervals at peak exercise (P<0.001) while the Fridericia and Framingham formulae led to significant shortening of QTc intervals at peak exercise (P<0.01). Conclusion: This study shows that the practical meaning of QT interval measurements depends on the correction formula used. In studies investigating repolarization changes (for instance in the long QT syndromes, congenital heart defects or in the evaluation of drug effects), the use of an ad-hoc selected heart rate correction formula is inappropriate as it may bias the results in either direction.

P395

Heart rate variability during sleep in healthy infants and infants with obstructive sleep apnea

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Heart rate variability (HRV) is increasingly used as a marker of cardiac autonomic activity. We examined the development of HRV during infants' sleep and HRV anomalies associated with obstructive sleep apnea (OSA). Polygraphic studies with ECG recording were performed in 587 healthy infants and 88 infants with OSA. Mean R-R interval, 5 time-domain HRV indices (SDNN, SDNNi, SDANNi, rMSSD, pNN50), and 5 frequency-domain HRV indices (spectral power in the very low (VLF), low (LF) and high (HF) frequency regions, total spectral index (TSI), and LF/HF), were determined over a 400min period of sleep, and separately for the periods of quiet sleep and those of rapid-eye movement sleep. Normal ranges were established by the Altman's method. Z-scores of were calculated for the ASO infants. All HRV indices were significantly correlated with the mean R-R interval and the age of the infants. High-frequency oscillations (HF, LF/HF) were correlated with the breathing rate

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

HRV-indices influenced by the vagal tone (rMSSD, HF) were significantly increased and those related to the sympathetic tone (SDNNi, SDANNi, VLF, LF/LF+HF) were depressed in OSA infants. Our data in a large cohort of healthy infants confirm a progressive maturation of the autonomic nervous system during sleep. They also suggest that vagal hyperactivity and sympathetic depression are associated with OSA. Our normal ranges may also be useful to examine the influence of other pathological factors on the autonomic control during polygraphic studies.

P396

Pediatric heart problems in Nepal and Japan

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Study was conducted to recognize the age, sex and pattern of pediatric heart problems in a children hospital each from Kathmandu and Tokyo. A year data (1996/97) of admitted children with heart disease were collected and analyzed. In Nepal, heart diseases were diagnosed with clinical examination, ECG, chest x-ray and echocardiography whereas hospital was well equipped with diagnostic facilities in Tokyo. Kawasaki disease without cardiac manifestations was not included. Also, an analysis of the catheterized children of the University of Tokyo hospital was performed. Among 5,848 admissions in Nepal, 122 (2.1%) had heart diseases. Of 122, 30.3% were <2 years. The male female (M:F) ratio was 1:0.74. Among them, 43.4% were diagnosed as rheumatic heart disease (RHD). Of 122, 6.6% died in the hospital. Among 1283 children in Tokyo, 9.8% had cardiac problems. Of studied 118, 32.0% were < 3 years. The M:F ratio was 1:0.87. Congenital heart disease (CHD) was observed among 95.6% patients. Ventricular septal defect, CHD associated with Down syndrome and atrial septal defects were major abnormalities. Two or more defect were observed in 51.7% patients. Of the total, 4.2% died in the hospital. Among the 90 catheterized children (M:F ratio 1:1.2) with various heart diseases, 60% were of < 3 years, and half of total patients had three or more cardiac anomalies. Of 90 patients, 62.2% were treated by cardiac repair operations. The mean and median age of repair was 2.8 year and 2.1 year, respectively. Early recognition and initiation of treatment of CHD in Japan and high mortality due to RHD in Nepal was observed a major difference. The management of CHD demands an early recognition of multiple anomalies.

P397

Cardiopulmonary exercise parameters in children with right heart volume overload: short-term effects of defect closure

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We assessed whether closure of a significant atrial septal defect (ASD) in asymptomatic children led to short-term changes in the cardiopulmonary function parameters at maximum exercise. Prospective study in ASD patients using complete cardio- pulmonary and metabolic function testing at baseline and maximum exercise (modified Conconi protocol on bicycle). An identical study was done in an age- and sex-matched normal population. 14 patients (median age 11.4 years) with a median ASD-size of 15 mm (10 – 20mm) and a median Qp/Qs of 2.2 were tested. Exercise capacity in ASD patients did not differ from 12 normals (N). Of all exercise parameters studied, ASD patients showed a lower max. oxygen consumption (median of 40.8ml/kg/min versus a median of 44.3 in N; p<0.05), a lower mean expiratory flow in small airways (median MEF 25 of 82% of predicted versus 102% in N; p<0.05), and a higher proportion of patients showing an increase in airway resistance at exercise (66% of ASD patients versus 10% of N, p<0.05). This proportion returned to normal at resting after defect closure, as did the other parameters that differed from N. Other short-term effects of closure were lower lactate serum level (p<0.05) and lower heart rate at max. exercise (p<0.05). ASD patients had a normal exercise capacity and only slight differences in cardiopulmonary exercise parameters compared to N. All parameters normalized as a short-term effect of defect closure.

P398

The left atrioventricular valve in ostium primum atrial septal defect: management strategy and surgical outcome

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Objective: To test the hypothesis that in patients with a partial atrioventricular septal defect (PAVSD), and a competent left atrioventricular valve (LAVV), sutures should be placed across the line of apposition of the superior and inferior bridging leaflets (septal commissure, SC) to prevent the development of regurgitation. Method: 152 children with PAVSD underwent surgical repair at the Royal Brompton Hospital between 1979 and 1999. The median age and weight at repair were 4 years and 15 Kg. 128 had normal chromosomes and 15 Down syndrome. The interatrial communication was closed using a pericardial patch in 69%. In 84% sutures were placed across SC partially to close the so-called, but incorrectly named, mitral valve cleft. Result: The overall hospital mortality was 2.6% (95% CI: 0.7, 6.7%), which did not differ statistically in the last 21 years. 21 patients (13.8%, 95% CI: 8.8, 20.1%) required reoperation, 19 of which were for LAVV regurgitation (LAVVR). Univariate analysis of risk factors for LAVV reoperation were young age, low weight, the severity of preexisting LAVVR, small size LAVV and the presence of a small interventricular (IV) communication. With each year increase in age at operation there is a 23% reduction in need for LAVV reoperation. The hazard ratio for the need for reoperation when preoperative LAVVR was moderate to severe was 4.7 times higher than with no to mild incompetence. 19 (15%) of 127 patients in whom the SC was sutured required LAVV reoperation but none of the 25 in whom the commissure was left alone (P=0.04). Conclusion: The hypothesis that in the absence of preoperative LAVVR it is necessary to place sutures on the SC has not been proven.

P399

Congenital coronary arteriovenous fistula in children

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Introduction: The incidence of coronary arteriovenous fistula (CAVF) is estimated to be 0.2 %. Although many children with CAVF are asymptomatic, heart failure, infective endocarditis and myocardial insufficiency may arise. Patients: Children with congenital AV fistula presenting to the National University Hospital over a 10 year period were reviewed. There were 6 patients (M = 3, F = 3) with mean age of diagnosis at 131±119 months. 4 were referred for evaluation of an asymptomatic cardiac murmur and 2 presented early with congestive heart failure. There were no arrhythmias or coronary insufficiency. Investigations: Diagnosis was confirmed on echocardiography in all. 3 fistulae arose from the right coronary artery and 2 from left coronary artery. All drained into the right ventricle except for 1 (RCA – RA). Cardiac catheterization was performed to delineate the course of the fistula and/or for treatment. The mean left-to-right shunt was 2.1 [1.1:2] and mean pulmonary artery pressure was 21 (±10) mmHg. Treatment: 3 patients underwent elective coil occlusion. This was performed using interlocking detachable coils in a delivery system with cricket catheter and Bisher wire. 3 patients underwent surgery, 1 at 3 months with good results, and the other 2 underwent surgery at 8.5 and 9 years respectively (before the technique of transcatheter closure was available). The latter had a residual leak which was subsequently coil occluded (Cook). Conclusions: All patients remained well on follow-up. With real-time imaging and color mapping, the diagnosis of CAVF can be readily made on echocardiography. Although surgical ligation of fistula is relatively safe, transcatheter closure with coils now offers an effective alternative.

P400

Value of Duke criteria for the diagnosis of infective endocarditis in children

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To explore the value of Duke criteria for the diagnosis of pediatric infective endocarditis. 50 patients with pediatric infective endocarditis who all underwent echocardiography, 12 of them surgically proven, were classified using Duke criteria. Same microorganism was detected in two or more than two separate blood cultures in 15 patients(30%), one positive blood culture in 10 patients(20%). Vegetation was detected by echocardiography in 39 patients(78%). 26 of them with oscillating vegetations, one patient complicated with valve perforation, one patient with new partial dehiscence of VSD's patch. Of 50 patients, 23 patients were classified as definite IE by Duke criteria, 12 patients met two major criteria, 9 patients had one major and more than three minor criteria. IE was rejected in one patient. In 13 surgically proven IE patients, 10 patients (38.5%) was currently classified as definite IE, 8 patients were misclassified as possible IE. 6 patients met one major and two minor criteria, 2 patients were associated with one major and one minor criteria. Negative blood culture occurred in 10 surgically proven patients, nonoscillating vegetation in 2 patients. The results of this study showed that the detection of vegetation using echocardiography had important

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

significance in the diagnosis of IE. vegetation should not be defined as oscillating intracardiac mass in pediatric patient. In patient with prior antibiotic therapy and typical echocardiographic findings, definite IE could be considered in the presence of one major and two minor criteria. Therefore, the sensitivity of the diagnosis of IE will be further improved

P401

Heart rate variability and heart rate responses to tilting in neonates.

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Heart rate variability (HRV) and heart rate (HR) responses following a 45° head- up tilt were measured in neonates to assess their autonomic function. The test was performed within the first week of adjusted age (postconceptional age = 40 weeks) in 7 healthy full-term neonates, 7 healthy preterm infants born 30–34 weeks of gestation, and 5 neonates under pharmacological therapy for heroin withdrawal. The test started when the baby was lying quietly with eyes closed. The ECG was then recorded during 5-min periods at baseline in the tilted position and after the bed was returned to the horizontal position. HR response profiles were characterized : sustained decrease (P1), or increase (P2), flat response (P3), increase followed by a decrease (P4) and decrease followed by an increase (P5) of HR. Three time-domain (SDNN, rMSSD, pNN50) and 3 frequency-domain HRV-indices (LF, HF, LF/HF) were simultaneously determined by 30-sec epochs. The tilt generally provoked a profile 1 (n=8), 3 (n=4) or 5 (n=4) response whereas return to the horizontal position was mostly associated with a profile 1 (n=5) or 3 (n=8) response. HRV-indices increased after the position was changed except in profiles 1, 4 and 5 in preterm infants. When compared to previous studies of the literature about response to tilting in older infants, our data suggest the immaturity of the neonate's ability to respond appropriately to cardio-respiratory challenges, especially in preterm infants.

P402

Cardiovascular follow up study of 70 patients with Williams syndrome

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The aims of the study were to asses the incidence, the age at onset of the symptoms and the overall outcome of different cardiovascular diseases in 70 patients with Williams syndrome (WS). At birth the median gestational age was 39 6 weeks and weight 2838 gm. 32 patients of 70 (46 %) were males and 6 (9 %) were twins. Cardiac symptoms were found in 33 of 70 (47 %) newborns with WS. In the follow up of median 16 years, 36 of 70 patients (51 %) with WS had structural heart defect. The ages at diagnosis and intervention of different heart defects are presented in the Table. Spontaneous recovery of heart defect occurred in 3 cases, 1 with supravalvular aortic stenosis (SVAS), 1 with supravalvular pulmonary stenosis (PS) and 1 with SVAS+PS. Operation or intervention were not needed in 10 patients with SVAS, 2 patients with SVAS+PS, 2 patients with PS and 2 patients with aortic valve defect (AS+AI). After operation or angioplasty, mild to moderate restenosis occurred in 2 of 7 children with SVAS, in 2 of 3 children with PS and in all of the 4 children operated on for hypoplastic aortic arch accompanied with hypoplastic pulmonary arteries (HAAPA). Cardiomyopathy (CMP) was evident in 9 of 70 cases (13 %), in 3 cases with SVAS (17 %), in one case with SVAS+PS (33 %) and in all cases with HAAPA (100 %). Two patients died (3 %); one child with HAAPA accompanied with CMP and one male adult with SVAS. Elevated blood pressure was evident in 21 of 43 patients older than 15 years (51 %). This study demonstrates, that extreme variability in severity and age of onset of cardiovascular involvement is a typical finding in patients with WS and that during the lifetime all the patients with WS, operated or not, should be followed by a cardiologist because of the risk of restenosis, developing CMP or elevated blood pressure.

P403

Sensitivity of clinical assessment of heart murmurs in term infants by pediatric house staffs

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Objective To determine the accuracy of physical examination in detecting congenital heart diseases in term infants. Design Diagnostic test. Material and method Term infants (GA more than 36 week) born at Phramongkutklao Hospital between July 1st,1999 and march 31th,2000 were examined by pediatric house staffs and pediatric cardiologists within the first week of life. To determine the definite diagnosis, echocardiogram was performed in all infants. The accuracy of physical examination was determined by comparing it with the echocardiographic finding and presented as sensitivity and specificity. Statistics chi-square test for categorical data and paired t-test for continuous data. Results Echocardiogram revealed congenital heart disease in 18 of 500 infants(3.6%). The physical examination by pediatric house staffs and pediatric cardiologists showed sensitivity of 38.9% and 94.4% and the specificity of 97.5% and 67.7% respectively. The first and second most common congenital heart disease in infants are Ventricular septal defect(44.4%) and patent ductus arteriosus(33.3%). Conclusion Prevalence of congenital heart disease is significantly high. Although echocardiography is the best technique to determine the definite diagnosis, most of congenital heart diseases can be well detected by skilled physicials. However, the value of physical examination of cardiovascular system should be emphasized in training general pediatricians.

P404

Thoracopagus conjoined twins – the Cape Town experience (1966–2000)

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Since 1966, 14 sets of thoracopagus twins have been seen at the Red Cross Children's Hospital Cape Town, South Africa. In 1 set of twins, cardiac structures were separate. In 3 further sets, the twins only shared pericardial sac. In the remaining 10 sets cardiac structures were shared. 5 sets were considered inoperable. Separation was performed in 5 sets with 4 children surviving. 2 hearts with venous sharing only were present in 3 of the 5 sets undergoing surgery. Nevertheless only 3 individuals survived. In 2 further sets, extensive sharing of ventricular structures was present. In these sets separation was attempted in order to save one individual only. One survivor died from an aspiration event 6 weeks after separation while the other is alive 3 years after the separation. Prolonged survival of either twin in a thoracopagus set with extensive ventricular wall sharing has not been previously described. Evaluation of thoracopagus twins requires synthesis of information available from all available modalities (ranging from clinical evaluation to angiography). The extent of the cardiac abnormalities may only be appreciated at the time of surgery. The decision to strive for the survival of one individual in a pair is only taken after extensive consultation.

P405

Relationship with defect area, defect area ratio and pulmonary-to-systemic flow ratio in ASD children

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Purpose) It has been known that the amount of the resultant shunting is usually not dependent on the size of atrial septal defect, but rather on the relative compliance of the right and left ventricles. Has the size of the defect any relationship with the shunting? Methods) The 52 patients with simple ASD were investigated by routine cardiac catheterization and 2-D-echocardiography (F.M= 27:26, mean age. 6.8 years old.) Oval fossa type was 49 and inferior type was 3. We calculated defect area/BSA and defect area ratio as atrial septal defect area / potential atrial septal area. Each area was calculated as ellipse and the diameter was measured in parasternal 4 chamber and subcostal parasagittal planes. Results) 1- Defect area/BSA was significantly correlated with pulmonary-to-systemic flow ratio (Qp/Qs) (Y = 1+ 0.006 X, r = 0.925, p<0.0001) 2- Defect area ratio was significantly correlated with Qp/Qs (Y= 1+ 6.44 X, r=0.928, p<0.0001) 3. Higher Qp/Qs than 2.0 could be examined that defect area was over 360mm2/m2 and defect area ratio was over 0.16. Conclusion) The size of the atrial septal defect was correlated with Qp/Qs in children. The measuring of the size of ASD is also important for the assessment of the hemodynamics. Defect area/BSA and defect area ratio might be useful for deciding the indication of surgical or catheter intervention without cardiac catheterization.

P406

Extremely high plasma bnp (>= 1000 pg/ml) indicates need for early intervention in children with cardiac disease

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https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

BACKGROUND: Brain natriuretic peptide (BNP) is known to reflect left and right ventricular pressure as well as volume overload. However, there are little data available concerning the clinical implication of high BNP in children with cardiac disease. PURPOSE: To investigate clinical outcome of patients with cardiac disease who show extremely high BNP (>= 1000 pg/ml). METHODS: From our patients' database, we identified patients who showed BNP >= 1000 pg/ml at initial evaluation. Based on the medical chart, we determined the diagnosis, initial presentation, and outcome of these patients. RESULTS: Among 226 patients who were admitted to our hospital under our service, 8 patients (3 boys and 5 girls) showed BNP >= 1000 pg/ml at initial evaluation. Four out of 8 patients were newborns and the other 2 patients were 6 months and 2 years old, respectively. Plasma BNP ranged from 1400 to 5700 pg/ml and was always higher than plasma ANP (380-1700 pg/ml). All but 1 patient presented with congestive heart failure. The diagnoses were Ebstein's anomaly with coarctation, truncus arteriosus complex, coarctation with dilated cardiomyopathy, single ventricle with unguarded pulmonary artery, aortic regurgitation caused by infectious endocarditis, and Down syndrome with patent ductus arteriosus, atrial septal defect and pulmonary hypertension. All patients, except for a patient with Down syndrome, required early intervention within a month; coarctectomy (2), balloon atrioplasty (1), Blalock operation (1), and pulmonary artery banding (1). All patients have survived with decrease in BNP and ANP with median follow up. CONCLUSION: Extremely high BNP (>= 1000 pg/ml) indicates needs for early intervention in children with cardiac disease.

P407

The influence of respiration and age on left ventricular diastolic filling pattern in normal Chinese children

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Objective: In order to evaluate the influence of respiration and age on left ventricular diastolic filling. Methods: Left ventricular diastolic function parameters of transmitral valvular diastolic filling pattern were assessed by echocardiography in 88 normal Chinese children. Doppler blood flow pattern obtained from transmitral valve presented positive two-peak curve, the first peak E occurring in early diastolic phase of ventricular diastolic filling was higher, and the second peak A resulting from atrial systole was lower. we measured the following variables: velocity indices-peak E and A velocity with which the peak E/A ratio was calculated, time indices-diastolic filling time, peak E acceleration time and R-R intervals in electrocardiogram, we calculated the following flow quantity indices: E area and A area(the area under the E and A portions of diastolic velocity-time integral), total area(total diastolic velocity time integral). Result: The results showed that respiration markedly influenced some parameters of left ventricular diastolic function. During inspiration ,velocity indices peak E and A velocity had a decrease by 2.4 and 8.0%, respectively. Whereas peak E/A ratio increased; flow quantity parameters E area/A area and total area fell by 3.5 to 10.0%; in time indices, diastolic filling time was slightly prolonged in inspiration, peak E acceleration time showed no change in inspiration and expiration. Age influence only few parameters in flow velocity and flow quantity indices, both in inspiratory and expiratory phase. But age markedly influence in diastolic filling time and R-R intervals in electrocardiogram. Conclusion: The influence of respiration and age on parameters should be considered in the evaluation of left ventricular diastolic function by Doppler echocardiography. We suggested that standardization for respiratory phases should be followed, that is left ventricular diastolic function parameters should be taken at end-inspiration and end-expiration, respectively.

P408

Cellular and humoral immunodeficiency in protein-losing enteropathy complicating congenital heart disease

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The immunologic profile of patients with protein-losing enteropathy (PLE) complicating congenital heart disease is undefined. We studied the lymphocyte subpopulation and immunoglobulin (Ig) pattern in this patient group. Six patients were studied at a median age of 15.6 (range 11 to 20) years. Protein-losing enteropathy was defined by clinical evidence of fluid retention, hypoalbuminemia (<25g/dL) and enteric loss of protein. The lymphocyte subpopulation was enumerated using flow cytometry while serum Ig levels were measured by nephelometric technique. The immunologic profile of the patients was compared to 6 controls matched for age and cardiac interventions. The cardiac diagnoses included complex cyanotic heart disease post Fontan procedure (n=3), post repair of tetralogy of Fallot (n=1), restrictive cardiomyopathy (n=1) and valvar pulmonary stenosis (n=1). In patients with PLE, the T lymphocyte (CD3+) count was significantly lower (300±196/mL vs 2070±1171/mL, P=0.017), both the helper/inducer lymphocytes (CD4+) (127±156/mL vs 927±377/mL, P=0.006) and suppressor/cytotoxic lymphocytes (CD8+) (129±49/mL vs 850±655/mL, P=0.057) were reduced with reversal of CD4+/CD8+ ratio (0.81±0.68 vs 1.64±0.89, P=0.027). Furthermore, IgG level was significantly reduced (5.12±2.84 g/L vs 12.5±1.58 g/L, P=0.005) and IgA level tended to be lower (0.36±1.37 g/L vs 2.50±0.60 g/L, P=0.095). In contrast, the B lymphocyte (CD19+) count (340±151/mL vs 685±427/mL, P=0.35), NK cell (CD 16+56+/CD3-) count (252±212/mL vs 276±251/mL, P=0.95), and IgM level (0.98±0.59 g/L vs 1.12±0.25 g/L, P=0.67) were similar between both groups. None of the patients developed opportunistic or severe viral infections. Abnormalities of both cellular and humoral arms of the immune system occur in patients with congenital heart disease complicated by PLE. Nonetheless, these abnormalities appear quantitative rather than qualitative in nature.

P409

Plasma natriuretic peptides in children with ventricular septal defect

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BACKGROUND: There is little data available concerning change of brain natriuretic peptide (BNP), compared with atrial natriuretic peptide (ANP), in children with ventricular septal defect (VSD). PURPOSE: To determine hemodynamic factors that control plasma concentrations of BNP and ANP and basal elimination of BNP and ANP in children with VSD. METHODS: Forty-nine consecutive patients with VSD (17 boys and 24 girls, age 4 months to 13 years) without ventricular outflow tract obstruction were enrolled. During cardiac catheterization blood samples were obtained from femoral vein, and plasma BNP and ANP were determined by IRMA. Hemodynamic variables were analyzed in terms of correlation with BNP and ANP. RESULTS: Plasma BNP significantly positively correlated with plasma ANP [ANP=2.1xBNP + 36 pg/ml, r=.827, p<.0001] and BNP never exceeded ANP in this patient series. Plasma BNP as well as ANP significantly positively correlated with pulmonary to systemic flow ratio (r=.659, p<.0001 and r=0.617, p<.0001, respectively), pulmonary to systemic pressure ratio (r=.746, p<.0001 and r=.771, p<.0001, respectively), total pulmonary to total systemic arterial resistance ratio (r=.620, p<.0001 and r=.677, p<.0001, respectively), total pulmonary artery resistance (TPR, r=.660, p<.0001 and r=.699, p<.0001, respectively) and pulmonary artery wedge pressure (r=.734, p<.0001 and r=.634, p<.0001, respectively). Importantly, plasma BNP >= 50 pg/ml and ANP >= 100 pg/ml identified children with TPR >= 10 wood units with a sensitivity of 71% and 88%, respectively, and a specificity of 92% and 76%, respectively. Conclusions: Plasma BNP and ANP reflect pressure and volume load to the pulmonary artery in children with VSD. Plasma BNP and ANP can be clinically useful to classify children who have high pulmonary artery resistance that requires early intervention.

P410

Congenital heart adolescent and teenager (CHAT) questionnaire - development of a disease-specific health status and quality of life instrument for congenital heart disease patients

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Background: With increasing survival and decreasing medical morbidity, issues of quality of life and health status are becoming increasingly important in survivors of congenital heart disease. While few generic measures exist, a disease-specific measure would be of greater use. Purpose: We sought to develop and validate a disease-specific measure of health status and quality of life in adolescents with congenital heart disease. Methods: From focus group interviews and review of existing measures, areas of interest and questions were developed to focus on domains including disease state, physical functioning, social health, schooling and employment, physical activity and disease concerns. The revised questionnaire was then completed by consecutive patients aged 12 - 18 years during routine outpatient visits. Results: Over a 4 month period, 76 cardiology outpatients (43 males) with congenital cardiac defects completed the CHAT. Previous cardiac surgery had been performed in 69%, with 29% on medication. Activity restrictions were applied in 17%. Learning disabilities were reported by 13%, behavioral problems 4%, held back in school 9% and educational assistance in 22% of patients. Self reported general health status was

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P411

Signal analysis of physiological and pathological murmurs in children

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Signal analysis of physiological and pathological murmurs in children El-Segaier M, Darcy L, Lukkarinen S, Nikkon P, Persson E, Sorsano L, Wetterl G, Lund Sweden A systolic murmur is common in children. Fewer than 1% have congenital heart disease (CHD). A method to differentiate physiological and pathological murmurs and evaluate the severity of the CHD would be helpful in outpatient care. Method: 88 children with murmurs: 26 without CHD, 35 with VSD and 27 with valvular aortic stenosis (AS) were investigated. Heart sounds and murmurs were recorded at standard auscultation points using Amicor, a digital recording computer program. The maximum gradient (Pmax) across the aortic valve was measured with Doppler echocardiography. To correct signal amplification bias the ratio (Q) between the murmur area and the whole systolic area in the tracing was calculated. The time-frequency analysis was done using the Choi Williams distribution. The pansystolic maximum (PM), frequency center of mass and maximum intensity of the murmur were determined. An algorithm to classify murmurs was developed. Result: The murmur spectrum of all children with AS was interpreted as pathological but three patients were interpreted as VSD. Two murmurs due to small muscular VSD were grouped as physiological. Two of the remaining murmurs due to VSD were grouped as AS. The incorrect grouping of VSDs as AS and vice versa was dependent on difficulty in determining the PM of the murmur. Correlation coefficient between Pmax and Q was significant: (r = 0.75, p <0.0001). Signal analysis separated physiological and pathological murmurs with 97% specificity and 80% sensitivity. Summary: The small muscular VSDs with limited clinical significance were not adequately grouped with our method. Signal analysis could differentiate physiological and pathological murmurs and determine the severity of AS.

P412

Plasma brain natriuretic peptide concentration level can be a convenient indicator of the severity of heart failure in pediatric VSD patients

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Background: According to recent reports, plasma brain natriuretic peptide concentration (p-BNP) level can be a predictor of the severity of heart failure in adult patients. The purpose of this study is to test whether p-BNP could be a reasonable indicator for operation in pediatric VSD patients. Methods: p-BNPs were measured in a total of 294 patients (252 non-heart disease patients (NHD) and 42 VSD patients (VSD)) ranging 1 m to 15 yrs. In all the VSD patients, we also measured Qp/Qs by cardiac catheterization. Results: In the NHD patients, p-BNP was 6.2 ± 5.4 pg/ml (mean ± SD) (ranging: below 4.0 – 22.3 pg/ml) and had no significant relation with age. p-BNP in the VSD patients was significantly higher (35.4 ± 24.8 pg/ml: p<0.01) than the NHD patients by Student t test. p-BNPs in the VSD patients had good correlation with Qp/Qs (p-BNP = 27.9 x Qp/Qs – 23.5, r=0.766, p<0.01). Conclusion: In the VSD patients, p-BNPs were widely varied and had good relation with Qp/Qs while p-BNPs in the NHD patients were below 15 pg/ml. In general, VSD patients with over 1.5 of Qp/Qs are required surgical operation. According to the present results, 20 pg/ml of p-BNP corresponds to 1.5 of Qp/Qs. Thus p-BNP would be a convenient predictor of the severity of heart failure and one of the indicators for cardiac catheterization and/or operative indication in the pediatric VSD patients.

P413

Infective endocarditis in children-incidence, pattern, diagnosis and management in a developing country

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Background: The pattern and outcome of children with infective endocarditis (IE) has changed in developed world. In our setup, patients (pts) are referred late, there is low yield of blood cultures and incidence of rheumatic heart disease (RHD) is yet high. Objective: Evaluate the clinical pattern, modify diagnostic criteria in our settings and determine outcome. Patients and Methods: All children with IE admitted to a single tertiary referral center (April 1996-March 2000) were analysed. The diagnosis was based on Duke's criteria. Minor criteria were expanded to include raised acute phase reactants (ESR&CRP) and presence of newly diagnosed or increasing splenomegaly. The pts were stratified as definite, possible and rejected cases. Clinically definite IE requires 2 major, 1 major and 3 minor or 5 minor criteria. Results: Of 2108 paediatric admissions, 52 were diagnosed as having IE. RHD was the underlying lesion in 34 pts (65%) while CHD in 27 pts (44%). One patient with myocarditis developed IE. Previous antibiotic treatment was given in 35 pts (56%) definitely. Blood cultures were positive in 29 pts (47%) while vegetations on echocardiography were present in 46 pts (74%). Surgery was undertaken in 5 pts and 6 left against medical advise. Of 62 pts with aortic valve involvement, 4 died (13%) and overall mortality was 13% (8 pts). Conclusions: The incidence of IE is 34/1000 hospital admissions in a tertiary paediatric cardiology referral center. Pts usually receive antibiotic treatment before reaching hospital, which is usually late. RHD is still the commonest underlying heart lesion (55% of all pts). Blood cultures are positive in less than 50% of cases and echocardiography is a more sensitive tool. Mortality is still high and aortic valve involvement in particular, carried poor prognosis.

P414

Adenosine in treatment of paroxysmal supraventricular tachycardia in children

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Adenosine in treatment of paroxysmal supraventricular tachycardia in children. Paroxysmal supraventricular tachycardia (PSVT) is the most common cardiac arrhythmia in childhood and adolescence. Conversion to sinus rhythm can be achieved by vagal manoeuvres, antiarrhythmic drugs, transesophageal overdrive stimulation or DC cardioversion. In the present study we analysed treatment of 44 paroxysms in 36 children during the period from 1995 to 1999. The objectives of the study were: (1) to determine the frequency of successful cardioversion using different methods of treatment, in particular adenosine, (2) adverse effects of adenosine treatment, (3) the dose of adenosine necessary for cardioversion, (4) the correlation between the method of cardioversion and the duration of paroxysm and (5) the frequency of starting with antiarrhythmic prophylactics after the episode of PSVT and it's success. The paroxysm of SVT converted spontaneously or using only vagal manoeuvres in 31,8% (14/44). The adenosine was successful in 85% (22/26). The majority of episodes were terminated by dose less than 0.2 mg/kg. There were no significant pro-arrhythmogenic effects using adenosine in doses exceeding the recommended doses. However, we observed brief apnea in neonate after adenosine dose of 1.6 mg/kg. There were no correlation between the method of cardioversion and the duration of paroxysm. b blocker, propafenone and digoxin were drugs most commonly used to achieve long term prophylaxis. Their success rate were 72,5% (29/40).

P415

28-years trend of infective endocarditis associated with congenital heart diseases, a single institute experience

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Several recommendations for prophylaxis of infective endocarditis (IE) are published, but their clinical impact has not been reviewed. And, microbiological profile causing IE has changed in Western countries, which is not studied in Japan. In order to elucidate these issues, we reviewed 183 cases of IE with congenital heart disease (CHD) between 1971 and 1998, which was evenly divided into 4 periods. The number of patients has not decreased, but patients < 15 years-old were 56% of all IE patients in the 1971–1977 period, and were only 20% in the latest 7-years period. There were 59 postoperative cases, which consisted of 18 cases with Blalock-Taussig anastomosis, 18 cases of extracardiac conduit and 4 cases with valve replacement or plasty. Microbiological profile, in contrast to data in Western countries, did not change during the last 28 years. The most frequently isolated bacterium was Streptococcus SP, which accounted for 59%. Preceding procedure or infection was confirmed in 48 patients (33%), which were associated with dental procedure in 38, and periodontal or periapical infection in 10 patients. In 3 other patients, severe atopic dermatitis was very likely the preceding cause. The decrease of children indicates that education of patients is effective to prevent IE, and the increase of adult patients suggests the need of continuous education to patients themselves. No increase in staphylococcus could be due to much less prevalence of people who does self-injection of drugs in our

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P416
Predictors of progression of mild pulmonary stenosis in children
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Background. Published data have suggested that children with isolated mild valvar pulmonary stenosis may experience progression even after several years of age. However, because standard life table analyses may be confounded by late identification of events, we investigated the risk of progression following a given observation, with the aim of identifying children at risk prospectively. Methods: Records were analyzed for 69 children initially noted to have mild valvar pulmonary stenosis (< = 30 mmHg by Doppler echocardiogram) between 1 day and 8.9 months of age (average 1.5 +/- 1.5 months). Follow up was for 20.1 +/- 10.4 months (172.9 pt-yrs). The outcome of interest was progression from mild obstruction to moderate or worse obstruction (echocardiographic or clinical assessment). Results: Progression to moderate or worse obstruction was observed in 9 children. Life table analysis demonstrated that 13.2 +/- 4.1% of children experienced progression by 4.6 months of age. No children rated mild after 4.6 months of age progressed subsequently. Multivariate analysis suggested that age at initial diagnosis may be a predictor of progression, but failed to reach statistical significance (HR=0.49 [0.23,1.04] per month of age, p=0.064). Conclusion: Of infants with isolated mild pulmonary stenosis, progression after 4.6 months of age is extremely unlikely. This type of information is of great use when counseling families and planning timing for follow up.

P417
Long term results after repair of isolated aortic coarctation in infants under 1 year of age
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Between January 1980 and December 1991 47 infants under 1 year of age underwent repair of the isolated Aortic Coarctation (CoA). Hospital mortality was 6,4%. There were no late deaths. The aim of the study was to evaluate long-term results after repair of the CoA early in infancy. 40 survivors (follow-up 91%) were reinvestigated between January 1998 and June 1999. 4 children (9,1%) developed recoarctation (reCoA). 85% children presented in NYHA class I. The function and development of the left upper limb were normal. The blood pressure at rest was between 15 and 95 percentile: mean 69 SD ± 23, increasing during treadmill from 2 to 68 torr, mean 34 SD ± 16. The pressure gradient between right upper and lower limb was -35 to 15 torr, mean -12 SD±12, during treadmill test -60 to 30 torr, mean -8 SD±20. Mitral valve diameter – mean Z value 1,12 (-1,7 – 13,3, SD±1,2), p < 0,00001. Aortic valve diameter – mean Z value 0,3 (-6,1 – +1,4, SD±2,3), p < 0,00005. LVDd – mean Z value 0,15 (-1,89 – +1,88, SD±0,8), p < 0,00001. LV mass – mean Z value 0,85 (-1,7 – +2,2, SD±1,58) p < 0,00012. LVPW thickness – mean Z value 0,9 (-4,61 – +1,64, SD±1,6) p < 0,00002. IVS thickness – mean Z value 0,45 (-5,03 – +3,56, SD±1,05), p < 0,0012. Development of the aortic arch segments was normal. Conclusions: Repair of CoA early in infancy provides satisfactory general development 15,6 years after surgery. The morphology and function of LV as well as development of the aortic arch are normal. Age at surgery below 3 months is the risk factor for reCoA.

P418
Potential eligibility, safety and cost savings of changing inpatient pediatric cardiac catheterization (pcc) to an outpatient procedure using a hypothetical us model of care
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Objective: To determine the number of candidates eligible for outpatient PCC based on patient selection, complications and cost using a hypothetical US model of care. The US model uses a 5-hour post catheter discharge whereas PCC are performed as a 2-day stay at the Swiss children's hospital. Analysis of economic efficiency and potential differences in reimbursement based on the existing Swiss insurance structure was performed. Methods: Hospital charts of all children (mean age 6.1 _ – 5.2 years) catheterized between January 1998 and December 1999 who met inclusion criteria for the hypothetical model were reviewed. Data collected included demographics, indications for catheterization, complications and time of occurrence. Cost data was compiled from hospital records and interviews with personnel. Results: Of 350 total cases, 53% (132 diagnostic, 77 interventional) children met inclusion criteria. Two complications (device dislocations) were discovered after the 5-hour discharge time in 2 children (1.1%). Additionally, 54 (30%) children received nursing interventions after the hypothetical discharge time. Cost differences between inpatient and outpatient catheterizations averaged $698. Current Swiss reimbursement methods for children with congenital heart disease allow itemized payments for outpatient procedures but flat per diem rates for inpatient ($502/day). Itemizing increases potential reimbursement by $3657 for a diagnostic catheter to $9867 for a radiofrequency catheter ablation. Conclusions: 53% of PCC at this Swiss children's hospital are potentially eligible for outpatient catheterization. Complications necessitating immediate intervention are rare and do not limit the safety of outpatient catheterization. The substantial rates of late nursing interventions must be reduced, requiring further investigation into possible changes to promote outpatient PCC. While average cost differences between inpatient and hypothetical outpatient PCC were modest, when the current reimbursement system in Switzerland is considered, hospital costs rise tremendously.

P419
Cardiovascular manifestations of hyperthyroxinemia in children with congenital hypothyroidism
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There are numerous reports in adults of the undesirable effects of thyroid hormone treatment on the cardiovascular system. These studies are based on patients with prolonged treatment with levothyroxine at dosages that inhibit thyroid stimulating hormone (TSH) and produce hyperthyroxinemia. At the Instituto Nacional de Pediatria, Mexico City, Mexico we studied a group of children with hypothyroidism and prolonged medical treatment. Objectives: 1) Identify the cardiac effects of prolonged treatment with levothyroxine in children with congenital hypothyroidism, 2) Determine if after prolonged treatment, the levels of levothyroxine continued to be elevated in the study group. Material & Methods: Forty children with congenital hypothyroidism detected with neonatal screening test, who had received treatment with levothyroxine during two years were included in the study group. We evaluated heart rate, QRS electrical axis, heart rhythm, corrected QT segment interval and R and S wave amplitude. A Doppler echocardiogram was performed on each patient to explore left ventricular wall thickness, ejection fraction, fractional shortening and cardiac reserve. Radioimmunoassay was performed to determine T4, T3, T4f, and T3f. Results: The study group was predominantly made up of thyroidism, followed by ectopic glands. Females were 77.5% and males 22.5%. All patients initiated treatment before 2 months of age and at the time of the study had completed from 25 to 120 months of treatment (median 48 months). T4T levels in patients with thyroidism and ectopic glands were 12.5 ± 2.9 ng/dl and 13.8 ± 2.1 ng/dl, T4f. levels were 2.5 ± 1.21 ng/dl and 2.0 ± 0.72 ng/dl, TSH 176 ± 34.7 ng/dl y 5.0 ± 1.0 ng/dl, TSH de 0.2 ± 0.9 _U/ml and 0.2 ± 1.2_U/ml. Left ventricular wall mass was found to be in accordance with age, gender and weight the latter in spite of biochemical hyperthyroxinemia. Conclusions: Children with congenital hypothyroidism and induced hyperthyroxinemia as a result of thyroid replacement therapy necessary for an adequate neurological development did not result in impairment undesirable secondary cardiac effects.

P420
Vascular rings in infants – review of imaging and treatment
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Fourteen children with diagnosis of vascular ring (VR) were hospitalized in the Department of Pediatric Cardiology Medical University of Gdansk in the period of 1990–1999. The age of patients at the moment of hospitalization was from 2 to 13 months. The main reasons for hospitalization were dyspnea in most of children and recurrent infections of the upper or lower respiratory tract. In two cases disturbances in swallowing were observed. There were no intracardiac defects in any of these cases. The diagnosis was carried out on the chest X-ray examination. In 11 cases it was confirmed by DSA. All children were qualified to be operated on. One child died on the third post-operation day. In spite of the operation on one case stridor and frequently upper tracts infections still persist probably this is due to tracheobronchomalacia. Twelve other children are feeling well and there are no signs of stridor or dispnea. One child died in the 12 month of life before the operation because of diffuse pneumonia. The diagnosis was performed on the base of DSA and intraoperation results and it was as follow: double aortic arch in 9 cases, right aortic arch with persistence innominate artery in two cases, left aortic arch

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

with anomaly of subclavian artery in two cases and the compression on oesophagus by PDA ligamentum also in one case

P421
Induction of protein-losing enteropathy after fontan-type of cardiac surgery by acute infection
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Aim. Children following Fontan type surgery might develop a post-operative protein losing enteropathy (PLE) with a substantial 10 years morbidity of >10% and a mortality of 80% among these patients. Although altered hemodynamics has been accounted for as a risk factor the etiology of PLE induction is still unclear. Single case examinations in non cardiac patients showed an influence of the immune system on PLE and its possible induction by infection. Methods: In a follow up study we examined 22 children who underwent Fontan surgery over a period of three years. The serum level of inflammatory mediators, adhesion molecules and complement factors was determined and a large panel of cellular immunodiagnostics was performed. One of the children developed PLE ten month after Fontan surgery. The child was hospitalised in a peripheral hospital with the suspicion for glomerulonephritis following Streptococcus infection (reduced serum protein level to >50%). One week later the patient suffered from Rotavirus infection and returned in our cardiac center (low protein level: 3g/l). Results: The laboratory analysis revealed a high CRP, leukocytosis, elevated IL-6 and -8 levels and a high titre of anti EBV-IgG. The patient had a striking lymphopenia with in particularly a dramatic loss of peripheral T and B lymphocytes. Following antibiotic therapy and albumin substitution the inflammation markers normalised to basal levels. However, the patient still had reduced IgG levels (>> 2500%) and lymphopenia with a massive reduction in particularly of T lymphocytes. Conclusion: Our data show for the first time an association of PLE with acute viral or bacterial infection. These results suggest the involvement of acute infection in PLE development. The selective loss of T-lymphocytes is yet unclear but might be due to auto-reactivity.

P422
An appraisal of cardiovascular changes in children with mucopolysaccharide disorders
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AIMS. To evaluate cardiac involvement, assess risk factors and mortality and define the spectrum of cardiac abnormalities with age in the different types of mucopolysaccharidoses (MPS). METHODS. Echocardiograms (M mode, two dimensional and colour Doppler flow mapping) were performed in 99 patients with MPS, age range from 1 to 49 years (median 10.3 years) between 1978 and 1999. RESULTS: Mitral regurgitation (MR) was detected in 29 patients (29%). MR was more frequent in type IH (38%), II (24%) and III (20%). 16 (16%) of patients developed aortic regurgitation (AR), seen mostly in type II (56%) and IV (24%). AR and/or MR was detected in 37 patients (odds ratio 2.05, 95% CI 1.0, p=0.05). Follow up echocardiograms were performed in 45% of patients of which 25 (56%) were abnormal and 20 normal. 13 (65%) developed a cardiac abnormality on subsequent echocardiogram which was statistically significant (p=0.002). Univariate binary logistic regression analysis performed for age of the patients at echocardiography as demographic predictor determined that mitral and aortic valve abnormalities showed a positive association with age. The correlation of age with AR was statistically significant when compared with other aortic valve abnormalities. 16 patients died during the time follow up in 1999: 25 year actuarial survival was 69.85%. At the second follow up at five years, 33[33%] patients had died, the actuarial survival dropping to 52.22%. Univariate analysis of risk factors showed that age at echocardiogram, MPS I and ejection fraction were significant risk factors for death. CONCLUSIONS. As cardiac lesions in MPS show progression with age with increasing mortality, it is recommended that all patients with MPS are followed up with serial echocardiograms to assess structural anomalies and ventricular function.

P423
Community acquired endocarditis in a pediatric population in the 1990's
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Background: Community acquired endocarditis (CAE) is a rare infection in children with congenital heart disease (CHD). To characterize the population at risk, the incidence of CAE at the Children's Hospital of Wisconsin during the 1990's was reviewed. Methods: The hospital database was reviewed for the diagnosis of CAE occurring as an outpatient without recent invasive procedures/indwelling catheters from November 1989 to May 2000. Results: 17 patients had 18 episodes of CAE. They ranged in age from 1 mo to 17 yr (mean 10.5+/-5.4 yr). 8 of the 17 patients had isolated mitral valve disease (47%) and 7 of these 8 patients (88%) had their initial diagnosis of CHD made at the time of presentation with CAE. 2 patients had a hinged aortic valve, 2 had a perimembranous VSD, 1 had aortic root valve disease, and the remainder had complex congenital heart disease s/p prior to the surgery. The most common organism was staph aureus, found in 9 of the 18 episodes. 7 episodes had an oral flora strep species. 1 had enterococcus faecalis and 1 had a group B strep (in a neonate). 10 of the 17 patients (59%) required urgent cardiac surgery within 6 weeks of presentation, two additional patients required late surgical valve repair because of chronic insufficiency. 9 of the 17 patients (53%) had significant complications, 5 had cerebral vascular accidents (CVA), 2 developed splenic abscesses, and 1 had renal failure. One of the patients with a pericardial abscess died of overwhelming sepsis; the remaining 17 episodes were successfully treated with antibiotics +/- surgery. Of the 8 patients with isolated mitral valve disease, 4 had significant embolic complications (3 CVA, 1 splenic abscess). Conclusions: CAE is a devastating illness in children with CHD. It frequently results in the need for urgent surgery, and significant morbidity is seen in the majority of children due to complications of the infection. Antibiotic resistance is not a complicating factor in therapy to date. Silent mitral valve disease appears to be by far the most common congenital abnormality in CAE in children, and mitral valve endocarditis is usually associated with significant complications and/or need for surgical intervention.

P424
Bronchial hyperreactivity after surgical correction of CHD. Influence of conventional mechanical ventilation (CMV)?
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Bronchial hyperreactivity (BHR) may potentially influence long-term evolution of patients after correction of CHD. To assess possible impact of CMV on lung or arterial pulmonary function (PFT) in patients with two cyanotic CHDs with contrasting preoperative pulmonary hemodynamics and in normal children after long-term CMV. Forty patients with TGA, 31 with TOF, long-term after repair (10.1±5.3 and 14.2±3.6 years, respectively) and 24 normal children (9.3±5.6 years) after severe craniocerebral trauma (CCT) were tested. Duration of CMV was 5.0±5.2, 1.7+-1.5 and 7.7±7.6 days, respectively. Lung volumes, elasticity, airway patency and acetylcholine (Ach) challenge tests were performed. Cumulative doses of 0.75 to 5.0mg Ach were used. Abnormal PFT was found in 75% (TGA), 58% (TOF) and 63% (CCT) patients, respectively. The most frequent findings in TGA were stiff lung (in 60%), in TOF and CCT hyperinflation (32% and 29% respectively). BHR was found in 70% of TGA, in 68% of TOF and in all CCT patients. Mean PD20 was 1.4±0.6, 1.1±0.4 and 1.1±0.5 mg Ach, respectively (NS). BHR was found in majority of both common CHDs and in all tested CCT patients. Possible consequences of conventional, especially long term CMV on the developing lungs could not be excluded. Development of symptoms of bronchial asthma in some of CHD patients should be considered. Supported by the Int. Grant Agency of the Czech Ministry of Health.

P425
Regression of pulmonary fistulae (PF): an "experimental" human model.
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Two patients with congenital heart disease developed PF after unintentional surgical exclusion of the hepatic venous flow from the lungs. Case 1: cor triatriatum and ASD operated at 2 yrs. Evidence of cyanosis at 10 yrs: O2 saturation: 70%, Hb: 21 g/dl. Pulmonary scintigraphy: intrapulmonary shunt. Cardiac catheterization: IVC to the left atrium, bilateral PF. At 10 yrs rerouting of IVC to the right atrium. Five months later normal O2 saturation and pulmonary scintigraphy. Case 2: biliary atresia, ASD, DORV. At 6 months correction of DORV, at 9 months liver transplant. Since 10 months cyanosis. O2 saturation 68%. Hb 17 g/dl. Pulmonary scintigraphy: intrapulmonary shunt. Cardiac catheterization: hepatic veins to the left atrium, bilateral PF. At 19 months rerouting of the hepatic veins to the right atrium. Two months later normal O2 saturation and pulmonary scintigraphy. Our data confirm that hepatic venous flow plays an essential role in preventing the pulmonary vascular bed from development of PF.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P426

The application of prostaglandins in neonatal cardiology

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The prostaglandins have made a revolution in saving children's lives in neonatal cardiology. The aim of this study was to evaluate Prostalgandin, first time administrated to neonatal cardiac pts in Bell. During December 1997 till June 2000 in Neonatal department of Pediatric Clinic in Sarajevo 1662 pts were admitted of which 17 (1.4%), age 4–5 days (45 min–20 days) with congenital heart disease, have received prostaglandin therapy. Central cyanosis was evident at admission in 15/17 pts, whose oxygen saturation was from 72% up to 76%. Diagnosis of cyanotic heart disease was made in 15/17 pts. PGE1 was administrated in 11/17 (the maintenance dosis of 0.025mcg/kg/min) and PGE2 in 6/11 (maintenance dose of 0.35 mcg/kg/hr). The duration of prostaglandin Th was from 2hrs up to 30 days, mean 12 days. The side effects of this therapy were present in 10 pts, 8/11 pts were operated and corrected abroad. 9/17 pts died. Conclusion: In pts with congenital heart disease whose survival is duct dependent, the availability and application of prostaglandins is compulsory.

P427

Should an intrathoracic ventricular diverticulum be treated?

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Intrathoracic diverticula of either ventricle are rare. Upon 3 own cases this review of the literature focuses on the necessity of surgical intervention. From literature 122 cases of true congenital intrathoracic diverticula are analysed. Clinics, ECG, X-ray findings and follow up with and without therapy. Reports of 86 left ventricular diverticula (LVD) and 36 right ventricular diverticula (RVD) were found. They are detected at each age ranging from prenatal diagnosis to geriatric patients. Most are clinically asymptomatic. There are no typical ECG-changes. Cardiomegaly is the most frequent X-ray finding. Operation was performed in 23 LVD and 8 RVD. Follow up of these did not show any difference from the untreated diverticula. Survival was idem as the rate of severe complications as rupture, rhythm disturbances or chest pain. Conclusion: Most congenital intrathoracic diverticula should be followed closely. Surgical intervention is necessary in case of complications.

P428

Unilateral absence of pulmonary artery. Report of four cases

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Most of the patients with absent pulmonary artery (APA) survive with few or no symptoms. We report four infants who presented early in life with congestive heart failure, cyanosis and severe pulmonary hypertension (SPH). The electrocardiogram had right ventricular hypertrophy and systolic overload. The x-ray as well as the nuclear pulmonary study showed asymmetric vascular margins with severe diminished perfusion of the affected lung. The echocardiograms showed SPH, without recognition of the abnormality. The cardiac cath showed APP right an two and left in the other one contralateral of the aortic arch. All of them had aortopulmonary vassels to the affected lung. One had precoz diuresis and underwent surgical ligation of the ductus and ligation of a large collateral artery with 'good' early postop, suddely developed right ventricular failure (RVF) and died without response to medical therapy. In another one the diagnosis was suspected clinically, confirmed on the cath she remained with SPH in RV until last seen, without surgery. In one the diagnosis was made on autopsy; a new born who was admitted with leptomeningitis and died without cardiac symtoms.

P429

Multiple aortic aneurisms in a new born. Case report

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New born (NB) referred because recurrent total collapsed of the left lung (LL) and respiratory failure (RF). On admission physical findings showed a well developed male, with mild RF. Chest x-ray with normal sized heart, opaque left lung. A bronchoscopy showed total extrinsic obstruction of the left bronchus without palpation. An echocardiogram showed a clear large image near the ductus that was believed it was a ductal aneurysm. Aortic angiograma showed three huge saccular dilatations, the first one at the level of the thorax, the second, above and below the diaphragm, and another one two centimeters above both renal arteries of the iliac arteries that later were recognized and followed up with the echo. He underwent resection of the aortic aneurysm that produced left bronchial obstruction with terminal terminal anastomosis. Medical treatment included plaquenil, antiagregants, antihypertensive drugs, cardiac and ventilatory support. Histology of the resected tissue showed abnormally diminished elastic cells. Because the extension and abnormality of the amount of elastic tissue any further surgery was denied. He survived for almost five months, discharged home remained several weeks without symptoms until he suddenly collapse and died. No autopsy was made.

P430

Interruption of the aortic arch associated to pulmonary valve stenosis. Case report

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Interruption of the aortic arch (IA) can be diagnosed alone or in association with several major congenital disease. It usually present in the new born period with congestive heart failure (CHF). We found only one report in which there was associates pulmonary valve stenosis (PVS). Male 19 day old with heart murmur, cyanosis, diminished brachial and absent femoral pulses. Chest x-ray with normal cardiothoracic index and diminished lung vascularity. EKG showed AQRS +150°, right ventricular hypertrophy with systolic overload. The echo showed IAA type B, large ventricular septal defect, severe PVS, small patent ductus arteriosus. The cath confirmed the diagnosis and showed large collateral vessels from the innominated artery which bypass the obstruction and connected to the descending aorta. He underwent a successful correction of IAA with a flap of left subclavian artery. At 3 months he cyanosis raised and treated with cyanotic spells, successful permanent dilatation of the pulmonary valve diminished the systolic gradient from 75 to 45 mm Hg. He was discharged and is doing well afterwards. Comment: It is very unusual to have a child in whom having IAA the main symptoms are related to the PVS as this case.

P431

Definitive repair of tetralogy of fallot (tof) during the first six months of life – comparison with older repair

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Early primary repair of TOF has been advocated to minimise effects of chronic hypoxemia and progressive right ventricular fibrosis. This study assessed the results of early versus late definitive repair. All infants aged six months or less undergoing primary repair of TOF from November 1995 to August 1999 at the Prince Charles Hospital were recalled at the time of surgery. This group was compared with a time-matched group of all patients undergoing definitive repair at greater than six months. 28 infants aged 1–6 months (median 4months) underwent primary definitive repair. This group is compared with 25 children age 7months to 13yrs (median 9.9months) who had definitive repair of TOF of which 4 had required an aorto-pulmonary shunt. Pre-operative haemoglobin and oxygen saturations were not significantly different between the groups. The median bypass times of 214 (standard error (SE) 5.8) vs 118mins (SE 7.0) and cross-clamp times of 56 (SE 5.3) vs 57mins (SE 7.7) in the infants and children respectively were not significantly different. The transventricular patch rate was 50% in the infants vs 60% in the children. There was no mortality in either group. Infants had a higher mean ventilation time of 64±34hrs vs 43±35hrs ($p=0.08$) and incidence of junctional ectopic tachycardia at 21% vs 4%. Otherwise early morbidity was comparable. Echocardiography demonstrated a residual small VSD in 21% vs 20% and mild-moderate right ventricular outflow tract obstruction in 25% vs 4% in the infants and children respectively. Moderate-severe pulmonary incompetence was seen in 64% of both groups. At follow-up (mean of 20±14 and 34±17months) in the infants and children, one of the children has decreased exercise tolerance. One patient in each group has required late surgery. This study supports early TOF repair.

P432

Echocardiographic and clinic long term follow-up evaluation of neonatal critic aortic valvar stenosis patients subjected to valvuloplasty

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https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Aim of the study: Analize the long term outcome of patients (P) with cavic aortic valvar stenosis (cavs) treated in the neonatal period by valvuloplasty. Methods: Thirteen cavs patients surviving neonatal valvuloplasty were evaluated clinically and by echocardiography. Doppler echocardiography (D-echo) evaluation was performed routinely in all cases. Residual aortic pressure gradient (APG) and aortic incompetence (AI) were evaluated in all cases. The follow-up time was 2.3 years ± 1.8 (ranging 0.5 to 6 years). Results: Nine P remained asymptomatic and four developed progressive signs of heart failure. At D-echo AI was mild in nine, moderate in two and severe in two. Patients with severe AI were subjected to surgical aortic valvuloplasty 1.2 and 5.8 years after initial procedure with good results. In another patient surgical mitral valvuloplasty was performed because of progressive mitral stenosis 3 years after aortic valvuloplasty. The mean APG at the last evaluation was 32 mmHg±13.2. There was no mortality in the series. Conclusion: At 2.3 ± 1.8 years follow-up 70% of the patients with CAVS remain asymptomatic, with mild AI and without evidence of significant recurrence of stenosis after successful valvuloplasty.

P433

Anomalous origen of the right artery from the ascending aorta. Report of three cases

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We report three cases: first a 3 months old female with severe pulmonary hypertension (SPH), on congestive heart failure (CHF). The cardiac cath showed anomalous origen of the right pulmonary artery (AORPA), she underwent surgery died infected with SPH on the early postop. Second case, was a 6 months male cyanosis he had complex transposition of the great arteries, large ventricular septal defect and AORPA. He underwent Mustard's repair and reimplantation of the right pulmonary artery (RPA). He is alive and doing well. Last one case: dysmorphic male 5 months old, with only mild feeding fatigue the echo showed that he had a large aortopulmonary window with AORPA and SPH, it was confirmed with cath, he underwent total repaired within the surgery another abnormality was found the right coronary artery arouse from the pulmonary artery. He had an uneventful post. Comments: This cases represent the broad congenital heart abnormalities that can be associated to AORPA. All of them were diagnosed very late, in spite two had symptoms since the beginning of their life. The surgery resolved two of them nicely, the first one died postop no autopsy was done.

P434

The echo finding of functional pulmonary atresia with severe tricuspid regurgitation in the neonate – differentiation from pulmonary atresia

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Background: Functional pulmonary atresia(FPA) must be distinguished from anatomic pulmonary atresia with intact ventricular septum(PA) to avoid any unnecessary treatments. The purpose of our study is to differentiate FPA from PA by the echo cardiography finding. Patients and methods: Four neonates with FPA, one neonate with Ebstein anomaly and three with tricuspid valve dysplasia were compared with four neonates with PA. All patients were treated with Prostaglandin E1 infusion. The following echo cardiography indices: LVDd (mm), LVDd%of normal (%), RVDd/LVDd, Tricuspid valve dimension (TVD)(mm), TVD%of normal (%), Tricuspid regurgitation(TR) grade, TR velocity (m/s), PG: TR velocity (mmHg), Pulmonary valve dimension (PVD)(mm), PVD%of normal (%), PDA (mm), PDA velocity (m/s), PG: PDA velocity (mmHg) were observed both in FPA neonates and in PA neonates. We defined PA pressure as (RP-PG: PDAvel). RV pressure as (PG: TR velocity+5mmHg. Rapressure: mmHg, and PAp/RVp ratio was also calculated. Results: Compared with PA groups, we found that in FPA neonates, RVDd/LVDd>0.6 (p<0.05), TVD%of normal>100%(p<0.05), PAp/RVp>0.55(p<0.01), TR velocity<4m/s (p<0.01). All these indices were significantly different from those in PA neonates. Conclusion: We can differentiate FPA from PA in echo cardiographic finding. According to the following echo indices: RVDd/LVDd >0.6, TVD%of normal>100%, PAp/RVp>0.55, TR velocity <4m/s, therefore we can reduced or quit the infusion of PGE1.

P435

Continuous blood pressure monitoring and pulse oxymetric measurements during exercise in children with and without open heart surgery

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In order to derive reproducible data for physical fitness of children treadmill, bicycle or other ergometric tests are necessary. The smaller the child the worse the result because of unreliability of pulse oxymetry and blood pressure measurements. Especially after open heart surgery a non invasive method of monitoring continuously blood pressure during exercise is desirable. In our study we examined 65 children with a newly developed continuous blood pressure determination system with a model based evaluation of pulse wave velocity and a photoplethysmogram using an artificial neural network. The method is described elsewhere. Additionally we measured oxygen saturation and its maximal decrease during exercise. There groups were subdivided: A" represented healthy children examined because of chest pain or fitness test. B" was derived from children with significant congenital heart disease (ASD, small VSD, PS, PR, TR, AS, AR) and C" was a group of children after open heart surgery for significant congenital heart disease. Excluded were palliative operations. Additionally diastolic blood pressure, heart rate and conventional measured blood pressure were monitored simultaneously. Intima blood pressure drops in beats and short peaks during effort which could not be registered with conventional methods were nicely observed. Conclusions: Using a newly developed non invasive continuous blood pressure measuring device in children during bicycle exercise tests, reliable values were derived. Children after open heart surgery delivered the same physical fitness than children with insignificant congenital heart disease and healthy children.

P436

Personal characteristics and social adaptation of school children and adolescents with congenital heart diseases (CHD)

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Introduction: The majority of children operated for CHD are chronic patients. Hence, the adequate care for these children should not only include pre-surgery medical diagnosis and therapy, but also at their social integration and rehabilitation. Goal: To assess the social awareness of children with CHD, and their physical and psycho-social functioning in everyday life. Patients and methods: The sample includes 97 school children and adolescents (both genders) with CHD and 93 healthy children of the same group. According to Doll's scale of social awareness we have estimated an individual's ability to function independently and as a part of a group. We have included the following parameters to assess the above: ability to dress and feed oneself, social skills, independency, hobbies and interests. The assessment of the influence of illness on daily physical and psychological well-being has been conducted by a questionnaire survey. Results: Children with CHD have worse performance in school (95% CI: 2.35–9.31) and higher incidence of exam retakes (95% CI: 1.53–2.56), participate less frequently in extra curricular activities (95% CI: 1.01–1.76), differ in their ability to communicate and interact socially (95% CI: 3.45–9.31), illness prevents them to continue education (95% CI: 1.18–2.14) and are over protected (mothers) in comparation to healthy children of the same age group (95% CI: 1.48–2.65). Conclusion: The children with CHD have more problems with social adaptation than healthy children. The reasons for that could be following: lack of self-confidence, bondage with parents, later achievement of financial independence, limitations in professional orientation because of CHD. Croatia is predominantly conservative environment and these results could be somewhat different to similar investigations performed in Western countries.

P437

Quantitative Judgement of exercise induced st-depression in children with aortic stenosis (AS)

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Some studies suggest that the endovascular adaptation to exercise can reveal the severity of AS especially for serial investigations in the young. 60 asymptomatic children, 9–17 years old, with AS underwent a controlled progressive bicycle exercise test. Their resting peak pressure gradients (PG) were 10–90 mmHg. The assessment of the ST-depression was classified by the score of Rautaharju et al.* The correlation of the ST-depression to PG at rest was

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

r = 0.40 (p < 0.05), during exercise r = 0.76 (p < 0.01). The sensitivity of significant ST-depression in relation to a PG > 50 mmHg was at rest 0,44 the specialty 0,98, during exercise 0,67 and 1.0 resp.. No patient had severe symptoms but one had ventricular ectopic beats. The results of the study show that exercise testing is useful for quantifying the severity of AS. It also shows that properly supervised exercise testing can be performed at minimal risk to children with significant AS. *Rautaharju, PM. et al. In: Frontiers of fitness, ed.. R.J. Shephard, Thomas. Springfild 1971

Hemodynamics and Physiology, Cardiac Function/Hemodynamics

P438

N/S

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To study alteration in content of myocardial intracellular Ca2+ due to acute pneumonia under both quiescent and contracting conditions, there to get a good understanding of the possible mechanism of heart failure secondary to acute pneumonia.

P439

The assessment of function of the heart ventricular block in normal human

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BACKGROUND AND METHODS While analyzing the heart performance as a whole [7-th All-Russia Congress of Anaesthesiologists and Reanimatologists. St Petersburg, 2000, p.54–56; 12th World Congress Anaesthesiologists, Montreal, Canada, 2000, p.14, 220, 284] we recognize three blocks in it located intrapericardially: 1. atrial" block – left (LA) and right (RA) atriums; 2. "aorta-pulmonary" block – aorta (Ao) and pulmonary artery (PA); 3. ventricular "three chambered" block (VB) consisting of a) left (LV) and b) right (RV) myocardial chambers, both with blood outflow into "aorta-pulmonary" block and c) "spongy" (venous) myocardial chamber with blood outflow (during the "common" systole – see below) through coronary sinus (CS) and Thebesian vein (TV) into "atrial" block. RESULTS & DISCUSSION: At the "common systole of" three chambered VB: 1. The following blood volumes are moved: a) Two – by blood outflow – from the "spongy" (venous) myocardial chamber into the "atrial" block; b) Two – by blood outflow – from RV & LV (their stroke volumes) into the "aorta-pulmonary" block. These volumes form an "overall" stroke volume of the "three-chambered" VB – an "overall" PV of VB. The outflow is effected through the "fixed" fibrous rings of a coronarius, tr. a. pulmonalis & bulbus aortae into the primary divisions of arterial ("aorta-pulmonary" block) and into the terminal divisions of venous ("atrial" block) systems. c) Two – by blood inflow – is mobilization from v. cava inf. et sup. & v. pulmonalis into the "atrial" block, by the so called "systolic membranous drawing in of blood" resulting from the drawing of the tricuspidal & mitral valves into the RV & LV chambers cavities at blood is driven out of them. The total volume of blood driven by myocardium of the "three-chambered" VB at the stage exceeds the "overall" PV of VB to the extent of venous blood volume mobilized into the "atrial" block. 2. Phase pressure levels are created (in mm Hg): – maximums in outflow vessels (accessible to examination by means of probing): – s.coronarius (8,6+0,2), tr.a.pulmonalis (22,2+0,9), bulbus aortae (107,6+2,6); – minimums in inflow chambers in the atria "block – "x-collapses" aD & aS. Maximum in s.coronarius (8,6+0,2) and minimum in aD ("x-collapse" aD=2,4+0,76) create the phase (systolic) endocardial veno-venous gradient (6,1+0,47) of blood flow out of the venous (extramyocardial) vessels of the "spongy" (venous) chamber of the "three-chambered" VB in aD. CONCLUSION: The "overall" systole of the "three-chambered" VB forms the basic regulating: 1. The blood inflows (mobilization) including the hepatic fraction into the "atrial" block in accordance with its outflow from the "three-chambered" VB. The "overall" systole of the VB of a current cardio-cycle prepares the next cycle. 2. Systolic synchronization of arterial and venous hemodynamics of the big and the small circles of blood circulation through the "aorta-pulmonary" & "atrial" blocks.

P440

Sanitometry gas pressure (pΣ=pO2+pCO2) in sinus coronarius, arteria pulmonalis and aorta blood in "normal" human

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P441

Abnormal cardiac recovery from physical exercise in patients after atrial correction of transposition of the great arteries assessed by ultra-fast MRI

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Background: Assessment of recovery from physical exercise has been used to demonstrate cardiac dysfunction. The purpose of the present study was to monitor changes in aortic flow during recovery from sub-maximal exercise in patients after atrial repair of transposition of the great arteries (TGA) using ultra-fast MRI. Methods: 8 atrially corrected TGA patients (26±5 years) and 10 healthy controls (25±5 years) were studied using a Philips MR scanner and a MR compatible bicycle ergometer. Based on 60% of peak oxygen consumption or individual sub-maximal MR-exercise level was calculated. Flow in the ascending aorta was measured at rest, with exercise and every 30 seconds after exercise during a period of 8 minutes. Results: From rest to exercise an increase in heart rate and aortic flow was significantly lower in the M/S patients. Heart rate: patients 71±8 to 116±12bpm (+64±13%) vs controls 65±7 to 123±7bpm (+93±21%, p<0.05). aortic flow: patients +91±39% vs controls +202±75%(p<0.05). During the recovery period heart rate decreased in a similar way in both groups. Immediately after cessation of exercise aortic flow showed an initial increase in both patients and controls. In the healthy subjects aortic flow decreased thereafter. In the patients, however, aortic flow remained elevated, resulting in a significant higher aortic flow, expressed as percentage difference from rest, at 4 to 6 minutes after exercise as

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

compared to the controls. Subsequently cardiac output of the patients also remained significantly elevated 4 to 6 minutes after exercise. Conclusion. Evaluation of cardiac function during recovery from sub-maximal exercise is feasible with ultra-fast MRI. Although heart rate recovery after physical exercise was normal, the observed delayed recovery of aortic flow and cardiac output in TGA patients after atrial correction indicate abnormal cardiac recovery from exercise.

P442

An experimental study on L-type calcium current of right ventricular myocytes in acute pneumonic juvenile rat models

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To probe the myocardial calcium- metabolism during heart failure subsequent to acute pneumonia, we recorded the L-type Ca^{2+} current through the right ventricular myocyte membrane of pneumonic juvenile rat models.

P443

Deterioration of the cardiac diastolic function after fontan operation in children, the bakuley center experience

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Objective: It is known that the hemodynamical assurance of physical loads after Fontan operation is impaired. This phenomenon is considered to happen due to the deterioration of cardiac systolic function mostly. Meanwhile, the diastolic function after physiological correction of complex congenital heart defects is much less examined. The purpose of the study was to investigate the cardiac diastolic function after Fontan procedure, and reveal its influence upon the late result of operation. Material and methods: 24 patients were examined 7–16 years (mean, 7.9±3.6 years) after Fontan operation (group I). The control group (group II) consisted of 12 healthy volunteers. To estimate physical capacity, the bicycle ergometry test was performed. The main hemodynamic parameters, such as cardiac index (CI), stroke index, left ventricular end-diastolic pressure, as well as velocity of increase of left ventricular pressure, were determined by means of mathematical analysis of impedance rheogram at rest and at every level of doses physical load from 0.5 to 2.5 wt/kg. Results: Physical capacity was higher in control group (2.2±0.3 wt/kg, vs 1.5±0.5 wt/kg, $p<0.01$). CI in group II was growing from 2.6±0.7 l/min/m2 at rest, to 6.3±2.1 l/min/m2 at the load of 2.0 wt/kg ($p<0.01$). The growth of CI was accompanied by gradual acceleration of heart rate, and the increase of left ventricular end-diastolic pressure, as well as the velocity of increase of left ventricular pressure. CI in group I was significantly lower at rest (3.4±0.9 wt/kg, $p<0.01$), as well as at every stage of physical load. The growth of CI was achieved mainly by the acceleration of heart rate only. Left ventricular end-diastolic pressure was already increased at rest up to 20.7±3.9 mm Hg, and showed the tendency for the decrease in exercise. Among the patients of I group, diastolic function was comparatively more favorable in those who were in NYHA class I. Conclusions: Cardiac diastolic function after Fontan operation is significantly impaired. Diastolic function is more favorable in patients being in higher functional classes. Limited realization of Frank-Starling mechanism, as well as decreased inotropic myocardial function, are the main causes of abnormal hemodynamical assurance of physical loads after Fontan operation.

P444

Cardiopulmonary functional evaluation during exercise in patients operated on by cavopulmonary anastomosis technique

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Aim: To identify the functional elements related to physical capacity in patients operated by the cavopulmonary anastomosis. Material and Methods: Eleven patients were evaluated, mean age of 10.7 years (7.10 to 14.8), all in functional class I in a postoperative mean time of 3.9 years (1.1 to 9.4). The congenital cardiac anomaly previously to the Fontan operation was tricuspid atresia in 6, single ventricle in 2 and transposition of great arteries in 2, being atrial fenestration present in 8 of them. Previous operations were performed in 9 patients. Blalock-Taussig in 7 and pulmonary artery banding in two. All patients presented oxygen arterial saturation above 90% except one patient with 70% value. Maximal cardiopulmonary exercise test evaluation was performed in threadmill in a Bake modified protocol, compared with a control group of 7 patients. Results: The heart rate median values in the anaerobic threshold and in the respiratory decompensation point were 132 bpm (107–160) and 165 bpm (141–190), respectively. The maximal oxygen uptake corresponding to the same periods considered before were 18.7 ml/Kg-1 min-1 (14.1–22.9) and to 26.7 ml/Kg-1 min-1 (20.8–30.6) respectively. The median duration of exercise was 14.4', anaerobic threshold reached by 5.8' and respiratory decompensation point by 10.7'. All these values were similar to those in the control group. The median maximal oxygen uptake (VO2 peak) was 27.9 (22.6–34) comparing to 35 (28–42.5) in the control group ($p<0.05$) and the maximal heart rate was 165 bpm and 188 respectively ($p<0.05$). There was systolic mediant pressure elevation of 25.7 mm in all patients operated on. Conclusion: The cardiorespiratory functional capacity is decreased in children at postoperative period of cavopulmonary anastomosis probably due to a lower peak cronotropic response.

P445

Therapy of protein-loosing-enteropathy, an unusual case

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Protein-loosing enteropathy is one of the major problems in patients after a Fontan-like palliation. There is no established therapy, not a known cause for this enteropathy. We think that unfavourable hemodynamics is one cause. We report such a case. M.R. had a single ventricle, L-TGA, mitralvalve atresia with a pulmonary valve stenosis. At the age of 1 year he received a bi-directional Glenn, a fenestrated total-cava-pulmonary-connection was completed at the age of 2 years and 2 months. Subsequently he developed sinus node dysfunction. His atrial-bradycardia was progressive, leading to minimal rates of 40/min. At the same time he had early supraventricular tachycardias causing maximum heart-rates of 135/min, effectively a rate of 65/min. His pulmonary resistance was increased to 4,5 WoodUnit 2. He developed a PLE, which was partially relieved by i.v. heparin therapy (protein from 3.5 up to 4.5mg/dl), the implantation of a pace-maker resolved the PLE even after stopping the heparin. He developed PLE 17 months later again, about 2 months after the fenestration was interventionally occluded. Reducing his PM-rate (at day, span 90–150/min to 85–145/min, night: 80/min to 55/min). His symptoms relieved only after Metoprolol (0,5mg/kg) was introduced. He did not tolerate any dose reduction. We believe that this boy once developed a PLE due to a significant bradycardia. The second time it was probably due to a mismatch of the systemic to pulmonary flow leading also to an elevated neurohumoral stimulation.

P446

Assessment of cardiovascular dynamics by pressure-area relations in pediatric patients with congenital heart disease

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Background: To separately quantify the ventricular contractility and loading conditions is particularly useful for better understanding cardiovascular dynamics in congenital heart disease (CHD) where abnormalities in chamber and loading properties may coexist and further that these may alter independently or simultaneously with disease progression and therapeutic intervention. The present study tested whether ventricular pressure-area (P-A) analysis can provide such quantitation among patients with various forms of CHD. We then assessed dual cardiovascular interaction in CHD using this method. We also tested the feasibility of this methodology in a simplified and less invasive form to further enhance its clinical value. Methods and Results: We constructed P-A loops during caval occlusion by transthoracic echocardiographic automated border detection (ABD) combined with ventricular pressure recordings in 67 pediatric patients with CHD and in 6 normal controls. Area measurements by ABD were highly reproducible ($y=1.0x-0.1$ for end-diastolic area, $y=1.0x+0.2$ for end-systolic area, $r=0.94$, $p<0.001$) and area changes during caval occlusion reflected volume changes ($r=0.87\pm0.06$, slope=0.98±0.11). The P-A data provided load-independent measures of contractility, which were consistently increased by dobutamine ($p<0.05$). End-systolic and arterial elastance individually quantified simultaneous changes in ventricular contractility and loading with milrinone infusion, and predicted net cardiac performance. The P-A analysis better characterized the ventricular contractile states under a variety of loading conditions in CHD, whereas predominant load-dependence of conventional indices confounded them. Furthermore, P-A relations were reasonably estimated from a single

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

beat (y=0.7hs+1.92, r=0.81) and from aortic pressure data during abdominal compression (y=1.06x+0.34, r=0.89). Conclusions: P-A analysis should provide a useful modality with which to assess cardiovascular dynamics in pediatric patients with CHD in more detail, and should thus help improve the management of patients with this disease.

P447

Systolic ventricular function in patients with failing Fontan converted to total cavo pulmonary connection (TCPC): early results

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Systolic ventricular function was assessed with transthoracic echocardiography (rest and stress) in 9 failing Fontan patients after TCPC associated with arrhythmias ablation. Mean age was 20.9±6.7 yrs, interval from Fontan operation 15.6±2.5 yrs, duration of follow-up 11.8±4.8 months. Preoperative ejection fraction (EF) at rest (R) and on effort (E) was 37±6.9% and 39.9±11.4%, respectively, with an average increment R-E of 7.5%. Ventricular dysfunction was present in 5 (R) and 4 (E) patients, respectively. After TCPC EF significantly improved both at rest (46.1±6.9%, p=0.049) and on effort (49.6±7.7%, p=0.011), with an average increment R-E of 10%. Myocardial dysfunction was present in 1 (R) and 1 (E) patients, respectively, being reversed in 4 (R) and 3 (E) patients. These preliminary results show that conversion of failing Fontan to TCPC promptly improves ventricular function, both at rest and on effort.

P448

Postoperative left ventricle function in patients with transposition of great arteries

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During August 1992 to August 1999 at Paediatric Clinic in Sarajevo 6 passage 10 hrs to 30 days, have been diagnosed to have D-TGA. First Group (n=4) had simple D-TGA. In Group II (n=4) pts D-TGA was associated with DILV and in 1/4, with DORV and subpulmonary artery stenosis. Anatomical correction of D-TGA-arterial switch has been performed in Group I, mean age 15.6 days (7–35). In Group I a palliative correction has been completed, mean age 4.7 months. Pts have been followed from 3–19 months postoperatively. The aim of this study was to evaluate LV function pre and postoperatively Using M and 2D echocardiographic techniques. LV function (FS) was measured in both groups of pts. LV function in pts post anatomical correction has returned to normal values faster, with statistically significant difference of p=0.02, than in pts post palliative-partial correction. Conclusion: Echocardiographically LV function in pts with D-TGA post arterial switch returned faster to normal values than in pts following the palliative-arterial correction.

P449

Mitral inflow (E) to mitral annular velocity (EA) ratio - a useful measure of LV preload in pediatric patients

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Many non-invasive indices of LV function have limited utility in pediatric patients as a result of their dependence on ventricular geometry and wall composition. As a step toward developing a load independent or load corrected measure of LV function which is independent of ventricular geometry and composition, we have looked at the Mitral Inflow Velocity (E) /Mitral Annular Velocity (Ea) ratio as a correlate of other measures of LV preload in a wide range of pediatric patients. In 36 patients (age range 6 months to 18 years) including normals, mitral and postop congenital heart defects (without significant anatomic or hemodynamic abnormality), and cardiomyopathy patients, doppler tissue imaging was used to assess Ea. In addition E, LV end diastolic dimension (LVEDD), LV volume and pulmonary capillary wedge pressure (PCWP) were assessed by standard methods. In this limited group E/Ea ratio is a correlate of LVEDD corrected for height ($R=0.73$, $P<0.01$). E/Ea also appears to correlate with PCWP as shown in adult populations however it remains small. In 5 patients we altered preload by IVC occlusion. The LVEDD decreased by 10% by a mean of 10% and the E/Ea ratio decreased by 14%. This response was seen in all cases. We suggest that the E/Ea ratio is a reasonable measure of LV preload.

P450

Hemodynamic monitoring by pulse contour analysis after cardiac surgery in infants and children

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The aim of our study was to optimize the minimal invasive management in children in the first 48 postoperative hours after cardiac surgery. We measured cardiac index (CI), intrathoracic blood volume index (ITBVI) and systemic vascular resistance index (SVRI) with PiCCO-technique (pulse contour cardiac output, PULSION) in children with a mean age of 49 +/- 32 months and a mean weight of 13 +/-4 kg. The continuous monitoring of these parameters led to precise administration of either catecholamines or fluid to maintain normal cardiac indices (3.5–4,5). There was a high correlation between CI and ITBVI ($r=0,9$) as well as CI and SVRI ($r= -0,8$). In contrast the central venous blood pressure was not suitable for measurement of deviation from normovolemia. PiCCO is a minimal invasive assessment of high clinical value for continuous monitoring of volume states and hemodynamics after cardiac surgery.

P451

A new index of total ventricular function-total ejection isovolume index (Tei index) in patients with complex univentricular heart

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The Tei index is a Doppler echocardiogram-derived index combining systolic and diastolic ventricular function. The index is defined as the sum of isovolumic contraction time (ICT) and isovolumic relaxation time (IRT) divided by ejection time (ET), being demonstrated as a useful index to estimate ventricular function in adults (Jpn. Circ. J. Cardiol. 1995;26:135–44). In patients (pts) with complex univentricular heart (UVH), the abnormal cardiac position in thorax and the poor image caused by an operative scar make a difficulty in accurate evaluation of the ventricular function (ejection fraction, EF) using the two-dimensional echocardiography. To evaluate the availability of the Tei index in pts with UVH, we compared the Tei index and the EF from cardiocatheterization. Twenty pts after right heart bypass operation were enrolled in this study. 16 pts after bi-directional Glenn procedure and 12 pts after total cavopulmonary connection. 10 males and 10 females, aged 1 to 21 yrs). The Tei index was easily obtained in all pts. The Tei index in pts with UVH was higher than that in normal children previously reported (0.40 +/- 0.12, n=30 vs. 0.72+/-0.02, n=81, p<0.05). There was a significant negative correlation between the Tei index in pts with UVH and the EF obtained from cineangiography ($r= -0.820$, $n=16$, $p=0.0001$). We conclude that the Tei index is a useful index of ventricular function in pts with UVH who have a narrow st.

P452

The value of n-HBDH and CTNT in diagnosis of congestive heart failure

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To evaluate the value of CTnT, α-HBDH and CK-MB in diagnosis of children congestive heart failure (CHF). Serum level of CTnT, α-HBDH and CK-MB were detected in 10 cases of grade II cardiac function, 22 cases of grade III cardiac function, 10 cases of grade IV cardiac function and 18 cases of normal children. In 42 cases of the CHF patients, 36 had CTnT positive and 34 had HBD increase, account for 90.48% and 80.95%, respectively. As contrast, CTnT was negative in all of the 18 cases of normal cardiac function, and only 4 of them had HBD increase, account for 22.22%. Positive rate of CTnT and elevation of α-HBDH in CHF group were significantly higher than that in normal group, with $p<0.05$. CTnT was positive in all of the grade IV and grade III patients. However, 66.67% of grade II patients had positive CTnT. Linear associate analysis indicated that CTnT was positive correlative with HBD. Electrocardiogram demonstrated ischemic changes of ST in 3 of the grade IV patients. Serum level of CTnT and HBD are high sensitive and specific biochemistry indexes for verifier of cardiac damage in CHF. We suggest that CTnT positive accompanied with HBD increased could be considered as guide line for judgement of cardiac damage.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P453

Hepatic venous flow in children with right ventricular

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The objective was to highlight the flow pattern through the hepatic vein (HV) caused by the restrictive physiology of the right ventricle (RVRL) after the correction of aetiology of Fallot (TF). Echocardiography was performed in 14 pts, mean age 14 yrs. Patients with RVRF (Group 1) were selected based on the presence of anterograde flow in the pulmonary artery (PA) during the atrial contraction. Other pts were considered as have normal diastolic RV function (Group 2). Pulsed Doppler recording was performed in the HV during respiratory variations. Maximal flow velocities during systole, early diastole and after the atrial contraction were measured, as well as velocity time integrals (VTI) of anterograde and retrograde flows (during inspiration and expiration). Restrictive PA flow profile was recognized in 4 patients (29%) aged 13–16 yrs, in whom the complete TF correction was done at the age of 11 months to 4 yrs. Moderate insufficiency of the PA was present in 3/4 of patients. Cardio-thoracic ratio was significantly lower in the Group 1 ($p<0.05$). Statistically significant difference was found in the VTI of total retrograde flow in diastole during the inspiration ($p<0.02$), i.e. it was highly increased in the group with RVRI. The ratio of anterograde flow in systole and diastole during inspiration in Group 1 was significantly higher ($p<0.03$) than in Group 2. There were no differences in velocities and time integrals of anterograde flow. Patients with RVRF, existing after the complete correction of TF, during the inspiration have increased retrograde flow through HV in diastole. They also had an increased ratio of anterograde systolic and diastolic flow.

P454

Left ventricular cardiac functions of Transient Tachypnea of Newborn (TTNB) with low cardiac output (CO)- Left cardiac diastolic dysfunction influenced respiratory distress

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Purpose. We found TTNB with low CO. Therefore we studied their left ventricular cardiac function. Subject. Cardiac function was assessed in 28 infants admitted to the neonatal intensive care unit. Group 1 consisted of 3 infants who were DTNB with low CO. Their COs were less than −1.9 SD for Group 2. Case 1: 3258g 37w4d. Case 2: 3212g 39w5d Down syndrome. Case 3: 3858g 35w4d. Group 2 consisted of control infants who were 25 very low birth weight infants. Methods. Cardiac function of both group was assessed by echocardiographies. Two dimension, M mode, Doppler mode, tissue Doppler imaging (TDI). We calculated the end systolic wall stress, rate-corrected mean velocity of circumferentiale fiber shortening, CO, left ventricular inflow velocity, velocity of the mitral annular motion. Group 1 was studied once a day from their admission to the improvements of their CO within Δ 1.0 SD for Group 2. Group 2 was studied 12, 24, 48, and 96 hours after birth. Result. The blood pressure of Group 1 was no hypotension. Their chest X-ray showed cardiomegary and pulmonary venous congestion. The contractilities between both Groups were no difference according to the force velocity relationship. The shunt flows of Group 1 were left to right shunts at their patient ductus arteriosus and interatrium. Their COs were showed lower than −1.0 SD. CO for A wave velocity of mitral valve of TDI at admission. And the improvements of their respiratory conditions were accompanied by the improvements of their relationship between A-wave and CO. Conclusion. We concluded their left ventricular diastolic dysfunction without contractility dysfunction caused their low CO, and their diastolic dysfunction influenced their respiratory distress.

P455

Echocardiographic evaluation of right ventricular diastolic function in the first month of life

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Objective. To investigate with a prospective study the RV diastolic function in healthy neonates in according to the fall of pulmonary resistances and PDA closure. Methods. 25 healthy full-term newborns were submitted to echocardiographic study at less than 12 hours of life (mean 3.2 h), at the third day and at one month of life. We evaluated TV peak velocities (E, A wave), their ratio, normalized peak filling rate (NPFR), deceleration time and RV Tei Index. The heart rate didn't change between first and second control (131±16 b/m vs 128±15) but was significantly higher (146±15) at one month. In no one baby was impossible to distinguish the waves. We averaged five consecutive beats and assessed PDA and PFO with color doppler. Results. E wave increased significantly in all controls (0,39 cm/sec ± 0,05 vs 0,43 ± 0,09 vs 0,49 ± 0,08 $p<0,0001$) as well as A wave (0,56 cm/sec ±0,07 vs 0,64 ± 0,08 $p<0,001$) but their ratio didn't change significantly. NPFR didn't change between first and third day but increased at one month (4,42 SV/s ± 0,63 vs 5,01 SV/s ± 0,67 $p<0,005$). Deceleration time was 121,8 msec ± 20,7 in first control vs 143,3 ± 24 ($p<0,001$) at second control and 124,2 ± 17,2 at one month ($p>0,005$). Tei Index was 0,39± 0,18 in the first day and normalized in third day. PDA was closed in all babies in the third day. PFO was patent with central shunt in all at one month. Conclusions: our data suggest that RV improves its compliance and performance early in third day of life (increase of rapid filling velocity and deceleration time, Tei Index normalization) confirmed by increase of NPFR at one month of life.

P456

Echocardiographic evaluation of left ventricle in neonatal and infant periode

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Background. During intrauterine life the volume stimulation of the left ventricle is low and its cardiac output in comparison to right ventricle is about 50%. Before and also after birth the right ventricle is dominant. Postnatally low – resistance placenta is excluded from the system circulation, leading to abrupt pressure elevation and on its further increase. The left ventricle responses to these changes with increase in its size and weight. Aim of study. The authors performed echocardiographic evaluation of healthy children from newborn periode to 6 month of age. Methods: 50 healthy randomly selected newborn infants born at full term were examined. Initial echocardiographic measurements of left ventricle (4 days of age) were compared with measurements at 1 month and then at 6 month of age. Results. The parameters characterizing the left ventricle (left ventricle end-end diastolic diameter, end-diastolic volume, cardiac output) increased significantly at 1 month and 6 months. Ejection fraction and fractional shortening did not change during the periode. Conclusion. Left ventricle parameters confirm postnatal left ventricle growth in healthy infants. The results are useful in interpretation of echocardiographic examination in newborns and infants.

P457

Efficiency of gas exchange during exercise after correction of cyanotic congenital heart disease

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Patients who underwent successful repair of cyanotic congenital heart disease may complain of exercise intolerance at heavier exercise intensities. To assess the efficiency of ventilatory gas exchange, alveolar ventilation was assessed and subtracted from the total exercise ventilation. The difference reflects the physiological dead space ventilation, which is a measure of ventilatory efficiency. Gas exchange was measured breath by breath with mass spectrometry and graded exercise testing was performed on a treadmill. Three groups of patients were studied: 33 patients with Fontan circulation, 34 patients with Fallot repair and 23 patients with atrial switch for TGA. The patients were compared to 16 normals of comparable age. During exercise the difference between total ventilation and alveolar ventilation was significantly ($p = 0.01$) larger in the patients compared to the normal values (difference varying from 12 to 48 %). The larger difference for alveolar to total ventilation reflects ventilation/perfusion mismatch, which may contribute to exercise limitation.

P458

Biventricular pacing in chronic heart failure and significant aortic stenosis

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Introduction. Biventricular pacing (bvp) represents a therapeutic option in pts with end-stage heart failure and left bundle branch block (LBBB). Severe aortic valve stenosis (AVS) is considered to be a contraindication for bvp up to now. Methods and Results. The 68-years old pt. presented with severe cardiac insufficiency after four myocardial infarctions leading to NYHA IV. Additionally a calcificated AVS grade III (opening area 0,8 cm2) had devel-

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

aped. The LV-EF was 20%, the PAPs 70 mm Hg and the LVEDD of 75 mm resulting in a MI III. We registered a LBBB with a QRS-duration of 170 msec. An ICD was implanted because of ventricular tachycardia, eight months ago. On the base of a generalized atherosclerosis the right internal carotid artery was occluded and the left 80–90% stenotic. On indication of biventricular stimulation, facing the high operation-risc, we firstly replaced the aortic valve by Hancock 25 mm after desobliteration of the left carotid artery under extracorporeal circulation (ECC). During the reperfusion period the implantation of one epicardial electrode at the lateral wall of the left ventricle was done followed by temporary pacing leads at the right atrium, at the apex of the right ventricle and again at the lateral wall of LV. Biventricular pacing was started immediately leading to the stopping of ECC. The implantation of the atrial electrode and the biventricular ICD-system connected with the remained RV-lead followed after 11 days. On day 20 after AVR the pt could be dentensioned fully mobilized and an improved cardiac situation (LV- EF = 35%, PAPs = 35 mm Hg, MI I°). Conclusions: In pts with severe aortic stenosis, chronic heart failure and LBBB biventricular stimulation can sufficiently be performed and should be started intraoperatively after AVR, still during extracorporal circulation, using bipolar epicardial electrodes fixed at the lateral wall of LV.

P459

The coronary hemodynamic influence of coronary aneurysm in patience after Kawasaki disease

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The coronary hemodynamic effect of coronary aneurysm after Kawasaki disease has not been fully elucidated. To clarify the influence on coronary blood flow, we analysed the relationship between the position of coronary aneurysm and coronary flow reserve (CFR) in patients after Kawasaki disease. Twenty-four patients who had undergone Kawasaki disease were enrolled in this study. The patients were divided into 4 groups by position of coronary aneurysm on left coronary artery. Group 1: no coronary aneurysm (n=12). Group2: aneurysm on left main coronary trunk (LMT) (n=12). Group 3: aneurysm on left anterior descending coronary artery (LAD) or left circumflex coronary artery (LCX) (n=7), and Group 4: aneurysms on LAD and LCX (n=1). We measured the CFR on left coronary artery. In patients who had aneurysm, the CFRs were measured at distal position to the aneurysms. The CFR were 3.7 +/- 0.36 in group1 and 3.3 +/- 0.25 in group 2. There was no statistical difference of CFR between group1 and 2. In group3, the CFRs were 3.0 and 3.2, although these in LMT were 3.8 and 3.9. The CFR in group 4 patient is 3.0 on LAD and 2.7 on LCX. Although the existence of aneurysm itself does not disturb the coronary flow, it can interfere the blood flow when a bypass e

P460

Contractile function in the systemic right ventricle: comparison with the normal right ventricle

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The reported reductions in global right ventricular (RV) function after atrial redirection procedures (Mustard and Senning operation) may merely reflect a normal response of the systemic right ventricle to its afterload. Indeed, little is known regarding the intrinsic, load independent, indices of RV function under these circumstances. We compared 15 patients with normal right ventricles undergoing routine coronary revascularisation with 14 late survivors of the mustard operation. The right ventricular end systolic pressure volume relationship (ESPVR), preload recruitable stroke work (PRSW), and end-diastolic pressure volume relationship (EDPVR) were obtained with a conductance catheter by preload reduction. Contractility was greater in the mustard group (ESPVR = 0.92 + 0.50, PRSW = 53.50 + 27.95) versus normals (ESPVR = 0.43 + 0.20 [p = 0.002]; PRSW = 15.08 + 6.45, [p < 0.001]), but there was evidence of reduced ventricular compliance (EDPVR = 0.36 + 0.09 versus normal EDPVR = 0.05 + 0.03, [p<0.001]). Thus contractility is supranormal in the systemic RV after atrial redirection procedures. Ventricular compliance is, however, reduced. Strategies to improve early diastolic relaxation flow, and late diastolic filling may be advantageous in the long-term treatment of these patients.

P461

The hemodynamic effects of carbon dioxide pneumoperitoneum in infants undergoing laparoscopy

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The purpose of the study is to investigate the hemodynamic effects of carbon dioxide pneumoperitoneum (CO2Pneumo) and different body positions in infants undergoing laparoscopy. Studies were done under general anesthesia and positive mechanical ventilation with ECG, phonogram, percutaneous CO2 and vital signs monitoring. Effects were evaluated in Trendelenberg, horizontal and reverse Trendelenberg positions with CO2Pneumo at intra-abdominal pressure of 0, 10 and 15mmHg. Doppler evaluation of pulmonary veins, SVC, mitral and tricuspid inflows and Acoustic Quantification study on both ventricles were done using TEE. Doppler evaluation of aorta and M-mode on left ventricle (LV) in short-axis for ejection time (ET), fractional shortening (FS%), LV wall stress (LVWS) and Vcfc calculation were done by TTE. Data was analysed by mixed linear regression analysis accounting for serial measurements. There were 13 boys and 2 girls, aged 3.19 ± 1.25 months. Different positions have no hemodynamic impacts. Increase in intra-abdominal pressure results in significant increase in diastolic and mean blood pressure (BP) ($p<0.01$), RV contractility ($p<0.01$) with decrease in RV peak filling rate (PR/FR) ($p=0.04$) and tricuspid inflow velocity time integral. Although LV preload was increased there were no significant effects on its systolic function. Hypercarbia causes significant increase in heart rate (HR) ($p<0.0001$), cardiac index ($p<0.0001$), LV peak ejection rate ($p=0.011$), FS% ($p=0.02$), Vcfc ($p=0.04$) and PR/FR of both ventricles ($p<0.01$). BP ($p=0.001$), DT ($p<0.04$), LVWS ($p=0.02$) end-systolic and end-diastolic areas of RV ($p<0.02$) and LV ($p<0.01$) are reduced. In conclusion, increase in intra-abdominal pressure causes decrease in venous return hence, reduce RV preload but increase BP by compressing on the abdominal aorta. By the various mechanisms seen, the normal hearts of these infants seem to tolerate and compensate well the hyperdynamic state caused by hypercarbia. However hypercarbia can be detrimental and have significant implications on cardiac patients with depressed function undergoing surgery by the route.

P462

Cardiac output measurement by transoesophageal Doppler ultrasound compared to clinical evaluation in the haemodynamic assessment of critically ill children

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Aim: To measure the cardiac output using transoesophageal Doppler (TOD) in mechanically ventilated patients in PICU and compare this to clinical assessment of haemodynamic values obtained in the same group of patients. Methods: 20 children were studied, age range from 2 to 192 months (median 32.5 months). The TOD transducer, emitting a 4-MHz continuous wave Doppler signal was introduced orally and advanced until the characteristic descending aorta waveform was obtained on the monitor (CODM II, Deltex Ltd Chichester, UK). Seven consecutive values of minute distance (MD) were calculated and the mean taken. Simultaneously the heart rate, mean blood pressure, central venous pressure and the variables such as base deficit (arterial blood gas analysis) and blood lactate were measured and the mean for 7 consecutive values was taken for each parameter. Following a fluid challenge, seven repeat pairs of measurements were made. Results: Scatter plot of the mean percentage difference of MD against the other variables showed that there was minimal degree of linearity between the heart rate, mean blood pressure, lactate level and base deficit for the difference pre and post fluid infusion. However central venous pressure percentage difference showed more marked negative linearity. Linear regression univariate analysis showed that there was no correlation between MD and heart rate, mean blood pressure, lactate level and base deficit. In the case of central venous pressure percentage changes there was a definite correlation but with borderline significance ($p=0.09$). Our TOD data showed consistent values with excellent reproducibility, confirming the accuracy of the technique. Conclusion: Clinical and laboratory assessment of haemodynamic status is not reliable in critically ill children. It is therefore important to have an accurate estimate of cardiac output using a non-invasive technique such as TOD which avoids the risks associated with pulmonary artery catheterisation.

P463

Relationship between right ventricular dysfunction and QRS duration in patients with repaired tetralogy of Fallot

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https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Background: After surgical repair, patients with tetralogy of Fallot (ToF) may develop right ventricular dysfunction due to pulmonary insufficiency. We evaluated the relationship between right ventricular enlargement, right ventricular function and ECG duration in patients with repaired ToF. Methods: 20 patients with repaired ToF followed-up in our department were studied. Right ventricular volumes were measured using Tc-99 angiocardiography. Right ventricular ejection fractions (RVEF) were calculated from end-diastolic and end-systolic volumes after correcting for body surface area. QRS duration was calculated on baseline ECG using DII and V1 leads. Mean (SD) age at surgery was 3.6 years. Mean follow-up occurred at 15.3 years. Results: Pulmonary regurgitation (PR) was present in all 20 patients(mild-to-moderate in 9, severe in 11). Patients with severe pulmonary regurgitation had a lower right ventricular ejection fraction (p<0.02) compared to patients with mild-to-moderate PR. Mean QRS duration was 148 msec. (range: 120–200 msec.). Mean right ventricular end diastolic and end-systolic volumes were 148 and 83 ml. The correlation between RVEF and QRS duration was: r = -0.66 (p<0.003). The correlation between RV end diastolic volume and ECG duration was r = 0.76 (p<0.0001). Conclusions: There is a significant correlation between RV end diastolic volume and QRS duration. The inverse correlation between RVEF and QRS duration is significant. This could be the link between RV dysfunction, QRS widening, and the increase in sudden death rate observed in patients with repaired ToF.

P464

The right ventricular restrictive physiology in repaired tetralogy of Fallot is associated with smaller respiratory variability

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To assess the relationship of biventricular diastolic forces on volume characteristics and influence of respiratory effect after repair of tetralogy of Fallot, we investigated 40 patients with- and without restrictive physiology(RP) of RV. The patients were studied 46.7±38.5 months after the operation and classified into 3 groups: Group I consistes 19 patients with RP and pulmonary valve(PV) not preserved, group II, 15 patients without RP and PV not preserved, and group III, 6 patients without RP and PV preserved(transannular patch not required). A simplified, indirect volumetric index(VI) indicating the severity of RV volume load was derived from 2D echocardiography and validated with the measured RV volume from biplane angiography in 31 patients. Doppler spectrals were obtained from superior vena cava, tricuspid valve, main pulmonary artery(MPA), pulmonary vein and mitral valve. The type of outflow tract reconstruction, VI, and biventricular diastolic function were compared and the following results were obtained: 1. 35% of patients have RP of RV and the incidence of TAP is higher in these patients(93.3% vs 38.9%, p<0.01). 2. VI is lowest in group III(0.79±0.12) followed by group I(1.01±0.36) and group II(1.35±0.16, p<0.01). 3. VTI ratio of total regurgitant/antegrade flow at MPA is larger(0.49±0.13 vs 0.68±0.18, p=0.05) and more dependent on respiration in group II, especially in expiratory phase. 4. The S/D ratio of pulmonary venous Doppler velocity and VTI was higher in group I compared to group II (0.93±0.19 vs 0.73±0.16, p<0.05). 5. The decreased S/D ratio of SVC Doppler velocity and VTI was also observed in group II and some of group III. We concluded that 1. RP is associated with less volume overload of RV and, in spite of the smaller reserve volume, smaller respiratory variability is associated. 2. The diastolic function and volume status of RV may influence the diastolic property of left heart.

P465

Blood flow of left anterior descending coronary artery in children with ventricular septal defect

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High frequency echocardiography offers a noninvasive approach for imaging left anterior descending coronary artery (LAD) blood flow from a transthoracic window. We applied this technique to study the effects of left ventricular (LV) volume overload on the size and flow of LAD, which have not been studied extensively in pediatric patients with congenital heart disease. The study groups consisted of 38 children with ventricular septal defect (VSD) and 15 normal children. Left ventricular mass (LVM), LAD cross-sectional area (CSA), and flow velocity were measured by transthoracic echocardiography. LVM was indexed for body surface area. The pulmonary to systemic flow ratio (Qp/Qs) was obtained by cardiac catheterization. Qp/Qs ranged from 1.2 to 3.1 (mean 2.0 ± 0.5). The mean LAD flow velocities, flow velocity integrals, and flow volumes were significantly higher in patients than in controls. In patients with VSD, LAD flow velocity, flow velocity integral, and flow volume correlated significantly with Qp/Qs. The ratio of flow volume to LVM did not differ between the two groups. In 8 patients with VSD, LAD flow velocity, flow velocity integral, and flow volume decreased after surgery. The present results suggest that patients with VSD have a higher resting coronary blood flow. LAD flow pattern is dependent on LV volume overload and its change after.

P466

Right ventricular myocardial dysfunction in children with ventricular septal defects-role for myocardial performance index?

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Background-Myocardial performance index (MPI)originally designed by Tei et al[1] has been shown to be load independent in children with congenital heart defects. Methods-We established our own normal values for MPI for the right and left ventricules based on 60 children with innocent heart murmurs (MPI-right: 0.208,95% CI: 0.183 - 0.232; MPI-left: 0.288,95%CI: 0.251 - 0.325). Seventeen patients with ventricular septal defects had the indices assessed preoperatively. Results-Left ventricular MPI was found to be within the 95% CI in all 17 patients studied. No correlation was found between the clinical need for ambulatory treatment and the values of left ventricular MPI. The values of right ventricular MPI was increased above the upper limit of normal(95%CI)in 14(82%)patients(mean MPI: 0.497,SEM: 0.062,range: 0.333–0.808). However no correlation was found between the values of right ventricular MPI in the 8 patients on ambulatory treatment and the remaining 11 asymptomatic patients(mean MPI: 0.474,SEM: 0.079 vs 0.416,SEM: 0.069,p>0.5). Conclusions-Right ventricular myocardial dysfunction as assessed by the MPI may be present in the majority of children with ventricular septal defects. Standard ambulatory diuretic treatment does not influence the incidence of myocardial dysfunction thus confirming the load independence of the index. [1] Tei et al. Circulation 1995;92:1-592.

P467

Clinical usefulness of color Doppler M-mode analysis in pediatric patients with heart failure

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Background: Diastolic function has recently been focused on in the field of cardiology, especially for the evaluation of patients with heart failure. However, since Doppler derived data such as E/A are strongly influenced by heart rate or general condition, this method is not useful for the evaluation of pediatric age group. We evaluated patients with this age group by color M-mode to clarify the clinical usefulness of this method. Subjects and Methods: Fourteen children (mean age: 9.4 years, 6 months to 15 years) with left ventricular ejection fraction less than 45% were enrolled in this study. LV-filling patterns of color Doppler M-mode were obtained by LV inflow in the apical 4- chamber or long axis view. Then, rate time difference between the occurrence of peak velocity at the mitral tip and in the apical region (TD) was calculated. We determined the maximal velocity point of color inflow using zero shift of Nyquist limit from around 100 cm/sec to 10 cm/sec after freezing the color M-mode view. The aliasing of color flow mapping subsequently appeared and the maximal velocity point was determined from the mitral tip to the apical region at each depth. Four patients died of heart failure and 2 exhibited severe conditions of NYHA III during three-year follow-up. The echo-derived data of these 6 patients (group A) were compared with rest of the 8 patients (group B). Results: There was no significant difference in LVEF, E/A, or deceleration time of the E wave between the two groups. On the contrary, TD was significantly prolonged (155.8±56.2 vs. 96±21.9 msec, p<0.05) in group A. Conclusions: TD is a very useful parameter in the evaluation of the prognosis of patients with heart failure in this age group.

P468

Pulmonary endothelial dysfunction does not contribute to pulmonary regurgitation late after tetralogy of Fallot repair

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Background: Cardiac lesions with high pulmonary blood flow may cause endothelial dysfunction in the pulmonary vascular bed. Previous studies have shown a relationship between increased pulmonary vascular resistance and the degree of pulmonary regurgitation in postoperative tetralogy of Fallot patients. The resulting high stroke volume in these patients may act in a way

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

similar to high flow lesions leading to pulmonary endothelial dysfunction and subsequently increase the burden on the right ventricle. The aim of the present study was to determine the impact of nitric oxide inhalation on pulmonary regurgitation as an indicator of pulmonary endothelial dysfunction late after tetralogy of Fallot repair. Methods: We studied eight patients (age: mean 12 years, range 5 – 22 years) with pulmonary regurgitation, but no pulmonary branch stenosis after repair of tetralogy of Fallot. Magnetic resonance velocity mapping (through-plane velocity encoding) was performed perpendicular to the main pulmonary artery. The velocity mapping was repeated after 15 minutes with nitric oxide in air (40 ppm) by face mask. Results: All patients had moderate to severe pulmonary incompetence (regurgitant fraction: range 0.26 to 0.51). The heart rate decreased from (mean +/- SD) 87 +/- 14 to 83 +/- 13 bpm ($p<0.06$) during the nitric oxide inhalation, although no significant changes were observed for the total cardiac output (7.5 +/- 3.0 vs 7.3 +/- 2.7 L/min) or the regurgitant fraction (0.37 +/- 0.07 vs 0.37 +/- 0.09). Conclusion: Nitric oxide inhalation has no effect on pulmonary regurgitation late after tetralogy of Fallot repair. These results suggest that endothelial dysfunction and elevated pulmonary vascular resistance are not part of the late pathogenesis of pulmonary regurgitation and right ventricle dysfunction in these patients.

P469

Coronary flow reserve in the newborn.

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Background: Recent studies have indicated that coronary flow reserve (CFR) is reduced in neonates with congenital heart defects. However, normal values for CFR in the anatomically normal neonatal heart are lacking. Methods: Eight mechanically ventilated newborn lambs, born near term, were studied during the first day of life. Average peak velocity (APV), and peak flow velocities in diastole (PFVd) and systole (PFVs) were measured in the proximal left anterior descending coronary artery before and after adenosine injection (140 and 280 ug/kg, intravenously) using an intracoronary 0.014 inch Doppler FloWire(tr) (Cardiometrics). CFR was defined as the ratio of hyperemic to basal APV. Measurements were made at arterial 100 saturation and after gradually lowering the fraction of inspired O2. Results: With the lambs in a hemodynamically stable condition and normal O2 saturation, mean (SD) CFR was 3.5 (1.9). The table shows results obtained by acute hypoxemia. Regression analysis showed a linear relation between O2 saturation and both log APV and log PFVd ($r = -0.89$ and -0.70 respectively), $p < 0.0001$, and a linear relation between O2 saturation and CFR ($r = 0.81$, $p < 0.0001$). Conclusions: CFR measured with intracoronary Doppler guide wire in newborn lambs is comparable to values reported for adults. This supports the interpretation of earlier studies indicating that neonates with congenital heart defects may have pathologically reduced CFR. Acute hypoxemia causes increased coronary flow velocity and therefore reduced CFR.

P470

A study of ventricular diastolic function after repair of tetralogy of Fallot

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Objective: to study the right and left ventricular diastolic function after repair of tetralogy of Fallot. Method: This is a case – controlled study involving 62 children, monitored from February to October 2000. The children were divided into two groups: 31 patients that underwent repair of tetralogy of Fallot, and 31 healthy subjects paired for age, sex and body surface. The mean postoperative time was 67 months. Echocardiographic values were obtained by the use of M -Mode and Doppler of the mitral and tricuspid valves. The tricuspid measurements were made according to the phase of the respiratory cycle. Results: The mean demographic variables of the patients were: age 112 months (+/- 39 months), weight 28.25 Kg (+/-13.50 Kg), body surface 0.98 m2 (+/-0.31m2). Seventeen patients were females (54,8%). The mean age at surgery was 36 months (+/- 26 months). The M-Mode measurements show the significant difference in diastolic diameter of RV and of the LA. Examining the mitral flow, we observed significant difference in E/A relation, integral velocity and at desaceleration time of E wave. Regarding tricuspid flow, there was no difference in inspiratory and expiratory measurements, among the patients, however as E/A relation, integral velocity and wave A velocity was significantly different compared to the control group. Conclusion: The measurements analyzed suggest that there is alteration in the early phase of the mitral diastolic flow and also in the last phase of the tricuspid diastolic flow, after postoperative of tetralogy of Fallot.

P471

Analysis of postoperative catheterism in tetralogy of Fallot

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Since the first total correction of tetralogy of Fallot 1954, surgical technique has been improved, but residual lesions are still frequent. Objective: to analyze the postoperative hemodinamic status after total correction of tetralogy of Fallot. Method: This is a retrospective analysis of 104 cases with total correction of tetralogy of Fallot. Patients were submitted to cardiac catheterization from January, 1992 to November, 2000. The mean age was 79 +/- 39 months, with 65 (62%) male patients. The mean weight was 27,000 +/- 12,320 g. Right ventricular abnormality, pulmonary artery alterations and residual defects were analyzed. Results: The mean postoperative time was 63.44 +/- 39.25 months. The mean systolic pressure of right ventricle was 47.82 +/- 23.23 mmHg. Median gradient of right ventricular tract obstruction was 11.0 mmHg (0 to 108 mmHg). Important stenosis was observed in the left branch of pulmonary artery of 9 patients and in the right branch of 7 patients. Twenty six (27.3%) cases had pulmonary hipertension. Four patients had residual ventricular septal defects. The right ventricle showed hypertrophy in 47 cases, with tricuspid regurgitation in 17 cases. Pulmonary insufficiency was minimal in 21, moderate in 16, and severe in 15 patients. Conclusion: Pulmonary artery stenosis and pulmonary hypertension are frequent in the postoperative course of tetralogy of Fallot. The right ventricle have been compromised in almost half of the cases.

P472

Ventricular end-diastolic volume from ejection fraction and stroke volume in adult pigs during IVC occlusion

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Purpose: Left ventricular end-diastolic volume (LVEDV) is important for the assessment of intraoperative systolic and diastolic LV function, but current methods are not practical for real-time analysis. LV ejection fraction (EF) and stroke volume (SV), measured easily and reliably, can be used to calculate LVEDV indirectly. Accordingly, this study was undertaken to validate the indirect determination of LVEDV during preload reduction by comparing it to LVEDV values derived from steady-state measurements. Methods: Five pigs (40—45kg) underwent median sternotomy and pericardiotomy. An ultrasound transit-time flow probe placed on the ascending aorta provided cardiac output (CO). A micromanometer provided LV end-diastolic pressure (EDP). Right-ventricular pressure, 2D-echocardiograms (2-DE) and ECGs were also measured. These measurements were obtained during the steady-state and during inferior vena caval (IVC) occlusion. SV was determined from CO and heart rate. EF was derived from short-axis 2-DE. Diastolic pressure-volume (P-V) curves based on SV/EF during preload reduction were compared to P-V curves generated from 2-DE EDV values derived at the steady-state from three 2-DE long-axis sections. Results: Correlation coefficients for linear regression and P-V relation analysis generally ranged from 0.70 to 0.99. The two methods for measuring LVEDV indirectly and directly generated comparable compliance curves. Conclusions: SV/EF is promising for measurement of LVEDV and may facilitate real-time determination of intraoperative changes in LV diastolic properties. Its accuracy and delay in these preliminary studies are acceptable and merit further investigation.

P473

Intraoperative changes in ventricular dimension, geometry and function in surgery for congenital heart disease

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We studied LV function in repair of atrial (ASD, n=16) and ventricular (VSD, n=13) septal defects and tetralogy of Fallot (TOF, n=10) with intraoperative two-dimensional echocardiography. Statistically significant ($p<.05$) changes are described. Preload from end-diastolic area (cm2, mean±SEM) increased in ASD from 7.7±.9 to 9.3±1 but decreased in VSD from 10.5±2 to 7.4±.5 and in TOF from 5.2±.5 to 4.4±.5. Geometry from the ratio of anterior-posterior to septal-freewall endocardial diameters (D2/D1) was more symmetric after ASD closure. Systolic function was assessed from peak wall stress (PWS), stroke area (SA), ejection fraction (EF%) and fractional shortening (FS). EF% (%) in ASD increased from 53±3 to 60±2 but decreased in VSD from 52±3 to 37±3 and in TOF from 64±3 to 50±4. SA (cm2) increased in ASD from 4.2±.7 to 5.4±.7 but decreased in VSD from 5.7±1 to 2.9±1 and in TOF from

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

3.22 ± 2.01 ± IFS increased along D2 but not D1 in ASD. Conclusion: Preload and systolic function increase after ASD repair and decrease following VSD and TOF repair. Diastolic LV symmetry improves after ASD repair. SA and EFs increases in ASD reflect improved diastolic septal mechanics since IFS increases along the D2 diameter. LV preload is the most important determinant of ventricular function postoperatively in this series. Contractility appears mildly depressed in postoperative TOF. Notable preload mediated LV contractile reserve may be present in postoperative VSD and TOF.

P474

Intraoperative changes in left ventricular compliance following ventricular septal defect repair in children

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PURPOSE: Immediate intraoperative effects of surgical correction of ventricular septal defects (VSD) on left ventricular (LV) dimension, symmetry, and loading may also contribute to changes in LV compliance. Using transesophageal echocardiograms (TEE) and LV pressure, we measured changes in LV compliance in six consented patients undergoing VSD repair. Patient age ranged from .56 to 2.1 years (mean 1.07 ± .25). METHODS: Ascending aorta and bicaval venous cannulation for bypass and 1:1 blood cardioplegia were used. TEE LV short axis (SS) sections, EKG, and calibrated LV pressure, using a 5F micromanometer inserted in the LV cavity via the apex, were simultaneously recorded during preload depletion prior to CPB and volume loading following VSD repair. LVP and EKG were also digitized using an A/D converter. Following beat to beat measurements by hand of LVP and LV SS end-diastolic area (EDA) at end-expiration, the relation between LVEDP and EDA was determined using the formula LVEDP = e^{bV}. The LV ventricular stiffness constants, a and b were compared pre and post repair to assess changes in ventricular compliance. RESULTS: The mean a pre-repair was 1.71 and post-repair was 2.22 (p^{-}ns). The mean b was 0.12 pre-repair and 0.10 post-repair (p^{-}ns). CONCLUSION: In this study, surgical correction of VSDs resulted in no significant change in LV compliance.

P475

Physiologic effects of induction and reversal of myocardial edema in vivo

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Purpose: Although myocardial edema is likely to cause physiologic abnormalities of the left ventricle (LV), the time course of this phenomenon is not well defined. This study will report the physiologic effects of induction and reversal of myocardial edema in the beating heart pig heart. Method: In conditioned anesthetized pigs, the coronary arteries were perfused for 50 to 60 seconds with diluted blood (hematocrit value 20% ± 2%, edema group, n=5), or whole blood (hematocrit value 28% ± 1%, control group, n=6) infused into the aortic root during aortic cross-clamping. After whole blood reperfusion, preload reduction by vena caval occlusion was used to define systolic and diastolic properties at 15-minute intervals. LV pressure, conductance, aortic flow, and two dimensional echocardiograms were recorded. Results: LV mass (wall volume) in the edema group increased significantly compared with that in the control pigs after cross-clamp removal. Mass returned to preperfusion levels after 45 minutes. The ventricular stiffness constant (b) increased significantly in the edema group versus the control group, returning to baseline by 30 minutes. The diastolic relaxation constant (τ) and base constant (a) did not differ between groups. There was no significant change in contractility. Conclusion: Increases in LV mass and diastolic stiffness induced by coronary perfusion with hemodiluted blood resolve after 45 minutes of whole blood perfusion in pigs. Myocardial edema is an important consideration in intraoperative assessment of LV diastolic properties and could cause antifarcal reduction in LV compliance.

P476

Perioperative changes in left ventricular function and geometry following atrial septal defect repair

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OBJECTIVE: The immediate effects of surgical correction of atrial septal defects (ASD) on left ventricular (LV) dimension and loading may also contribute to changes in LV compliance. Using transesophageal echocardiograms (TEE) and a left ventricular (LV) micromanometer, we studied these changes intraoperatively in consented patients (n=7) undergoing ASD repair. METHODS: Ascending arch and bicaval venous canulation for bypass and 1:1 blood cardioplegia were used. Geometry was assessed from the ratio of septum-freewall to anterior-posterior endocardial diameters (D1/D2). Systolic function was assessed from area ejection fraction (EFa) and fractional shortening along D1 and D2. Preload was measured from end diastolic area (EDA). Diastolic function was assessed using simultaneously recorded LV slices and TEE, LV EDP, and EKG during preload depletion prior to CPB and volume loading following CPB. RESULTS: Data are tabulated as mean±SEM. CONCLUSION: LV EDP increased, and systolic function indicators improved following ASD repair. Diastolic and systolic LV symmetry were restored. No significant change was seen in ventricular compliance, despite a significant increase in septal to freewall diameter.

P477

Validation of left ventricular end-diastolic volume from ejection fraction and stroke volume

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OBJECTIVE: LV end-diastolic volume (EDV) is important for the assessment of intraoperative systolic and diastolic LV function, but present methods are cumbersome for real time analysis. LV ejection fraction (EF) and stroke volume (SV), measured easily and reliably, can be used to calculate EDV indirectly. Accordingly, this study was undertaken to validate the indirect determination of EDV during the steady state and during preload reduction. METHODS: Five pigs (42–45 kg) underwent median sternotomy and pericardiotomy. An ultrasound transit time flow probe on the ascending aorta provided cardiac output (CO). A micromanometer provided LVP. RVP. 2D-echocardiograms (2-DE) and ECG were also measured. After data recording in the steady state, hearts were arrested and excised for measurement of post-mortem pressure-volume (PV) curves. SV was determined from CO and heart rate. EF was determined from short axis 2-DE. EDV calculated from SV/EF was compared to values derived from PV curves and LVEDP and to measurements based on three long-axis sections from 2-DE. RESULTS: Data are tabulated and illustrated below. Correlation coefficients for linear regression analysis generally exceeded 0.90. CONCLUSIONS: SV/EF is promising for measurement of LVEDV and may facilitate real time measurement of intraoperative changes in LV diastolic properties. Its accuracy and utility in these preliminary studies are acceptable and merit further investigation.

P478

Noninvasive quantification of left-to-right shunt in 50 pediatric patients by phase-contrast cine magnetic resonance imaging. a comparison with invasive oximetry

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Background: Blood flow can be quantified non-invasively by phase-contrast cine magnetic resonance imaging (PC-MRI) in adults. Little is known about the feasibility of the method to quantify left-to-right shunts in children with congenital heart disease. Therefore we sought to evaluate PC-MRI in a larger pediatric population with a simple left-to-right shunt lesion. Methods and Results: In 50 children (mean 6.2 years, range 1.1–17.7) with an atrial or ventricular level shunt, blood flow rate in the great vessels was determined by PC-MRI and the ratio of pulmonary to aortic flow (Qp/Qs) compared with Qp/Qs by oximetry. We found a difference of 2% and a range of -20% to + 26% (limits of agreement), (mean±2SD). In another seven children with congenital heart disease but no cardiac shunting (mean 7.9 years, range 1.3–17.7), Qp/Qs by PC-MRI was 1.02 (SD±0.06). No difference between systemic venous and aortic flow volume was found (range -1.7% to +2.0%, n=17). Blood flow through a ventricular atrial septal defect as assessed by PC-MRI (n=24) overestimated the shunt compared with the difference between pulmonary and aortic flow. The mean difference between three repeated PC-MRI measurements in each location was 5.2% (SD 4.0, n=52.2), demonstrating good precision. The interobserver variability was low. Accuracy of PC-MRI was confirmed by in vitro experiments. Conclusions: Noninvasive determination of Qp/Qs by PC-MRI in children with a simple left-to-right shunt lesion is quick, safe and reliable as compared with oximetry and may replace invasive or x-ray-based methods.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Systemic venous flow can be quantified by PC-MRI, whereas through-plane shunt measurement within an atrial septal defect is inaccurate.

P479

Comparison of the Echocardiographic methods for pulmonary to systemic blood flow ratio (Qp:Qs) estimation

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Objective: to assessed the accuracy, reliability and safety of different echocardiographic methods for Qp:Qs calculation in comparison with scintigraphic Qp:Qs calculations Patients and Methods We evaluated 36 patients with ASD secundum, median age 5.6 years (2.5 to 16 years). In all patients Qp:Qs was calculated using 1) maximal flow velocity through the mitral and tricuspid valves (V TV-MV), 2) mean flow velocities through the mitral and tricuspid valves (vM TV-MV), 3) mean flow velocities through pulmonary and aortic valves (vM AV-PV) and compared them with standard scintigraphic method. Max velocity (V) was determined by Doppler measurement of the max flow velocity through the mitral (VMV) and the tricuspid valves (VTV). Mean flow velocity (v M) is calculated by integrating the area under the Doppler curve using standard ECHO software. The heart valves diameters were measured in four-chamber (MV,TV) and parasternal (AV,PV) ECHO views, during their maximal extension (MV, TV during diastole and AV, PV in systole). Valve area (VA) were calculated as circle surface (pr^2). The Qp:Qs were calculated as: 1)Qp:Qs (V TV-MV) = (TVA X VTV) / (MVA X VMV), 2)Qp:Qs (v M TV-MV) = (TVA X v M TV) / (MVA X v M MV) and 3)Qp:Qs (v M AV-PV) = (AVA X v M AV) / (PVA X v M PV). Linear regression was used for statistical analysis. Results: We found the best correlation between scintigraphic and ECHO Qp:Qs calculations using V TV-MV (Qp:Qs = 0.3 + 0.8 X Qp:Qs-V TV-MV; r = 0.96; p< 0.01). ECHO Qp:Qs calculations using vM had worse correlation (vM TV-MV r=0.82, vM AV-PV r = 0.78, p<0.05). V TV-MV had also the highest specifity and sensitivity among all ECHO Qp:Qs calculation methods (Table 1). Conclusions: The most accurate echocardiographic method for Qp:Qs estimation is the method using maximal flow velocity. It had better correlation, specifity and sensitivity for detecting haemodynamic important ASDs than echocardiographic calculation using vM.

P480

Velocity of flow propagation, although influenced by heart rate and maturational changes can detect diastolic dysfunction in children

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Background: Velocity of flow propagation (Vp) has the potential of being influenced by age and heart rate (HR). Our objective was to determine accuracy of Vp in detecting diastolic dysfunction in children of varying ages and HR. Methods: We studied 97 normal children (age: 1 day-18years) and 8 children (age 3.8-10 years) with severe dilated cardiomyopathy (DCM). In DCM patients M-mode fractional shortening was less than 15 % (range 9-14%). Vp was calculated from slope of first aliasing velocity of color M-mode Doppler during early filling. Deceleration time (DT) of mitral E wave was measured from its peak to baseline as an additional noninvasive index of diastolic function. Results: Vp tended to decrease with increasing age & increase with increasing HR. However, correlation of Vp with age and HR was weaker compared to DT, suggesting Vp was less influenced by age and HR. (See table). Despite the influence of age and HR on Vp, it detected diastolic dysfunction easily in DCM group, showing much lower values of Vp in DCM versus controls (See figure). To reduce the influence of age and HR, Vp was measured in a younger group (0-3 years) and an older group (>3-18 years) of children. Even when children with DCM (ages ranging from 0.8 to 10 years) were compared only with those in older age group (>3-18years), Vp remained significantly reduced. Conclusions: Despite a weak correlation with HR and maturational changes in age, Vp accurately detected diastolic dysfunction in children of varying ages and HR. Further studies on patients with more subtle degrees of diastolic dysfunction, will help to establish the usefulness of Vp in children.

P481

Effects of continous ambulatory peritoneal dialysis on left ventricular dimensions and systolic functions

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To investigate the effects of continuous ambulatory peritoneal dialysis (CAPD) on left ventricular dimensions and systolic functions, we followed up sixteen children with end-stage renal failure with echocardiography for 12 months. The study population consisted of nine girls and seven boys. Their mean age at the entry of the study was 11.3 + 2.2 years (range: 7-14 years). The aetiology of the end-stage renal failure was vesicoureteral reflux in seven, chronic glomerulonephritis in five, pyelonephritis due to urolithiasis in two and unknown in two patients. Echocardiographic studies were performed before the beginning of the CAPD program and 3rd, 6th, 9th and 12th months of CAPD. End-diastolic and end-systolic diameters of the left ventricle, the thickness of the interventricular septum (IVS), and the left ventricular posterior wall (LVPW) were measured by M-mode echocardiography. Ejection fraction (EF), fractional shortening (FS) and left ventricular mass index were calculated according to guidelines of American Society of Echocardiography. At the end of follow-up period (12 months) there was no statistically significant changes in any of the parameters. At the beginning of the study the mean left ventricular end-diastolic diameter was 40.8 mm, end-systolic diameter was 24.9 mm, the thickness of IVS was 6.7 mm, the thickness of LVPW was 7.3 mm. EF was 68 %, FS was 39 % and LVMI was 105.9 g/m2. At the end of follow-up left ventricular end-diastolic diameter was 41.6 mm, end-diastolic diameter was 24.6 mm, the thickness of IVS was 7.6 mm, the thickness of LVPW was 6.9 mm. EF was 70 %, FS was 38.9 % and the LVMI was 98.8 g/m2. In conclusion, CAPD seems to be effective for preserving left ventricular mass and systolic functions in children with end-stage renal failure.

P482

Effects of coronary inflow and outflow pressures on left ventricular function: implication for the fontan operation

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Increased right atrial (RAP) and subsequently coronary venous pressure after Fontan operation may alter coronary perfusion and cause left ventricular dysfunction. The matching between coronary perfusion pressure (CPP) and myocardial contractility and the influence of an elevated RAP were investigated in a canine model (n=8) with pressure-controlled perfused coronary arteries. Left ventricular hemodynamic variables were measured by a pressure-volume conductance catheter, the slope of the end-systolic pressure-volume relationship (Ees) was calculated. First, Ees was assessed under normal conditions and under Fontan circulation at a CPP decreased stepwise from 240 to 45 mmHg. Then, Ees was assessed at CPP=90, 75 and 60 mmHg with a stepwise increase of RAP from 9 to 21 mmHg. The relationship between CPP and Ees could be described by biphasic J-shaped curves which were nearly identical in normal hearts and under Fontan circulation. While above a critical CPP (72+/-8 mmHg vs. 81+/-8 mmHg; n.s.) the changes of CPP did not affect Ees, below this level the decrease of CPP resulted in a progressive decrease of Ees. The progressive increase of RAP did not influence Ees at CPP=100 mmHg, led to a moderate decrease of Ees at CPP=75 mmHg and a severe decrease at CPP=60 mmHg (see Fig., *p<0.05). In conclusion, Fontan circulation per se does not impair the preload-contractility relationship. The effects of RAP are dependent on actual CPP: at a lower CPP with net perfusion pressure (CPP-RAP) below the critical CPP, an increase of RAP results in a subsequent decrease of contractility.

P483

Coronary flow-velocity dynamics in congenital heart disease

Hamaoka K., Tajirma K., Kawakita A., Oka E., Ozawa S., Yamamoto Y., Iwaoki N., Sakata K., Hirano T., Shiraishi I., Ito T.
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Coronary Flow-Velocity Dynamics in Congenital Heart Disease. Hamaoka K., Tajirma K., Kawakita A., Oka E., Ozawa S., Yamamoto Y., Iwaoki N., Sakata K., Hirano T., Shiraishi I., Ito T. Division of Pediatrics, Kyoto Prefectural University of Medicine, Kyoto, JAPAN. Myocardial ischemia in the hypertrophic heart is clinically important as an exacerbation factor to progressive myocardial damage. In order to assess the myocardial ischemia in congenital heart disease (CHD) with significant myocardial hypertrophy, we examined the coronary flow-velocity dynamics using an intracoronary Doppler guidewire at rest and during the ATP-induced hyperemic response (coronary flow reserve: CFR) in 36 patients (mean age: 4.0+/-3.8 years) with CHDs (TOF 13, TGA 1, PA 1, DORV 1). The data were compared with the age-matched normal data previously reported. In the coronary flow-velocity waveforms, a significant reverse-flow pattern was noted in 5 patients (31%) of 16 in the LAD and in 8 (50%) in the RCA. Furthermore, in the younger

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

group under 5 years, significantly higher or lower values for diastolic-to-systolic velocity ratio (DSVR) were noted in 7 patients (58%) of 12 in the LAD, and in 8 (67%) in the RCA. In the older group over 5 years, abnormal DSVRs were noted in 3 patients of 4 in the RCA, but only one in the LAD. In the younger group, significantly lower values for CFR were noted in 6 patients (57%) of 12 in the LAD, and in 9 (75%) in the RCA. In the older group, a reduced CFR was noted in 2 patients of 4 in the LAD, but only one pre-operative patients except three post-operative patients in the RCA. This study show that abnormalities in the coronary microcirculation contribute to the pathophysiologic effects in CHD.

P484

Blood pressure changes after repair of aortic coarctation

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There is no proven cause for the common problem of systemic hypertension after the repair of aortic coarctation. We hypothized that wave-reflection and resistance changes are partially responsible for this development. We examined 36 patients (mean age 9,9 years) (Goal without significant re-coarctation performing an echocardio-graphy and MRD. We did in all an echocardiography and an ergometry (ergo). We calculated the stiffness of the ascending aorta (Aoa). We obtained in 20 of these patients a 24-hour-holter blood-pressure (BP) monitoring. Values above the age appropriate 95 percentile were thought to be hypertensive. Systemic hypertension was more often at night (systolic 18/20, diastolic 8/20) than during daytime (systolic 12/20, diastolic 2/20). The day-to-night changes were smaller than normal. The BP-amplitude was increased (tab). 15 patients had an exercise induced systolic hypertension, often combined with a diastolic hypertension. There was no correlation between age of coarctation repair, op-technique, stiffness of Aoa, time since operation or sex and blood-pressure, day-BP (+/-STD) night-BP (+/-STD) ergo (+/-STD) Norm 47,7 (4) mmHg 52,1 (6,5)mmHg 101,8 (24,7)mmHg Coa 59,4 (10,2)mmHg 57,2 (7,8)mmHg 137,8 (35) mmHg p <0,0001 0,009 <0,0001 We found a pathologic bloodpressure (hypertension or increased BP-amplitude) in nearly all patients. This finding fits a model of wave-reflection and changed resistance, other influences have also to be kept in mind in individual patients.

P485

Influence of afterload on the midwall stress-velocity relationship

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We wanted to study the differences between endocardial and midwall (mv) stress-velocity relationships to assess contractility at low afterload. In 12 piglets (5–6 weeks) afterload was manipulated by aortic balloon occlusion (BO) and sodium nitroprusside infusion up to 3 mg/kg/min (SNP). The relationship between velocity of circumferential fiber shortening (VcFc) and end-systolic wall stress (ESS) was linear above 50 g/cm2 (VcFc = 1.1627–0.0018ESS); with a much steeper slope below 50 g/cm2 (VcFc = 1.9993–0.0156ESS). The relationship between mVcFc and ESS was linear above 30 g/cm2 and almost afterload independent (mVcFc = 0.6572–0.0004ESS); but below 30 g/cm2 the slope became again steeper (mVcFc = 1.0395–0.0122ESS). Conclusion: if using midwall shortening indexes, the stress-velocity relationship becomes relatively afterload independent over a wide and clinically relevant range. However, as with endocardial shortening indexes, the slope of the midwall relationship becomes steeper at low afterload.

P486

Influence of ventricular morphology on aerobic exercise capacity in patients after the Fontan operation

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Objectives: To investigate the influences of ventricular morphology, hemodynamics, and clinical findings on exercise capacity in patients after the Fontan operation. Background: Determinants of exercise capacity after the operation remain unclear. Methods: Peak oxygen uptake (PVO2) was determined in 135 patients by exercise test and compared to hemodynamics and clinical findings. Patients were divided into 3 groups based on ventricular morphology: those with a right ventricle (group RV), a biventricle (group BV), and a left ventricle (group LV). Results: PVO2 was 58 ± 11% and correlated with peak heart rate ($p < 0.05$). Patients with atrioventricular valve regurgitation (AVVR) or hypoxia exhibited a low PVO2. After excluding such patients, although PVO2 did not correlate with hemodynamics, it correlated with age at the Fontan operation and daily activity ($p < 0.001$). PVO2 was higher in group LV (64 ± 9%) than in groups RV (57 ± 10%) and BV (53 ± 11%) ($p < 0.01$), and an inverse correlation between PVO2 and age at operation was demonstrated only in group RV ($p < 0.05$). Lower daily activity, groups RV or BV, AVVR, and hypoxia were associated with a lower PVO2, while a higher daily activity and being group LV were independent predictors of a higher PVO2 ($p < 0.05$). During 4.2 years follow-up a decrease in peak heart rate was related to that in PVO2 ($p < 0.05$) and group RV showed a decrease in PVO2 ($p < 0.01$). Conclusions: Ventricular morphology, daily activity, AVVR, hypoxia, and heart rate response are related to exercise capacity. Early Fontan operation may be beneficial in terms of exercise capacity, especially in group RV patients.

P487

A new technique to localize and quantify left ventricular outflow tract (LVOT) obstructions using a single arterial entry

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We describe a new technique to assess various types of LVOT stenosis using a single arterial puncture. A 6F long sheath is inserted percutaneously via the femoral artery to the ascending aorta. A 4F pigtail or multipurpose catheter with a straight guide wire (GW) is introduced through it and GW is manipulated to enter the left ventricle (LV). The sheath and catheter are then slid over it into LV. The catheter and the side arm of the sheath are connected to equidistant transducers to record simultaneous pressures. With continuous pressure recording, the sheath is slowly 'pulled back' millimeter by millimeter so the aorta over the catheter held stationary in LV. The catheter records LV pressure while the sheath records pressure curves from every millimeter of the LVOT to localize the obstructions and measure the gradient. Similarly the sheath can also be 'pushed in' to LV over the catheter recording pressures. These maneuvers are repeated to obtain diagnostic tracings without arrhythmias. Angiograms are then performed. Our technique was successful in all 6 patients (age 4–18 years, weight 12–60 kg) we studied without complication. The average peak gradient was 85 mm Hg (range 50–120). The pressure pattern was diagnostic of subvalvar stenosis in 4 (2 discrete tubular stenosis, 1 with additional valvar stenosis and 1 hypertrophic cardiomyopathy) and valvar stenosis in 2. We conclude that our new technique using a long sheath and a single femoral arterial puncture can localize and quantify LVOT obstructions precisely and repeatedly. This avoids additional arterial puncture or transseptal catheterization to obtain simultaneous pressures.

P488

Exercise tests with measurements of oxygen saturation; a method to differ between intrapulmonary and intracardially right to left shunts in patients with fontan circulation

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BACKGROUND. The development of increasing cyanosis by time in patients with Fontan circulation is a well-known complication and often leads to a significant morbidity. The cause can either be an intracardial right to left shunt and/or a pulmonary arteriovenous malformation (PAVM). Many of these patients may have almost normal saturation at rest, but drop markedly during exercise. MATERIAL. Twenty of the surviving patients operated between 1980–1991 were investigated in a follow up study. There were 10 women and 10 men. The age at the investigation was 16,9 yrs (11,6–31,2) and 12,0 yrs (4,6–14,2), respectively. The mean follow up time was 12,6 yrs. METHOD. All patients underwent cardiac catheterization and pulmonary angiography as well as bubble contrast echocardiography with selective injections of Haemaccel into the right and left pulmonary artery and into the TCPC-tunnel or the right atrium in the Fontan patients. Simultaneous TEE was done to detect the microbubbles. They also performed an exercise test with measurements of oxygen saturation. In 14 patients a catheter was introduced in the brachial or radial artery and in 6 patients we used percutaneous pulse oximetry. RESULTS. In 9 patients we found PAVM on bubble contrast echocardiography. Six of them also had an intracardial leakage. In 8 patients we found only leakage intracardially. When we compared the drop in saturation during exercise between these patients, we found a significant difference in patients with PAVM compared to patients without. Patients with PAVM had significant lower oxygen saturation both at rest 88% vs 95%, p<0,01 and during maximal exercise 78% vs 89%, p=0,01. CONCLUSION. Exercise tests with measurements of arterial saturation may indicate

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

the use of the right to left shunt in patients with Fontan circulation. This is important in the decision making.

P489

Are estimated indices of pulmonary vascular resistance (PVR)

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Purpose: To assess risk stratification for Fontan using PVR calculated from measured oxygen uptake (VO2) compared to PVR from predicted VO2, and other haemodynamic data. Method. 35 patients with a bidirectional glenn (BDG) underwent cardiac catheterisation prior to Fontan (wt 5.4–51.7kg, age 0–12yrs). PVR was calculated using directly measured VO2 (Delta-trac metabolic monitor). Cases were stratified into high-risk, PVR >4 wu.m2 (n= 6); moderate-risk, PVR 3–4 (n=6); low-risk, PVR <3 (n=23). This was also done using PVR calculations from predicted VO2 formulae by Lindahl, Lundell and LaHaye et al. respectively. Hemodynamic data, such as transpulmonary gradient (TPG) and pulmonary arterial pressure, were also investigated as alternatives. Results. Predicted VO2 values were consistently higher than measured (mean differences +20%, +57% & +23%), leading to an underestimation of PVR with mean difference from -0.62 to -1.57 wu.m2, consequently misclassifying between 5 and 9 of the 12 moderate or high risk patients as low-risk. No other hemodynamic data could reliably separate low-risk from high-risk subjects. TPG >7mmHg was 100% specific for elevated PVR, but only 33% sensitive (2/6 pts). Conclusion: In assessing risk of Fontan failure in patients with BDG, all predictive VO2 formulae lead to serious underestimation of true PVR. u/m2, and other measured data, including TPG, cannot be used as a reliable PVR surrogate.

P490

Increased angiogenic growth factor levels in cyanotic congenital heart disease

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Background. Previous studies demonstrated that expression of angiogenic growth factors is impaired in hypoxic models. However, little is known about these factors with cyanotic congenital heart disease. The purpose of this study was to examine the relationship between plasma levels of angiogenic growth factors (vascular endothelial growth factor (VEGF) and hepatocyte growth factor (HGF)) and the severity of cyanosis. Method. The study included 65 patients with cyanotic congenital heart disease and 61 normal controls. Age ranged from 1 day to 40 years (median 1.2 year) in the cyanotic group and from 5 days to 31years (median 4.8 years) in the control group. Mean systemic oxygen saturation was 80.6 7.3% in the cyanotic group and 98.1 0.5% in the control group. Plasma VEGF and HGF were measured using an enzyme-linked immunoassay. Results. In the control group, VEGF at the neonatal period was significantly elevated 215.5 150.5pg/mL, then rapidly decreased within 3 months after birth. After 3 months of age, VEGF level remained as a plateau. In contrast, such age dependency was not found in HGF. Although VEGF and HGF levels were not different between the cyanotic and control groups within 3 months after birth, the VEGF level in the cyanotic group after 3 months of age was significantly elevated compared to the control (149.10b vs 67.23pg/mL, p<0.0001). Moreover, the VEGF level was significantly negatively correlated with oxygen saturation (y=440.6–3.53x, R=0.67, p<0.0001) in cases more than 3 months old. In contrast, no correlation was found between HGF level and oxygen saturation, or between VEGF and HGF level. Conclusion: Although physiologically unelevated VEGF in the neonatal period is rapidly decreased under normal oxygen saturation, a higher VEGF level persists if systemic hypoxia is present. These findings may influence the development of systemic to pulmonary collateral arteries in patients with cyanotic congenital heart.

P491

Pressure recovery and pressure gradients in stenotic outflow tract lesions: a simultaneous doppler and catheter correlative study in pediatric patients

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Purpose. Despite good correlation in general, Doppler-catheter gradient relation shows substantial discrepancies, contributed mainly by pressure recovery phenomenon. Pressure recovery magnitude is highly dependent on stenosis geometry. We prospectively studied the effect of pressure recovery on Doppler-catheter gradient relation across the spectrum of outflow tract stenotic lesions in pediatric patients. Methods. Simultaneous double blind Doppler and catheter pressure gradients were prospectively measured pre- and post-intervention in 51 consecutive patients (age:median 12.2 mo, range 1–224 mo, wt:median 7.5kg, range 2.8–72kg) with isolated stenosis of aorta(n=19), aortic(n=17) and pulmonary(n=17) stenoses. They were compared before and after correcting for recovered pressure. Pressure recovery(aortic stenosis) was derived by 4V2 x [AVA/AOA x(1-AVA/AOA)], where aortic valve area(AVA) by continuity equation and ascending aortic cross-section area(AOA) were calculated. Pressure recovery for pulmonary stenosis was similarly derived. Result: Doppler peak instantaneous and mean gradients correlations with corresponding catheter gradients and peak-to-peak gradient for aortic(r= .84, .79, and .86 respectively) and pulmonary (r= .94, .89, and .92 respectively) stenoses were significantly (p< .05) higher than for aortic examination(r= .71, .75, and .71 respectively) due to less overestimation. Predicted recovered pressures for Doppler gradients for aortic and pulmonary stenoses compared well with observed Doppler-catheter gradient discrepancies(r=.93 and .94 respectively). Correlations between all Doppler and catheter gradients in aortic and pulmonary stenoses improved significantly after correcting Doppler gradients for recovered pressure(r=.84 vs r=.94, p< .05). Conclusions: Doppler predictions of catheter pressure gradients are more accurate for isolated stenotic valvular lesions than for stenotic vessel like aortic coarctation due to less pressure recovery. Incorporating recovered pressures in Doppler gradients significantly improves its correlation with catheter gradients in valvular stenotic lesions. Both observations are relevant in clinical management and in decision making for intervention for outflow tract stenotic lesions.

P492

The efficacy of pulmonary artery banding in CAVSD of Down's syndrome

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OBJECTIVE. Pulmonary artery banding (PAB) is frequently applied to CHDs of Down's syndrome as for pulmonary obstructive vascular disease progress rapidly. The efficacy of PAB is evaluated with the cardiac catheterization results of complete atrio-ventricular septal defect (CAVSD). MATERIAL: 15 patients of CAVSD completed intra-cardiac repair (ICR) were divided retrospectively into 3 groups: primary ICR/ Down's synd. (Gr-A), PAB&ICR/ Down's synd (Gr-B), and PAB&ICR/non- Down's synd (Gr-C). Each group contained 5 patients. PABs were underwent at averaging 4.5 month, primary repairs were at 11.8 month, and second-look ICRs were at 22.5 month. These cardiac catheterizations were performed before each surgical treatment and after ICR. RESULT: Comparing Gr-B and Gr-C, the value of Pp/Ps and Rp/Rs were significantly higher in Gr-B before ICR. In 2 patients of Gr-B, the values of Rp/Rs elevated after PAB. Although these data normalized after ICR, the Rp/Rs level was significantly higher in Gr-B. In Gr-A, the data were effectively decreased and normalized after ICR excluding the Rp/Rs. The value of Rp/Rs after ICR was not deferred from Gr-B. CONCLUSION: PAB prevents the progression of the pulmonary obstructive vascular disease effectively. But in Down syndrome, stricter follow up and earlier ICR is necessary even after PAB.

P493

Efficiency of monitoring the near-infrared spectrophotometry of the brain during pediatric cardiopulmonary bypass

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OBJECTIVE. Neurological events during cardiopulmonary bypass (CPB) remain a well-recognized complication of pediatric cardiac surgery. Monitoring of cerebral oxygenation during operation may be useful to diagnose cerebral hypoxia. Near-infrared spectrophotometry (NIRS) is one of method for non-invasive monitoring of cerebral oxygenation and hemodynamics. We measured regional cerebral oxygenation using NIRS in pediatric cardiac surgery. METHOD: We studied 8 children (mean age 33 months) undergoing cardiac surgery monitored NIRS (INVOS 5100 cerebral oximeter) during operation. Hemodynamic parameters were measured at each stage (pre-CPB, during CPB, after CPB). Lactate, glucose and hemoglobin saturation was sampled from artery and SVC and compared with

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

during CPB. RESULTS: mean NIRS data was 45.0 point in pre-CPB, 33.2 point in CPB before aorta clamp, 37.5 point in CPB after aorta clamp, 39.7 point in CPB after aorta de-clamp and 47.4 point after CPB. Cerebral oxygenation during CPB was significant decrease compared with pre-CPB ($p<0.01$). Acute cerebral oxygenation decrease changeful was shown when each cannulation was performed. Lactate and glucose were increase from during CPB and decrease after CPB. Lactate and glucose in SVC were no significant change compared with artery. Hemoglobin saturation in SVC was decrease during aorta clamp. CONCLUSIONS: The observations of cerebral oxygenation suggest the timing of hypoxic brain injury during CPB in pediatric cardiac surgery. The most changeful time of cerebral oxygenation was when especially each cannulation was performed. Therefore, We have to be careful to perform cannulation and the monitoring of regional cerebral oxygenation in pediatric cardiac surgery will be helpful for preventing cerebral hypoxia.

P494

Determination of exercise tolerance in cyanotic congenital heart disease

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The arterial oxygen saturation is an important determinant of exercise tolerance (ET) in cyanotic congenital heart disease (CCHD) but it is not clear whether there are other factors. This study was to correlate several factors to ET in CCHD. We performed exercise test in 66 patients, whose age ranged from 11 to 40 (m=22) years, by a ramp loading cycle ergometer and anaerobic threshold (AT) was determined. The percutaneous oxygen saturation (SpO2) at rest was linearly correlated to AT ($r=0.45$). Expected AT at any given SpO2 was defined as AT=0.354*SpO2 at rest -17.9. We divided the patients into 3 groups, measured AT was higher than upper limit of 95% CL (confidence limit) of expected AT (group A), near expected AT (group B), lower than the expected AT (group C). Ejection fraction, pulmonary artery hemodynamics, hematocrit and mean corpuscular volume were not significantly different among the 3 groups. SpO2 at AT was 69+/-9% in group A, 72+/-11% in group B, and 76+/-6% in group C (NS). The differences of SpO2 between at rest and at AT were 16+/-6% in group A, 13+/-7% in group B, and 8+/-5% in group C. The differences in group A and group B were significantly greater than that in group C ($p<0.01$ and $p=0.04$, respectively). Minute ventilation was largest in group A (m=31.9 l/minute) and smallest in group C (m=19.6 l/minute). In conclusion, exercise capacity in CCHD is largely determined by rest SpO2 as reported before, and taking this into account, ventilatory capacity and ability to increase SpO2 during exercise are also very important factors. This study implies the importance of regular exercise to maintain physical ability in CCHD patients.

P495

Left ventricular hypertrophy in systemic hypertension in children

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Background: The aim of this study was to determine left ventricular function in different patterns of left ventricular hypertrophy and geometry in children with systemic hypertension. Methods: The study was performed in 52 children aged from 6 mo to 18 years with mild to moderate hypertension. Control group consisted of 30 healthy children. From m-mode echocardiography posterior wall thickness, interventricular septum, left ventricular mass, left ventricular mass index, relative wall thickness, end-diastolic volume, end-systolic volume and end-systolic wall stress were calculated. Results: We confirmed statistically significant differences in LVM between control group and in patients with systemic hypertension. On the basis of complete cardiological investigation in children with hypertension we classified hypertensive cardiomyopathy. Based on the echocardiographic left ventricular mass index and relative wall thickness patients were classified into normal geometry(15 children) concentric remodeling(4 children), concentric hypertrophy (16 children), and eccentric hypertrophy (17 children). End-systolic, end-diastolic and stroke volumes were highest in eccentric hypertrophy and lowest in concentric remodeling. End-diastolic volume/body surface area was highest in eccentric hypertrophy and lowest in concentric remodeling. The highest values of end-systolic stress (afterload) were noted in normal geometry and the lowest ones in concentric remodeling. Total peripheral resistance was highest in concentric remodeling and lowest in eccentric hypertrophy (2801 vs 1683dyn x cm-5). Ejection and shortening fraction remained within normal limits. Mean systolic ejection rate was highest in eccentric hypertrophy and lowest in concentric remodeling. Conclusions: On the basis of echocardiographic analysis hypertensive cardiomyopathy in children is divided into four groups.

P496

A method of vascular impedance spectrum analysis for identifying the location and magnitude of wave reflection in the aorta

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Objective: An accurate description of cardiovascular physiology depends on an understanding of the role of wave reflections in ventriculoarterial interaction. There is continued debate in the literature concerning the location and magnitude of reflections in the arterial vasculature. We present an alternative approach to investigating these wave reflections. This technique utilizes the Fourier transform of the input impedance to yield the ideal impulse response of the arterial system in the time domain. We demonstrate theoretically and with computer simulations that the technique allows the determination of locations of significant reflection sites and the magnitude of the reflected waves. The usefulness of the technique is demonstrated in a newborn piglet model and in children with Hypoplastic Left Heart Syndrome (HLHS) following aortic reconstruction. Methods: Four newborn piglets underwent cardiac catheterization. Baseline instantaneous pressure, flow, and electrocardiographic data were measured in the ascending, thoracic, and abdominal aorta. Subsequently, a balloon catheter was used to occlude the aorta at a known distance from the aortic valve to create an artificial reflection site. The data obtained was fit to our theoretical model and the magnitude and location of reflection sites were predicted. Following the piglet study, we applied the analysis to catheterization data obtained from children with HLHS (n=3). Results: Using this technique we were able to accurately predict the location of the imposed reflection site in the piglet model. Applying the same analysis to the study of children with HLHS yielded reflection sites consistent with those predicted. Conclusions: This analysis may be better suited for studying reflections in the human arterial system than traditional analysis. It may be useful for studying the vascular physiology in children with congenital heart disease.

P497

Endothelin-1 can influence the early outcome in neonates with hypoplastic left heart syndrome after the Norwood procedure

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Purpose: The operative outcome of the Norwood procedure for hypoplastic left heart syndrome (HLHS) is still not satisfactory and the mortality rate is particularly high in early postoperative period. Careful ventilatory and pharmacological modulation of the ratio of pulmonary to systemic vascular resistance and optimization of the heart work are the crucial elements of the perioperative management. Endothelin-1 (Et-1) is the most potent vasoactive peptide, which can influence both pulmonary and systemic circulation including coronary circulation in an age dependent fashion. Method: In a prospective study we analyzed the perioperative plasma Et-1 concentration in 24 neonates with HLHS after the Norwood procedure. Blood samples were collected simultaneously with hemodynamic assessment before the operation, during cardiopulmonary bypass, 2, 6, 12 and 24 h postoperatively. Plasma Et-1 levels were measured in arterial blood by radioimmunoassay. The results were compared with the controls (12 neonates with transposition of great arteries (TGA) after the arterial switch operation, where the plasma Et-1 level was assessed according to the same protocol). Results: The peak plasma Et-1 levels were observed 6–12 hours postoperatively in both groups. The mean plasma Et-1 level was significantly higher in neonates with HLHS as compared to children with TGA (46.89 pmol/l versus 29.09 pmol/l, $p<0.05$) and the maximal level was significantly higher in HLHS neonates (93.33 pmol/l versus 56.03 pmol/l, $p<0.05$). Conclusions: High levels of Et-1 may affect the early postoperative course in children with HLHS after the Norwood procedure probably due to imbalance between pulmonary and systemic resistance.

P498

Skeletal muscle oxygenation evaluated by NIRS during exercise in patients after Fontan operation

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Aim and Subjects: The anaerobic threshold (AT) of patients after Fontan operation gradually decreases with aging. We investigated skeletal muscle oxygenation using a near infrared spectroscopy (NIRS), respiratory function and AT in 15

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Fontan patients whose age ranged from 12 to 34 years. Method: After evaluating vital capacity (VC), each patient underwent cardiopulmonary exercise test on a cycle ergometer using ramp protocol to evaluate anaerobic threshold, and NIRS was performed on the quadriceps femoris throughout the exercise. Result: The value of AT was correlated negatively with patient's age (r=0.64), and positively with %VC (r=0.80). %VC was negatively correlated with age. A sudden increase in deoxy-hemoglobin fraction on NIRS during exercise, detected in 12 patients, preceded to AT point in time by 81 sec in an average. The ratio of oxy-to deoxy-hemoglobin at AT point did not show any significant correlation with AT itself and age of patients. Comments and Conclusion: The decrease of %VC with aging is one of the major factors of low AT of older patients. The lack of relation between AT and the state of muscle oxygenation may suggest that there is biochemical and physiological adaptation in the skeletal muscle in Fontan patients.

P499

Total primary occlusion as a major criterion in the ductal detachable coil closure

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Introduction: In the last years Coils has been adopted as the preferable device for small-medium ductal occlusion. Detachable Coils diminished the risk of undesired embolizations maintaining a high rate of success and a low procedure and fluoroscopy time. Although residual shunt: a rare and might spontaneously close in the follow-up, its appearance is still a major of concern. Since April 98 our policy was to attempt total occlusion whenever possible, and without regards of the number of Coils required. 59 patients underwent ductal occlusion with this policy (group A) and its results are compared with 99 previous patients (group B). Material and Methods: age (years), ductal anatomy, procedure and fluoroscopy time (FT and TT), Coil number (CN) and rate of complications are compared between the two groups. Also residual shunt in group B is repeated. Medium stalles ductus diameter was 1.8 mm in both groups. Results: Table. Residual shunts: in group A: 3; in group B: 38% immediate, 26% 24 hours, 12% 6 months. Complications: major (Group B), 2 Coil embolization, 1 hemolisis. M: none related to anesthesia or vascular access. Conclusions: 1) primary total Occlusion of the ductus may be a proper approach to the ductal closure procedure with Detachable Coils. Therefore FT, TT and rate of complications are similar to the classical approach. 2) More studies are required to identify which patients might benefit of waiting for spontaneous closure of the residual post-Coil shunt.

P500

Effect of the surgical modification on the flow characteristics of systemic veins and pulmonary artery after Fontan operation – comparision by direct intravascular Doppler measurements

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The effect of the surgical modification and the influence of respiratory effort on the systemic venous and pulmonary arterial flow patterns were investigated by examining 21 patients with Fontan circulation (atriopulmonary connection APC: 9, Total cavopulmonary connection TCPC: 12). A new method of direct intravascular Doppler measurements was employed to get Doppler spectrals at SVC, IVC, hepatic vein(HV) and the pulmonary artery(PA) with a Flowtap system(Medtronics, USA) under simultaneous respirometer recording. Flow characteristics including maximum(Vmax) and minimum velocity(Vmin), velocity-time integral(VTI), pulsatility index(PI), net antegrade flow integral(NAFI) and respiratory variability index(RVI) were compared and the following results were obtained: 1. SVC flow patterns: APC group has higher PI(0.99±0.44 vs 0.42±0.1, p<0.05) and is more frequently associated with reversal flow(80% vs 10%, p<0.01). Vmin is higher in the TCPC group(0±14 vs 20±5cm/s, p<0.01). 2. IVC flow patterns: velocities and PI were higher in the APC group(1.34±0.07 vs 1.02±0.18, p=0.05). RVI of NAFI is higher in the TCPC group(0.42±0.08 vs 0.56±0.12, p<0.05). 3. HV flow patterns: TCPC group has higher NAFI especially in the inspiratory phase(4.3±1.7 vs 16.1±6.3, p<0.01). RVI of NAFI is also higher in the TCPC group(0.93±0.27 vs 1.25±0.19, p<0.05). 4. The difference of RVI between the two groups was not observed in the PA. We concluded that 1. TCPC is more dependent on respiration but the respiratory effect is reduced in the PA. 2. TCPC has higher NAFI which indicates a more efficient circuit and the advantage is augmented especially in the inspiratory phase. 3. The pulsatility by atrial contraction in APC does not significantly support the PA flow. 4. APC is associated with higher velocity and flow change in the IVC and HV, and the surveillance of the long-term effect on splanchnic circulation is warranted.

P501

Cardiac pathophysiology in a transgenic model of hypertension is due to elevated vascular impedance

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Background. Despite the advances in pharmaceutical treatment for hypertension, the morbidity and mortality of this disease remains high. The transgenic TGR (mRen2) 27 rat is a valuable model for the study of hypertension. This monogenetic model expresses a profound dysregulation of renin-angiotensin system leading to malignant hypertension. Our aim in this study is to evaluate the nature of the cardiac pathophysiology in this model in relation to the peripheral vascular resistance, vascular impedance (pulsatile) and external left ventricular power. Methods: Seven rats from each group were studied. Echocardiographic data included LV end diastolic diameter and posterior wall thickness, LV end systolic diameter and LV mass. After an abdominal incision, the abdominal aorta was isolated and skeletonized. A Millar pressure sensor was inserted intravascularly. The aortic blood flow was measured by transonic flow probe. The flow and pressure waves were digitized and analyzed by Fourier series to obtain vascular impedance, peripheral vascular resistance and LV external power. Results: LV mass and LV wall thickness were significantly higher in TGR rats. No significant difference in LV end diastolic diameter and shortening fraction. Pericardial effusion occurred in 4 of 7 TGR rats but not in the controls. The characteristic impedance and LV external power were significantly higher in TGR rats. The peripheral vascular resistance and LV systolic function were not significantly different between the two groups. There was a correlation between the increase in LV mass and the increase in the characteristic impedance, the LV external power and the LV wall thickness. Conclusion: The vascular characteristic impedance is the major determinant of the cardiac changes in this model of hypertension, while the peripheral systemic resistance has no significant role in the cardiac pathophysiology. The finding of pericardial effusion in transgenic rats is significant and might be due to decreased lymphatic drainage secondary to increased interstitial pressure. This rat model can be very useful in future therapeutic studies to reverse the end organ changes.

P502

Characterizing a neonatal porcine left-to-right shunt model

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A neonatal animal model was developed by stenting the patent ductus arteriosus (PDA) shortly after birth. The purpose of the study was to characterize the physiological effects of the porcine neonatal left to right shunt model. The study involved two groups of newborn pigs: Group 1 (n=6) Stented PDA (L-R shunt) and Group 2 (n=8) Unstented PDA (Control). Group 1 underwent the insertion of a stainless steel Palmaz stent (J&J), dilated to 4 mm at diameter in the PDA by a transvenous approach within 24 hours after birth. Daily weights, respiratory and heart rates were collected for 2 weeks, at which time the LV dP/dt and L-R shunt was measured using the indocyanine indication dilution method. Echocardiography measuring left ventricular systolic and diastolic dimensions (LVIDs, LVIDd) with calculations of shortening fraction (FS) was used as an index of function for all pigs. The data was expressed as means ± SEM and analyzed using ANOVA repeated measures and unpaired t-test, with P<0.05 considered significant. Over the 2 week period, Group 2 showed a significant greater increase in percent weight gain, 60.6 ± 3.8%, P<0.001, as compared to Group 1 with 50.2 ± 1.4%. Heart and respiratory rates showed equivalent significant mean differences, P<0.001; with 226.8 ± 2.7 beats per minute for Group 1 and 208.7 ± 3.8 bpm for Group 2, and 95.2 ± 7.2 breaths per minute for Group 1 and 63.7 ± 3.2 breaths per minute for Group 2 respectively. The dye curves in Group 1 showed a mean Qp/Qs of 2.3/1 ± 0.2. The mean LVdP/dt was 988.4 ± 87.2 in Group 1 and 730 ± 153.8 in Group 2. Echocardiographic data including LVIDd, LVIDs and FS all showed significant differences (P<0.05) between the stented and control groups. LVIDd in Control Group 2 measured 19.8 ± 1.5 cm as compared to Stented Group 1 which measured 30.1 ± 1.5 cm. Concomitantly LVIDs showed similar differences, with Group 2 measuring 14.4 ± 1.0 cm and Group 1 measuring 25.9 ± 1.2 cm. The Group 1, the L-R shunt greatly reduced shortening fraction to 15.6 ± 1.7% as compared to 35.2 ± 3.5 % in

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Group 2 We conclude that significant physiological changes result from L-R shunting in the stented pulmonary PDA are reflected in weights, respiratory and heart rates, echocardiography data and hemodynamic values and as such will be an excellent model with the pleura and pericardium intact for pharmacological studies of heart failure in the young

Genetics Basic Research, Genetics Clinical Research

P503

Hypoxic hearts: effective myocardial protection provided by an anti-apoptotic cell-permeable peptide

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Activation of JNK, a mitogen-activated protein-kinase, is responsible for apoptosis in several models of cellular damage. We studied the effects in isolated rat hearts exposed to hypoxia-reoxygenation injury of JNK1, a recently synthesized peptide containing TAT-10-aminoacid and 20-aminoacid minimal JNK1 Akt- substrate (oxygen content =100%), hearts were exposed to 60 minutes hypoxia (oxygen supply =10% of the baseline), followed by 20 minutes reoxygenation (oxygen content =100%). Coronary flow remained constant (=15 ml/min) through the experiment. Three groups of hearts were evaluated: controls without peptide (n=8), controls perfused with a solution of TAT only (n=6) and hearts treated with JNK1 (1 mM given over 2 minutes) (n=12). Myocardial function were evaluated as % recovery of pre-hypoxic values (mean±SD). TUNEL assays were utilized to identify on tissue slides apoptotic cells, calculated as number of TUNEL-positive cell nuclei/(number of TUNEL positive cell nuclei + number of total cell nuclei)X100. No functional difference was found between control groups without and with TAT peptide. Pooled controls (n=14) were compared with the hearts (n=12) treated with JNK1. The recovery of diastolic as well systolic function of JNK1 treated hearts was significantly better than controls. LVEDP = 135 ± 17% vs 28% = 42% ($p<0.005$), -dp/dt = 78 ± 3% vs 64 + 5% ($p<0.05$), +dp/dt = 92 ± 4% vs 66 ± 5% ($p<0.001$) and LVDP×HR = 85 ± 4% vs 63 ± 4% ($p<0.001$). Over 90% of apoptosis cells were demonstrated by TUNEL assays in untreated hearts vs 5% of apoptotic cells in JNK1-treated hearts. JNK1 provides evident anti-apoptotic properties and effective myocardial protection against hypoxia-reoxygenation damage in isolated perfused rat hearts. This cell-permeable peptide could have a substantial impact in the management of cyanotic heart malformations.

P504

Cervical origin of the subclavian artery – a specific marker for monosomy 22q11

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Anomalous origin of the subclavian artery from the common carotid artery has been termed 'cervical origin of the subclavian artery' (COSA). The purpose of our study was to investigate the association between COSA and monosomy 22q11. In our 2 centers 151 patients with conotruncal malformations were tested for monosomy 22q11. 43 pts. were positive, 68 pts. were negative (interrupted aortic arch (IAA): 11 pos./17 neg.; Ts-var.: 3 pos./46 neg.; tetralogy of Fallot: 16 pos./8 neg.; truncus arteriosus: 5 pos./17 neg.). Among the 43 children with monosomy 22q11 COSA contralateral to the aortic arch was present in 9 pts (21%): 5 pts. had IAA, 3 pts. pa vsd, and 1 pt. had tetralogy of Fallot. COSA wasn't found in any patient without monosomy 22q11. According to our study cervical origin of the subclavian artery (representing maldevelopment of the 4th aortic arch) is rather frequent in children with monosomy 22q11 especially in pts. with IAA. Since we did not find COSA in any patient without monosomy 22q11, COSA appears to be a specific marker for the chromosomal anomaly.

P505

Abnormalities of the subclavian arteries and monosomy 22q11 in children with interrupted aortic arch (IAA)

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In pts. with monosomy 22q11 (M22q11) IAA is usually found in type B, representing unilateral maldevelopment of the 4th aortic arch. We reviewed our pts. for additional abnormalities of the subclavian arteries (SA). M22q11 was present in 11/28 children with IAA. All pts. with M22q11 had IAA type B. Abnormalities of the SA contralateral to the aortic arch were present in 8/11 children with M22q11. Cervical origin of the SA: 5 pts., aberrant SA: 2 pts. and dextrapositio of the SA: 1 pt. Only 2/17 pts. without M22q11 had abnormalities of the SA. The difference between children with M22q11 and the other pts. was significant regarding the total number of SA abnormalities ($p=0.003$) and regarding the presence of cervical origin of the SA ($p=0.002$). Abnormalities of the subclavian artery contralateral to the aortic arch are frequent (73%) in children with IAA type B and M22q11 suggesting bilateral impairment of the 4th aortic arch development. Cervical origin of the SA appears to be a specific anomaly in children with this microdeletion and IAA.

P506

Endothelial nitric oxide synthase gene polymorphism is positively associated with development of kawasaki disease

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BACKGROUND: Nitric oxide (NO) formed by endothelial constitutive NO synthase(eNOS) mediates endothelium-dependent vasodilation and antithrombotic action, exerting vascular protection against acute inflammation. Recent reports suggested positive association of eNOS gene polymorphism with essential hypertension and coronary heart disease in adults, indicating genetical susceptibility to heart and vascular diseases. We examined eNOS gene polymorphism in patients of Kawasaki disease and tried to relate this polymorphism to the development of Kawasaki disease and coronary artery aneurysm. METHODS: Blood samples were obtained from patients (54 cases) who had a history of Kawasaki disease with (10 cases) or without (21 cases) coronary aneurysms. The presence of coronary aneurysms was confirmed by two dimensional echocardiography or selective coronary angiography previously performed. Control blood samples were also obtained from healthy volunteers (51 cases). According to the methods previously established, we extracted genomic DNA from blood cellular components by guanidine thiocyanate method. The extracted genomic DNA fragments were amplified by the polymerase chain reaction (PCR) to determine the eNOS genotype. The oligonucleotide primers we used were primers flanking the 27-bp direct repeat region in intron 4 of eNOS gene previously reported to be linked to vascular diseases in adults. The PCR products were separated by electrophoresis to determine the eNOS polymorphism. RESULTS: The frequency distribution of the eNOS genotype for healthy controls was: b/b(46/51, 90%), a/b and a/a(5/51, 10%), respectively. eNOS genotype distribution among the patients of Kawasaki disease was: b/b(22/31, 71%), a/b and a/a(9/31, 29%) with a significant($p<0.05$)higher incidence of the a allele in Kawasaki disease than healthy control. However, there was significant difference of frequencies of the a allele of eNOS genotype between patients with (4/10, 40%) and without (5/21, 24%) coronary aneurysm in Kawasaki disease. CONCLUSION: This study suggests a genetical contribution of eNOS polymorphism to the development of Kawasaki disease.

P507

Genotypic and phenotypic comparisons between long QT syndrome and Brugada syndrome

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Background: Long QT syndrome (LQTS) and Brugada syndrome are two distinct human hereditary cardiac diseases known to cause ventricular tachyarrhythmias (torsade de pointes) and idiopathic ventricular fibrillation, respectively, which can lead to sudden death. Five mutant LQTS genes have been identified: KCNQ1 (LQT1), HERG (LQT2), SCN5A (LQT3), KCNE1 (LQT5), and KCNE2 (LQT6). Brugada syndrome is characterized by an electrocardiographic pattern consisting of right bundle branch block with ST-segment elevation in leads V1 to V3 without prolongation of the QTc interval. Mutations in SCN5A, encoding the subunit of cardiac sodium channel protein, are responsible for both syndromes. In order to compare the genotypic and phenotypic findings in both syndromes, we genetically screened SCN5A. Methods and Results: Two Japanese patients who were suspected of having LQT3 and three patients with Brugada syndrome were examined after informed consent. Autosomal sequencing of the sodium channel

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

blocker, procainamide (500mg IV), induced ST-segment elevation in the right precordial leads in the 3 patients with Brugada syndrome. Using single strand conformational polymorphism (SSCP), the 28 exons of *SCN5A* were analyzed and aberrant conformers were directly sequenced. We have already reported one of the LQT3 patients. This patient had a R1023Q mutation, and was treated with mexiletine. A novel missense mutation (G1742R) was identified in one patient with Brugada syndrome, who needed implantation of an ICD. DNA sequencing confirmed a G-to-A transition, leading to amino acid substitution of glycine for arginine 1743, located in the loop between transmembrane segments 5 and 6 of domain IV. Conclusion: In our study, each identified mutation of LQTS and Brugada syndrome was located on the same domain IV of the cardiac sodium channel gene (SCN5A). However, these mutations had divergent ECG phenotypes and clinical manifestations.

P508

Familial congenital heart disease

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The aim of this study was to estimate the family predisposition for congenital heart diseases. Although there has been a great progress in diagnosis and treatment of congenital heart disease their epidemiology is not known exactly yet. The recognition of the genetic factors is very important for genetic consulting. The analysis comprised 1600 patients who were under our care in the years 1990–1999. Ninety eight families were selected. These families were characterised by more than one family member who suffered from the congenital heart disease – altogether in 280 cases. Accordingly 4 kinds of families were selected: Group I– isolated congenital heart defects (60 families-143 cases), Group II – concomitant heart defects (20 families 50 cases), Group III- hypertrophic cardiomyopathy (5 families-12 cases), Group IV– genetic syndromes such as Long QT syndrome, Marfan syndrome, Holt-Oram syndrome, Noonan syndrome (13 families-75 cases). The analysis allowed us to estimate the risk of having next child with congenital heart disease and to select most endangered families. The emphasis was put on the necessity of broad genetic consulting especially of young patients who are planing to set up their own families. Molecular genetic diagnosis has been performed in group II and IV.

P509

Concurrence of supravalvular aortic stenosis and peripheral pulmonary stenosis in four generations of a family

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Background: Knowledge of the risk of occurrence of congenital heart defects in offspring of individuals with a congenital heart defect is important for genetic counselling. Isolated supravalvular aortic stenosis (SVAS) commonly is an autosomal dominant trait, it may also occur in the Williams syndrome (WS). Peripheral pulmonary stenosis can occur in the same individual with familial isolated SVAS. Methods and results: We describe seven affected individuals in one family: one had isolated SVAS, two had isolated PS and four had SVAS and PS. Two of them died before cardiosurgery: one as a small infant and one as a 62 year old man. Molecular genetic diagnosis was performed in each of them. Conclusions: Based on this family and review of literature, we suggest that SVAS is a form of arterial dysplasia encompassing PS in its spectrum, and the family history should be obtained very carefully even in neonates.

P510

First cases of familial arrhythmia,from antenatal sinus node dysfunction to atrial fibrillation in the elderly

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A few authors have been working on the genetic localization of rhythm abnormalities, e.g. atrial fibrillation (AF) or conduction abnormalities, e.g. bundle branch block or atrioventricular block. We report a particular pedigree where some patients presented with prenatal sinus node dysfunction, then nodal bradycardia and finally AF. Six out of twenty three family members were identified. Two of them (II5, III2) had to be implanted a pacemaker. The results of our investigations include: ECG, exercise testing, echocardiography and ambulatory ECG. This new entity is inherited in a dominant autosomal manner. This is the first ever report of a rhythm disorder involving four generations, with two prenatal manifestations of the disease and showing a progression from sinus node dysfunction to slow AF frequency. DNA-testing is under way to confirm or infirm a linkage on chromosome 10, as previously reported.

P511

Factor v 1691g-a and prothrombin 20210 g-a mutations in children with intracardiac thrombosis: a prospective study

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We performed this study to determine the association between intracardiac thrombosis and hereditary causes of thrombophilia, including the Factor V 1691 G-A and Prothrombin 20210 G-A mutations. Over a 3-year period, genetic risk factors were evaluated in 13 consecutive children (mean age 6.27±5.44) with intracardiac thrombosis, diagnosed cross-sectional echocardiography. Thrombi were localised in the left heart in four patients and right heart in nine patients. All children had predisposing factors for thrombus formation: Ventriculoatria. shunt for hydrocephalus (n=3), indwelling catheter for chemotherapy (n=5), cardiomyopathy (n=2), sepsis (n=1), hemicystinuria (n=1), Tetralogy of Fallot (n=1). Six of the 13 chidren with intracardiac thrombosis had heterozygote for FactorV 1691 G-A mutation. Three of these 6 children with FactorV 1691 G-A mutation had ventriculoatrial shunts for hydrocephalus, two children had cardiomyopathy and one had sepsis. Our patients did not carry the Prothrombin 20210 G-A mutation. In conclusion: we recommend that Factor V 1691 G-A mutations should be investigated in all cases of intracardiac thrombosis irrespective of whether or not another predisposing factor is identified.

P512

The role of the insertion/deletion (i/d) polymorphism of the angiotensin converting enzyme gene in left ventricular mass in children with congenital heart disease

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Background: The Insertion/Deletion (I/D) polymorphism of the Angiotensin Converting Enzyme (ACE) gene has been related to left ventricular mass (LVM) in adults. The association of the ACE genotype with LVM in children with congenital heart disease (CHD) is unclear. Methods: We studied 57 children: aortic stenosis (AS) n=9, coarctation (COA): n=11, ventricular septal defect (VSD) n=22, normal children (NL): n=15. The mean age was 5.4+/-8.0mo (AS), 5.2+/-5.0mo (COA), 6.1+/-9.6mo (VSD), 8.4+/-8.6mo (NL). The ACE genotype was determined by polymerase chain reaction. LVM indexed to the body surface area (LVMI) was determined echocardiographically. The relationship between diagnosis (Dx), LVMI and ACE genotype was determined with analysis of variance. Results: The ACE genotype distribution in the entire cohort matched previous studies: DD: 33%, DI: 47%, II: 20%. The mean age was: 8+/-18mo (DD), 35+/-49mo (DI), 20+/-52mo (II). DD children were younger than DI and II children (p=0.1). LVMI in normals was consistent with previous reports. LVMI was increased in all groups with CHD compared to normals: AS: 94.7+/-33 g/m2, COA: 63.5+/-29 g/m2, VSD: 81.2 +/- 14.7 g/m2 vs NL: 48.2+/-7.2 g/m2 (p<0.0001). LVMI was significantly higher in the DI genotype vs the DD and II genotype in AS (p=0.01) and COA (p=0.03). No relationship between ACE genotype and LVMI was found in the VSD or NL groups. Conclusion: Pressure and volume overload in children with CHD result in increased LVMI. Significantly higher LVMI was present in DI but not in DD children with AS and COA. DD children were younger and thin, the shorter duration of pressure overload may explain the lack of an association between LVMI and DD genotype. The role of the ACE genotype on LVMI in CHD remains unclear. Further studies are warranted.

P513

Analysis of five candidate genes in twenty-three patients with heterotaxia

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Introduction - The molecular mechanisms for the intricate left-right asymmetry at which a growing number of genes and cellular processes have been implicated have not been clarified. To assess their roles in the left-right axis development in humans, 5 specific genes were studied: ZIC3, zinc finger protein of the cerebellum (an X-linked transcription factor), Lefty A-B, a transforming growth factor (TGF)-b family of cell-signalling molecules (chromosomal location in 1q42), ACVR1B, the gene for human activin

receptor type IIB (chromosomal location in 3p22). WNT11, a WNT member of secreted signaling protein (chromosomal location in 11q13.5) and UVRAG UV radiation resistance associated gene (chromosomal location in 11q13). Materials and Methods:- We studied 25 patients, 12 females and 13 males (from 2 months to 45 years old) with typical left-right axis malformation, 10 with asplenia syndrome and 15 with polysplenia syndrome. We performed single-strand conformation polymorphism, direct sequencing of the polymerase chain reaction and restriction length fragment polymorphism, looking for mutations, and making genotypical and phenotypical correlation. Results:- We found a novel mutation of the ACVR IIB gene in one patient at position +7 of intron 2. The patient exhibited visceral heterotaxia with polysplenia and cardiac anomalies, such as single right ventricle and pulmonary stenosis. We also found a mutation of the WNT11 gene in another patient at position 1209 of a non-coding region. This patient exhibited heterotaxia with polysplenia, endocardial cushion defect, atrial septal defect and pulmonary stenosis. None of these mutations were found in 300 control subjects. Conclusion - Candidate genes involved in left-right axis development display mutations only rarely in humans.

P514

QT interval changes during face immersion in cold water in patients with LQTS1 and LQTS2

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Gene-specificity for electrocardiographic T wave morphology and for specific triggers for arrhythmic events have been described in the congenital long QT syndrome (LQTS). Facial immersion in water has been shown to prolong QT in non-familial LQTS. The objective of the study was to identify patients with LQTS1 or LQTS2 who might be at risk of cardiac arrhythmias during diving or swimming. 29 patients (15M; median age 14 years) with LQT51 (N=8) or LQTS2 (N=21) underwent facial immersion in warm (25 degrees C) and cold (10 degrees C) water, with continuous ECG recording. Six of 8 patients with LQTS1 and 8/21 patients with LQTS2 were using beta-blocker medications. There was no significant difference in age, resting heart rate, resting QT interval or QTc between the LQTS1 and LQTS2 patients. There was a significant decrease in heart rate during cold water immersion for LQTS1, and during both warm and cold water immersion for LQTS2 patients. The QT interval in LQTS1 increased by 36 +/- 17 msec (mean +/- SEM; p=0.01) during cold water immersion, and by 10 +/- 7 msec for LQTS2 (p=0.2). When comparing beta blocker users and non users with LQTS2, there was a significant difference in QT change during cold water immersion (-10 +/- 11 msec for users versus 22 +/- 8 for non users; p=0.03). Non-sustained ventricular arrhythmias occurred during warm and cold water immersion only in LQTS2 patients, none of whom had had a clinically documented ventricular arrhythmia related to diving or swimming. None of the LQTS1 patients who had experienced near-drowning while swimming had an inducible arrhythmia. QT interval increases significantly during cold water immersion in LQTS1 patients. LQTS2 patients not using beta-blockers show a small but significant increase, while in LQTS2 patients using beta-blocker the QT interval tends to decrease. Facial immersion in water is not sensitive for identifying LQTS patients at risk for clinically significant arrhythmias.

P515

Familial presentation of balanced translocation t(7;14) with split in elastin region

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Supravalvular aortic stenosis (SVAS) is the least common form of fixed aortic stenosis. In about 20% of cases there is present familial form of SVAS without characteristics of Williams syndrome. Sporadically there are report of balanced translocations with split in elastin region. About two-thirds of patients have additional cardiovascular abnormalities, including pulmonary stenosis, coarctation, patent ductus arteriosus, and mitral insufficiency. First patient came to us because of the presence of a systolic murmur. Echocardiography revealed SVAS with high gradient (80 mmHg), intracavitary stenosis of left ventricle outflow tract (gr. 40 mmHg), and bilateral peripheral pulmonary stenosis (gr. 40 mmHg), with mitral insufficiency grade I. Child had normal mental development, without characteristics of Williams syndrome. Heart catheterization confirmed echocardiographic findings, with narrowed segments in LVOT and supravalvular region. Right ventriculogram showed diffuse narrowing of both pulmonary artery. Child was operated due to SVAS and LVOT by Doty procedure and resection of LVOT stenosis. Several patients in his brother, which also had systolic murmur, and echocardiographycally almost identical findings as first. LVOT obstruction. On catheterization findings were equal, except stenosis at origin of left common carotid artery. Child was also operated by Doty procedure. The conventional chromosome analysis of two brother and mother by high resolution banding unexpectedly revealed a balanced translocation between chromosomes 7 and 14. Subsequent FISH analysis with WSCR and centromeric 7 probes showed that the break point has split the elastin region. The mother and her two sons carry the same translocation. Mother was echocardiographically evaluated, but no significant stenosis were registered. The karyotype of brothers is: 46,XY, t(7;14)(q11.23;q12) als t(7;14)(D7Z1+;EL N+)mat.

P516

Familial truncus arteriosus : a possible autosomal recessive trait

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Truncus arteriosus (TA) is an uncommon type of congenital malformation. It accounts approximately 1% of all congenital heart diseases. The defect occurs sporadically but the prevalence of 22q11 deletion among patients with TA and conotruncal defects is well known and is estimated at 8 - 17 %. Genetic studies of babies born with TA is done routinely looking for microdeletion of 22 or 10 chromosome abnormalities. We present 6 cases of TA that were born in 4 closely related families. Their full genetic studies were normal. Family No.1 had 2 affected babies (1 male and 1 female) who were born in 1989 and 1995 respectively. They have 4 other normal children. Family No. 2 had 2 affected children (1 male and 1 female) born in 1981 and 1984 respectively. They have 3 other normal children. Family No 3 had one affected male infant born in 1998. Family No.4 had one affected male infant born in 1998 and another healthy male child. All families are cousins and the parents of all the affected children are double cousins. The data shows that TA may be due to a mutant gene that we were not able to identify. This gene may be transmitted as an autosomal recessive trait. A multifactorial inheritance can be another possibility. Amplification of this defect may have occurred due to cousin marriages.

P517

Postmortem molecular analysis of SCN5A defects in sudden infant death syndrome

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Background. The causes of sudden infant death syndrome (SIDS) remain elusive. Occult long QT syndrome (LQTS) has been raised as a diagnostic consideration. Since patients with LQT3 have an increased frequency of cardiac events during sleep and with the recent case report of a sporadic SCN5A mutation in a near-SIDS infant, the cardiac sodium channel gene, SCN5A, has emerged as a candidate gene for SIDS. Methods: 95 cases of SIDS or possible SIDS were identified by the Arkansas State Crime Laboratory between September 1997 and August 1999. Necropsy tissue was collected prospectively and frozen in 90 cases. Genomic DNA was extracted and subjected to SCN5A mutational analyses by PCR amplification, denaturing high performance liquid chromatography and DNA sequencing. Missense mutations were incorporated into the human heart sodium channel alpha subunit, transiently transfected into HEK cells, and characterized with the whole cell patch clamp technique. Results: 4 of 95 cases of SIDS possessed SCN5A mutations. A 6 week old male had an A997S missense mutation in exon 17. A 9-base pair intronic deletion 4395+17—25delACCTGAGGG was present in a 2-month old male and a 2-month old female. Finally a 3-month old male had an R1826H mutation in exon 28. These 3 distinct mutations were absent in 200 controls (400 chromosomes). Functionally, the R1826H-SCN5A mutant channel was characterized by slower recovery, negative shift in voltage inactivation, and a 2–5 fold increase in late sodium current (n = 5). Conclusion: This study represents the first molecular autopsy for cardiac channelopathies in a prospective population-based cohort of SIDS. 4.3% of this SIDS cohort had an identifiable SCN5A channel defect and suggests that mutations in cardiac ion channels may provide a lethal arrhythmogenic substrate in infants at risk for SIDS.

P518

Predictors of ventricular dysrhythmia in patients with Marfan syndrome

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Sudden death without aortic dissection occurs in Marfan syndrome. The

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

frequency of arrhythmias in these patients remains poorly defined. We reviewed clinical data, echocardiograms, ECGs and Holters where available (30) in 60 patients with Marfan syndrome. Age at diagnosis was 10.9 (0–51) years with follow-up of 6 (1–16) years. There were 2 arrhythmogenic deaths and no deaths from aortic rupture. Aortic root replacement was performed in 4 (7%). Medication for prophylaxis of aortic dissection was used in 47 (80%). Aortic root enlargement was present in 55 patients (95%) measuring 130+/-21% predicted. MVP was present in 31 (53%) with associated mild MR in 15 (25%). Mild AI was present in 5 (9%) and moderate AI in 2 (3%). LV dilation was present in 32 (54%) with a mean LV z-score of 2.2+/-1.7. ECG revealed QTc prolongation in 9 (15%) patients. Ventricular ectopy was present on Holter in 15 (50%) and noted to be frequent in 7 (25%). Ventricular couplets were present in 7 (25%) and ventricular tachycardia in 4 (14%). Patients with frequent ventricular ectopy had larger LV z-scores (3.8 vs. 1.8, p= 0.01) and a greater incidence of aortic insufficiency (33% vs 0%, p= 03). Age, duration of follow-up, QTc, medication, MVP and MR were unrelated to ventricular arrhythmia. In regression analysis LV z-score was the only independent predictor of arrhythmia. Patients with a larger LV had a greater QTc (r=.48, p=.001). The 2 parent deaths occurred at a mean age of 15.1 +/- 6.6 years in patients with significant LV dilation (z scores 3.7 & 5.1) and documented VT on Holter. There is a high incidence of minor LV abnormalities with associated ventricular ectopy in patients with Marfan syndrome. Holter monitoring should be employed for routine follow-up.

P519

Holistic molecular genetic (HMG) medicine in Williams syndrome

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[Introduction] Williams syndrome is a developmental disorder showing characteristic facial features, congenital heart defects (mainly supravalvular aortic stenosis and peripheral stenosis), visuo-spatial features, a typical cognitive profile, low birth weight, short stature and infantile hypercalcemia. Williams syndrome patients have microdeletion of chromosome 7q11.23. We applied holistic molecular genetic (HMG) medicine in patients with Williams syndrome to clarify the molecular genetic pathogenesis of congenital and hereditary heart disease. Patients are hospitalized and receive holistic care for one week. The results are analyzed discussed by all physicians and laboratory technicians and used for each patient's daily life, including education and protection from disease. [Materials and Methods] We examined 16 WS patients whose age ranged from 5 to 20 years. Of the cardiovascular findings, supravalvular aortic stenosis was seen in 12 of 16 patients. These 12 patients also had peripheral pulmonary stenosis (PPS) (7/12, 58%). Ventricular septal defect was seen in 2 patients, multiple pulmonary stenosis with PPS in 1, and mitral regurgitation with patent ductus arteriosus in 1 patient. [Results] All patients had hemizygosity of chromosome 7q11.23. Three patients, who had a smaller deletion, did not show the typical facial features, or an outgoing personality and their birth weight was within the normal range. Eleven patients (69%) had high cholesterol levels, including highly oxidized LDL and lipoprotein (a) which are closely related with atherosclerosis. These findings as well as abnormal elastogenesis indicate that patients with Williams syndrome show a high susceptibility to adult cardiac vascular disease. [Conclusion] HMG medicine, based on molecular genetic diagnosis, will prevent disease in the early-phase of the disease. HMG medicine may become a very significant system of medical care in the 21st century.

P520

Can maternal MTHFR polymorphisms increase the risk for conotruncal cardiac defects in the offspring

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The etiology of conotruncal cardiac defects (CTCD) involves multiple genetic and environmental factors. Homozygosity for C677T mutations of 5, 10 methylenetetrahydrofolate reductase (MTHFR) gene that reduces MTHFR activity and increases homocysteine plasma level, is associated with an increased risk of neural tube defects and Down syndrome. Also the A1298C mutation results in a decreased MTHFR activity. Because of the common embryonic origin of the cells involved both in the neural tube closure and heart septation processes, we retained that maternal MTHFR genotype could be associated with the development of CTCD in the offspring. By RFLP analysis, we studied 68 mothers of affected children and 100 mothers of healthy children as controls. Allele frequency distribution, in Hardy-Weinberg equilibrium, was no different between cases/controls. Heterozygosity for 677T was found in 38% of the case mothers versus 44% of controls and homozygosity in 9% of case mothers versus 13% of controls (P<0.01). There was not significant difference between genotype frequency distribution for 1298C mutation (38% and 12% in case versus 50% and 8% in controls in heterozygosity and homozygosity respectively). The distribution of the combined 677/1298 polymorphisms was different between the 2 groups (P<0.05). The 677TT genotype conferred a 1.77 fold increased risk (OR= 1.77, 95% CI: 0.2[4.39) and 1.24 fold for 1298CC (95% CI 0.88/1.34). The risk increased to 2.80 fold (95% CI: 0.50/10.91) for 677CC/1298CC genotype, to 2.22 (95% CI: 1.48/10.27) for 677CC/1298AC and to 3.33 (95% CI: 0.68/15.18) for 677CT/1298AA. Data are not statistically significant probably for the small sample size. Mothers with 677CT genotype showed a 2 fold decreased risk (P<0.05, OR=0.45, 95% CI=0.21/0.94). These preliminary data indicate that a 677TT/1298AA mother could have an higher risk to

P521

Echocardiographic evaluations in girls with Turner syndrome without evident heart disease

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Since January 1990 to July 2000 all girls with Turner syndrome during or prior to growth hormone treatment given in our institution were studied by echocardiography on a routine basis. The results of 43 consecutive asymptomatic parents without previous treatment for heart disease are reported. The median age at examination was 11.4 years (3.3–18.4 years). A structurally normal heart was found in 30/43 patients (70%). Bicuspid aortic valve was found in 9 (21%), two having dilatation of the ascending aorta. Mild valvular aortic stenosis was found in 3 (7%), one of which also had a mild coarctation of the aorta. In one case partially anomalous pulmonary vein return was detected. The mean z-score for the diastolic thickness of the interventricular septum (mean z-score -.55, 95% CI: 1.1–2.6) and for the left ventricular mass ASE (mean z score 0.9, 95% CI: 0.3–1.5) were significantly increased. Using Spearman's rank order test a positive correlation, although not statistically significant (p=0.056), was found between duration of growth hormone treatment and diastolic thickness of the interventricular septum. Left ventricular diastolic and systolic diameters as well as the diastolic thickness of the posterior left ventricular wall were not significantly different from reference values. M-mode measurements of the left ventricular dimension did not differ between girls with myocardially normal and abnormal hearts. There was also no correlation between karyotype and cardiac findings. In conclusion structural heart abnormality was found in 30% of girls with Turner syndrome presumed to have normal hearts. A possible correlation between septal hypertrophy and duration of growth hormone treatment is in conflict with earlier findings warranting further investigation.

P522

Absent pulmonary valve has different genetic causes and different anatomical associated malformations: Combined study of 40 patients

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In 40 consecutive pts (10/1974–01/2000) with total or partial absence of the pulmonary valve (APV) and severe pulmonary insufficiency the clinical phenotype and cardiovascular anomalies were assessed. Also, 14 of 22 surviving pts were examined by a clinical geneticist for dysmorphology. After informed consent in 23 pts caryotype and analysis for 22q11 microdeletion by FISH (D22S75) were carried out. In addition, 14 probands and their siblings and parents were genotyped at 6 polymorphic loci of the 22q11 region (D22S42, D22S941, D22S944, D22S264, D22S311, D22S539). The ductus arteriosus could be assessed in 36 pts. 3 pts had a persistent duct. In 2 of them a deletion of the long arm of chromosome 18 (18q-) was shown. In the other 33 pts the duct was absent in 24, abnormal in 5 and present in 4. All had conotruncal defects (26 typical Fallot). Standard caryotyping in 18 pts was normal. The total group of 20 pts with normal karyotype were further classified according to clinical phenotype. G1 (normal): 13 pts, G2 (with one or more of the following features: facial anomalies, learning disability, cleft palate, hypocalcemia, immune defect): 7 pts. In G1, 13 had FISH and 5 genotyping. In G2 all 7 had FISH and genotyping. None of the G1 pts had a microdele-

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

tion, but 5 of 7 G2 pts had a microdeletion (in 2 it was detected only by genotyping). Comparison of the group with microdeletion vs no microdeletion revealed important differences: multiple or subaortic VSD (60%/0%), left ascending and descending aorta (100%/67%), high aortic arch (20%/0%), abnormal aortic branches (60%/14%), conotruncal (non-pharyngeal) anomalies (60%/20%). In conclusion, APV has at least 2 genetic causes. 18q- and 22q11 microdeletion with different associated cardiovascular anomalies. In APV pharyngeal arch anomalies (III, IV, VI) are good predictors of 22q11 microdeletion. Genotyping can detect microdeletion not detected by FISH. Some cardiovascular and non cardiovascular anomalies are also suggestive of microdeletion in APV.

P523

Multiple cardiac tumors in fetuses and neonates as a predictor of Tuberous Sclerosis

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Tuberous sclerosis (TS) is a multi-system disease with variable clinical manifestations that may have potentially devastating effects on the nervous system. Although prenatal genetic testing for TS is not currently available, the presence of prenatal or neonatal cardiac tumors may be an early marker for the diagnosis. Previous case reports have described fetuses with multiple cardiac tumors who have been subsequently diagnosed with TS. Other reports have described fetuses and infants with single tumors without TS. We performed this study to determine the predictive value of multiple versus single tumors for the diagnosis of TS. We reviewed data from four centers of 51 patients diagnosed with cardiac tumors on fetal (n=28) or neonatal (+1 month; n=23) echocardiograms. We analyzed cross-sectional follow-up data to determine which patients had subsequently been diagnosed with TS. Three pregnancies were terminated. Of the remaining 48 patients, 39 had multiple and 9 had single tumors. Among the patients with multiple tumors, 36 (92%) were subsequently diagnosed with TS. The other 3 did not have clinical or radiological signs of TS at the time of follow-up. Among patients with single tumors, only 1 (11%) had TS. The positive predictive value for TS of multiple cardiac tumors on fetal or neonatal echocardiography was 92%. The negative predictive value for TS of a single tumor was 73%. By Fisher's exact test, patients with multiple tumors were significantly more likely to have TS than those with a single tumor ($p<0.001$). These findings demonstrate that a fetus or neonate with multiple tumors has a significant probability of subsequently being diagnosed with TS. This data will aid in counseling parents and monitoring disease manifestations.

P524

Clinical characteristics of adult Marfan patients who fulfill the revised Gent criteria

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Introduction: Marfan syndrome (MFS) has well recognized clinical manifestations making the application of diagnostic criteria possible. Little is known about the patterns of clinical abnormalities in patients fulfilling established criteria. We have sought to characterize the musculoskeletal, integumentary and ocular abnormalities in patients fulfilling the revised 1996 Gent criteria. Methods: Patients referred to the University of Toronto Congenital Cardiac Centre for Adults with a possible diagnosis of MFS were prospectively recruited over a 1-year period. Patients were assessed in a standardized fashion using the Gent criteria and formal ophthalmology reports were reviewed. Patients were classified as 1. Definite MFS, 2. Possible MFS, or 3. Not MFS. Cardiovascular endpoints were aortic dissection, aortic root dilation (AD), and mitral valve prolapse (MVP). Results: 71 Patients, mean age 33 (SD 14) years (range 15 to 76), comprising 38 males (54%) were recruited. Of these, 48 (68%) had definite MFS, 13 (18%) possible MFS and 10 (14%) were excluded as having MFS. Of the 48 with MFS (age 38, SD 13 years, range 18 to 72), 5 (10%) had aortic dissection, 38 (79%) aortic root dilatation and 41 (85%) had MVP. Clinical features are summarized in the table. TABLE HERE. Cardiothoracic musculoskeletal manifestations in MFS occur commonly (85%). The most frequent musculoskeletal features include a high arched palate, dural ectasia, pectus excavatum and the characteristic facies. Lens dislocation may occur in up to two-thirds of patients.

Informatics/Internet

P525

On-line educational resource for grown-ups with congenital heart disease (GUCH): emphasis on imaging modalities and complex surgical procedures

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Grown-ups with congenital heart disease (GUCH) represent a rapidly expanding population. Existing Internet resources for GUCH have primarily focused on family education, promotion of particular institutional programs and databases but have not targeted education of specialists directly involved in care of these complex patients. Evaluation and surgical treatment of these patients require sophisticated imaging and complex surgical procedures often difficult to understand except by specialists performing the procedures themselves. A multidisciplinary team of a pediatric and adult cardiologist, a cardiac surgeon, imaging specialist, and a medical illustrator and web designer have joined to create an educational Web-site for the clinician or trainee dealing with evaluation and surgical management of GUCH patients, using the case-study methodology. Relevant clinical history, X-rays catheterization data, EKGs, CT scans, and MRI's are displayed, including real-time echocardiogram and angiograms. Surgical procedures are described by text, drawings and when available, intra-operative photographs and videos. The Web-site contains a logically organized comprehensive list of congenital heart defects based on physiologic patterns with textual explanations and illustrative images. A menu of surgical procedures is used to describe procedures with text and illustrations, all of which can be cross-referenced to individual cases. A glossary is added for difficult terminology and a relevant reference section is included. The site can be used easily and freely by medical practitioners and trainees connected by the now more widely available high-bandwidth connections (T1, DSL or cable-modem). We have created an Internet-based education resource which is demonstrated to be ideally suited to assemble and display a variety of sophisticated imaging studies and surgical techniques in a simple, well organized way, for the complex evaluation and treatment of GUCH patients.

P526

Internet based environmental and genetic research: the wisconsin pediatric cardiac registry (WPCR)

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Background: Congenital heart defects are the most common birth defects encountered in Wisconsin, occurring in approximately 5–8 of every 1000 live born children, with 400–600 new cases each year. The etiology of most congenital cardiac abnormalities is unknown. The Wisconsin Pediatric Cardiac Registry is a registry of children born with a congenital heart defect in the state of Wisconsin, beginning January 1, 2000. Methods: Cardiologists within the state are asked to identify patients presenting with a structural congenital heart defect confirmed by echocardiogram, cardiac catheterization, surgery or autopsy and notify the Registry Coordinator via toll free phone line, fax, or e-mail. The Registry Coordinator asks the family to participate at one of three levels: 1) registration, 2) registration and questionnaire, or 3) registration, questionnaire and DNA blood sampling. The questionnaire is comprehensive, consisting of 155 questions of genetic and environmental data. It can be completed in print form or on the Internet in 2 hour's time. DNA blood sampling is done on patients with conotruncal abnormalities, Elastin anomaly or hypoplastic left heart syndrome. Parents and siblings are also asked to submit to a DNA blood sample. Pregnancy and pre-pregnancy additives, provided on the questionnaire, are mapped on a computer. Conclusion: Since January 1, 2000, almost 300 families have registered. Half have completed participation and 95 DNA samples have been drawn. The accumulation of epidemiological data will be useful in studying patterns of disease. Also, the use of a database on the Internet will provide a method to database, organize, search, evaluate and analyze diagnostic and therapeutic outcomes and allocate resources within the state of Wisconsin. Questionnaire data entry via an Internet based Oracle database represents a novel way to perform epidemiological data collection and research.

P527

Pedihear, an internet discussion group for pediatric cardiology professionals

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https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Since 1994 more then 9000 messages have been transmatted via PediHeart, a free Internet discussion group with over 1,500 members world wide. Purpose: We proposed to study PediHeart's 1 Influence on clinical practice 2 Educational value 3 Quality and reliability of postings Methods: Questionnaires to all members. Results & Conclusions: The response rate was 14 to 21% in the past. Further questionnaires will be used for updated data. Past responses indicated that: 1 PediHeart is useful for clinical practice. 33% of respondents asked for help in the management of their patients and 67% of these found the answers useful. Seventy percent considered that PediHeart had a positive influence in their clinical practice. 2 PediHeart has educational value: 80% of answers. 3. The quality of discussion is high. The most active members, responsible for >30% of postings, have vast experience (20+yrs) and are highly qualified (academicians, authors, reviewers & editors).

P528

Application of the information approach to modeling the market of medical cardiology services

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The purpose of the present research is the creation of information model of the medical cardiology services market taking into account the mutual influence within the city, which is divided into districts. The development of information model is made using methods and techniques of the system analysis and in particular, techniques of structurization of the purposes and functions of systems, information approach of A. A. Denisov. The basis of model is the set of dependences for determination of i type medical services supply of category d of the population of city district k, with allowance of interference of districts Hidk. The dependences take into account the number of medical establishments (ntipsl), quantity of medical staff posts (fipti) and their city regional distribution (Sqiksl) and categories of the population (Xdipksl), population of d services (nk), and its distribution into categories (Kdk). Formula: Hidksipl=((Xdipksl * Siipksl * Jqipl)/(nk * Kdk)), i=1 n, k=1,m, d=1,t, s=1,n; p=1,r. I=1, m ips. Now the market of cardiology medical services of St. Petersburg is characterized by: - High degree monopolization: children's cardiological help is rendered mainly by state and municipal medical establishments (97%), the private medical establishments make 3% from the number of rendered services; - Non-uniformity of the children's population of districts by consultative-diagnostic cardiological help, the deviations are 15–52% from an average level. In condition of limited budget financing it is expedient to enlarge a share of private medical establishments for alignment and maintenance of necessary level of supply of the population by cardiological help in various districts of the city and formation of competitive environment in the medical services market.

P529

Use of artificial intelligence for outcome prediction after palliation of cyanotic congenital heart disease

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Objective: Neural Network (NN) is a type of artificial intelligence that tries to simulate the human thinking process and proved to be reliable in the field of pattern recognition and prediction of outcome. Our objective was to use the NN technology in the prediction of outcome after palliation shunts for cyanotic congenital heart disease. Methods: We initially conducted a retrospective study using the data of 250 patients (pts) who had a Blalock-Taussig shunt(BT) to develop a back-propagation type of NN to predict the mortality depending on the preoperative demographics. These included: age, sex, weight, diagnosis, oxygen saturation, preoperative need for prostaglandin, mechanical ventilation, balloon atrioseptostomy and urgency of operation. The mortality data were entered as pattern (output) for the NN to be trained on. From this we constructed a NN model with an output = prediction of the risk of mortality expressed as a fraction of one. The discriminative accuracy of the NN was estimated using the Receiver Operating Characteristic Curve(ROC). The area under the curve(AUC) was 0.871. Then prospectively, this network was used to predict the outcome of 59 cases of BT shunts. Results: With a threshold >0.5 for NN output there were 10 cases identified as high risk. In comparing the prediction to the actual mortality (9 deaths), the calculated sensitivity of the NN model was 0.88 and specificity of 0.96. Conclusion: Artificial intelligence technology can play an important role in risk stratification and outcome prediction in pediatric cardiac surgery.

P530

Web information about the pediatric cardiology

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Parents of child with congenital heart disease are seeking more and more information about the disease and therapeutic methods. In the last of 20th century, the internet became a great supporter of such parents and they could easily reach to the medical information resources on the web. There will be a lot of pages on the web, but we did not know how deep or how prevalent they are. So we searched the Web by two kind of search engines to get the appropriate information about congenital heart disease for parents of their sick child. We used one category search engine and one robot search engine. 1) Congenital heart disease' was searched by category search engine in English and Japanese. We checked the whole searched pages and compared the contents between two languages. 2)A hundred and forty-six medical terms were selected from the index of 'Moss and Adams Heart disease in infants, children, and adolescents' and were searched by the robot search engine on the web. The top 50 pages searched for each term were evaluated if they were suitable for parents. 1) We reached to 24 web pages in English and 13 in Japanese. In English several integrated pages were detected but in Japanese 100% pages were written about individual patient. 2) We could get enough information about 137 terms (95%) out of 146 terms on the Web. Only 7 terms (5%) were thought to be inappropriate because the term was used for the other meaning or special interest was paid for about that term. Internet is already a useful tool for the parent of child with congenital heart disease in English-speaking countries, and it will be in Japan in the future.

P531

Internet in developing fetal cardiology

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Prenatal cardiology is not well developed in Poland. Less than 10% neonates in referral centers are admitted after prenatal diagnosis. To improve this situation www pages for perinatology, including prenatal cardiology, sponsored by Danone Foundation, were developed. The goal of this paper is to present our www pages and most important discussion subjects from the mailing list. All necessary information concerning risk of having a baby with congenital heart disease and pictures of the normal fetal examination, normal fetal heart and most common lesions could be seen on the page. Additionally, discussion mailing list was organized for professionals interested in perinatal problems. Obstetricians, neonatologists, pediatricians, pediatric cardiologists and other doctors oriented in this subject could be members of the discussion group. We hope that www pages combined with mailing list is a good way for training and improving prenatal diagnosis in our country. Obstetricians, especially those who performed fetal scan at the first and second level would be able to receive quick consultation about their every day problems. The address of web site is: www.cbr.waw.pl/prenatal. Contact e-mail address is: dangel@cbr.waw.pl

P532

ECMO cannulation: an Internet-hosted training model

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A review of 300 consecutive Extracorporeal Membrane Oxygenation (ECMO) patients revealed a high incidence of vascular cannulation complications, in particular when carried out by newly appointed junior staff. Aim: To develop a process to minimise ECMO cannulation complications. Methodology: A novel internet-based training model for ECMO cannulation is part of a training programme for incoming registrars. Results/Analysis: of the ECMO experience after this protocol was introduced has encouraging trend towards fewer cannulation problems. Medium-term results will be presented. Conclusion: Apart from the advantages of convenience, universal accessibility, visual appeal and cost-effectiveness, our tool has been acceptable to the trainees and was valuable in guiding them through a new technical procedure without endangering patients. It is feasible that the model can be applied to: (a) train a team setting up an ECMO program, remote from a referral location (b) instructing surgeons and intensivists in carrying out other complex technical procedures.

P533

Computerized interpretation of the pediatric electrocardiogram based on newly established normal limits

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https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Interpretation of pediatric electrocardiograms (ECGs) is complicated by the strong age dependency of the criteria. Computerized interpretation could be helpful to assist the pediatric cardiologist. We wanted to develop such a computer program. For this we had to establish up-to-date normal limits, because previous studies that assessed normal limits had their imperfections: ECGs were recorded with relatively low sampling rate, ECG measurements were done manually or normal limits were presented for only a limited set of parameters. ECGs from 1992 healthy Dutch children were recorded, from which normal limits were established for all clinically relevant ECG measurements. Additionally, two pediatric cardiologists independently interpreted 1719 ECGs and rated the certainty of each abnormality on a four-point scale. When their interpretations differed, a third pediatric cardiologist arbitrated. This set of ECGs was divided in a training set of 1197 ECGs and a test set of 642 ECGs. Our normal limits showed clinically significant differences with previously established normal limits, especially in R- and S-wave amplitudes in the precordial leads. Based on the normal limits and the training set, diagnostic rules were formalized in an iterative process using expert interviews and an automatic learning algorithm. The resultant rules were evaluated on the test set. The table shows the performance for the major diagnostic categories: right ventricular hypertrophy (RVH), left ventricular hypertrophy (LVH), and right bundle branch block (RBBB). The newly established normal limits suggest that diagnostic criteria for the pediatric ECG need to be adjusted. A computer program has been developed and validated for the automatic interpretation of pediatric ECGs. The performance of the program appears to justify its use in a clinical setting.

Myocardial Preservation and Cardiopulmonary By-pass, Perfusionists and Cardiopulmonary By-pass

P534

Pulsatile flow enhances regional myocardial blood flow during and after hypothermic cardiopulmonary bypass in a neonatal piglet model

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Purpose: The objectives of this study were: 1) to investigate the effects of pulsatile versus nonpulsatile perfusion on myocardial blood flow during and after hypothermic cardiopulmonary bypass (CPB) and 2) to quantify pulsatile and nonpulsatile pressure and flow waveforms in terms of the energy equivalent pressure (EEP) for direct comparisons. Methods: Ten piglets (mean weight: 3 kg) underwent: pulsatile (n=5) or nonpulsatile (n=5) perfusion. After the initiation of CPB, all animals were subjected to 15 minutes of core cooling (rectal temperature, 25°C), followed by 60 minutes of hypothermic CPB, 10 minutes of cold reperfusion, and 30 minutes of rewarming. During CPB, mean arterial pressure (MAP) and pump flow rates were maintained at 40 mmHg and 150 ml/kg/min, respectively. The aorta was cross-clamped at 25°C for 60 minutes. During pulsatile CPB, a pump rate of 150 bpm and a stroke volume of 1 ml/kg were maintained. Regional blood flows were determined with a radiolabeled microsphere technique. The results (see table) are expressed as mean ± standard error (ml/100 g/min). Normothermic CPB: Hypothermic CPB: After Rewarming: After CPB: Flow: P: NP: P: NP: P: NP: P: NP: LVBF: 202±25: 122±20: 101±16: 150±51: *276±48: 140±12: *271±10: 130±14: RVBF: 184±14: 126±26: 118: ±35: 150±62: *279±57: 150±22: *315±48: 137±22: *P< 05 vs. NP; LVBF, left ventricular blood flow; NP, nonpulsatile; P, pulsatile; RVBF, right ventricular blood flow. The average increase in pressure (from MAP to EEP) was 10%±2% in the pulsatile group and 1% in the nonpulsatile group (P< 0001). The average increase in extracorporeal circuit pressure (ECCP) (from ECCP to EEP) was 33%±10% in the pulsatile group and 3% in the nonpulsatile group (P< 0001). Conclusions: These results suggest that pulsatile flow generates significantly higher energy and enhances myocardial blood flow during and after hypothermic CPB in the piglet model.

P535

Comparision of troponin-T release in infants with cyanotic and acyanotic heart disease following cardiopulmonary bypass surgery: does cyanosis cause more myocardial injury?

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Comparison of troponin-T release in infants with cyanotic and acyanotic heart disease following cardiopulmonary bypass surgery. Does cyanosis cause more myocardial injury? Ozan, O., Barth, J., Parsons, J.M., Dickinson, D.F., Gibbs, J.L., Wilkinson, K.G., Leeds, United Kingdom. We aimed to define preoperative, postbypass values of troponin T in children with cyanotic and acyanotic heart disease, and its correlation with operative, postoperative recovery variables and outcome. 74 children aged 1day to 13 years undergoing cardiopulmonary bypass were prospectively studied. Blood samples were taken after anesthetic induction, 4 hours post-bypass, then at regular intervals for a further 120 hours. Preoperative, peak and final levels of troponin T were compared between cyanotic and acyanotic patients. Troponin T showed higher values preoperatively in cyanotic and sick infants. It peaked at 4 hours, declined gradually over 48 hours, but remained detectable at 120 hours even in patients with uncomplicated recovery. Younger age, cyanosis and decreased acute outputs were all correlated with higher postoperative values. Of the two infants who died, one showed highest preoperative value 0.98mcg/L and a peak value of 14.98mcg/L, and in the other levels continued to rise beyond 48 hours. Elevated levels of Troponin T above 5mcg/L after 24 hours was associated with a longer and complicated postoperative recovery. There was no significant difference in postoperative recovery, duration of ventilation, and hospital stay between cyanotic and acyanotic patients. Preoperative detection of elevated serum troponin T levels may allow to identify high risk infants. Serum troponin T levels greater than 5 mcg/L at 24 hours postoperatively may indicate complicated recovery.

P536

Is elevated protein s-100 beta level reliable in diagnosing cerebral injury following cardiopulmonary bypass surgery in infants?

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Is elevated protein s-100 beta level reliable in diagnosing cerebral injury following cardiopulmonary bypass surgery in infants? Ozan, O., Barth, J., Parsons, J.M., Dickinson, D.F., Gibbs, J.L., Wilkinson, K.G., Leeds, United Kingdom. We studied reliability of serum protein s-100 beta in detecting cerebral injury following cardiopulmonary bypass surgery in infants and older children with cyanotic and acyanotic heart disease. 74 patients aged 1 day to 13 years were included in the study. Blood samples were taken preoperatively, after anaesthetic induction, 30 minutes postbypass, then at regular intervals for a further 120 hours. Levels were correlated with operative data, postoperative recovery variables and outcome. The elevated serum levels of protein s-100 were detectable preoperatively in all patients. Infants and cyanotic patients showed higher preoperative values compared to older children regardless of outcome. S-100 beta levels peaked at 30 minutes after completion of bypass and declined to preoperative values within 72 hours. There was no significant correlation between postoperative s-100 beta levels and operative data or postoperative recovery variables at any time. No cut-off value of s-100 beta at any time has been found to be predictive of postoperative neurologic outcome. Even elevated levels above 2mcg/L after 24 hours was not associated with a longer or complicated postoperative recovery. Two patients died and two patients had neurologic deficit but s-100 beta levels failed to provide predictive or prognostic information. Although cyanotic patients showed sustained and higher postoperative values compared to acyanotic patients, the differences did not reach statistical significance. Preoperative detection of elevated serum s-100 beta levels may occur in infants and young children. Postoperative elevated levels even greater than 2 mcg/L beyond 24 hours did not also correlate with neurologic outcome.

P537

Percutaneous arterial perfusion in pediatric cardiac surgery

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Objective: To assess the suitability of percutaneously introduced arterial perfusion (PCAP) in pediatric cardiac surgery. Patients and Methods: PCAP was used at 7 operations of the aortic arch and/or intracardiac anomalies for 6 patients. Age: 19days-3years (4 neonates, 2 infants), weight: 3,6–9,2kg, BSA: 0,17–0,43m2. All PCAPs were accomplished on an elective basis at reoperations (4 early, 2 late), at primary repair in 1. 5F and 6F regular or 3and 3arm ECMO cannulae were inserted via percutaneous puncture of the femoral artery using Seldinger-technique. Patients were fully heparinized and connected to the heart-lung machine, then chest was opened. Statistical analysis computed PCAP data with those of previous perfusion of the same patient using traditional arterial cannulation (trad). Results: No morbidity or mortality attributable to PCAP occurred. The duration of the extracorporeal

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

riac lesion (PCAP: 118,8(55–229), crad: 136.5(95–178)min. p=0,98) and flow rate (PCAP: 280–1200, crad: 350–1200ml/min, p=0,94) did not differ. PCAP vs. traditional perfusion exhibited significant difference in line pressure/flow rate (PCAP: 188,79; crad: 81,96 Hgmin/L/min, p=0.0003) and line to perfusion pressure drop/flow rate (PCAP: 118.81 crad: 32,71 Hgmm/L/min, p=0,0028). Having accomplished the operation the cannulae were removed with pressure dressing. No limb perfusion complications occurred and pulsatile flow could be detected in all. Conclusions. Theoretically, PCAP has less complications than a cut down method in neonates and infants. Introduced on elective basis in selected reoperations median sternotomy and arterial cannulation are made safe and easy. As primary repairs it may obviate the risks of addressing a diminutive ascending aorta. The arterial cannula is well away from the operating field, therefore, PCAP may diminish the need for the total circulatory arrest. All these should translate in a broader indication for its use. Specially designed cannulae should overcome high line to-perfusion pressure drop.

P538

The predominance and transient immune suppression following cardiac surgery with cardiopulmonary bypass in children

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Aim: In children CPB surgery induces substantial release of the immunosuppressive Th2 cytokine IL-10. Yet no studies are available verifying if Th2 response and IL-10 release are due to CPB or surgical trauma alone. It is not clear if Th2-response is correlated with post operative complications as capillary leak or post-cardiotomy syndrome. Method: Serological and immunological analysis of children (age: 3–16yrs.) undergoing surgery with (n=50) or without CPB (20, control) with otherwise similar surgical data and medication was performed. Pre-, peri- and post-operative blood samples were analyzed by ELISA, immunophenotyping, intracellular (IC) cytokine expression and semi qPCR for cytokine mRNA. Results: Shift of T-helper cells to Th2 phenotype, IL-10 secretion and elevated IL-10 mRNA levels were specific to CPB surgery. CPB patients showed significant increase of serum IL-10 with maximum values at the end of surgery (peak: 20–900pg/ml, ANOVA: p<0.01). 1 to 3 days after surgery increased IgE/IgG2 and IL4/IL-Ny ratio was detected. IFNγ serum level and mRNA expression decreased. Control patients did not sequester IL-10 and had no Th2 shift. In CPB patients who developed PCS Th2 response and IL-10 secretion was significantly increased (p<0.001). The source for IL-10 are most probably not circulating monocytes as their expression of activation markers (e.g. HLA-DR, CD14) and IC IL-10 levels decreased during surgery. In contrast, T-cells had steady an elevated IC IL-10 level. Conclusion: Surgery with CPB induces immunosuppression and humoral immune response. This is in part due to Th2-cells as a source for IL-10. The shift of the immune system to the Th2 response correlates with postoperative morbidity and resembles to immune sequel after sepsis (immune paralysis) or an allergic response. These findings are in agreement with the result that patients at risk for PCS exhibit a Th2/allergic predisposition.

P539

The prognostic value of troponin T and troponin I in children after repair of complex heart defects in cardiopulmonary by-pass

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Aim: Troponin T (TnT) and troponin I (TnI) are sensitive and highly specific indicators of cardiac muscle damage. The aim of the study was to assess the diagnostic value of TnT and TnI in children after correction of complex congenital malformations. Methods: Fifty-eight patients (aged from 0.01 to 16.58 years, mean 4.03) undergoing the correction of HLHS, TGA, DORV, TAPVD, TOF, A-V canal and single ventricle-type defects using single dose crystalloid cardioplegia were prospectively recruited. TnT and TnI were measured before and after operation (6, 12, 24, 72 hours) by enzyme immunoassay. The concentration values of the markers were correlated with intraoperative parameters (cardiopulmonary bypass time- CPB, aorta cross-clamp time, type of hypothermia, dose of cardioplegia) and postoperative parameters (ventilation support time, postoperative inotropic support, intensive care stay time). The statistical analysis was based on the Mann–Whitney test, multivariate logistic regression analysis and the log-rank test. Results: A postoperative release of TnT and TnI was noted with the peak at 6 hours after cross-clamp release. Significant differences in TnT and TnI concentration values were noted in association with all the investigated parameters except the dose of cardioplegia. Factors significantly affecting the total amount of TnT and TnI (area under the curve, r2=0.59 and 0.49) were CPB time (p=0.0001; p=0.0002, respectively) and type of hypothermia (p=0.0024; p=0.0078). Significantly elevated TnT and TnI values were noted in patients who required prolonged ventilation and inotropic support. The critical values for TnT and TnI were determined 72 hours after cross-clamp release as 4.5ng/ml (p<0.00001) and 48ng/ml (p<0.00001), respectively. Conclusion: TnT and TnI are sensitive predictors of cardiac muscle damage following operations of complex heart malformations in children, as well as a useful prognostic indicator for difficulty of recovery.

P540

Comprehensive proteomic and genomic expression analysis of cardiopulmonary bypass

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Introduction: To date, there has been no exhaustive analysis of the molecular changes associated with cardiopulmonary bypass (CPB). A thorough, prospective analysis of the genome and proteome should complement candidate-based techniques to define the set of molecular alterations associated with CPB. In addition, these studies may provide insights into general mechanisms of tissue injury, hypoxia, and inflammation. Methods: We isolated the right atrial appendage from pigs placed on CPB with cross clamp and hypothermic arrest at the following time points: pre-CPB, immediately post-CPB, 2 hours post-CPB, and 2 days post-CPB. Human right atrial appendage samples were snap-frozen from patients pre-CPB or immediately post-CPB. Proteins were electrically focused and separated by SDS-PAGE. Matched gels from each time point were silver stained and compared by visual and computer-based methods to identify differentially expressed proteins. Mass spectroscopy was used to identify novel proteins. RNA expression pattern analysis was performed using Affymetrix U95B chips, using a replicated study design to minimize the statistical limitations of multiple comparisons. The same replicated study design was used for both protein and RNA expression analysis. Results: Analysis of RNA levels for over 6000 genes revealed a subset that was clearly differentially expressed. Most of these genes have not been previously implicated in ischemia-reperfusion signaling and these represent new targets for further study. Proteomic analysis offered a different perspective since protein expression patterns do not necessarily correlate with RNA expression patterns. Over 1000 protein spots were digitally captured and aligned. Intensities calculated from matched spots. Conclusions: Together, genomic and proteomic approaches identify a more complete set of the molecular changes than those identified by candidate based approaches and offer complementary insights into the cellular response to cardiopulmonary bypass.

P541

Antioxidative status of the cpb-prime used for neonates

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A prime solution with a high antioxidative and iron binding capacities may reduce the oxidative damage associated with cardiopulmonary bypass (CPB). We investigated how preparation of prime solution with preserved red blood cells (RBC) and either albumin solution or fresh frozen plasma (FFP) affects the antioxidative properties of the prime solution. The prime solutions were prepared with either pasteurized human albumin (ALB prime) or FFP (FFP prime). The total antioxidant capacity was measured with the Total Radical Antioxidant Parameter (TRAP) assay and with the Ferric Reducing Ability of plasma (FRAP) assay. The individual scavengers, vitamin C, sulfhydryl groups, uric acid and total protein were measured before, during and after the prime preparation. Malondialdehyde (MDA) was measured as a parameter for lipidperoxidation. The efficiency of glutathione (GSH) recycling in the RBC as an index of RBC antioxidative capacity was determined and the presence of prooxidative free hemoglobin / heme (Hb/heme) and non protein bound iron (NPBI) was measured. Both prime solutions showed no TRAP value and a low FRAP value. Vitamin C was only found in the FFP prime. Both prime solutions contained sulfhydryl groups and uric acid in low concentrations. After the procedure RBC showed a less efficient GSH recycling than before the preparation. In both primes concentrations of prooxidative Hb/heme increased. However only in the ALB prime NPBI was detectable. We showed that prime solutions based on either albumin or FFP had very low antioxidant capacity. During the preparation of the prime the ability of the RBC to recycle

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

glutathione decreases, and the RBC releases Hb/heme and NPBI. A diminished antioxidative capacity of RBC and the presence of prooxidants in the prime solution may increase oxidative stress in neonates undergoing CPB.

P542

Systemic inflammatory response syndrome related to cardiopulmonary bypass and its modification by high dose methyl prednisolone: a controlled clinical trial

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Objective: To investigate the safety and efficacy of high dose methylprednisolone (MP) in modifying the systemic inflammatory response in cardiopulmonary bypass (CPB) in children undergoing cardiac surgery for congenital heart disease. Patients and Methods: Thirty children with congenital heart disease undergoing CPB are randomly assigned to two groups: group 1 received MP 30 mg/kg by an intravenous infusion of 30 minutes and group 2 received MP 2 mg/kg intravenously before the onset of CPB. Postoperative clinical parameters were recorded, serum IL-6 and 8 levels, acute phase reactans and blood biochemistry were determined serially for both groups. Results: In both groups plasma IL-6 and 8 levels were elevated above the preoperative levels at 2 and 24 hours after declamping. The peak levels were obtained at 2-hour samples. The difference between two groups in terms of postoperative IL-6 and 8 levels was not statistically significant. CRP levels and polymorphonuclear leucocyte counts, postoperative core temperature, extubation time, period of stay in intensive care unit, oxygenation indices and biochemical parameters of patients did not significantly differ in two groups. Only one patient in group one had elevated liver enzymes, blood urea nitrogen, and creatinine in the postoperative period. No significant complications were observed due to treatment with high dose MP. Though postoperative interleukin and CRP levels indicated a systemic inflammatory response in our patients clinical picture was apparently affected in only one patient and she was in the high-dose MP group. Conclusion: CPB induces a systemic inflammatory response which is associated with an increase in neutrophil count, CRP, IL-6 and 8 levels. High dose (30 mg/kg) MP was not superior to low-dose MP (2 mg/kg) in blunting the systemic inflammatory response to CPB in pediatric patients undergoing open heart surgery.

P543

Could high cardiopulmonary pump flow prevent neurological impairment in pediatric non-blood open heart surgery?

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Neurological impairment, at least partly ischemic in origin, has been issue of infants undergoing non-blood cardiopulmonary bypass. Controversy continues about the effect of temperature, pH, Hb, pump flow, pulsatile and nonpulsatile perfusion and autoregulation on the brain. To promote non-blood-transfusion surgery in the smaller infant, to grasp the safe hemodilution limit for their cerebral metabolism is the point. To know the actual scattering of cerebral metabolism during pediatric cardiac surgery, we have recorded regional cerebral Hb index (HbI) and brain oxygen saturation (rSO2) simultaneously using a specially-made-near-infrared-spectroscopy- system (TOS96, Tostec Co. Ltd.) since 1997. We employed low-pump flow (70–100 ml/kg/min, at venous side, pulsatile) from 1997.6 to 1998.9 in 115 pediatric cases. Then we changed it to high-flow (120–160 ml/kg/min.) after 1998.10 in 155 cases. We tried to keep their lowest Ht higher than 15%. To study the effect of pump flow on cerebral metabolism in cases undergoing non-blood cardiopulmonary bypass, 50 cases' records each were analyzed in detail retrospectively. The product of rSO2 and HbI (rSO2*HbI) sensitively and significantly decreased soon after the initiation of cardiopulmonary bypass. In the low-flow group, rSO2*HbI stayed in low level and two of five cases whose lowest rSO2*HbI were under 30% of control (before pump, on NLA anesthesia) showed intermittent neurologic impairment postoperatively. In the high-flow group, HbI tended to stay low but increase rSO2 recovered soon, so that rSO2*HbI kept higher than 30% of control in all 30 cases. Its up-down scattering was smaller than low-flow group and no neurological impairment was seen in high-flow group. The rSO2*HbI difference in the two groups was completely invisible from all other conventional monitoring and laboratory investigations. High-pump-flow worked to shift up the bottom line of cerebral rSO2*HbI, suggesting an superiority regarding intraoperative hemodiluted cerebral metabolism.

P544

Paediatric myocardial metabolism after open heart surgery

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Myocardial metabolism was studied in 27 children who underwent repair in our institution. Fuel preference was determined by comparing metabolite concentrations between arterial and coronary sinus blood (AV-diff) after discontinuation of cardiopulmonary bypass. Patient median age was 7.5 months(1–74) and weight was 7.9 kg(3.7–23.0). Diagnoses were VSD(13), tetralogy(5), AVSD(3), ASD secundum(2), aortic stenosis(1), papvd(1) and complex(4). Median bypass time was 80 min(21–187), aortic cross clamp time 44 min(8–116), and inotropic support duration 1 day(0–2). There were no intracranial coronary artery problems identified, and no hospital mortality. AV-differences for substrates are shown in the table. Put Table Here, Please. There was a significant correlation between ischaemic time and release of lactate ($p<0.01$). Conclusions: Uptake of fatty acids, ketone bodies and oxygen suggests ongoing oxidative phosphorylation. Some dysfunction of the citric acid cycle was found, especially in the patients with longer ischemic times demonstrated by abnormal lactate metabolism. However, the whole population showed an uptake of glutamate with alanine release, evidencing anaplerotic replenishment of citric acid cycle intermediates by anaplerosis. Post-bypass insulin resistance would account for unimpaired uptake of glucose, as well as contributing to the abnormal lactate metabolism by inhibition of pyruvate dehydrogenase. This study demonstrates myocardial fuel preference after uncomplicated heart surgery in children, and the experimental setup may be used to compare protection strategies.

P545

Safety of aprotinin use and re-use in pediatric cardiothoracic surgery

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Objectives: We sought to determine the incidence and impact of hypersensitivity and anaphylactic reactions to the serine protease inhibitor aprotinin in children undergoing cardiothoracic surgical procedures, including those employing deep hypothermic circulatory arrest. Methods: In this retrospective review of our entire experience with aprotinin (n=646), 519 cases of first-time exposure, 101 cases of second exposure, and 26 cases of third or higher exposure were reviewed. Reactions to aprotinin were classified as mild (skin whirl or response to an intradermal test dose injection-Type A) or severe (development of severe, otherwise unexplained hypotension during the administration of loading dose – Type B). Case records of patients sustaining a reaction were reviewed to assess the impact of the reaction on the patients' subsequent outcome, as well as survey the strategies implemented to manage the reaction. Results: Reactions occurred in 4 of 519 first-time exposures (0.8 %), 3 of which were type A and 1 of which was type B. In second-time exposures, there were reactions in 3 of 101 (3.0%) patients, 1 of which was type A and 2 of which were type B. In third-time or higher exposures, 2 of 26 exposures resulted in reactions (7.7%), of which one was type A and one was type B. Patients were more likely to sustain a reaction to aprotinin on re-exposure than on first exposure ($p<0.05$ by Chi-square analysis), although the rate of reaction on initial or subsequent exposure was low. In only one of the nine cases with observed reaction to aprotinin was the drug ultimately not used. In the other eight cases, modifications in the rate of administration and/or the timing of the loading dose allowed us to proceed with the use of aprotinin. No significant adverse sequelae were attributed to the use of aprotinin. Conclusions: The risk of reaction to aprotinin is low in children undergoing cardiothoracic surgical procedures, even with multiple exposures to the medication. Reactions are more likely with re-exposure. When reactions occur, management strategies for discontinuous delivery of the medication allow for safe usage of the drug.

P546

The effects of modified ultrafiltration on respiratory function after the cardiopulmonary bypass

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We evaluated the effects of modified ultrafiltration (MUF) on respiratory function of patients with congenital heart disease and increased pulmonary

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

blood flow, who underwent intracardiac repair without blood transfusion. Thirty-two consecutive patients, less than 10kg in weight, were divided into two groups; 16 with MUF (group M) and 16 without MUF (group C). We calculated the arterial/alveolar oxygenation ratio (a/APO2) and oxygenation index (OI). They were compared at the end of cardiopulmonary bypass (before MUF in group M)-[1], at the end of operation (after MUF in group M)-[2] and at the first examination of blood gas analysis at ICU-[3] respectively. There was no statistical significance between two groups in age at operation, cardiopulmonary bypass time and minimum hematocrit during operation. a/APO2 were [1]0.675, [2]0.735 and [3]0.764 in group M, [1]0.712, [2]0.775 and [3]0.648 in group C respectively. In group M, a/APO2 before and after MUF did not differ, but that at ICU improved comparing to that before MUF (p<0.05). On the other hand, a/APO2 showed no change in group C. At ICU, a/APO2 of group M was significantly better than that of group C (p<0.05). There was no significant difference in OI between group M and group C. In conclusion, MUF improves oxygenation ability after the cardiopulmonary bypass by decreasing the postoperative physiological shunt which was refl

P547

Development of a cardiopulmonary bypass system with extremely low priming volume for the pediatric open heart surgery

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When considering bloodless open-heart operation for the pediatric patients, priming volume of the pump system is key to success. If extreme hemodilution is neglected, organ damage may occur. We have developed a pump system with an extremely low priming volume to minimize hemodilution. <Methods> A separated type heart-lung machine was used by which the system could be freely laid out. To reduce the distance between clean operative area and pump system, we used a sterile sheet with tubes piercing it. The smallest applicable oxygenator and arterial filter were selected according to patients' size. Between Dec. 1999 and Oct 2000, the system was used for 28 cases (VSD,ASD, TOF). Body weight was 4.9 to 17.0 kg (mean 8.9 +/- 3.9 kg) and age was 5 months to 6 yrs (mean 1.5 +/- 1.4 yrs). <Results> The priming volume for the patients weighing under 6.5 kg was reduced to 160 ml (130 ml without an arterial filter). For the patients weighing between 6.5 and 9.5 kg, the priming volume was 190 ml and for 9.5–17 kg patients, the volume was 220ml. No case was given homologous blood transfusion. Hematocrit and platelet count before bypass, during bypass (minimum value), and after bypass were 33.1+/-4.6%, 25.5+/-6.3x104/mm3, 21.4+/-4.2%, 15.8+/-4.9 x104/mm3, and 29.6+/-5.0%, 20.3+/-7.2 x104/mm3 respectively. No organ damage was noted in this series. Not only we could further develop the system and smaller size with 147 ml (112ml without an arterial filter) was realized. By this newer system, 3kg baby could be operated without blood transfusion. <Conclusions> We have developed extremely low priming volume system which could broaden application of bloodless cardiac operation for the small pediatric patients.

P548

The effects of volume and pressure hypertrophy on myocardial adenosine triphosphate (ATP) level and function in children with cyanotic heart disease

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Objective: The effect of volume and pressure overload hypertrophy on myocardial ATP levels has been studied in animals. It is not clear whether these findings are applicable to humans. This study was designed to determine if volume and pressure hypertrophy affect myocardial ATP re-function and clinical outcome. Methods: 14 children were recruited, the volume overload children(group 1:n=6)were children with VSD with dry right ventricular muscle bundle(VSD/RVMB). The pressure overload children(group 2:n=8)were children born with tetralogy of Fallot and had a preoperative saturation>90% (Pink TOF). ATP was measured from RV biopsies taken a)pre-cathemia,b)15 min of ischemia,c)end-ischemia,d)15 min reperfusion. Ejection fraction(EF)was measured by echocardiography. Results: The preoperative age, weight and saturation were similar in both groups. The total pump time(137 vs 102 min, p=0.02)was longer in group 2. The ATP values did not differ significantly between the two groups at each interval a)21.7 vs 21.9,b)18.6 vs 16.3,c)11.3 vs 15.4 and d)12.6 vs 15.8; NS). The pre and post op EF were also similar(67 vs 66%, 68 vs 67% respectively, NS), as was the length of inotropic support(34 vs 29 hours; NS). The postoperative ventilatory support tended to be longer in group 2 (8 vs 66 hours, p=0.1). Although not significant, both the mean ICU stay (41 vs 116 hours, p=0.21)and mean hospital stay were longer in group 2 (17 vs 36 days). Conclusions: In cyanotic children it seems that volume and pressure overload effects on ATP and function are similar and yet the clinical course is prolonged in children with pressure overload hypertrophy. This suggests that the difference seen in clinical outcome is not explained by the ATP levels or mechanical performance of the heart.

P549

Neuroprotective effect of deep hypothermia in a piglet model of cardiopulmonary bypass with circulatory arrest

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Purpose: We evaluated the mode of neuronal cell injury in neonatal piglets with respect to deep hypothermic circulatory arrest (DHCA) duration and the possible neuroprotective effects of hypothermia. Material: Sixteen neonatal piglets (age < 10 days, weight 1.9 +/- 0.5 kg, f=m) were included in this study. The animals were divided in 3 groups: control with sham operation (n = 6), DHCA for 60 min (n = 5), DHCA for 120 min (n = 5). All animals were anaesthetized, intubated and mechanically ventilated. After rewarming of perfusion, the animals were monitored for 6 – 8 hours prior to sacrifice. The brains were immediately removed for histological and immunohistological studies. Neuronal cells (560 +/- 20) were counted in sector CA1 of the hippocampus of each animal. Sectors CA1 – CA3 and dentate gyrus were examined for necrotic and apoptotic neurons. Results: The main preliminary results were the quantitative differences of cell injury including the perivascular astroglial cells according to duration of cold ischemia. Neuronal cell damage was predominantly found in sector CA4. By prolongation of ischemia up to 120 min the amount of necrotic cells in the CA4 region increased dramatically from 0.5% to 89%. Additionally, necrosis in the CA1-3 regions occurred and apoptotic cell death in the dentate gyrus reached 20%. Conclusion: Deep hypothermic homogeneous systemic perfusion prior to circulatory arrest for less than 60 min is an effective neuroprotective method when total circulatory arrest for blood-free surgical procedure is required.

P550

Cardiopulmonary support with extracorporeal membrane oxygenation for postcardiotomy low output syndrome in children

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Although outcome of surgical repair for congenital heart disease has improved, postcardiotomy low output syndrome(LOS) still remains major cause of operative death. Extracorporeal membrane oxygenation (ECMO) has been used as a life supporting device for such a situation as well as in adult population. In this study, we reviewed our experience of ECMO for treating early LOS in children. During the 8 years period since 1992, 15 patients (under 15 years old) were placed on ECMO for cardiac surgery in our institute. We introduced heparin-coated(HC) ECMO circuit instead of non-coated(NC) one in 1996, and 10 patients were treated since then. With HC anticoagulation was limited to maintain ACT to 150 sec, whereas 300sec or more with NC circuit by high dose heparin infusion. They underwent definitive repair operations in 9 and palliation in other 6. Indications for initializing ECMO were low oxygen saturation caused by unbalanced pulmonary blood flow after palliative operation in 6, severe ventricular dysfunction in 8 and postoperative PH crisis in 1. Nine of 15 patients (3/5 in NC 6/10 in HC) were able to be weaned from ECMO after 2.5–12 days support and 7 were long term survivors. After introducing HC circuit, all the patients placed on ECMO for ventricular dysfunction were successfully weaned from it. In contrast, no patient treated for hypoxia after palliation could be removed ECMO. Among the 7 patients who were survived, no major complication was observed in 5 with HC, whereas either 2 with NC had neurological deficit. Postoperative bleeding of patient placed on NC was significantly higher than that on HC. We conclude that ECMO, especially with HC, is useful in pediatric population for ventricular failure after definitive repair as well as in adults.

P551

Heart failure impairs vasomotor functions of the mesenteric bed after cardiopulmonary bypass

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https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

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Background: Mesenteric dysfunction is a rare but severe complication after open heart surgery which may be aggravated by coexistent heart failure. The aim of this study was to investigate the effect of cardiopulmonary bypass (CPB) on intestinal vascular endothelial and smooth muscle function in a canine model of heart failure. Methods: In 6 dogs volume overload heart failure was created by femoral artery–vena cava shunt, 5 healthy animals served as control. Heart rate (HR), mean arterial pressure (MAP), mesenteric blood flow (MBF) and mesenteric vascular resistance (MVR) were measured before and after 90 minutes of cardiopulmonary bypass. Reactive hyperemic response and the response to acetylcholine (ACH) and sodium-nitroprusside (SNP) were expressed as percent change of MVR. Results: Before CPB, baseline hemodynamics (MAP: 125±5 vs. 117±10 mmHg, MVR: 0.96±0.09 vs. 0.99±0.17 mmHg•min/ml), reactive hyperemia (-53±5 vs. -53±2%) and response to ACH (-41±3 vs. -55±6%) and SNP (-68±4 vs. -56±4%) did not differ significantly. In both groups, 90 minutes CPB led to a similar significant drop of MAP (61±17 and 51±16 mmHg, respectively, p<0.05 vs. baseline) and also of reactive hyperemia (-10±5 vs. -29±15%, p<0.05 vs. baseline). After CPB, response to ACH (-22±9 vs. -42±9%, p<0.05) and to SNP (-14±4 vs. -58±7%, p<0.002) showed a more pronounced decrease in the heart failure than in the control group. Conclusions: The development of heart failure per se does not attenuate mesenteric vasomotor function. However, CPB induces some impairment of mesenteric endothelial- dependent vasodilatative response and deeply damages its endothelial independent function in heart failure animals. This phenomenon may have an impact on the higher incidence of mesenteric complications in young patients with manifest heart failure.

PS52

Evaluation of six pediatric cardiopulmonary bypass pumps during pulsatile and nonpulsatile perfusion

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Purpose: This study evaluated six pediatric cardiopulmonary bypass (CPB) pumps in terms of energy equivalent pressure (EEP) during CPB with pulsatile and nonpulsatile perfusion in a neonatal piglet model. The EEP is the ratio between the area beneath the hemodynamic power curve and the area beneath the flow curve. Methods: Thirty-nine piglets, with an average weight of 3 kg, underwent CPB with a hydraulically driven physiologic pulsatile pump (PPP, n=7), Jostra HL 20 pulsatile roller pump (Jostra-PR, n=5), Stockert S III pulsatile roller pump (S III-PR, n=6), Stockert S III mast-mounted pulsatile roller (S III-Mast-PR, n=7), Stockert S III mast-mounted nonpulsatile roller pump (S III-Mast-NP, n=7), or Sarnicare CAPS nonpulsatile roller pump (CAPS-NP, n=7). After initiation of CPB, the piglets were subjected to 20 minutes of hypothermia, 60 minutes of deep hypothermic circulatory arrest (DHCA), 10 minutes of cold reperfusion, and 40 minutes of rewarming. In all groups, the pump flow rate was 150 ml/kg/min, and the mean arterial pressure (MAP) was 45 mmHg. During pulsatile perfusion, the pump rate was 150 bpm, and the stroke volume was 1 ml/kg. The following results were obtained during normothermic CPB (15 minutes on pump): Pump % Change (from MAP to EEP) % Change (from ECCP to EEP): PPP: 13±3*, 46±13*; Jostra-PR: 5±1**, 13±2**; S III-PR: 4±1**, 9±1**; S III Mast PR: -0.3±1.6 & 2±5; S III-Mast-NP: -0.2±1.8 & 1.3±1.1; CAPS-NP: 1.4±0.7, 17.6±7.6. *P<0.05, PPP vs. other groups; **P<0.05, Jostra-PR & S III-PR vs. other groups. ECCP = extracorporeal circuit pressure. The results obtained during hypothermic CPB and after rewarming were identical to those obtained during normothermic CPB. Conclusions: Pulsatile flow with the PPP, Jostra PR, and S III-PR pumps produces higher hemodynamic energy, which may improve vital-organ perfusion during CPB.

PS53

Pulsatile versus nonpulsatile perfusion and vital organ blood flow using a new pediatric cardiopulmonary bypass pump

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Purpose: The objectives of this study were twofold: 1) to investigate the effects of pulsatile perfusion (with a new pediatric pump) and the effects of nonpulsatile perfusion (with a conventional pediatric pump) on cerebral, renal, and myocardial blood flow and on regional cerebral oxygen saturation (rSO2) and 2) to quantify the pressure and flow waveforms in terms of hemodynamic energy, using the energy-equivalent pressure (EEP) formula, for direct comparison. Methods: Fourteen piglets (mean weight: 3 kg) underwent perfusion with either a new pulsatile pediatric pump that has a miniature roller head (n=7) or with a conventional nonpulsatile pediatric pump (n=7). After initiation of CPB, all animals were subjected to 25 minutes of core cooling (rectal temperature: 18oC), followed by 60 minutes of deep hypothermic circulatory arrest (DHCA), 10 minutes of cold reperfusion, and 40 minutes of rewarming. Blood flows to vital organs were assessed with radiolabeled microspheres, and rSO2 levels were assessed with near-infrared spectroscopy. Results: The pulsatile and nonpulsatile groups had no significant differences in cerebral, renal, and myocardial blood flow at any of the experimental stages. DHCA significantly decreased the vital-organ blood flow in both groups (p<0.001). The post-CPB rSO2 was higher in the pulsatile group (45.3%±1.5% vs. 38.8%±2.6%; P<0.05). The average change in arterial pressure (MAP) (from MAP to EEP) was -0.38±1.6% in the pulsatile group and 0.17%±1.8% in the nonpulsatile group (P=NS). Conclusions: The pulsatile and nonpulsatile groups had no significant differences in hemodynamic energy, and CPB with DHCA impaired cerebral, renal, and myocardial blood flow in this pediatric model. Therefore, not all pulsatile roller pumps generate sufficient energy to provide adequate blood flow to vital organs.

PS54

Effects of temperature and circulatory arrest on cerebral blood flow during and after pulsatile cardiopulmonary bypass

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Purpose: The objective of this study was to investigate the impact of deep hypothermic circulatory arrest (DHCA) versus hypothermic cardiopulmonary bypass (HCPB) on global and regional cerebral blood flow (CBF) during pulsatile perfusion in a neonatal piglet model. Methods: The piglets were divided into DHCA (n=6) and HCPB (n=6) groups. A pediatric physiologic pulsatile pump was used in all experiments. The HCPB group was subjected to 15 minutes of cooling, followed by 60 minutes of HCPB, 10 minutes of cold reperfusion, and 30 minutes of rewarming. The DHCA group underwent hypothermia for 20 minutes, DHCA for 60 minutes, cold reperfusion for 10 minutes, and rewarming for 40 minutes. In both groups, the pump flow (150 ml/kg/min), pump rate (150 bpm), and stroke volume (1 ml/kg) remained constant. The aorta was cross-clamped for 60 minutes at 18oC for DHCA and 25oC for HCPB. A radiolabeled microsphere technique was used to measure the global and regional CBF (ml/100g/min). Results: The CBF decreased by 45% during deep hypothermia at 18oC (before DHCA) compared to hypothermia at 25oC (106±8% vs. 56±6%; P<0.05). After rewarming, the global CBF decreased by 45% more in the DHCA group than in the HCPB group (87±15% vs. 48±7%; P<0.05). Fifteen minutes after the end of CPB, however, the global CBF was reduced by only 25% more in the DHCA group than in the HCPB group (56±11% vs. 42±8%; P=NS). The blood flow pattern in the right and left hemispheres, cerebellum, basal ganglia, and brainstem was similar to that of the global CBF. Conclusions: The impairment of CBF caused by the use of DHCA may be minimized with pulsatile perfusion, which rapidly increases CBF recovery after CPB with DHCA.

PS55

A reusable training circuit for cardiopulmonary bypass

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At the turn of the millennium, perfusion teaching programs were faced with a significant difficulties. The number of students in pediatric perfusion training had increased and more importantly the number of pediatric open heart procedures had decreased due to a variety of reasons. In addition surgeons preferred an experienced perfusionist for complicated procedures. Hence they could barely satisfy the minimum requirements of pediatric cases established by the teaching programs. The idea of 'a teaching circuit' that could reproduce and simulate cardiopulmonary bypass was designed. The trainee is able to manipulate the cardiopulmonary bypass circuit according to patient's responses, perform perfusion related maneuvers in establishing and maintaining haemodynamic validity. The aim of the study was to design a circuit to simulate initiation of bypass for paediatric and if required, adult patients, maintain stability during varying clinical situations and then to wean off cardiopulmonary bypass. The circuit also provides the trainee an opportunity to learn perfusion related techniques i.e. cardioplegia delivery, ultrafiltration and modified Ultrafiltration. Suction, venting lines and cell saver can be

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

adapted to this circuit, according to the establishment protocol. The equipment used was reusable and non sterile, and is divided into a patient component and perfusionist component. The patient side, when completed by an experienced perfusionist, could reproduced clinical features such as changes in arterial blood pressure(systolic/diastolic) and central venous pressure variation that will affect the arterial pressure. It is our belief that this circuit will be an invaluable tool in teaching perfusion students, cardiac surgery residents and other disciplines related to perfusion. To our knowledge, there is presently no dedicated, reusable and disposable circuit for perfusion training that would allow them to practice perfusion related techniques before being exposed to clinical situations.

P556

Extending the capability of mechanical cardiopulmonary support: rapid, portable, flexible

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We designed a novel cardiopulmonary support (CPS) unit to overcome the limitations of conventional extracorporeal life support (ECLS) circuits. This unit allows rapid deployment (less than 2 minutes), antegrade pulsatile augmented venous drainage for excellent cardiac decompression, extreme portability (including medical transport helicopters) and heparin-bonded components allowing support without systemic heparinization. This new circuit has allowed us to extend the capabilities of CPS. Prospectively collected data over a five-year period was evaluated for risk and outcome variables. Forty-three children (age 1 day to 14.7 years) underwent CPS support (mean time 52 hours) with 25/52 (44%) surviving to hospital discharge and 38/52 (73%) surviving to decannulation. Overall, 34/52 (65%) were supported after heart surgery. Support was initiated on 14 patients in the OR (3/14, 21% survived) and on 20 patients perioperatively in the Cardiac ICU (9/20, 45% survived). Patients initiated in the OR or immediately postoperatively received no heparin until bleeding was controlled. Mean time of CPR on patients initiated on support emergently for cardiopulmonary arrest in the ICU was 12 minutes. Six patients were initiated on CPS as an outlying institution and transported by helicopter with 2/6 (33%) surviving. Two patients were initiated on support emergently in the cardiac cath lab and both survived. Eight patients underwent cardiac catheterization on support (5/8, 63% survived). 4/8 had interventions on support (3/4, 75% survived). Six patients underwent preoperative support and all survived. One patient was converted from conventional ECMO to CPS without anticoagulation for support during surgery to control exsanguinating pulmonary hemorrhage. Rapid deployment capability portability and decreased requirement for anticoagulation has allowed the rescue of otherwise unsalvagable children with the CPS system. We believe it is the method of choice for cardiopulmonary support in children.

P557

A quality control/quality improvement (QC/QI) program for pediatric perfusionists

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A perfusion QC/QI program has 4 interwoven aspects: 1) Risk equalization: All cases are ranked by risk category, weight, pump time and cross-clamp time. They are assigned to each perfusionist to ensure equal exposure to similarly ranked cases. 2) Outcome assessment: Specific end-points include defibrilation rate, post-MUF hct, blood use, ECMO use, 1st and 2nd anion gap values, length of stay, patient charges and mortality. Cases such as end-point falling outside a ± 2 SD range are examined for the cause of the variation. 3) Performance assessment: Each perfusionist performs case reviews on the other perfusionists monthly to ensure on-going compliance to protocol and to identify variations in technique that cause out of range end points, both good and bad. Techniques leading to improved end points are adopted as protocol and those leading to worsening end points are abandoned. 4) Evaluation: Cases subcategorized by weight and risk category are compared to earlier subcategory patients to further identify areas needing improvement. This QC/QI program has guided improvements in modified ultrafiltration and recalcification with a correlative decrease in post-MUF hct's and decreases in length of stay, patient charges and mortality. This program can also be used to identify and rectify non-perfusion related variations such as seasonality associated mortality.

P558

Design of pediatric bypass circuit permits reduction in circuit volume to optimize hematocrit in infants.

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A circuit designed for flows of 800 ml/min or less has a non-variable component (380 cc prime volume) and a variable component (350 cc prime volume) totaling 340 cc prime volume. Non variable components include an oxygenator, bubble trap, arterial/venous loop and associated tubing. Variable components include a hard shell venous reservoir, cardioplegia set, a hemoconcentrator and associated tubing. Reduction in circuit volume through manipulation of variable components can decrease the operating volume from 340 cc to 190 cc. This is achieved by draining the 150 cc volume of the variable components back into the venous reservoir and using concurrent hemoconcentration to transfer the red cells into the non-variable component, optimizing the hematocrit prior to the end of CPB. Typically, the hematocrit can be increased about 8% using this variable technique. This is comparable to hematocrit increases after modified ultrafiltration (MUF) and gives the perfusionist the option of optimizing the hematocrit without adding additional red cells or using MUF.

P559

Affect of early recalcification on morbidity after cross-clamp removal during pediatric cardiotomy.

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Quality control evaluation of perfusionist performance on patient outcome detected differences between 2 perfusionists within similar patient populations (GC: n = 55 vs. JK: n = 31). The most differences ($p<0.05$) were between the 1st anion gap (AG in mEq/L) after surgery (GC = 14.7 ± 4.6 vs. JK = 11.7 ± 3.6), the subsequent AG @ 12 hrs post-op (GC = 11.2 ± 4.4 vs. JK = 8.6 ± 2.1), the length of stay (LOS) in days (GC = 9.3 ± 6.1 vs. JK = 6.8 ± 4.6), and the patient charge (GC = $68,580 ± 42,341 vs. JK = $50,069 ± 27,765). Case reviews demonstrated a difference in timing of recalcification after cross-clamp removal (GC = 5 min vs. JK = 2 min). A review of 247 cases using delayed recalcification showed a LOS = 10.1 ± 16 min/mary = 6.6% and ECMO use = 3.0%. A review of 178 cases using early calcification showed a LOS = 8.0 ± 10.5, mortality = 3.4% and ECMO use = 1.7%. Based on the improved patient outcomes of JK, early recalcification became standard procedure. Low ionized calcium (iCa) may reduce reperfusion damage after aortic cross-clamp removal. Typically, recalcification is done after a period of reperfusion by blood with a low iCa. While iCa is a known mediator of reperfusion injury the optimal period of reperfusion may be very short as the detrimental effects of iCa may be blunted if administered while the heart is still cold. Without adequate iCa availability, the contracting heart lacks vigor and may be subtlety damaged by dispersion during rewarming.

P560

Vacuum assisted venous drainage (VAVD) in pediatric cardiotomy

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VAVD offers several advantages to the pediatric perfusionist and cardiac surgeon. VAVD eliminates air locks and permits the use of smaller venous cannulae and lines than with gravity drainage. In selected cases, VAVD eliminates the need for tourniquet placement around vena cavae thus reducing dissection and bleeding, particularly in redo cases. VAVD also gives the perfusionist some active control of the venous return by manipulation of the suction applied to the venous line. Potential advantages that are currently undergoing evaluation include a reduction in the weight cut-off for clear prime, reduced MUF time, increased post-MUF hct and reduced blood donor exposure in a circuit designed for VAVD use. Potential disadvantages include the generation of air microemboli under vacuum that could pass through the oxygenator and bubble trap and the possible over pressurization of the cardiotomy and venous reservoir. However no emboli have been detected by the bubble monitor and careful visual inspection of static high points in the circuit have failed to detect the coalescence of air emboli. A whistling safety valve alerts the perfusionist to the danger of over pressurization. The advantages of VAVD out weigh the disadvantages making it an important adjunct to perfusion strategy during pediatric cardiotomy.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P561
Zero balance ultra filtration reduces exogenous substrate loads in blood primes for pediatric pump primes and post pump transfusions: a prospective study
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Objective: A prospective randomized pilot study was performed on 15 children undergoing heart surgery to determine what effect Zero Balance Ultra Filtration (Z-Buff) would have on reducing the substrate loads associated with banked blood primes. Methods: The extra corporeal circuit was primed with 600cc of Plasma Lyte 'A', one unit of packed red cells, and one unit of fresh frozen plasma. Utilizing the hemoconcentrates present in the circuit for Modified Ultra Filtration, the circuit prime was hemoconcentrated down to the 20Hct level in the cardiotomy reservoir. Then a total of 400cc of Plasma Lyte 'A' was slowly infused into the circuit, with simultaneous removal of crystaloid via the hemoconcentrator, to maintain the 20Hct level in the reservoir. Upon completion, 100–150 cc of the Z-Buffed reconstituted whole blood was removed for ultrafiltration post pump. Pre and post Z-Buff glucose, potassium, urea and lactate levels where measured. Results: Initial results show that a more physiologically correct pump prime was achieved with a significant decrease in the substrate load. This allowed for a more metabolically stable patient upon weaning from the circuit. Further analysis of the data will determine if Z-Buff impacts upon morbidity and mortality for children, particularly neonates undergoing complex heart surgery. Conclusions: It is possible to achieve a more physiologically correct pump prime using the Z-Buff technique. How Z-Buff reduces morbidity and mortality will be determined with further analysis with a larger study population.

Pulmonary Hypertension, Pulmonology

P562
Potential obstructive structure of peripheral bronchi in the fetal rat displaying sacralogy of Fallot characterized by an absent pulmonary valve
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In cases of Tetralogy of Fallot (TOF) with absent pulmonary valve (APV), markedly enlarged pulmonary arteries compress main stem bronchi resulting in respiratory failure. Moreover, intrapulmonary bronchial compression by intrapulmonary arteries has been reported during the neonatal period. Consequently, the possibility of potential obstructive structure of peripheral bronchi in a fetal rat lung model of TOF exhibiting APV was investigated. Bis-diamine was administered to virgin Wistar rats. The animals were sacrificed in the near-term period and the fetuses were immediately fixed. Following diagnosis by sectional study, examples of TOF displaying APV were chosen exclusively. Histologic study of the lung by serial section was conducted and compared with controls. The medial thickness of the hilar pulmonary arteries was not significantly different in comparison to controls as expressed in terms of percent wall thickness as per our previously reported method. Radial alveolar count and bronchial generations counts revealed hypoplastic changes evident at the acinus level. No changes were apparent at the bronchiolus level. Computer-aided three-dimensional graphic reconstruction of arteries and bronchi demonstrated, in instances of TOF with APV, markedly enlarged arteries near the hilar level. Pulmonary arteries and bronchi displayed orderly branching much like a tree in the case of controls. The pulmonary arterial pattern was parallel to the bronchi. In contrast, bronchi exhibited orderly branching in cases of TOF involving APV. However, arterial branching occurred in a radial pattern which was not in parallel with bronchi. These branching patterns are thought to be potential obstructive structures which may appear as respiratory fail.

P563
Association of nitric oxide dose and methemoglobin levels in patients with congenital heart disease and pulmonary hypertension
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Methemoglobin (MH) is produced during metabolism of inhaled nitric oxide (NO), and high MH levels may impair oxygen transport. Our study purposes were to determine the relation of NO dose to MH in patients (PTS) with congenital heart disease (CHD) and pulmonary hypertension (PHTN), and determine factors contributing to higher MH levels. Demographics, MH levels, NO dose and duration, fraction of inspired oxygen (FiO2), and bypass times of all PTS treated postoperatively with NO from 1995 to 2000 were reviewed. Seventy-five PTS, ranging from 0.01 to 20.2 years (median=0.4 years), were treated on NO doses of 0.5 to 80 ppm (median=20ppm). NO duration ranged from 0.5 to 858 hours (median=96 hours). Average bypass time was 179 min. MH levels had a direct linear relation to NO dose. For each ppm increase of NO, MH levels increased by 0.03 to 0.05 ($p<0.001$). PTS of younger age ($p<0.05$) or on higher FiO2 ($p<0.001$) were likely to have higher MH. NO duration, bypass time, and weight did not affect MH. Seven PTS had MH levels >6%, all had NO doses of >50ppm. NO dose has a direct linear relation to MH levels in postoperative PTS with CHD and PHTN. For each ppm increase of NO, MH increases by 0.03 to 0.05. Younger age and higher FiO2 are contributing factors in increased levels. MH levels >6% only occurred in PTs who received >50ppm of NO, therefore, close monitoring is indicated for these PTS.

P564
Plasma arginine, citrulline, nitrite/nitrate and cyclic guanosine monophosphate in children early after cardiac surgery
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Objective: Increased pulmonary vascular reactivity is frequently observed in infants after surgery for congenital heart defects with pulmonary hypertension. We examined the role of endogenous arginine, citrulline, nitric oxide (NO) and cyclic guanosine monophosphate (cGMP) production in postoperative period. Patients: Group A: 15 infants aged 1 to 17 months (median 4 months) with severe pulmonary hypertension and left to right shunt; Group B: 7 children aged 2 to 15 years (median 6 years) after closure of atrial septal defect; Group C: 9 children aged 1 month to 3 years (median 6 months) after closed heart procedures. Method: plasma arginine, citrulline, nitrites and nitrates (NO_x) and cGMP were determined by high pressure liquid chromatography before and at 2-4 hours after operation and on the postoperative Day 1, 2 and 7. Results: all patients survived and were discharged from the hospital. In Group A inhaled nitric oxide was used prophylactically in 9 patients (20 ppm for 1 to 9 days (median 4 days). Increased pulmonary vascular reactivity was observed in 5 pts. All pts in Group B and Group C had uneventful postoperative course. Plasma cGMP, NO_x and citrulline increased (peak: Day 1) whereas arginine decreased (nadir Day 1) significantly early after surgery in all groups. There was no difference between groups in measured plasma variables at any sampling point. There was no correlation among cGMP, NO_x, arginine and citrulline. Conclusion: metabolic pathway arginine – nitric oxide – cGMP is activated in children early after cardiac surgery. Cardiopulmonary bypass had no additional effect on nitric oxide and cGMP production.

P565
Amlodipine for the treatment of pulmonary arterial hypertension in children
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Six children aged 1–16 yrs (mean 9.5 yrs) with pulmonary arterial hypertension (PAH) were treated with amlodipine in addition to digoxin and diuretics. PAH was primary in 4 and was secondary to shunt lesion (patent ductus arteriosus) in 2 cases. All patients were receiving maximally tolerated dose of nifedipine (30–90 mg daily) and were not optimally controlled. Amlodipine (instead of nifedipine) was empirically started and dose was gradually increased (5–15 mg daily). All patients symptomatically improved and clearly preferred amlodipine to nifedipine. Objective evidence of improvement (control of congestive heart failure, improvement in exercise tolerance, or in systolic time intervals) were present in five. In conclusion, our preliminary observations suggest that amlodipine seems preferable to nifedipine in the treatment of PAH. Larger controlled studies are warranted.

P566
Echocardiographic oxygen / tolazoline test in infants operated for a – v canal or VSD with pulmonary hypertension
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Use of echocardiographic oxygen and/or tolazoline test could be useful in predicting postoperative pulmonary hypertension. 28 infants (mean

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

3.3 months) with complete or partial a-v canal and symptoms of pulmonary hypertension underwent total surgical repair without prior cardiac catheterisation. Methods: 1/ direct PA/Ao press. indexes were measured at the theatre before and after correction; 2/ Echo - measurement/estimation of PA/Ao press. indexes, estimation of basic Qp/Qs index, after oxygen respiration and/or nifedipine. Results: Direct PA/Ao press. index preop: 0.60, postop: 0.43. Echo values: preop. PA/Ao press. 0.65, Qp/Qs index: basic 3.0, after oxygen (+38.8%), after nifedipine (+30%). Analysed: I gr.-high preop PA/Ao press and >0.6 (N 22): a/ postop AP/Ao press 0.49, b/ Echo-derived PA/Ao press 0.67, c/ Qp/Qs basic 2.57, d/ Qp/Qs post O2 inh.(+34.4%), e/ Qp/Qs post nifedipine (+29%). II gr. - very high preop PA/Ao press. >0.8 (N 7): a/ 0.66, b/ 0.76, c/ 2.8 d/ +33.1%, e/ +28%. III gr - high postop PA/Ao press >0.6 (N 5): a/ AP/Ao postop 0.82, b/ 0.77, c/ 2.1, d/ +22.2% e/ +18.2% IV gr-low postop PA/Ao ind. < 0.4 (N 14) a/ 0.65, b/ 0.59, c/ 2.8, d/ +14.4% e/ +15.4%. Conclusions: 1/ Increase of Qp/Qs in O2 / nifedipine test: ~ 44% means good and ~ 22% bad prognosis for postop. PA press. 2/ Qp/Qs index by itself, and preop PA/Ao press ind. has small prognostic value in infants with a-v canal. 3/ echocardiographic oxygen/nifedipine tests are of equal value for prediction of pulmonary hypertension in infants with CAV.

P567

Chronic effects of orally administered prostacyclin on pulmonary hypertension with associated congenital heart defects

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It is reported that intravenously administered prostacyclin is useful to secondary pulmonary hypertension (PH) with congenital heart defects. To elucidate the chronic effects of orally administered prostacyclin on secondary pulmonary hypertension with congenital heart defects, 10 patients administered with orally prostacyclin were included. 4 patients were after total correction (CAVC in 2, VSD in 1, TOF in 1) and all were with Down syndrome. Other 4 patients were associated with cyanotic heart diseases. Three of them were after bidirectional cavopulmonary shunt (BCPS) for PA/IVS, SV and Ebstein anomaly respectively and 1 was no surgical treatment for TOF/PA with MAPCA. Remaining 2 patients were Eisenmenger syndrome with ASD and PDA. The dosage of orally administered prostacyclin was 1.5 to 2.0 mg/kg/d, and follow-up time was from 4 months to 3 years and 4 months (average, 1 year 10 months). Results: 2 of 4 patients with PH after total correction, who had been orally administered prostacyclin more than 2 years, showed the significant decrease of PA pressure evaluated by a doppler examination. In 1 patients with SV after BCPS, the shunt from SVC to RA decreased and Qp/Qs increased after administeration of oral prostacyclin, and as result Fontan operation has been performed successfully. Other patients had no effect. These results suggested that orally administered prostacyclin for a long time might be useful for secondary pulmonary hypertension, in especially associated with congenital heart defects.

P568

The experience with primary pulmonary hypertension at children's hospital of wisconsin: can we predict who will respond to vasodilator therapy?

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Purpose: To determine whether children with primary pulmonary hypertension (PPH) have specific characteristics that favor a response to vasodilators. Methods: All children diagnosed with PPH at Children's Hospital of Wisconsin over an 8-year period underwent standard work-up followed by cardiac catheterization and acute drug testing (n=15). Pulmonary artery (PA) pressure, wedge pressure, thermodilution cardiac output, mixed venous oxygen saturation, transpulmonary pressure gradient, systolic PA to systemic pressure ratio, and pulmonary vascular resistance index (PVRI) were measured at baseline and with acute drug testing (inhaled nitric oxide 80 ppm, intravenous prostacyclin, and oral nifedipine). Beneficial response during acute drug testing was defined as a 25% decrease in PVRI and/or a 25% increase in cardiac index. Based on this, patients were classified as either acute responders or non-responders. Results: Baseline measurements were not significantly different between groups; however, the responder group was younger at time of diagnosis. All patients were started on continuous intravenous prostacyclin, coumadin and digoxin. Non-responders were listed for lung transplantation. Because of symptomatic right heart dysfunction, one non-responder underwent balloon atrial dilation during the acute drug study. Follow-up catheterization data was performed in the majority of patients. Two patients required lung transplantation prior to the acquisition of follow-up data. At follow-up catheterization, acute responders continued to demonstrate lower than baseline measurements and lower values than those achieved during acute vasodilator testing. In non-responders, hemodynamic measurements were significantly decreased from the baseline study despite lack of response to acute drug testing. Conclusions: Younger age is associated with a lower likelihood for acute response to vasodilators. Long-term prostacyclin therapy may provide benefit to patients who do not respond acutely. Atrial septal decompression and lung transplantation continue to be important therapeutic adjuncts for this patient population.

P569

Chronic and continuous measurement of mixed venous oxygen saturation and pulmonary artery pressure in patients with pulmonary hypertension: effects of exercise

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We describe the novel use of a 4-French oximetric catheter in the continuous measurement of mixed venous oxygen saturation and pulmonary artery pressure (PAP) in the non-surgical, non-postoperative setting. A 4F thermistor catheter was placed into the pulmonary artery of two patients with pulmonary hypertension after an acute drug study in the catheterization lab. Patient one was a 7 year-old girl with primary pulmonary hypertension who had been on continuous intravenous prostacyclin for six years. PAP was now normal and prostacyclin therapy was being discontinued. Patient two was an eight year-old boy with pulmonary veno-occlusive disease in whom chronic intravenous prostacyclin was being initiated after a favorable acute response to inhaled nitric oxide and prostacyclin in the catheterization lab. His baseline PAP was suprasystemic. In each patient, the indwelling catheter was maintained for 72 hours in the ICU with continuous monitoring of heart rate, systemic saturation, blood pressure, mixed venous oxygen saturation, and PAP. An exercise bicycle was placed in the ICU room and the same hemodynamics were measured during 10 minutes of exercise. In patient one, it was reassuring to note that exercise-induced cardiac output did not result in significant elevation in PAP or a drop in systemic saturation. However, in patient two, the increase in cardiac output with exercise was associated with suprasystemic PAP as well as a significant drop in systemic oxygen saturation, even while on prostacyclin. Patient one continues off prostacyclin and is doing well. Patient two was maintained on prostacyclin and listed for lung transplantation. The described technique is a useful method to continuously measure mixed venous oxygen saturation and PAP and to measure the effects of exercise or other interventions in an at-risk population in a more chronic setting.

P570

Improved endothelial cell function following intentional hemodilution in Eisenmenger syndrome

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Since endothelial cell dysfunction may significantly impact on the progression of arterial remodeling in pulmonary hypertension, we decided to measure the circulating levels of endothelial cell markers in 10 patients (5 female) with Eisenmenger syndrome (ES) and examine how those indexes might be changed by improving hemorheological conditions. Patients were aged 20 to 50 years and had arterial oxygen saturation (SaO2) of 75–90%. Plasma levels of von Willebrand factor (vWF), thrombomodulin (TM) and tissue type plasminogen activator (t-PA) were measured by ELISA. In contrast to decreased levels of TM (11.4 ± 9.6 ng/mL), plasma levels of vWF (135 ± 55 U/dL) and t-PA (23.4 ± 11.0 ng/mL) were increased in patients (reference values, 17.1 ± 2.7 ng/mL, 95 ± 21 U/dL and 4.4 ± 0.2 ng/mL respectively). Regression analysis showed no influence of the hematocrit on the variables under investigation in the baseline condition. In spite of that, lowering of hematocrit by means of therapeutic hemodilution (Hd) was followed by a significant improvement of vWF and TM levels as follows. The lack of correlation between the hematocrit and biochemical indexes in the baseline condition suggests that improvement of endothelial function following hemodilution may be related to factors other than final hematocrit itself. Anyway, correction of hemorheological abnormalities in ES appears to be beneficial from the pathophysiological point of view.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P571
Therapy with inhaled iloprost in children with severe pulmonary hypertension: first observations
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Efficacy of aerosolized iloprost has been demonstrated in adults with severe pulmonary hypertension (PH). Only limited data are available for children. We report our first experience with inhaled iloprost in three young patients with severe PH. Diagnoses were primary PH (patient 1, 12 year old), secondary PH after correction of an atrioventricular septal defect and Down syndrome (patient 2, 9 year old), secondary PH due to untreated patent ductus arteriosus (patient 3, 15 year old). Prior to initiation of therapy patients underwent echocardiography and cardiac catheterization including i.v. prostaglandin testing. Inhalation therapy with aerosolized iloprost (50 ng/kg/min 6 times) was carried out with an ultrasound nebulizer. Control cardiac catheterization and echocardiography were performed after 3 to 9 months. All patients presented with systemic or suprasystemic pulmonary arterial pressure (PAP) and resistance (Rp) in echocardiography and cardiac catheterization. Initial testing with i.v. prostaglandins and oxygen resulted in decreased Rp (25 - 50 %), PAP (0 - 35%) and increased cardiac index (CI) (20 - 34%). Control catheterization in patient 1 and 3 showed effects comparable with those under i.v. prostaglandin for CI (+17 % and +20 %), PAP (-17 % and +7 %) and Rp (-20 % and -29 %). In addition the immediate effect of iloprost inhalation was demonstrated in patient 2 (CI +20 %, PAP -16 % and Rp -32 %). Iloprost inhalation was carried out without problems and was well tolerated. Patients and parents reported of stable or improved physical condition. Two of the patients are listed for transplantation. Therapy with inhaled iloprost is a feasible and well tolerated option for children with severe PH. It is appropriate for bridging to transplantation as well as for palliation.

P572
Does inhaled nitric oxide improve survival in operated congenital heart disease with severe pulmonary hypertension?
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Inhaled nitric oxide (INO) was ordered for management of residual pulmonary hypertension in 24 children following surgical correction of their underlying heart defects. Morphological diagnoses included ventricular septal defect (n=7), obstructed total anomalous pulmonary venous drainage (n=4), atrioventricular septal defects (n=2), single ventricle (n=1), transposition of the great arteries (n=8), and double outlet of the right ventricle (n=2). Age ranged from 15 days to 14 months (median 5 months). INO was used elective in 22 patients when ratio of mean pulmonary arterial pressure and mean arterial pressure exceeded 0.5. In the remaining 2 patients nitric oxide was used only to evaluate pulmonary hypertensive crisis. Twenty two patients showed initial response to therapy and pulmonary artery pressures dropped significantly. Of the patients with direct pulmonary artery pressure monitoring, a pulmonary artery to systemic artery pressure ratio below 0.3 on prolonged therapy was associated with a survival ratio of 4/6, that between 0.3 and 0.5 with a survival ratio of 3/4. Three out of four of the patients with sustained echocardiographic and clinical response also survived to hospital discharge. All patients who showed lack of response (n=2), tolerance (n=1), or dependence (n=6) to use of inhaled nitric oxide died. In addition all 5 patients who had pulmonary hypertensive crisis died. A regime of successful resuscitation utilizing nitric oxide. Thus excluding one neurological death and one preoperative, out of 22 patients, only 9(41%) survived. Though inhaled nitric oxide is effective in lowering pulmonary pressure, it does not appear to improve survival following repair of congenital heart disease with severe pulmonary hypertension. A randomized trial between use and non-use of INO is warranted to determine its exact role in influencing survivorship in the patient with residual pulmonary hypertension following surgical repair.

P573
Quantitative analysis of pulmonary vascular disease in coarctation with ventricular septal defect
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In the study, we morphologically analyzed pulmonary vascular disease in cases of coarctation with ventricular septal defect (VSD). The materials were obtained from 11 cases of coarctation with VSD, ranging in age from 1.5 month to 8 years (mean age of 32 months). Twenty three cases of simple VSD (mean age of 75 months) were used as the control group. The thickness of the media of small pulmonary arteries and arterioles of pulmonary vascular disease (PVD) were determined morphometrically and comparative analysis was performed between the two groups. Positive correlations were observed between the medial thickness and pulmonary arterial peak pressure in each group. The medial thickness of the coarctation group was significantly greater than the VSD group at the same blood pressure level. Though there was no significant difference in PVD values between two groups, the mean value of PVD in VSD group was higher than that in coarctation group. These results suggest that thicker media of small pulmonary arteries in coarctation with VSD may suppress the development of PVD.

P574
Right ventricular function in severe pulmonary hypertension due to congenital or acquired heart disease: Is there a difference?
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BACKGROUND: The long term prognosis of patients with severe pulmonary hypertension (PHT) due to congenital heart disease (CHD; eg Eisenmenger) is much better than with acquired severe PHT (AHD; eg pulmonary emboli) or primary PHT (PPH). The reason for this survival difference is not clearly delineated, but as right ventricular (RV) failure is a common cause of death, we prospectively assessed RV function in patients with PHT due to different etiologies. METHODS+RESULTS: In 42 patients with PHT (29 with AHD, 6 with PPH; 7 with CHD) and 12 controls, complete echocardiography was done including fractional area change (FAC; normal >25%) as a marker of RV function, tissue Doppler imaging of the right ventricle (RV TDI systolic Sa and early diastolic Ea annular velocities). Also, levels of natriuretic peptides ANP (normal: 20-60ng/l) and BNP (normal: <10ng/l) were measured. The results are shown in the Table - Despite comparable pulmonary artery pressures as estimated from the systolic pressure difference between RV and right atrium (RV/RA), RV function was worse and levels of ANP/BNP more increased in primary PHT than in patients with severe PHT due to CHD. TDI measurements were not helpful to discriminate between PPH and PHT due to congenital heart disease. CONCLUSIONS: These results show that primary PHT and other acquired PHT are much more detrimental to RV function than comparable PHT due to CHD. This helps to explain the survival advantage of Eisenmenger patients compared to those with primary PHT.

P575
Oral sildenafil - a novel therapy for primary pulmonary hypertension
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To assess whether phosphodiesterase 5 inhibition improves exercise capacity and quality of life in primary pulmonary hypertension, we treated two Patient 1, a 4 year old, presented with dyspnoea, cyanosis and low patients with oral sildenafil (VIAGRA(r)), cardiac output. She refused to intubate and travelled in a wheelchair. At cardiac catheterisation pulmonary systolic pressure was 90mmHg with a simultaneous systolic pressure of 55mmHg. The mean right atrial pressure was 12mmHg. Nitric oxide and incremental doses of prostacyclin produced no change in haemodynamics or cardiac output. Maintenance prostacyclin was reduced and oral sildenafil instituted. One year later her quality of life remains improved, with an exercise capacity of 160m during a 6 minute walk. Patient 2, a 28 year old, presented with a 3 year history of worsening dyspnoea and was unable to walk 100 yards without rest. At cardiac catheterisation the systolic pulmonary pressure was 128mmHg with a simultaneous systolic systemic pressure of 126mmHg. He declined both transplantation and prostacyclin therapy, and oral sildenafil therapy was commenced. Maximal oxygen consumption was 15.2ml per kilogram per minute (predicted value, 42.9) after seven minutes of exercise. Five months later he regularly enjoys hourly periods of aerobic exercise. Maximal oxygen consumption has increased to 20.3 ml per kilogram after 1 minute of exercise. No side effects have been reported. Sildenafil is a selective and potent inhibitor of phosphodiesterase 5, increasing cellular levels of cGMP potentiating vascular relaxation. Phosphodiesterase 5 is abundant in lung tissue, and sildenafil may minimise the systemic side effects encountered with calcium antagonists. Sildenafil therapy in primary pulmonary hypertension appears to improve exercise capacity and life quality, and may be a useful adjunct to, or delay the need for, prostacyclin or transplantation.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P576

Atrial septal defect with failure to thrive in infancy: hidden pulmonary vascular disease?

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Introduction: Although usually asymptomatic, secundum atrial septal defects (ASD's) may occasionally cause heart failure with failure to thrive in infancy. Such cases may be resistant to medical therapy. We describe 6 infants (in whom 5 had additional extracardiac pathology) who underwent surgical closure with variable outcomes, and highlight the need to exclude other causes of symptoms, including primary pulmonary vascular disease. Patients and Methods: Between 1995 and 1999, 90 children underwent secundum ASD repair at Guy's Hospital at median age 4 years (range 3 months to 16 years). Four were under 1 year at the time of surgery. Two other infants underwent ASD repair at other centres, whose care was subsequently transferred to Guy's Hospital. All 6 cases presented with heart failure and failure to thrive, which was resistant to medical therapy. They were assessed by echocardiogram and cardiac catheterisation, and underwent surgery at a mean(sd) age of 6.8(3.4) months and weight of 5.4(0.9)kg, without complication. Results: On echocardiogram, all cases had evidence of a significant left to right shunt with elevated pulmonary artery pressures. At catheter, mean Qp/Qs was 2.7(0.2)1:1, and mean pulmonary artery pressure 60% systemic. Mean pulmonary vascular resistance was 2.1(0.5)U.m2. The child with an isolated ASD is now completely well following surgery, but there was no significant improvement in the other cases, with persisting pulmonary hypertension. Of these 2 were found to have significant lung pathology. 1 had left pulmonary vein stenosis requiring further surgery, and 1 had primary pulmonary vascular disease. Conclusion: Surgical closure of secundum ASD in infancy may improve symptoms and weight gain when no other cause for pulmonary hypertension is found, but other causes should be excluded, and the possibility of primary pulmonary vascular disease considered.

P577

The role of lung biopsy in adults undergoing corrective surgery for left to right shunts associated with pulmonary hypertension

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Background: Adult patients with left to right shunts and pulmonary hypertension (PHT), and equivocal hemodynamic data of reversibility may benefit from lung biopsy. Predicting reversibility of pulmonary vascular obstructive disease (PVOD) under these circumstances may guide appropriate management. Aims: To assess the role of lung biopsy in the surgical management of these patients. Methods: All patients > 18yrs who have had open lung biopsy and corrective surgery in the presence of PHT were identified from the surgical database. Hospital records were reviewed for pre and post surgical data. Results: 20 patients (9 males) with mean age 44 ± 9 years (range 34 to 66 years) were identified. Nine patients had ASD, 5 VSD, 2 PDA, 2 TGA and 2 patients had Tetralogy of Fallot. Heath Edwards (HE) PVOD grades were: II-1, II-8, III-7, IV-3. After surgery patients were followed for a median 9.5 years (range 0 to 20). Important pre and postoperative characteristics are summarized in the table. Two patients died, 1 perioperatively and the other 14 years after surgery secondary to pulmonary arterial dissection. TABLE HERE. Eleven patients had improved PA pressures. They tended to have lower HE grade (I & II), and had lower perioperative pulmonary vascular resistance. Conclusions: Pulmonary arterial (PA) pressures, aortic saturations and QpQs improve significantly after corrective surgery. Lung biopsy provides useful adjunctive information in the management of these patients.

P578

Endothelin antagonism: hemodynamic effect in severe chronic pulmonary hypertension

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Endothelin-1, a powerful vasoconstrictive peptide, has been implicated in the pathophysiology of pulmonary hypertension (PH). Objective: The purpose of this study was to evaluate the effect of short-term administration of an ETA receptor antagonist (BQ-123) in pts with severe chronic PH. Methods: The study group consisted of 26 pts (NYHA II-IV) aged 29.4±15.4 years with severe chronic PH and systolic pulmonary artery pressure (PAP) 102.7±27.5mmHg. Eight pts had primary PH, 4 pts PH associated with autoimmune disease, 3 pts primary PH in conjunction with an atrial septal defect, 5 pts PH after correction of congenital heart defects and 6 pts PH due to uncorrected congenital heart disease. All pts underwent continuous infusion of BQ-123 for 60 min at 200 nmol/min/kg. The right atrium with hemodynamic evaluation before the infusion, at 30 and 60 min of the infusion and 30 min after the end of the infusion. Data were analyzed with ANOVA for repeated measures. Results: Significant improvement was noted on systolic PAP (sPAP), mean PAP (mPAP), transpulmonary gradient (TPG), pulmonary vascular resistance (PVR), pulmonary cardiac index (Qp), and effective cardiac index (Qeff). When studying the pts with primary or autoimmune PH, we observed additionally a significant increase in systemic cardiac index ($p<0.05$). Systolic aortic pressure and pulmonary vascular resistance fell significantly but remained within normal limits. No adverse effects were noted. Conclusion: Short term administration of the ETA receptor antagonist BQ-123 significantly improves hemodynamics in pts with severe chronic PH. New avenues for further research on endothelin antagonists as treatment options in this prognostically poor patient population need to be pursued.

P579

Long term outcome after correction for congenital vascular compression of trachea and main bronchi

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We examined 21 patients aged 9 – 19 years who had suffered from a significant tracheal or bronchial obstruction by vascular compression in infancy. Cause of the obstruction was a double aortic arch (n=11), right descending aortic arch with ductal ligament (n=1), pulmonary sling (n=3) and truncus brachiocephalicus (n=16). 10 patients suffered an acute life threatening event, 10 needed mechanical ventilation preoperatively. Corrective surgery took place during infancy, in two cases in the third year of life. On presentation a thorough clinical examination was followed by ECG, echocardiography, X-ray of the thorax, lung function and exercise testing with monitoring of blood pressure, oxygen saturation and capillary blood gas analysis. At the time of examination 10 patients complained of exercise related symptoms. 12 patients had skeletal or muscular anomalies of the thorax. Cardiac function as measured by echo was regular in all patients. 8 patients had abnormal lung function with limited peak expiratory flow, 4 showed a variable degree of restriction. No obstructive dysfunction was observed. 8 patients had a limited exercise capacity, 6 showed an in- or expiratory stridor on exercising. 3 patients needed treatment for skoliosis. Despite severe and in some cases life threatening symptoms in infancy only 4 patients demonstrated exercise related symptoms on examination. In conclusion even severe stenosis of trachea and main bronchi by vascular structures has a good prognosis when surgical decompression is carried out early in life.

P580

Plastic bronchitis after Fontan operation: treatment with percutaneous creation of fenestration

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Objective: To report recovery from plastic bronchitis after Fontan operation following transcatheter fenestration and to elucidate the underlying pathophysiology. Background: Plastic bronchitis is an increasingly rare and potentially fatal complication after Fontan operation and is characterized by formation and expectoration of bronchial casts. Etio-pathogenesis of this condition is unclear and the role of elevated pulmonary venous pressure, increased central venous pressure and endobronchial lymphatic leakage has been proposed. Treatment options are limited and several different treatment modalities have been tried with only limited success. Patient and Methods: A 3 1/2 years old boy with recurrent acute episodes of severe cough accompanied by expectoration of pentranchial and mucoid bronchial casts four weeks following Fontan operation. Conservative therapy including chest physiotherapy, nebulylitic, steroids, antibiotics and bronchoscopic cast removal had no beneficial effect. Intervention: Transcatheter atrial fenestration was performed achieving reduction in the right atrial mean pressure by 2 mmHg and reducing arterial saturations by 12%. Result: Symptomatic recovery from plastic bronchitis maintained for 2 2 years after the procedure. Discussion & Conclusion: Possible mechanisms for bronchial cast formation in patients with cyanotic congenital heart defects are "mucin loosing bronchopathy" in response to elevated central venous pressure or "endobronchial lymphatic leakage" in patients with pulmonary lymphatic abnormalities. Reduction of central venous pressure by percutaneous creation of a fenestration is effective

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

in patients with 'mucin loosing bronchopathy' Ligation of the thoracic duct was shown to be effective in patients with lymphatic bronchial cuts in the setting of pulmonary lymphatic abnormalities. Cardiac transplantation should be reserved for refractory cases while pulmonary lobectomy or pulmonary artery embolization procedures can detrimental for fontan haemodynamics

P581
Stroke in a young man with cystic fibrosis after lung transplantation
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This case report describes an unusual cause of stroke after lung transplantation and subsequent patient management. The patient is a 23-year-old male with cystic fibrosis (CF) who developed progressive pulmonary dysfunction and underwent a successful, non-essential bilateral lung transplant. On the 6th post-operative day he presented with seizures and right-sided hemiplegia. Computerized tomography showed an ischemic area in the left brain suggestive of embolic stroke. Extensive work-up ruled out deep vein thrombosis or other obvious underlying causes for stroke. Trans-esophageal echocardiogram (TEE) documented a patent foramen ovale (PFO) with right to left shunting noted by contrast. Heparin was started. The patient was taken to the catheterization laboratory for placement of a PFO occlusion device. Pre-device placement TEE demonstrated a small thrombus adherent to a central venous line and extending into the right side of the atrial septum. A 25 mm CardioSEAL device was placed for PFO occlusion. Because of the thrombus, as little manipulation as possible was performed. Device placement occurred uneventfully. After device deployment, right to left shunting was no longer present and the thrombus was absent. The central line was removed. The patient underwent an uneventful recovery with return of full neurologic capacity, and was discharged from the hospital one week later on enoxaparin. Retrospective analysis of this patient's data suggests that his systemic vasoconstrictors and oxygen requirement were out of proportion to his degree of lung dysfunction. Though a preoperative echocardiogram may not have changed this patient's management, knowledge of the PFO would have allowed for closure at the time of transplantation. We recommend echocardiography with contrast in CF patients who are oxygen-dependent, hypoxemic and in whom transplantation is being contemplated. Patients with documented PFO and right-to-left shunting should undergo closure at the time of transplantation.

Radiologic Science/Mass Data Storage

P582
The use of Electron beam ct in total correction of tetralogy of Fallot
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Objectives To assess the accuracy of electron beam CT and the technique of three dimensional reconstruction in the diagnosis of Tetralogy of Fallot and its application in the total correction. Methods: Total correction of Tetralogy of Fallot was performed in 20 consecutive patients. With the help of electron beam CT, the pathological and hemodynamic changes were compared before and after operations. Results: The accuracy of electron beam CT in the diagnosis of Tetralogy of Fallot is 100%. Three-dimensional reconstruction images can present the deformation exactly and completely and can be observed in multi-angle and any layer. Conclusions: Electron beam CT is a precise and reliable application in the diagnosis of Tetralogy of Fallot. With the help of three-dimensional reconstruction of electron beam CT, a standardized method in the total correction of Tetralogy of Fallot can be provided.

P583
Abnormality of left ventricular sympathetic nervous function assessed by (123)I-metaiodobenzylguanidine imaging in patients with vasovagal syncope
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Cardiac and systemic autonomic nervous function may be impaired in patients with vasovagal syncope (VS). No reports, however, have described sympathetic nervous function of the left ventricle (LV) in VS patients. OBJECTIVE: To asses the LV sympathetic nervous function in patients with VS using (123)I-metaiodobenzylguanidine (MIBG) imaging of the heart. DESIGN: Prospective comparison of (123)I-MIBG imaging results in 30 VS patients (6 boys, 24 girls, mean age 12.2) during a 45 min 60 degree head up tilt test (HUT). 20 min and 4 hr imaging was carried out following injection of 80 mBq (123)I-MIBG. Specific (123)I-MIBG uptake was assessed as the cardiac to mediastinal activity ratio in the delayed image. Results were compared between the two groups. RESULTS: HUT was positive in 13 patients and negative in 17. Specific (123)I-MIBG uptake, assessed as the cardiac to mediastinal activity ratio in the delayed image, was significantly higher in the tilt positive group than in the tilt negative group ($p<0.05$). CONCLUSION: Patients with tilt test positive group have significant sympathetic nervous stimulation of the LV myocardium as a result of paradox reflex of Bezold Jarisch due to orthostatic hypotension and sympathetic overactivity.

MAY 29 Time: 14:00–15:30

Session 4
Drugs/Vasodilators/Receptor Blockers, Medical Management and Drugs

P584
Hormonal treatment of cardiac surgical complications
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Background-Chylothorax after pediatric cardiac surgery is a well recognized complication. It usually occurs late and is associated with significant morbidity, increased hospitalisation and psychological effects on patients and parents. Octreotide (Sandostatin; Novartis Pharmaceuticals), a synthetic somatostatin analogue, increases splanchnic arteriolar resistance and decreases gastrointestinal blood flow and thus secondarily reduces lymph flow. Methods: We evaluated the role of octreotide infusion in addition to standard therapy with fat free diet and medium chain triglyceride based milk formula in four consecutive patients with this complication after cardiac surgery. The mean age was 2.5 years with a male to female ratio of 3:1. Three children had complex cardiac anatomy which precluded a biventricular repair and were at different stages of staged palliation. The fourth child had tetralogy of Fallot. All children had high systemic venous pressures in the first 72 hours post surgery (18–24mm Hg). Intravenous octreotide infusion was started on the first week after diagnosis and continued for 4 days after the drains were removed. The dose varied between 1–4micrograms/kg/hour. Results: Intravenous octreotide infusion combined with medium chain triglyceride diet led to resolution of chylothorax within 5 days in all four patients. Conclusions: Octreotide represents an effective, non-invasive and safe adjunct to the conventional treatment of chylothorax after cardiac surgery. It can markedly reduce the requirement for chest drainage with its associated benefits.

P585
The use of angiotensine-converting enzyme inhibitors in the newborn. A five-year experience.
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The use of angiotensine-converting enzyme inhibitors (ACEI) in newborns has been preconized, but small experience in this age group and fear of its secondary effects have restricted their prescription. Aims: Evaluation of the effects of ACEI in newborns in a Pediatric Cardiology Center, from January 1995 to December 1999. Methods: Retrospective analysis of 20 newborns treated with ACEI, with a median age of 9.5 days, 12 being male (60%). Twelve babies had a gestational age of 37 weeks or more, birth weight ranging between 2000 and 4030 gr (median: 2620 gr.). All had cardiac diseases, with heart failure and or systemic hypertension. Seven newborns were submitted to surgery prior to the beginning of ACEI. Median age at initiation of ACEI was 18.5 days. Ten patients received Captopril (initial med. dosage = 0.6 mg/Kg/day; maximum: 1.35 mg/Kg/day); 10 received Enalapril (initial med. dosage: 0.18 mg/Kg/day; maximum: 0.28 mg/Kg/day). Duration of treatment with Captopril ranged from 2 and 108 days (median: 26) and with Enalapril between 2 and 32 days (median: 6.5). All patients received diuretics and in some digitalis was added. Results: Treatment was suspended in one newborn treated with Enalapril. In 4 it was reduced (three on the Enalapril

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

group and one in the Captopril group), due to hypertension. Impaired renal function was recorded in one and no serum electrolyte abnormalities were noticed. There were 5 deaths: 3 after surgery and two from septic shock. Ten of the 15 surviving patients had good results with ACEI. Conclusions: In this group, good results were achieved from the use of ACEI with improvement of congestive heart failure and no serious side effects. Although the small number of patients, we could notice better tolerance with Captopril.

PS86

Absence of coagulation factor abnormalities (protein c, protein s, antithrombin III) after the fontan procedure and its modification

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Background: Previous publications (Gramme-Dykues AH, Lancet 1998;336:1086;–1090 – Jahangiri M, J Thorac Cardiovasc Surg 1997;113:689–93) suggested coagulation factors abnormalities, principally low levels of protein C, as contributing factors to thromboembolism in children who underwent the Fontan operation. However, those studies compared patients data with the normal range of values established in adults. Methods: We analyzed the coagulation status in 16 children (age 4–14 y, mean 8.5 y) more than a year after a Fontan procedure. Their data were compared with the values in normal children, previously published by Andrew M (Blood 1992;80 1998–2005) and with the value of 16 age-matched normal children (controls). Results: Six patients had low levels of protein C (< 60%) with reference to adult normal values. However all patients had normal levels of protein C, protein S and antithrombin (AT) when their data were compared with the values reported in normal children. Their values of coagulation factors were also similar to the values measured in age-matched the normal children (p=0.40): KONDAN NL CHILDREN NL ADULTS min mean Max min mean max Min max prot C 53% 62% 95% 45% 69% 92% 63% 111% prot S 58% 86% 117% 41% 78% 114% 66% 144% AT 82% 96% 121% 74% 100% 126% 84% 128%. Conclusions: One year after Fontan-type surgery there is no deficiency in protein C, protein S and AT, when the values of coagulation factors are compared with the values of age-matched normal subjects. The mechanisms and causes of the thromboembolism observed after the Fontan procedure remains unclear.

PS87

Efficacy of Irbesartan versus Atenolol for treatment of hypertension in paediatric patients: double blind randomized prospective study

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Objective: To assess the safety and hypertensive activity of the highly selective angiotensin II receptor (AT1) antagonist Irbesartan (IRBE) in comparison to that of atenolol in paediatric patients with hypertension (seated systolic blood pressure(SeSBP) and seated diastolic blood pressure (SeDBP)>95%percentile of age). Design and methods: In a double blind study 30 patients aged 5 months to 18 years were randomized to IRBE (2mg/kg) or Atenolol (1mg/kg) once daily. Doses were doubled at week 4 for SeSBP>35% or SeDBP>20% of age specific normal blood pressure. After a 4 week placebo lead in period SeSBP and SeDBP were measured 24 hours post dose and after 8 weeks of active drug treatment. Efficacy was evaluated by determining the change from baseline as trough SeSBP and SeDBP and the proportion of patients normalized. Safety/tolerability was assessed by adverse events and by patients on a specific-symptoms questionnaire, and by clinical and laboratory evaluations. Results: Both treatments lowered BP from baseline with no significant difference between treatment groups with respect to efficacy. In patients with doubling doses at week 4, IRBE or Atenolol produced further BP lowering. Nonparametric testing revealed a change of SeDBP of 12 mmHg (p<0.001) and SeSBP of 9 mmHg (p<0.001). BP normalized in 60% of patients with IRBE versus 46% with Atenolol. Adverse effects were lower in the IRBE group. No patient had to be withdrawn from the study in either group. Conclusion: In the first controlled study in paediatric patients, Irbesartan was shown to be an effective and well tolerated antihypertensive treatment in single daily dose. Effectivity is comparable to Atenolol, lowering of blood pressure is dose dependent in both groups.

PS88

Own experiences with thrombolytic therapy with rt-pa in children with peripheral vessels thrombosis and pulmonary embolism

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This report is the continuation of our previous presentation in 1997(Hawaii) and 1999(Barcelona) concerning thrombolytic therapy with rt-PA(Acrylise) in children with thromboses in the course of CHD. Now we present our experiences with rt-PA with Acrylise in 12 children aged 14 days to 17 yrs with peripheral veins (p.v.), arterial thromboses (p.a.t.) and pulmonary embolism (p.e.). The thromboses were diagnosed by echo-dopplers or cardiac catheterisation and angio). Indications for rt-PA were as follows: 1. P.v. or p.a. thromboses after diagnostic (N= 4), after interventional catheterisation (N= 4). 2. P.e. in children with cyanotic CHD (N= 4). In peripheral vessels thromboses doses of Acrylise were 0.01 – 0.1 mg/kg/h. In 7 pts Acrylise was given i.e. p.v., in 1 pt directly to cardiotid vessel (two boluses each 0.04 mg). In 2 pts with p.e. Acrylise was given directly to pulmonary artery in two boluses (0.05 and 0.1 mg/kg) and later was continued in systemic vein during 1– 10 days. In 2 pts only to systemic vein (0,05– 0,1 mg/kg/h) during 2–7 days. Results: complete resolving of thrombi were achieved in 6 pts (75%) with peripheral vessels thromboses, in 2 pts (50%) with p.e., partial in 2 pts (1 pt with p.v. and 1 pt with p.a.t.), no resolving in 3 pts. Conclusions: 1. Rt – PA (Acrylise) in doses 0.03–0.1mg/kg/h is very effective and safe thrombolytic drug in children with vessels thrombosis. 2. The best effect of rt was achieved when Acrylise was given in boluses directly to occluded vessels.

PS89

Effectivity of Bisoprolol treatment in children with hyperkinetic heart syndrome

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Background: Authors evaluate the use of Bisoprolol in 52 children, aged 1 mo to 18 years. That study was performed in children with symptomatology of criteria for minimal circulatory hyperkinesis. Methods: The study included 32 children with various forms of arrhythmias, 20 patients with essential form of hypertension. The patients were investigated by noninvasive methods at rest at baseline and following 24 hours and 6 weeks treatment. The diagnosis was documented on electrocardiograms and echocardiographic analysis in each case and confirmed by Holter studies in some children with cardiac arrhythmias and 24 h blood pressure monitoring in some children with systemic hypertension. The effective dose of Bisoprolol ranged between 0.05 mg and 0.1 mg/kg/day. Results: The Bisoprolol treated patients responded favourable as expressed by improved electrocardiographic and hemodynamic parameters. Bisoprolol decreased heart rate to a significant extent. The reduction in heart rate during the treatment with Bisoprolol was 23 beats/min. The reduction of systolic blood pressure was 23 mmHg and reduction of diastolic blood pressure was 14 mmHg. Bisoprolol produced a significant lowering of the cardiac output, end-diastolic volume, stroke volume and the mean systolic ejection rate, thus indicating a general decrease in contractility. In both groups of the patients with symptomatology of minimal brain, Bisoprolol considerably reduced the heart work. Conclusion: Results show that in many children with systemic hypertension fulfilled criteria on hyperkinetic syndrome, high blood pressure and hypokinesis can be controlled on a beta – blocker alone. In patients with prolapse mitral valve which are associated with various forms of tachyarrhythmias, Bisoprolol had a beneficial effect on cardiac arrhythmias and on hyperkinetic syndrome.

PS90

Percutaneous endoscopic gastrostomy (peg) feeding in patients (pt's) with congenital cardiac disease

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A retrospective study comparing complex cardiac pt's who underwent PEG and pt's with similar problems who were fed orally, to determine the effect of PEG on growth parameters. From Jan 1995 to Nov 1999, 37 cardiac pt's (22 f, 15 m) underwent PEG. The procedure was performed at a median 259 days (range 1 – 858). 35 pt's (94.6%) required nasogastric tube nutritional supplementation prior to gastrostomy. The population is subdivided into 3 groups: I) 14 pt's (37.8%) with O2 saturation less than 95% at the time of the PEG, II) 18 pt's (48.6%) with O2 saturations more than 95%, III) 5 pts (13.6%) with major multisystemic problems. We compared the 3 groups with 3 control

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

groups matched for age, sex, syndrome, systemic problem, heart disease, surgical or interventional procedures and follow up. We evaluated the variations of the body weight expressed as Z score before PEG and at a median follow up of 291 days (range 19 – 1481). The Z-score for weight improved in pts undergoing PEG nutritional supplementation (I): 57%, II): 68.9% and III): 80%. The median Z-score improved significantly in each study population group and decreased in each control group who did not benefit from nutritional support. We have demonstrated that the PEG procedure is safe and aids nutritional supplementation of cardiac pts who have an increased metabolic rate and difficulty achieves adequate voluntary oral calorie intake.

P591

Cardiac output (CO), peripheral vascular resistance (PVR) and atenolol therapy (AT) in hypertensive children after aortic coarctation repair (ACr).

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CO and PVR are the main hemodynamic parameters (HP) that establish the blood pressure (BP) values. Aim of the study: to assess the effect of AT on these BP determinants. We studied 12 children (P) after ACr, mean age 15.2 years, divided into two groups: A (nP not on antihypertensive therapy, 3m, 3f) and B (6P on AT; 4m, 2f), both the P groups had the same age (respectively 15 vs 15.5 yrs) and BSA (respectively 1.41 vs 1.55 sq.m., p=n.s.). They performed an exercise testing (ET) on the treadmill; parameters recorded at rest (r) and at peak (p) exercise: time of exercise (TE), heart rate (HR), BP, CO (rebreathing method), cardiac index (CI) and PVR. Student's t-test was used to compare the data of the two groups. P in group B, compared to the group A, showed not significant differences for the same TE (10 vs 9.9 min), the lower HRr (97 vs 109 bpm) the higher COI and COp (5.2 vs 4.9, 10.4 vs 8.8 L/m'), the higher PVRr and lower PVRp (1497 vs 1411; 806 vs 881 dine*sec/cm5). higher SBPr and SBPp (132 vs 129, 180 vs 155 mmHg), the same DBPr and DBPp (72 vs 69; 84 vs 84 mmHg); significant differences were found for the group B only for the lower HRp (155 vs 174 bpm, p=0.03). Our data suggest that AT normalizes resting HR and BP but the HP are the same of P not on AT and not so in the healthy condition (Pediatric Cardiology 2000; 21: 6 pag 590, n 14)

P592

Application of home oxygen therapy to children with congenital heart disease

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Although home oxygen therapy (HOT) is well established as a treatment in adult patients with chronic lung disease (CLD) associated with pulmonary hypertension (PH) or severe cyanosis. Therefore, we investigated the clinical course of children underwent HOT and evaluated the effect of it on these cardiac patients, and compared with pulmonary hypertension without cardiac disease. Patients: Twenty-four patients underwent HOT were included, primary or porto-pulmonary hypertension in 6 (PH); PH associated with CHD in 7 (CPH; post-total correction in 5, Fontan type in 2) cyanotic heart disease in 9 (CyD) and heart failure after Fontan operation in 2 (CHF). The averaged age and percutaneous oxygen saturation at the beginning of HOT in each group were 15.2, 4.0, 7.8, and 6.7 years old, and 98, 91, 73, 92% respectively. Results: In PH, 3 patients were already dead and other 3 were still on without significant improvement of symptom. In contrast to PPH, 5 patients in CPH who were after total correction could stop the use of HOT because of the improvement of residual PH (duration of use: 1.2+/-0.4 yr). However, it was an clinical effect on both patients with Eisenmenger. In CyD, 2 patients were dead during HOT. In other 2 patients, HOT was discontinued because of the following favorite operation. In this group, 6 of 9 patients (67%) felt the improvement of clinical symptoms such as head ache and chest pain. In CHF, one patient could stop its use because of clinical improvement. These results suggest us that HOT in various CHD associated with PH or cyanosis may be useful for the improvement of clinical symptom.

P593

Prospective evaluation of cisapride on corrected QT interval in infants

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Purpose: To evaluate the effect of cisapride on QTc interval. Methods: We prospectively studied infants receiving cisapride (0.8 mg/kg/day) for clinical indications. Infants of any gestational age were included. Infants with baseline corrected QT interval (QTc) > 450 were excluded. Electrocardiograms were obtained at baseline and at 3, 5, 7 and 14 days after initiation of cisapride. QTc >= 450 was considered prolonged. Independent variables included gestational and postnatal age, birth weight, medications inhibiting cytochrome P-450 3A1 enzyme system, use of cimetidine, ranitidine, or theophylline, serum protein, albumin, AST, ALT, direct bilirubin, electrolytes and calcium. Results: Fifty infants (6 term, 44 preterm) completed the study; none developed arrhythmias. Baseline QTc (401 +/- 20) showed no difference for gestational age. There was no correlation between prolonged QTc and baseline QTc or any study variables. Fifteen infants (15/50, 30%) had prolonged QTc (PQT) at some time following cisapride initiation. 35 did not (35/50, 70%) (NPQT). There was no difference between the two in baseline QTc or study variables. However, we noted that infants with QTc >= 2 SD above the baseline QTc on day 3 were more likely to subsequently develop prolonged QTc (p < 0.0001). Timing: monitoring discontinued cisapride in 5 PQT infants (5/15, 33%, highest QTc 526), all 5 infants subsequently normalized QTc. The remaining PQT infant (10/15, 67%, highest QTc 494) continued receiving cisapride without increase (QTc 463) normalizing QTc by 14 days of treatment. Conclusion: QTc on three days following cisapride initiation may predict subsequent development of prolonged QTc. Many infants receiving cisapride may develop prolonged QTc, however most will undergo QTc normalization by day 14 despite continued cisapride use. Cisapride (0.8 mg/kg/day) may be safe to use in a NICU setting.

P594

The role of capillary whole blood monitoring in children on oral anticoagulation with CoaguCheck-Plus

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Objective: to determine the accuracy, reliability, safety and acceptance of a whole blood prothrombin time/international normalized ratio (PT/INR) monitor (CoaguChek-Plus) in children on oral anticoagulant therapy. Study design: capillary PT/INR was measured using the portable coagulation monitor CoaguChek-Plus (CCP) and compared with venous PT/INR assayed by Thromborel S. We compare 180 INR measurements from 105 patients on oral chronic anticoagulation therapy. Patients were assigned to one of three groups: group A: CCP-INR < 2.0, group B: 2 < CCP-INR > 3.5; and group C: CCP-INR > 3.5. Results: A good correlation was found between INR of CCP and Thromborel S in whole group (r = 0.845, p < 0.05) and group B (r=0.858, p<0.05) with sensitivity of 0.95 and specificity of 0.93. But poor correlation was found in group A (r=0.768, p=0.05) and no correlation in group C (r=0.501). We also calculated specificity and sensitivity of CCP-INR assessment. We found high sensitivity in group A (0.96) but lower in group C (0.85), while specificity of CCP was very low in group C (0.68) and group A (0.77). CONCLUSION: In children on oral anticoagulation capillary whole blood INR measurement with CCP is reliable and accurate when compared with standard venous measurement only if INR is inside the therapeutic ranges (group B). But CCP-INR show no correlation with standard venous INR measurement when INR > 3.5 and very poor correlation with low specificity of test when INR < 2. We conclude that INR measured with the CoaguChek-Plus monitor did not appear to be suitable for the management of children on chronic anticoagulant therapy.

P595

[51]Cr and [131]I radioisotopes as useful tools for assessing the circulatory overload in Eisenmenger syndrome

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Accurate measurements of circulating blood volumes are essential for proper management of patients with Eisenmenger syndrome (ES). Using [51]Cr and [131]I as radiotracers, we determined the red cell volume (RCV), plasma volume (PV) and total blood volume (TBV) in 21 ES patients (10 female) aged 6 to 53 years, all potential candidates for therapeutical hemodilution. Measured volumes were increased in patients as compared with values predicted on the basis of body weight: RCM 3,282 ± 1,200 vs. 2,324 ± 1,026 mL, p < 0.0001; PV 2,297 ± 659 vs. 1,594 ± 534 mL, p = 0.0001; TBV 5,549 ± 1,577 vs. 3,919 ± 1,445 mL, p < 0.0001. TBV was differentially characterized as a function of body weight in patients vs. controls (r^2 = 0.44, p = 0.0011 and r^2 = 0.97, p = 0.0004, respectively). Successful hemodilution planned on the basis of measured volumes was followed by a significant improvement in hematocrit, RCV and TBV seven days after treatment. Thus,

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

proper management of hyperviscosity and circulatory overload in ES may be planned on an individual basis taking into account blood volumes measured directly using appropriate radioisotopes.

P596
Clinical effects of Colforsin daropate hydrochloride in pediatric cardiac surgery
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Colforsin daropate hydrochloride increases cAMP in a cell to activate directly the adenyl cyclase which is synthase of cAMP existing in cell membrane not via receptor and has a positive inotropic action and a vasodilatory action. This unique agent is recently available in Japan. The purpose of this study is to examine clinical effects of Colforsin daropate hydrochloride after open heart surgery for congenital heart disease. Eight children underwent open heart surgery for congenital heart disease. Main diagnosis was VSD in 5 and DCRV in 3. The average age was 8.8 years. Colforsin daropate hydrochloride was administered intravenously at a rate of 0.25 g/kg/min for 1 hour after operation. Hemodynamic changes were measured before, after, 2 hours after and 12 hours after its administration using Swan Ganz catheter. The list of hemodynamic changes was shown below. Significant arrhythmia was not recognized during its administration. Colforsin daropate hydrochloride decreased pulmonary vascular resistance index (PVRI) and increased cardiac index (CI) and HR, while it decreased systemic vascular resistance (SVRI) and BP within acceptable ranges. Colforsin daropate hydrochloride may be a useful agent after pediatric cardiac surgery with pulmonary hypertension.

P597
Continuous intravenous furosemide in haemodynamically unstable children after cardiac surgery
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Objective: The commonly used continuous IV furosemide dosing schedule after cardiac surgery in children is largely empirical and may not be optimal. This may even be more outspoken in children after cardiac surgery who are haemodynamically unstable, and in whom transient renal insufficiency may occur. A study was performed to get an impression which clinically applicable measures may be used to design a rational scheme for continuous IV furosemide therapy in children after cardiac surgery. Subjects and methods: Twelve paediatric patients (5F/7M, age 0–33 weeks) post-cardiac surgery who were to receive 3 days of continuous IV furosemide treatment, were included in an open study. Blood and urine samples were taken for furosemide, creatinine and electrolyte levels and total urinary output was measured. Furosemide in blood and urine was measured using HPLC. Results: The mean starting dose of continuous IV furosemide was 0.093 (±0.016) mg/kg/hr and was increased to 0.175 (±0.045) mg/kg/hr on day 2, and changed to 0.150 (±0.052) mg/kg/hr on day 3. Infusion rates were increased from day 1 to day 2 in 10 cases, and decreased from day 2 to day 3 in 3 cases. Urinary furosemide excretion rate was inversely related to serum creatinine levels. Discussion and conclusion: This study extends the observation of the beneficial effects of continuous IV furosemide to children who are haemodynamically unstable after cardiac surgery. As the furosemide effects are dependent on renal function, it can be hypothesized that the dosing schedule may be optimized. Contrary to the currently used dosage schedule in which the dose of furosemide is gradually increased over time, it may be more rational to start with a higher dose and adapt this dose (downward) guided by the observed effect (urine output).

P598
Preoperative use of inderal in patients with Tetralogy of Fallot does not effect postoperative inotropic score
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Beta-blockade is used in some patients with Tetralogy of Fallot as prophylaxis against hypercyanotic spells prior to surgical repair. Beta-blocker therapy has been linked to catecholamine resistance in postoperative patients. The aim of this study was to determine if preoperative inderal therapy had an effect on postoperative variables in patients with Tetralogy of Fallot. Methods: From November 1996 to June 1999, 53 patients underwent surgical repair of Tetralogy of Fallot at our institution. Preoperative, intraoperative and postoperative variables were retrospectively reviewed. Results: Fourteen of the 53 patients with Tetralogy of Fallot were treated as an outpatient with inderal prior to surgical repair. Nine of the 14 patients were placed on inderal secondary to hypercyanotic spells, 2 of the 14 patients required preoperative intubation and inotropic support. Intraoperative variables were similar in the inderal group and the non-inderal group. There was no difference in postoperative inotropic scores upon arrival in the CICU (mean ± SD 5±2 vs. 5±3, p>0.05, inderal vs. non-inderal) or 12 hours later (8±5 vs. 7± 5, p>0.05 inderal vs. non-inderal). Four patients (2/14 inderal patients and 2/39 non-inderal patients) required temporary pacing in the early postoperative period. The postoperative length of intubation (63±40 vs. 47±52, p>0.05, inderal vs. no inderal) and hospital stay (8 ± 5 days vs. 8±3 days, p>0.05, inderal vs. no inderal) was similar in both groups. Conclusions: Preoperative inderal therapy was not associated with an increased postoperative inotropic requirement. Postoperative length of intubation and hospital stay was not prolonged in patients on inderal therapy.

P599
Enalapril in infants with congestive heart failure secondary to a left-to-right shunt
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This study was undertaken to investigate the effects of enalapril on the clinical and hemodynamic findings in infants with intractable heart failure secondary to a left-to-right shunt. In 18 patients aged 2–28 (mean 10±7) months with heart failure secondary to a left-to-right shunt refractory to at least one month of therapy with digoxin and furosemide, enalapril 0.5 mg/kg/day was added as treatment. Clinical and echocardiographic findings were assessed before and 15–67 days after the initiation of enalapril therapy. The liver size and dyspnea (interaction score) decreased significantly (p=0.03 and 0.02, respectively). An increase in body weight in 61%, and decrease in respiratory rate in 55%, heart rate in 67%, cardiothoracic ratio in 78% of the patients were observed, but the differences were not significant. Qp and pulmonary artery pressure decreased significantly (p=0.02 and 0.01, respectively); Qp/Qs decreased in 44%, increased in 16% and did not change in 11% of the patients, the overall decrease in Qp/Qs was not significant. A decrease in Qp/Qs and wall stress was noted in 72% of the 8 patients in whom improvement in clinical findings was observed. No differences in the echocardiographic measurements of LV size, volume, mass, meridional wall stress, LA size, CI, systolic and diastolic functions of the heart could be documented. No side effects or changes in biochemical and hematologic parameters were detected during treatment. Enalapril may serve as a useful adjunct to conventional therapy and a bridge to operation in patients with severe heart failure secondary to a left-to-right shunt awaiting surgery.

Embryology/Developmental Cardiology, Morphogenesis/Morphology

P600
Evolution of aortic arches from fish to humans
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Throughout evolution, the 4th and 6th aortic arches in lower vertebrates have been either paired or right-sided, whereas in mammals these arches became left-sided due to the unique placental hemodynamics and bronchopulmonary expansion. The systemic arterial duct, essential in fetal mammals, was displaced by the right aortic arch filling the restricted space above the unilateral right eparterial bronchus. With no left eparterial bronchus, the arterial duct, aortic arch and descending aorta gravitated to the left. Where a fetal systemic arterial duct is not essential, as in tetralogy of Fallot or continued truncal interruption, a nonobstructive right aortic arch continues to occur. With normal cardio-respiratory anatomy, a right arterial duct remains viable with cervical search extension. Otherwise, right aortic arch becomes obstructive as it connects to a left arterial duct behind the esophagus, or through patency of the left dorsal root (Komstedt 1). Right or double aortic arch in humans is viewed as reversion to morphology of nonplacental vertebrate aortic arches.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P601
Differential expression of the cardiac alpha-actin gene in neural crest ablated chick embryo
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Hemodynamic abnormalities in the chick embryo have been documented after neural crest ablation. Ventricular dilation may contribute to malformation of outflow tract. To explore the relation of neural crest ablation to depressed contractility, suppression subtractive hybridization was used to discover messages decreased or no expressing in outflow tract tissues with or without cardiac neural crest cells. Cardiac neural crest was ablated by electric stimulation in 51 chick embryos, 48 chick embryos were as control. A subtractive library was constructed and cDNA array was used to identify significant difference, then sequencing was done. 261 clones were constructed. 83/261 clones were identified to be significantly decreased or no expressed by cDNA array. 68 clones were sequenced. 18 kinds of cDNA fragments were identified. 7/68 were known as Gallus cardiac Alpha-actin gene. The results of this study indicate that ablation of cardiac neural crest could cause decreased expression of cardiac Alpha-actin gene which might be responsible for depressed contractility.

P602
Echocardiographic studies on cardiac functions in the mice with cx43 gene defects
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Connexin 43 (Cx43) gap junction gene is one member of the connexin multigene family recently shown to be crucial in heart morphogenesis. This study was to characterize heart defects which may arise in the Cx43 knockout mice (Cx43KO), transgenic mice overexpressing Cx43 (CMV43), and transgenic mice expressing Cx43/b-galactosidase fusion protein (FZ). In utero Doppler echocardiography was carried out to evaluate the mouse embryonic cardiac abnormalities in these mouse lines. The studies showed that cardiac defects in Cx43KO, CMV43 and FZ mice were mainly associated with the obstruction of right ventricular outflow tract and abnormal myocardial development as reflected by the increased peak systolic ejection velocity (PSV) and by the monophasic diastolic inflow pattern respectively. Echocardiographic findings were consistent with the morphological changes including pulmonary stenosis and right ventricular hypertrophy or enlargement. We concluded that Doppler echocardiography is of importance in the study of heart defects in mice with the gain or loss of Cx43 function.

P603
Secundum atrial septal defect due to malformations of the septum primum or secundum: implications for transcatheter closure
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The morphogenetic concept that secundum atrial septum defects (ASDII) are the result of a malformed septum primum (valvula foraminis ovalis, VFO) cannot explain all morphological variations. Components of the atrial septum were studied in normal heart specimens (n=22, age 0–43 yrs) and with an ASDII (n=58, age 0–49 yrs). Placement of an Amplatzer Septal Occluder (ASO) was estimated in each case. Normal specimens (closed/open foramen ovale) showed little variation of the superior limbus (SL) forming the muscular rim of the oval fossa from anterio- to posterosuperior. The VFO with variable thickness formed the floor of the oval fossa without a muscular rim. In 50 specimens (group I) the ASDII was due to malformation of the atrial septum primum with normal size/position of the SL, group Ia (n=11) with an absent VFO and lack of posteroinferior rim; group Ib (n=17) with a large ostium secundum (OS) causing a central defect with sufficient rims; group Ic (n=19) with one or multiple fenestrations in the VFO in addition to the OS causing variable defects, group Id (n=3) with a posteriorly positioned OS with lack of posteroinferior rim. Hitherto undescribed, 8 ASDII's (group II) were the result of a small/absent SL (septum secundum) lacking the superior rims. The size/position of the VFO was normal in this group. Stable ASO placement (40%) was possible in 27/50 of group I and none of group II. Conclusion: Different types of ASDII's can be discriminated based on malde-velopment of either atrial septum primum (VFO) or septum secundum (superior limbus), which is important in view of transcatheter closure techniques.

P604
A 3d echocardiographic study of the postnatal changes of right and left
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A 3D echocardiographic study of the postnatal changes of right and left ventricular volumes and contractility in normal term neonates. To evaluate the postnatal changes of right and left ventricular volumes and contractility, we performed serial 2D and 3D echocardiographic examinations in 21 normal full term neonates at 0–6, 24–48, and 48–72 hours after birth. Dynamic 3D echocardiographic images of LV and RV were obtained from a transthoracic apical view while moving the probe freely by hand, with acquisition gated to control for ECG and respiration (EchoScan, Tomtec). Results: Heart rate did not change significantly during the study period. LV end diastolic volume correlated significantly with the diameter of ductus arteriosus (r^2 0.48, $p<0.05$). LV end-diastolic volume decreased significantly one day after birth from 1.41±0.16 to 1.27±0.15 ml/kg, corresponding to the closure of ductus arteriosus. LV end-systolic volume also decreased significantly on day 2 and day 3 with associated increase in LV ejection fraction from 54±2 to 59±2 and 62±2%. RV end-diastolic volume was 1.40±0.18, 1.30±0.14 and 1.25±0.16 ml/kg, and did not change significantly during the period. RV end-systolic volume decreased progressively from 0.69±0.1 to 0.57±0.09 ml/kg, and RV ejection fraction increased progressively from 51±2 to 59±3 on day 3. Both LV and RV output decreased steadily during the study period. Our results of 2D echocardiography for all parameters of ventricular size and function correlated strongly with that of 3D, but the mean values were generally higher in 2D measurements. We conclude that our 3D data provide a useful reference for the interpretation of ventricular function in the early neonatal period.

P605
Distinctive differences in the morphology of the right ventricular outflow tract between tetralogy of Fallot (ToF) and ToF + pulmonary atresia + patent ductus arteriosus in autopsied hearts
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Purpose: To examine the morphology of the right ventricular outflow tract in autopsied infants at National Children Hospital, 2 years old or younger who died as a consequence of Tetralogy of Fallot (ToF) or ToF + pulmonary atresia (PA) + patent ductus arteriosus (PDA). Subjects and Methods: The heart of 5 infants with ToF (Group A) and 6 with ToF + PA + PDA (Group B), who had not been operated, was examined and measured using the measurement method employed Berdon et al. in 1975. The average age, height, weight, body surface area, infundibular length (a), length of the right ventricle (b), aortic outflow tract (c), coronal septal width (d), pulmonary outflow tract (e), circumferences of the aortic external orifices (f), circumferences of the pulmonary external orifices (g), and coronal septal length (h), were determined and the ratios of a/a+b, b/a+b, c/a+b+c and g/f were calculated. Analyses were performed using Mann-Whitney's test, and a P value of < 0.05 was considered statistically significant. Results: The average values of parameters a, g, h, a/a+b, b/a+b and g/f significantly differed between the two groups. The infundibular septal length in the right ventricular was proportionally smaller in Group B than in Group A; the length of the conus septum tended to be shorter in Group B, and the diameter of the right ventricular outflow and that of the main pulmonary artery were narrower in Group B. Conclusions: There were distinctive differences in the morphology of the right ventricular outflow tract between the 2 groups.

P606
Immunolocalization of tenascin in the heart of the developing rat embryo
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Busulfan administration to pregnant rats induces cardiovascular anomalies including tetralogy of Fallot and truncus arteriosus communis. These congenital anomalies are clinically known to be often associated with anomalous coronary arteries. However, the pathological mechanisms of anomalous coronary arteries associated with these congenital defects have not been fully established. Tenascin-X, a member of extracellular matrix proteins, is suggested to be involved in the formation of the vascular channel

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

by epicardial cells. In order to determine whether bis-diamine induces abnormal coronary vascular development, we examined the expressional patterns of Tenascin(TN)-X in the hearts of the developing embryos from bis-diamine-treated mothers using immunohistochemical methods. A single dose of 200mg of bis-diamine was administered to pregnant Wistar rats at 10.5 days of gestation. The embryos were removed on 11, 12, 13, 14, 15, 16 and 20 days of gestation (ED), and used for morphological analysis of coronary arteries and immunohistochemical study using anti-TN-X. An anomalous or hypoplastic left coronary artery were detected in 92% of bis-diamine treated embryos at 20ED. The TN-X was first detected in the 11ED normal heart. In the 12ED normal heart, the TN-X was expressed in the epicardium and atrial and ventricular myocardium. During cardiac development, TN-X expression was localized to the epicardial cells that migrate into the myocardium, forming vascular channels. However, the immunolocalization of TN-X to the epicardial cells and vascular channels were not observed in the hearts of bis-diamine treated embryos in each developmental stage. This study indicated that bis-diamine induced abnormal coronary arterial development by disturbing coronary vascular channel formation from epicardial cells.

P607

The right ventricle in hypoplastic left heart syndrome

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In a series of hearts with hypoplastic left heart syndrome (HLHS) the interventricular septum (IVS) are variable in size. Sometimes the left ventricle (LV) and thus the IVS cannot be identified by gross dissection. In other cases the length of the septum can be measured by approximating the apex of the LV against the right side of the septum. The morphology of the septomarginal trabeculation (ST) and other muscular bundles of the right ventricle (RV) was studied in 25 hearts with hypoplastic LV and various combinations of aortic and mitral valvar anomalies, with or without ventricular septal defect. In hearts in which the apex of the LV approximated closely to the RV, the ST was usually adherent to the septum in the usual way. In hearts in which the LV and thus the septum could not be identified, remodelling of the ST was sometimes excessive, producing anatomic obstruction of the right ventricular outflow tract. A spectrum was present between these extremes which was deemed to be associated with the length of the septum and the degree of hypoplasia of the LV. Anatomically remodelling of the ST may have a secondary effect on the tricuspid valve (TV) and the right ventricular outflow, which can influence the outcome of surgery for HLHS. It is likely that such hypoplasia of the LV and the septum is present before delamination of the TV. Remodelling of the RV and its outflow may also be anticipated relative to the degree of hypoplasia of the septum but irrespective of other intracardiac anatomy.

P608

Different patterns of abnormal cerebral metabolism in neonates with various congenital heart diseases detected by in vivo localized 1h MR spectroscopy

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Introduction: The underlying mechanism for growth retardation and/or abnormal neuro-developmental outcome in children with congenital heart disease (CHD) has been explained mostly by the postnatal effects however the prenatal effects of altered hemodynamics in the fetus have not been investigated. In this study, we designed a study to evaluate our hypothesis that abnormal fetal circulation due to CHD affects the fetal brain metabolism as well as growth retardation. We evaluated the cerebral metabolism of neonates with different types of CHD: complete TGA (TGA), pulmonary atresia with VSD (PA), and coarctation of the aorta with VSD (COA) groups. Methods: Seventeen TGA, 12 PA, 7 COA, and 15 age-matched normals (ages = 3 ~ 20 days) were evaluated. Localized 1H- MR Spectroscopy was performed on the parietal white matter (PWM) and occipital gray matter (OGM) of the brain to calculate the [NAA/Cr], [Cho/Cr], [mI/Cr] and [NAA/Cho] ratios. Results: The [Cho/Cr] (1.44 ± 0.14, $p < 0.05$) was highest, and the [NAA/Cho] (0.61 ± 0.08, $p < 0.05$) was lowest for TGA group, in PWM and OGM, respectively. For COA group, the [Cho/Cr] (1.25 ± 0.13) and the [NAA/Cho] (0.73 ± 0.10) was not significantly different from those for the normal controls. For PA group, the [Cho/Cr] (1.34 ± 0.15) and the [NAA/Cho] (0.71 ± 0.09) were between TGA and COA groups. Conclusion: This is the first study to demonstrate that unfavorable screening of the fetal circulation and hemodynamics due to CHD can result in the deranged fetal brain metabolism and abnormal body growth. The degree of altered cerebral metabolism seems to follow the amount of oxygen content in the brain circulation determined by the reversed streaming (TGA), no streaming (PA), or normal streaming (COA) of blood flow.

P609

Ventricular aneurysm or diverticulum? Clinical differential diagnosis

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Intrathoracic ventricular aneurysms and diverticula can be differentiated by several criteria. Ventricular diverticula are always congenital, while aneurysms can be acquired as well. Histological examination show that a true congenital diverticulum is always composed of epicardium, myocardium and endocardium, while an aneurysm may either show a defect or absence of muscle. Morphologically a diverticulum is considered to have a narrow connection with the ventricle, which is wider in aneurysms (2.5). While diverticula contract synchronously with the ventricle, aneurysms show a paradoxical expansion (1,6). When first detected by angiography or echocardiography it is impossible to decide whether an intrathoracic outpouching of the ventricle is congenital or acquired (extrathoracic outpouchings must have at least a diaphragmal defect and are therefore always congenital). It is also impossible to assess the histological layers without an open biopsy. The width of the connection with the ventricular cavity as a diagnostic criterion to distinguish between diverticula and aneurysms is arbitrary due to lack of precise definition. The contractility of the diverticulum during the ventricular systole is the only objective parameter. Comparison of the time-phases outpouching of the ventricle simultaneously with the true ventricle indicates that it is a diverticulum, if it expands paradoxically during systole an aneurysm has to be considered.

P610

Morphological quantization and three-dimensional reconstruction of human embryo hearts for virtual model validation

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Three-dimensional reconstructions of human embryo hearts from synthetic images are extrapolated from two dimensional representations. To validate virtual volume geometry, we reconstructed 10–13 WOA embryonic hearts in three dimensions. Hearts of human embryos were obtained by selective collection during voluntary terminations. Cardiac blocks were fixed in a 10% formaldehyde solution, then were included in paraffin and entirely sliced from the apex on a microtome in serial sections 10 µm thick. One slide out of 10 received topographic coloration with H. E. S. (Hematoxylin, eosine, safranin). Digital images were obtained by optical microscopy. Images were pre-processed (noise reduction, contrast enhancement and semi-automatic segmentation). A voxel volume was reconstructed by piling up sections and resizing image after image. Intermediate sections were computed by interpolation to obtain isotropic volume rendering. Cardiac volumes were studied with the display tool showing internal and external areas of the heart. It was thus possible to know the size, thickness and precise position of the various of cardiac structures: ventricles, atria and large vessels, in terms of both external and internal morphologies. This new reconstruction and visualisation method makes it possible to validate models based on synthetic images. It can be used regardless of the size of embryonic hearts. So its implementation at earlier stages of embryogenesis will provide a clearer view of cardiac development.

P611

Causes of aortic root dilatation in patients with tetralogy of Fallot (TET) prior to repair

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Aim: To explore the relative contributions of a developmental defect Vs abnormal hemodynamics in aortic root dilatation (root dil.) of TETs. Do 'pink' TETs with less cyan, aortic flow have less root dil.? Do cyanosed TETs get more root dil. after a shunt? In Fig 1, M mode root in mm (y axis) in 147 studies of 99 pre-op TET <1y was plotted with the normal data of Henry et al, both expressed as a polynomial relationship to age in years (x axis). In 23 pts, root indexed to BSA (y axis) was plotted against arterial O2 Sat. (x axis) (Fig 2). In 11 TETs studied 3m before and 12m after shunt, additional non age related dilatation was a mean of 46%. Put Fig 1 here. Put Fig 2 here.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Conclusion:- Ebstein dil. in TETs is a developmental defect present from birth. It bears a minor relationship to crano-caudal blood flow prior to correction.

P612

Four-dimensional virtual pictures: a new method for understanding cardiac embryology at 35 min

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Teaching embryology is a difficult task as it requires to understand the representation and the movement of many complex phenomena. In classical embryology textbooks, this evolution is usually described by means of drawings between which it is difficult to imagine the spacial and temporal links, which are in fact the keystone of descriptive embryology. We have synthetized the bibliographic data of different textbooks of embryology. Every stage of heart evolution, according to this synthesis, has been modelled by a group of experts in cardiac embryology. Three-dimensional objects in synchronized pictures have been modelized for each anatomical structure involved in the different stages of human heart developement : fecundation, development of trilaminar germ disc, formation and folding of the primitive heart tube, morphogenesis of the heart chambers and valves, development of the aorta and the pulmonary artery. An animation wholly realized in virtual pictures, describing every process of heart development, from fecundation phase on the last cardiac events has been realized. This animation, completed by two explicative video sequences describing the different meanings and meanings involved in heart evolution. The object is realized in 3-D for this animation can be seen thanks to an open GL viewer directly integrated. Besides access to a data base including several echocardiography video sequences is possible and allows to get used to the real forms of acquisition and holography references are recorded. We have realized a 3-d produced an animation on the human embryologic development to demonstrate that using virtual pictures in teaching improves substantially the comprehension of complex phenomena. The contributions of the dynamic point of view represents a new teaching means.

P613

The morphologic nature of non-committed ventricular septal defects in specimens with double outlet right ventricle

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Biventricular correction of double outlet right ventricle (DORV) with non-committed ventricular septal defect (NC-VSD) is increasingly feasible. NC-VSD excludes a close and direct relationship of the VSD to the semilunar orifices without further anatomic definition. From the morphogenetic stance, the VSD in DORV opens into the outlet of the RV, which implies proximity to a semilunar orifice, unless structures in the outlet preclude this. We propose to call this variant a "non-directly-committed VSD", in keeping with the Leiden terminology. A true NC-VSD excludes all forms of outlet VSD. We examined the Leiden collection of hearts with DORV focusing on the location of the VSD and the space between it and the semilunar orifices with the aim defining the morphologic variants comprising the different forms of NC-VSD. Of 67 specimens, there were 55 in which the VSD was committed to one or both semilunar orifices. In 8 specimens the non-directly-committed VSD opened into the outlet portion of the RV, the distance between the VSD and semilunar orifice being extensive due to a broad anterior infundibular fold (VIF) or long outlet septum. There were 4 specimens with true NC-VSD. An atrioventricular septal defect without extension to outlet in 3 cases and an isolated inlet VSD were found in one case. the VIF and outlet septum having fused to form a structure similar to a supravenous solar crest. The surgical implication is twofold. the tricuspid valve is interposed between the NC-VSD and the semilunar orifice. The non-directly-committed VSD opens into the outlet of the RV, the relationship to the semilunar orifice being influenced by the extent of the VIF and outlet septum.

P614

Immunolocalization of cardiotrophin-1 and bromodeoxyuridine (BrdU) in embryonic rat hearts

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Cardiotrophin-1 factor capable of inducing cardiomyocyte proliferation, was detected in embryonic mice heart. However the role of CT-1 has not been fully established in the cardiac development. In order to examine the role of CT-1 and cardiomyocytic DNA synthesis in an embryonic heart, we performed immunohistochemical studies using CT-1 antibody and bromodeoxyuridine(BrdU) flash labeling in 10–17 embryonic day(ED) rat heart. BrdU was intraperitoneally administered 3 hours before sacrifice, and the heart was fixed in 4% paraformaldehyde for 12 hours and then embedded paraffin. Serial sections cut at 5 mm were incubated with CT-1 and BrdU antibody. Weak CT-1 expression was first detected in the epicardium at 10ED. CT-1 was expressed in the atrial ventricular and outflow myocardium at 12ED, which continuously seen until 14ED. At 15ED the outer layer of the ventricular wall showed less CT-1 expression than the septum or ventricular trabeculae, which was more obvious at 17ED. Although immunolocalization of BrdU positive cells were well coincident with that of CT-1 positive cells in the early embryonic days, less number of BrdU positive cells were seen in the ventricular trabeculae than the outer layer of ventricular wall in the later developmental stages. These findings suggested that CT-1 might induce early cardiomyocytic DNA synthesis and subsequent proliferation.

P615

The septomarginal trabeculation as a marker of the ventricular septal defect-great vessel relationship in double outlet right ventricle. A morphologic study

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The septomarginal trabeculation (SMT) is a characteristic structure of the right ventricle (RV) and a useful landmark for the morphologist. In hearts with double outlet RV (DORV), the SMT may help the surgeon to delineate the relationship of the ventricular septal defect (VSD) to the great vessels. Objectives: the recognition and intactness of the SMT and whether the relationship of the limbs of the SMT to the outlet septum is predictive of the relationship of the VSD to the great vessels. Method: morphological examination of specimens with DORV focusing on the SMT, the ventriculo-infundibular fold (VIF), the VSD and outflow tracts. Outflow tracts were designated antero-posterior if the anterior limb of the SMT was closely related to outlet septum, and side by side if the VIF and posterior limb of the SMT were closely related to outlet septum, according to M.V. de la Cruz. Results: In 44 of 67 specimens examined, the SMT was recognizable and intact. There were 27 specimens with antero-posterior outflow tracts (23 subaortic and 4 doubly committed VSDs) and 16 with side by side outflow tracts (16 subpulmonary VSDs). In one case the outlet septum passed through the VSD to the left ventricle. In 14 specimens the SMT was not intact but remained recognizable. Eight could be designated as having antero-posterior outflow tracts, and 6 side by side outflow tracts. In the remaining 9 specimens the SMT was not recognizable. In 44 of the 67 specimens other congenital abnormalities were present, including all cases in which the SMT was not recognizable. Conclusion: For the surgeon a recognizable SMT is a useful marker of the VSD-great vessel relationship. For pre-operative assessment 3-D echocardiography may be helpful in delineating the SMT.

P616

Time-lapse study with a high speed video camera in the early embryonic chick heart for a better understanding and visualization of cardiomorphogenesis

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The fascinating dynamic process of heart development is gaining a renaissance in capturing the imaginations of developmental biologists and clinicians. But understanding and visualizing cardiomorphogenesis from the early heart tube to the four chambered organ still remains a complex developmental process including the steps of looping, convergence, alignment, wedging and septation. Time-lapse studies (TLS) are widely used to present developmental phenomena and provide a useful tool for a better understanding of intricate processes. They give motion pictures that compress long developmental time periods. With this study we present a time lapse movie on the development of a stage 14 chick heart in shell-less culture over 10 hrs. Methods: At stage 10 the intact chick eggshell content was transferred in a hexagonal polystyrene weighing boat on a petri dish with water and incubated at 37°C. Twenty four hrs later, at stage 14, the chick embryo was filmed every 30 min for 10 sec with a digital high-speed video camera (HSVC) mounted on a stereomicroscope over 10 hrs. Images were transferred from BAYER to TIFF and later to JPEG files to edit and create a movie with Quick Time Player (QuickTime Version 4.1.2) upgraded to

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

QuickTime Pro. Each 30 min event shows one heart cycle, 20 events (2400 image frames) demonstrate a continuous short movie of heart development over 10 hrs. Results/Conclusion. TLS with a HSVC could provide a new tool for better understanding and visualization of cardiomorphogenesis. The produced short movie will be presented at the meeting. Future studies are planned to employ the method in different experimental settings which lead to malformations of the heart to visualize the events of dysmorphogenesis step by step and 4dm heart development over 24, 48 and 72 hrs

P617
Aortic arch anomalies in congenital heart disease
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Aim To study the incidence of aortic arch anomalies in CHD and describe the variations. A retrospective analysis of 375 consecutive angiograms which had arch views, performed between Jan 1998-Jan 2000 were analysed. Age range was 20days-36 years. Male Female ratio was 1.6:1. RI aortic arch was seen in 45 (12%) cases. Of these, 30 had conotruncal anomalies. (TOF 18 (41%), VSD with pulmonary atresia 8(17%), DORV 4(8.8%), and 10 others. Six patients, (1.3%) of the right arch and 45 patients (13.6 %) of left arch had branching anomalies. Aortic arch branching anomalies included common origin of all arch vessels (3), common origin of first 2 vessels- 28(61%), separate origin of 4 arch vessels-11 (23%), abnormal origin of retroclavian artery (4), abnormal L subclavian (3)- all with a high cervical right arch. There were 2 patients with double aortic arch - one of whom had a VSD and another, truncus arteriosus. Majority of the abnormalities were in the VSD pulmonary atresia and the TOF groups. Conclusion Aortic arch branching abnormalities are commonly encountered with congenital heart disease, especially with conotruncal defects, and require careful evaluation, prior to surgery

P618
Immunolocalization and function of focal adhesion kinase and paxillin during cardiac looping of chick embryo
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To understand the mechanism of normal cardiac looping and myofibrillogenesis, we have reported 3D observation of myofibril formation, cell-cell and cell-matrix adhesions, and tyrosine phosphorylation during looping of whole-mounted chicken embryonic heart by using confocal microscopy (Shiraishi et al., Anat Embryol). We found that the looping simultaneously progresses with formation of myofibrils and that arrangement of myofibrils develops in close association with changing cell- cell and cell-matrix adhesions. In this study we performed 3D observation of focal adhesion kinase (FAK) and paxillin immunohistochemical localization before and during looping (4- to 15-somite stages). We also perturbed the function FAK by treating embryos with antisense oligonucleotides or FAK-related non-kinase (FRNK). Before myofibrillogenesis (4 to 7-somite stages), faint and particulate pattern of FAK and paxillin was randomly expressed at the luminal of the inner myocardial cell layer facing the cardiac jelly. During initial stages of myofibrillogenesis (8 to 9 somite stages), fibrillar pattern of FAK and paxillin appeared at terminal of stress fiber-like thick actin bundles which were recognized as an initial stage of myofibrillogenesis. During active stages of myofibrillogenesis (10–13-somite stages), paxillin was also localized at Z-bands of immature striated myofibrils, while expression of FAK decreased. At the 14–15 somite stages or later, the expression of both FAK and paxillin diminished especially at sites of mature striated myofibrils. Treatment of chicken embryos with antisense oligonucleotide against FAK or microelectroporation with FRNK cDNA inhibited circumferential myofibrillar alignment in the inner layer, resulting in abnormally ballooned heart tube. These results suggest that paxillin is involved in the initial arrangement and sarcomere formation, while FAK is involved in the initial arrangement of myofibrils, and that FAK is essential for the normal myofibrillar alignment in the inner cells and the n

P619
The first echocardiographic, angiographic and pathology description of the Topsy-Turvy heart
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Topsy turvy heart is a unique form of cardiac malposition with 180 degree rotation of the base-apex axis. We report the first integrated echocardiographic, angiographic description with pathology specimen correlation (exhibited) of this unique cardiac curiosity. The position of the heart in the thorax is pathognomonic. The apex of the heart is directed cranially and pointing to the left shoulder. The great arteries arising out of the heart just above the diaphragm are directed caudally. There is no ascending aorta, it descends into the abdomen soon after arising from the heart. The arch branches are long and ascend the thorax. The pulmonary artery branches are almost at the level of the diaphragm. Intracardiac anatomy shows situs solitus of the atria with concordant atrio-ventricular and ventriculo-arterial connections. The systemic and the pulmonary veins enter the respective atria normally. Patent foramen ovale was present. The left atrium lies inferior and to the left of the right atrium. The right ventricle (RV) inflow is superior (and to the left) of the left ventricle (LV) and the RV outflow posterior and to the left of the LV. The LV outflow is directed posteriorly and inferiorly. The ventricular septum is intact. Both the outflow tracts point inferiorly. The pulmonary artery is to the left and anterior of the aorta. A large aorto-pulmonary window is encountered. The proximal aorta, which develops, gives off the arch branches, which ascend from the diaphragm to the neck. The dilated pulmonary artery divided into the branch pulmonary arteries almost at the level of the diaphragm. The topsy turvy heart, a unique variety of cardiac malposition, may more appropriately be referred to as the base-apex inversion" of the heart."

P620
Congenital absence of the anatomic left atrium with mitral valve atresia (MVA) and supra-a developmental complex
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We describe 2 cases of complete absence of the anatomic left atrial chamber in situs solitus with no identifiable left atrial appendage or venous pouch associated with MVA and TAPVR. DORV with subaortic VSD, hypoplasia of the LV and HDA were present in each case. The TAPVR was infradiaphragmatic in one case and associated with severe valvar and subvalvar PS with hypoplastic PAs. In the other case the TAPVR was to the coronary sinus with obstruction and associated with juxtaductal coarctation of the aorta with severe tubular hypoplasia of the aortic arch and bicuspid AV with AS. We believe the association of TAPVR and early presence of MVA provides the substrate for the lack of development of the anatomic LA. Although rare we can anticipate this developmental complex to be observed in mitral valvar atresia, double inlet ventricle, other abnormalities of ventricular-arterial connection (main pulmonary atresia or interrupted aortic arch) or LA isomerism.

P621
Histology of the aorta, and aortic root dilatation in adults with tetralogy of Fallot
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BACKGROUND: Aortic root dilatation -with or without aortic regurgitation- is a well-described feature in patients with Tetralogy of Fallot and pulmonary atresia or atresia, contributing to late morbidity. Increased aortic stroke volume, particularly in patients with Pulmonary Atresia is thought to be the main pathogenic mechanism. We hypothesized that intrinsic abnormalities of the aorta are also contributory to aortic dilatation. We evaluated histologic changes in the aortic wall and its possible relationship with aortic dilatation seen in adult patients with Tetralogy of Fallot. METHODS: We examined clinical, echocardiographic and histologic data of all adult cases of Tetralogy from our cardiac morphology database. Aortas were studied by light microscopy using haematoxylin-eosin, elastic van Gieson and alcian blue stains. Cystic medial necrosis, fibrosis and elastic fragmentation were classified from 0=absent, 1=mild, 2=moderate and 3=severe. RESULTS: Nine cases of Tetralogy (5 pulmonary atresia, 3 pulmonary stenosis) who died at a median age of 45 (range 20–57) years were identified. Cystic medial necrosis changes > 2 were present in 3 (33%) patients (1 with pulmonary atresia, 2 with previous repair). Aortic root diameter on last echocardiogram in these 3 patients was greater (4.5±0.5 vs 3.7±0.1cm, P=0.02) compared to the remainder. Eight patients had also fibrotic changes > 2 in the aorta (6 of them with Elastic fragmentation > 2). Cystic medial necrosis changes, found also in the ascending and descending thoracic aorta, were not related to age at death, presence of pulmonary atresia, previous repair and degree of aortic regurgitation. CONCLUSIONS: Cystic medial necrosis changes, similar to those seen in patients with Marfan syndrome, are common in adult patients with Tetralogy of Fallot. These changes were seen in association with a larger aortic root on echocardiogram, and were not related to age at death and to different morphological or surgical substrates. Assessment of aortic root size should be part of the routine follow-up of adult patients with Tetralogy.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P622

Persistent fifth aortic arch – an ignored and underestimated disease

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Persistent fifth aortic arch with systemic-to-pulmonary connection is an extremely rare congenital cardiovascular malformation. Less than 10 cases were reported in the literature. All previous reported cases have either been cases of pulmonary atresia or aortic atresia, and the existence of a fifth aortic arch was a benefit to the underlying great vessel anomaly. We report two cases of persistent fifth aortic arch with systemic-to-pulmonary connection. In our two cases, there is a huge vessel (the same size of ascending aorta) arising from the distal ascending aorta just beneath and opposite the origin of the innominate artery and reuniting at the superior margin of the pulmonary trunk. Our two cases did not have associated pulmonary atresia or aortic atresia, and the large persistent fifth aortic arch resulted in a large left-to-right shunt with severe pulmonary hypertension and heart failure. The first case received successful surgical repair and is doing well now. The second case expired before surgical repair and the post mortem findings will be showed and discussed. We report these two cases because the size, the associated anomaly and the hemodynamic event are complete different from the reported cases in the literature.

P623

The first septal perforating artery in common arterial trunk: anatomical relations to the ventricular septal defect and potential damage during surgery

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The surgical correction of common arterial trunk (CAT) sometimes requires enlargement of the ventricular septal defect (VSD), in order to connect properly the new aorta to the left ventricle. Since the resection must be done at the antero-superior margin of the VSD, a potential risk exists on the septal branches that originate from the anterior descending coronary artery. Methods: By anatomical dissection we studied the relations of the first septal artery and VSD borders in necropsy hearts from 11 patients with CAT (mean age = 3 months). We measured: a) the shortest linear distance between the first septal branch (FSB) and the inferior border of the VSD, b) the distance from the anterior margin of the VSD to the epicardial surface, c) the smaller and larger VSD diameters. Results: The FSB took origin from the proximal third of the anterior descending coronary artery in 8 anatomical specimens and from the medial third in the remaining three. Its distance to the VSD border varied from 0.22 to 1.10cm (mean=0.67cm). Cases showing a proximal origin of the FSB showed a shorter mean distance to the VSD border when compared to those presenting distal origin ($p<0.01$). We found a significant negative linear correlation between the product of the VSD diameters and the distance from the FSB to the VSD border ($r = -0.65$; $p=0.03$). Hearts presenting the FSB running less than 0.5cm from the VSD border had a more extensive anterior ventricular septum than those running further distant, although the difference was not significant. In conclusion, the risk of damaging the FSB in CAT seems to be greater in cases presenting larger VSDs. However, the presence of a well developed antero-superior ventricular septum should also be considered as a protector of risk.

P624

The prenatal origins of human pulmonary veins

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The origins of the intrapulmonary veins were investigated in serial reconstructions of the lungs of 36 human fetuses aged between 28 and 140 days gestation (MRC Human Embryo Collection, UK) and 3 newborn infants. The initial appearance, identification and specification of endothelial cells was assessed by expression of CD31 and of ephrinB2 and EphB4, thought to be markers for premature arterial and vein. Muscularisation of veins was tracked by expression of alpha-smooth muscle actin. At 28 days gestation the mesenchyme of the new lung bud contained a primary capillary plexus (PCP). By 34 days a PCP around the medial surfaces of the bronchi connected with the atrial cavity via definitive, pervaporive extrapulmonary veins. The PCP was continuous between the pulmonary veins and pulmonary arteries developing against the luminal airway wall. Subsequently, intrapulmonary veins formed by coalescence of the PCP in the mesenchyme, midway between the branching airways, lengthening as gestation advanced. From 45 days gestation all new pulmonary veins and arteries expressed EphB4 transiently. From 56–96 days only the peripheral 3–4 generations were positive, with additionally from 105 days to birth the capillary bed. By contrast ephrinB2 was expressed only in intrapulmonary arteries. From 44 days gestation smooth muscle cells differentiated from the mesenchyme in hilar veins, and from 56–95 days from endothelial cells in the PCP. Thus in human lungs pulmonary veins originate by vasculogenesis from the lung mesenchyme, as do pulmonary arteries. EphB4 expression in endothelial cells is not specific for intrapulmonary veins, unlike other species. Muscularisation of veins occurs from one primary source, the mesenchyme. The definitive venous connection to the heart is established by 5 weeks gestation, when blood may circulate through the pulmonary vasculature. British Heart Foundation supported.

P625

The potential for vascular ring in congenitally corrected transposition of the great arteries

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Background: Congenitally-corrected transposition of the great arteries (CCTGA), atrioventricular (AV) and ventriculoarterial (VA) discordance, is an uncommon anomaly. In patients with situs solitus (SS) and right aortic arch (RAA), or situs inversus (SI) and left aortic arch (LAA), tracheal compression may result from a vascular ring and would influence surgical planning. Purpose: This study determines the incidence of right and left aortic arches in CCTGA and SS or SI, respectively, and finds the potential for a vascular ring. Methods: Retrospective review of imaging studies of patients with CCTGA at the Hospital for Sick Children, Toronto, from 1970 to 1999. Patients with SS or SI, AV discordance, and VA discordance or pulmonary atresia (PAT) were included. Patients with situs ambiguous, single or ambiguous AV connection, single ventricle, ambiguous VA connection, or double outlet ventricle were excluded. Anatomical data included situs, location and morphology of great arteries, arch sidedness and branching pattern. Results: Sixty-six patients fulfilled the inclusion criteria, aged 1 day to 18 years. Fifty-eight patients had SS of whom 6 had PAT. 5 had RAA, of whom 3 had PAT. Of the 5 patients with SI, 2 had PAT. None had a LAA. In patients with CCTGA and 2 great arteries, the incidence of RAA with SS or LAA with SI is 2/56 (3.6%) but in those with CCTGA and PAT, the incidence is 3/10 (30%). Conclusion: In patients with CCTGA, the potential for a vascular ring is much higher in those with PAT than in those with 2 great arteries.

P626

The spectrum of cardiovascular anomalies in the zebrafish embryo

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Over the last 8 years several mutagenesis screens have been carried out using the zebrafish (Danio rerio), a particularly tractable organism for the study of early embryonic development. Fecundity of this species, optical clarity of the embryos, and the anticipated completion of the zebrafish genome project by the Sanger Center in October 2002 are among the major advantages of studying cardiovascular morphogenesis, including pattern formation in this system. Two developmental processes which play an essential role in the correct patterning of the cardiovascular system are: (a) formation of the left-right body axis, and (b) migration of particular cranial neural crest subpopulations to the ventral end of the brain tube. Although past screens did not achieve saturation, more than a dozen zygotic recessive mutations with left-right phenotypes have already been identified, and these can be grouped into two classes: (1) heterotaxy, and (2) randomization to situs solitus or situs inversus totalis. Attempts to order the genes in which these mutations occur into a molecular pathway will be discussed. Finally, in the last year, my lab has been participating in a large-scale 4-generation ethylnitrosourea mutagenesis diploid screen led by Dr. Mary Mullins (Dept. of Cell and Developmental Biology, Univ. of Penn. School of Medicine) and members of her laboratory. The goal is to isolate novel zygotic and maternal-effect mutations with left-right and cardiovascular phenotypes. Using whole-mount in situ hybridization with riboprobe for the transcription factor AP2, we are additionally screening for mutants with loss or alteration of the pre-otic or post-otic migratory streams of cranial neural crest cells. The results so far (1000 mutagenized genomes screened) will be presented.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P627
Injury to the Recurrent Laryngeal Nerve during congenital heart surgery because of anatomical variations of the aorta
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Both the right and left recurrent laryngeal nerves usually enter the chest on their way back to the larynx. The usual anatomic approximation of the left and right subclavian arteries becomes extremely important not only in the location of the patent ductus arteriosus but also in the avoidance of injury in the ligation of the patent ductus arteriosus. This situation is also true in the placement of a Blalock-Taussig shunt. However the location of the recurrent laryngeal nerves varies according to the anatomic variations of the aortic arch and the takeoff of the subclavian arteries. Knowledge of the course of the nerve may be helpful in the avoidance of injury to these nerves. However, other causes or mechanisms of injury to the recurrent laryngeal nerves which are beyond the responsibility of the congenital cardiac surgeon are considered.

P628
Coronary artery anatomy in (S,L,L) hearts: implications for surgical management of atrio-ventricular discordance
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Introduction: The advent of double switch and Senning-Rastelli procedures for the treatment of atrio-ventricular discordance with L-looped ventricles has made definition of the coronary artery anatomy in these hearts important. Previous studies have suggested a consistently inverted coronary arterial pattern, such that the right ventricular coronary arises from the left posterior sinus (sinus 3) and the anterior descending and circumflex arteries arise from the right anterior sinus (sinus 2). Methods: A morphologic study was conducted of the coronary arterial anatomy of all heart specimens in our registry with segmental anatomy (S,L,L) (situs solitus, ventricular L-loop, and levoposition of the aorta), two well developed ventricles and atrio-ventricular valve and septal anatomy to permit anatomic biventricular repair. Results: There were 20 specimens collected between 1965–93. Patients ranged in age from 1 day to 25 years. Ten (50%) had transposition of the great arteries and 2 (10%) had double outlet right ventricle. Of these, one had a single coronary artery that arose from the right anterior sinus and criss-crossed anterior to the anterior descending artery arising from the right ventricular coronary artery while the circumflex arose alone, directly above the intercoronary commissure. Three specimens had eccentric ostia, with one left ventricular coronary artery originating directly above the intercoronary commissure. Eight (40%) of the total had right ventricular aorta with pulmonary atresia. Of these, one had both ostia from the left posterior sinus, with both eccentrically entered. None of these specimens had additional coronary anomalies that would complicate placement of a right ventricular to pulmonary arterial conduit. Conclusion: Six (30%) of the specimens had coronary artery abnormalities that could have complicated, but not necessarily precluded, anatomic surgical repair of atrio-ventricular discordance.

P629
Cardiovascular phenotype screening applied to murine embryos: non-invasive approaches
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Rapid, accurate, and affordable methods to screening developing murine embryos for abnormal cardiovascular phenotypes are now essential due to the rapid expansion of murine models of cardiovascular diseases. However, standard techniques used to assess mature cardiovascular function are not easily applied to reproducibly identify abnormal cardiac structure and/or function in the developing myocardium. Cardiovascular screening in the murine embryo requires careful attention to several critical elements. Standardized breeding and timing of gestation are essential. Inter-strain variation in both growth and hemodynamic function are important. Depending on the anesthetic technique, maternal sedation for hemodynamic study can produce maternal hypothermia, hypoxia, and hypercarbia. Intraperitoneal sedation can produce inflammatory reaction while inhalational anesthetics can cause cardiac depression and alter systemic vascular resistance. Determining accurate embryo position for comprehensive longitudinal studies is both time-consuming and technically challenging. Most importantly, screening for changes in embryonic cardiovascular structure and/or function requires the measurements of cardiac and vascular hemodynamics and multiple sites in each embryo. In contrast to the fetal or neonatal heart, embryonic heart rate is NOT always altered despite significant changes in global function. Likewise, some measures of blood flow, such as peak velocity or velocity-time-integral are not always altered despite depressed cardiac function. It is worth noting that there is significant functional variability in normal murine embryos, in part related to uterine position. This may explain why numerous published studies that have investigated altered murine genotypes have contained both false-negative and false-positive results. Finally, the equipment and expertise required for accurate embryo phenotype screening is substantial. Thus, rapid, accurate, and affordable cardiovascular phenotype screening on genetically targeted mice will require technical improvements in animal isolation, data acquisition, expanded access to core facilities, and most importantly, definition of the critical hemodynamic parameters that regulate both structure and function in the developing myocardium.

P630
Pulmonary vein stenosis: a morphologic spectrum with puzzling pathology
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Pulmonary vein stenosis with anatomically normal connections is a rare and poorly understood disease of unknown pathogenesis and variable morphology. Patients present and die in infancy with progressive hypertension and cardiac failure. We reviewed 5 autopsied cases with pulmonary vein stenosis (PVs). Age at diagnosis ranged from 4 hours to 8 months and at death from 7 hours to 6 weeks after diagnosis. The diagnosis was made in one on clinical suspicion and confirmed in all at autopsy. One patient had intraoperative insertion of 4 stents in 4PVs. 4 weeks after successful transcatheter insertion of one stent in the left lower pulmonary vein. There was evidence of luminal proliferation on histological sections at the left lower pulmonary vein distal to the stent (only 4 weeks after insertion). Obliterative neointima in the PVs secondary to the presence of stent has not been reported yet. In all 5 patients no evidence of pleuritis or pleural or lung lesions in the lungs despite clinical presentation of moderate to severe pulmonary hypertension. The morphology of the removed PVs showed the already reported bilateral tubular hypoplasia, hourglass constriction at ventricular junction, bilateral multiple short hypoplastic extrapulmonary PVs. We report a new morphological variety where each pulmonary vein enters the left atrium as normal but each is fed by multiple tiny hypoplastic extra pulmonary veins. We conclude that a fourth morphological type of PVs exists. Pulmonary vascular disease due to PVs in patients less than 18 months seems to be of a different nature than those with left to right shunt. The etiology of PVs is unknown, triggering factors should be considered.

P631
The effects of stress on congenital heart disease in rat embryos.
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The purpose of this pilot study was to investigate the effects of a complex noise stress factor (fundamental frequency: 400 Hz, IBSPL100, side band frequency: 2000Hz-4000Hz, dBSPL95) on a critical stage of cardiac development. In this study we used 12 female and 5 male healthy rats fed by a standard diet and lived in well standard conditions. Newborns from the first and third pregnancy were used as control. On the second pregnancy, mothers were under stress from the 2nd day to the 20th day of pregnancy, 3 times per day, 3 minutes each time. After birth, we took EKGs from newborns then sacrificed them and studied their heart by serial sections and H&E staining under a light microscope (100x10 and 40x10). We had 192 newborns in group 1 (studied 120), 153 in group 3 (studied 110), but in group 2 there were 49 newborns. Ten were eaten by their mothers and 39 were studied. The incidence of congenital heart disease (CHD) in the control group was 4%. In group 2, CHD significantly increased to 17% ($p < 0.001$). Abnormalities included: DGV, IVC, VSD, ASD and TF. Under stress, abortion increased and fewer newborns were delivered. Stress can be a teratogen through 1) physiologic cellular death in embryo, 2) neuroendocrinology, vasculogenic, and corticosteroid effects on embryo's heart, and 3) uterine circulation.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Epidemiology/Outcomes Research

P632

Hepatitis b and c following repair of congenital heart-disease in children using the heart-lung-machine

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The incidence of transfusion associated hepatitis (hep) has decreased since all blood-donors are tested for hep (hep B 1972, hep C 1990 in Austria, 1991 in Germany). How high is the prevalence of hep B and C in patients with congenital heart-disease after repair with the heart-lung-machine? Between 1970 and 1997 967 patients (mean age 3.7 years) with congenital heart disease were operated on using a heart-lung-machine. They were operated in either the German heart-centre Munich (DHZM) or Innsbruck (IBK). 217 were lost to follow up. We contacted 750 parents and 439 replied. We tested all for hep B and C. None had any other transfusion or clinical symptoms of an active hepatitis. There is a positive correlation between the transfused units of blood and the incidence of hep C but hep B. The amount of units of blood per operation decreased with time. The prevalence of a transfusion associated hep C and hep B decreased since the screening for hepatitis started. All our patients with hepatitis showed either little progression or even recovery.

P633

Prevalence and patterns of congenital heart disease in rural Pakistan

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PURPOSE - Prevalence of congenital heart disease (CHD) is well established in most of the developed countries where childbirth is obligatory in hospitals and allied facilities. In Pakistan the situation is reverse where most of deliveries take place in homes by traditional birth attendants therefore true prevalence of CHD in our population is unknown. In rural Pakistan almost 100% children are born at home hence the figures are absolutely unknown. This study is an attempt to answer this question. SUBJECTS & METHODS - During a cross-sectional survey of rural population belonging to major ethnic groups living in three provinces of Pakistan to determine the prevalence of rheumatic heart disease CHD cases were calculated as a by-product. 9476 subjects of all ages were screened. 24 sociodemographic variables recorded. Auscultation and short physical examination performed for initial screening and final diagnosis confirmed on M-mode/2D/Doppler. SUMMARY OF RESULTS - 29 Patients identified with pure CHD another 7 patients had mixed CHD & RHD with total of 36 cases. Overall prevalence for CHD was 3.7/1000. The commonest lesion in acyanotic was bicuspid aortic valve followed by ASD. In children the most frequent anomaly was VSD. CONCLUSION - Apparently this prevalence rate is less than reported elsewhere because data for stillbirth, autopsies are not available. Very high infant mortality also favours high prevalence for CHD in this setting. However these figures represent an overall picture of CHD in a community where medical facilities are lacking.

P634

Epidemiology of congenital heart disease (CHD) - Croatian CHD study-interim analysis

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Introduction. Malformations of the heart and great arteries today belong to the most frequent congenital anomalies detected during the first year of the life. Most recent population-based epidemiological studies of CHD were reviewed by Terence indicating prevalence ranging from 3.5‰ to 13.7‰. In our study CHD were examined according to Clarks pathogenetic classification. Goal. The primary goal of the study was to evaluate the incidence of CHD in Croatia, to classify observations according to Clark classification and to compare our results with those in the literature. Patients and methods. The study population consisted of all the children born between 1995 and 1999 in Croatia, who were seen by local pediatric cardiologists. Altogether 100.736 newborns were surveyed until now, which represents 40% of children with CHD. Results. During the observed period we followed 1417 patients with CHD. The overall incidence rate (1.41%) was slightly higher than found in the general population, as reported in the literature. When analyzed according to Clark classification we found intracardiac blood flow defects to be the most common ones (58.7%), followed by extramesenchymal tissue migration defects (16.6%), cell death abnormalities (9.3%), extracellular matrix abnormalities (4.7%), undifferentiated defects (3.3%), abnormal situs and looping defects (1.7%) and abnormalities of targeted growth (0.1%). Combined defects were found in 5.2% patients. The most common CHD was ASD II (18.6%), followed by perimembranous VSD (14.2%), muscular VSD (9.2%) and stenosis of pulmonary artery (8.8%). Conclusion. The purpose of the study was to evaluate the incidence of CHD in Croatia with the final goal of encourage better planning and treatment of the patients. Also, the studies of CHD which represent the leading cause of infant mortality from the congenital defects benefit from advances in understanding of the biology of cardiac development characterized in Clarks mechanistic classification.

P635

Effect of surgery on growth and body weight in patients with ASD and VSD, resp.

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The aim of the study was to analyze the effect of surgical atrial defect closure on height and weight development in 2 groups of children with spontaneous left-to-right shunt. The ASD group (n=38) were older (mean 5.37 yrs) and asymptomatic, the VSD group (n=18) were younger (mean 0.85 yrs) and had symptoms. The following data were collected before, 3 and 12 months after surgery: standard deviation scores of height (H-SDS) and weight (W-SDS) according to Prader (1989), body mass index (BMI) and percent of weight expected for height (WEH). H-SDS, BMI and WEH increased in all pts. However, VSD pts showed decreased SDS (not signif.) before and after surgery. Nutritional status improved in both groups. ASD pts showed marked catch-up growth. Those with smaller shunts showed weight values above the expected mean for age at 12 months postop. Surprisingly, VSD patients were not only more retarded before but equally so after surgery. The reasons remains unclear from our data.

P636

Predictors of developmental outcome following neonatal cardiopulmonary bypass

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Few studies have adequately assessed the impact of early cardiac surgery, using cardiopulmonary bypass techniques, on the developmental outcome and quality of life of infants with congenital heart disease. This ongoing prospective study is assessing these outcomes on a large group (approx. N=125) of infants who undergo a palliative or corrective procedure, using cardiopulmonary bypass under the age of 2 months. Infants are assessed at 8, 12, and 24-months. Primary outcomes include cognitive and language development, gross and fine motor skills, neurological status, hearing, vision, behaviour, and impact on the family. The poster will present this research program, highlighting important variables to include and factors to control for. Preliminary data on the 1-year follow-up visit will be presented with an emphasis on predictors of good versus poor outcome and the extent to which early intervention is needed to prevent disability.

P637

Mid-term hemodynamic comparison of extracardiac conduit and intra-atrial lateral

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Previous researches have been described that the extracardiac conduit total cavapulmonary connection (EC-TCPC) provides superior hemodynamics compared with the intra-atrial lateral tunnel procedure (LT-TCPC) in complex univentricular heart disease. However the clinical advantage of EC-TCPC in late postoperative course is still controversial. The purpose of this study was to compare the mid-term hemodynamics in 59 patients after EC-TCPC and 64 patients after LT-TCPC and foresee the long-term result of EC-TCPC. All patients enrolled in this study have undergone TCPC at our institute in 1991–1999. The age at TCPC is 6.1 (1–15) years, mean (range). They have been treated with ACE inhibitor, aspirin, low-dose warfarin and underwent catheterization study 3.1(0.5–8) years after TCPC. There was no difference between EC-TCPC and LT-TCPC on cardiac index (3.4 vs 3.6 l/min/m mean vs mean), central venous pressure (10.7 vs 9.8 mmHg) and atrioventricular regurgitation grade (0.75/IV vs 0.63/IV). TCPC using a synthetic graft has a supposed disadvantage of thrombosis. In fact, plasma level

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

ml d-dimer) was slightly higher in EC-TCPC than in LT-TCPC (0.39Å}0.05 vs 0.27Å}0.02 E g/ml, p<0.05). However, no patients had thrombus formation or thromboembolism. Furthermore, we investigated plasma levels of type A and B natriuretic peptides (ANP and BNP) to estimate the extent of atrial and ventricular wall stretch, respectively. Both plasma levels of ANP and BNP were significantly lower in EC-TCPC than LT-TCPC (40Å}3 vs 97 Å}6 pg/ml, p=0.0005, 16Å}2 vs 31Å}6 pg/ml, p=0.01, respectively), suggesting that EC-TCPC causes less wall stretch in not only atrium but also ventricle than LT-TCPC. These data suggest that EC-TCPC may provide superior long-term hemodynamics and progress compared with LT-TCPC though both TCPCs apparently provide acceptable

P638

Heart tumors in children

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Heart tumors in children are extremely rare, belong to the least frequency encountered neoplasms in children. Echo examination allows for quick and non-invasive detection of abnormal masses in heart. The knowledge of them is based on collections of case reports rather than large cohort studies. The material of this study is 15 cardiac tumors in children diagnosed at the echocardiography in the period 1977 – 99: 13 primary: myxoma, hamartoma, fibroma and rhabdomyoma; and 2 metastatic. Malignant tumors were detected in 5 cases. First symptoms were heart campondy in 3 cases, heart failure in 2 children and cardiac arrhythmia in 2. In 6 cases heart murmur was the only one symptom. 7 children died. Permanent therapeutic success was achieved only in the case with left atrial myxoma after operation. 8 patients are still in follow-up. Despite major progress in cardiac surgery prognosis in cases of cardiac neoplasms in children is still grave and uncertain.

P639

Evaluation of the incidence of congenital heart anomalies based on the echocardiographic examination

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The frequency of congenital heart anomalies has been estimated at 0.5–1.2%. Recent studies suggest that the true incidence may be higher, because of use more sensitive diagnostic tools, in particular echocardiography. For this reason, we have performed study based on an echocardiography of every child in the available population. All babies born during one year in our hospital were pooled for the study. Physical and echocardiographic examinations were performed in 1498 newborns before fourth day of life. Anomalies were observed in 458 children (30.6%). Atrial septal anomalies and PDA were the most frequent one. These children underwent control examination. Persisting pathologies were seen in 96 children (6.5%). In most cases, closure of ductus arteriosus or septal defect occurred in the follow up. Conclusions: The incidence of congenital heart anomalies in our population is higher than reported in the literature. The most common were ASD and PDA; in the majority of cases spontaneous resolution occurred. Echocardiography allows detection of small, asymptomatic defects. Differential diagnosis of ASD and PDA in the neonatal period may be difficult.

P640

Strong hypercoagulability in patients after bidirectional glenn procedure

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Thrombosis is one of major complications in patients after Fontan operation. Though the staged Fontan operation via bidirectional Glenn procedure (BDG) has been recently spread for patients with complicated single ventricle hearts, there was little data on the coagulable state in patients after BDG. We investigate the hemodynamic parameters and the plasma levels of the regulatory factors of coagulation and fibrinolysis in 32 patients who had undergone the staged Fontan operation. All patients had been taking warfarin-Na, aspirin and ACE-Inhibitor after BDG. There was no difference of PT-INR levels between at post-BDG and post-Fontan. Data and * indicated the mean values (post-BDG vs. post-Fontan) and p<0.05, respectively. <Hemodynamics> Cardiac Index was higher at post-BDG than post-Fontan (4.2 vs. 3.6 l/min/BSA*, respectively), while SaO2 was lower (87 vs. 94 %*, respectively). There were no difference between both the states on SVC pressure, SV-EF and AV valve regurgitation grade. <Coagulable state> PIC-EH+2 was higher at post-BDG than post-Fontan (0.82 vs. 0.34 nmol/L*). Plasma PIC levels at both states were higher than normal subjects(0.21 vs. 1.02 micg/ml). Plasma thrombomodulin level was lower at post-BDG state than post-Fontan one (1.9 vs. 2.6 FU/ml), while plasma PAI-1 level was higher at post-BDG state than post-Fontan one (57 vs. 20 ng/ml*), suggesting vascular endothelial cells and platelets are activated. These data suggest that the patients at post-BDG were have stronger hypercoagulability than at

P641

Pediatric bacterial endocarditis: does delay in diagnosis influence outcome? A 20 year national survey in the netherlands

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Bacterial endocarditis (BE) is associated with high morbidity. Mortality in the pediatric population has hitherto been ascribed to late diagnosis and hemodynamic compromise due to acquired cardiac lesions. The aim of this study was to assess the impact of time from onset of symptoms to diagnosis of BE on subsequent complications and outcome, in the current era. A case note study of all pediatric patients with proven BE (using Duke's criteria) and presenting to one of the 8 tertiary referral centers in the Netherlands between January 1 1980 and December 31 1999 was undertaken. Patients were divided into 2 categories: those with previously diagnosed congenital heart disease (CHD) and those without CHD. Patients were further subdivided into two groups, those in whom BE was diagnosed within 30 days of onset of symptoms (early diagnosis) and those in whom BE was diagnosed >30 days after onset of symptoms. 121 children were identified to have had BE (132 episodes) during the study period; 98 with CHD and 23 without CHD. Presenting symptoms were fever and malaise in both groups. 12 patients without CHD had a new heart murmur. The interval to diagnosis of BE was <30 days in 71/98 patients with CHD, and in 17/23 without CHD. 33/96 patients with CHD and 8/23 without CHD had undergone a potentially septic surgical procedure in the 90 days preceding onset of symptoms. Complications (cerebral emboli, abscesses, acquired intracardiac shunts, valve dysfunction, mycotic aneurysms) occurred during 30 BE episodes, 23 in patients with CHD and 7 in patients without CHD; 15/23 complications in the CHD group and 6/7 complications in the non-CHD group occurred >30 days from onset of symptoms. Cardiac surgery in the acute phase of BE was performed in 25 patients, 16 with CHD (12 diagnosed at <30 days after onset of symptoms) and 9 patients without CHD (6 diagnosed at <30 days). There were 6 surgery related deaths (5 in patients with pre-existing CHD), all in patients who were diagnosed early. In the current era, in the majority of pediatric patients the diagnosis of BE was established early. Major complications also occurred early. A delay in diagnosis did not by itself influence the outcome.

P642

Lipoprotein profil in children with family history of premature coronary heart disease

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The purpose of this study was to determine the lipoprotein profil in children with family history of premature coronary heart disease (CHD). The study included 40 children (42.5% boys and 57.5% girls) aged 3 to 18 years: risk group (RG) -whose one of their parents suffered from myocardial infarction, diagnosed according to WHO in age <=55 years and 40 children - the control one (CG)- without family history of CHD who were matched according the age and sex. Fasting plasma concentrations of total cholesterol (TC), high-density lipoprotein cholesterol (HDL-C), low-density lipoprotein cholesterol (LDL-C) and triglyceride were measured. Total cholesterol/HDL-C (FR) and LDL-C/HDL-C(IA) were calculated. Levels of TC, LDL-C and TG were higher in children RG (4.78;2.89; 1.10 mmol/l) than in children CG(4.39;2.39;0.94 mmol/l) but statistically there was no difference (p>0.05). The mean HDL-C level was significantly higher (p<0.05) in CG(1.61 mmol/l) than in RG(1.34 mmol/l). The mean levels of IA and FR were significantly higher (p<0.01) in RG(2.33;3.81) than in CG(1.02;2.95). We identified children with abnormal lipoprotein profil: TC>=5.17 mmol/l had 21.21% children of RG and 12.5% CG. LDL-C>=3.36 mmol/l had 33.3% children in RG and 2.5% CG. TG>=1.48 mmol/l 23.33% RG 1.5% CG. HDL-C<=1.17 mmol/l had 40% RG and 6% CG. It is very important

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

to determine lipoprotein profile in children with family history of premature coronary heart disease. These children would be early detected, inspected and counseled, in order to prevent premature development of atherosclerosis.

P643

Comparison of the health outcomes of surgical and device closure of paediatric atrial septal defect

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This is an on-going study comparing cost, clinical outcome, family satisfaction and functional status of children undergoing surgical or device closure of secundum atrial septal defect (ASD) at the Royal Children's Hospital, from May 1st 1999. Data shown includes patients recruited to April 30th 2001. Selection for device closure or surgery depends on the cardiologist and parental preference, after discussing the options. Clinical details are documented, hospital admission costs assessed and a parental questionnaire completed on discharge and after 6 months. Non-parametric statistical methods are used and results expressed as median (25th – 75th centile). Analysis of the year shows that 13 children underwent surgical closure and 25 device closure with an Amplatzer septal occluder. The two groups were comparable in terms of age, size of ASD and body surface area. Procedure times and hospital stay were significantly longer for surgical patients [169 mins (145 – 175 mins) versus 80 mins (70 – 110 mins)] and [79.5 hours (76 – 90.5 hours) versus 29 hours (29 – 30 hours) p<0.01]. No child in the device group required intensive care or blood products. There was no difference in the complication rate. The median post-operative pain score, amount and duration of analgesia use and convalescence time was greater for surgical patients. No parent regretted their choice of procedure for their child. Theatre costs for surgical and device closure were similar. However nursing, laboratory and pharmacy costs occur based on a significantly greater total cost for surgical repair [Aus$12,841 ($11,475 – $13,669) versus Aus$9,860 ($9,450 – $10,640); p<0.01]. In conclusion, in our hospital, device closure involves a shorter hospital stay, and causes less discomfort and less disturbance to family life. The total cost is significantly less for device closure than surgical closure of ASD.

P644

Screening for congenital heart diseases of newborn babies using echocardiography

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To clarify the true incidence of congenital heart diseases, we studied all newborn babies of our hospital by echocardiography on the 2nd day after birth. From Oct/1985 to Dec/1999 echocardiographies were performed on 7,370 newborn babies by pediatric cardiologists and doppler method was used for detecting shunts and abnormal flows. We also examined whether we could notice the presence of heart murmur and femoral pulse if abnormal findings were recognized we managed in some cases and reexamined the babies on the 1st month of age. In 244 cases(3.31% of total) we detected abnormal findings. VSD was found in 134 cases(1.82% of total) but 97 cases(70.2% of VSD) were muscular. IAS shunt(atrial septal defect >= 4mm) was 74. PS(doppler maximum flow >= 2m/sec) AS 5, CoA 2, DORV 2, ToF 1, TAPVC 1, and Coronary AV fistel 1. Sixty percent of VSD closed at 1 year of age. Over 90 % of IAS shunt could not detected at 1 year of age. Among examined cases, cardiac surgeries were performed in 10 cases. VSD was 3, DORV 2, CoA 2, PDA 1, ToF 1, and TAPVC 1. From our experience we concluded, 1)70% of VSD was muscular and over 60% of VSD closed at 1year of age. 2)It is difficult to differentiate ASD from PFO and some cases will grow the size of atrial septal defect. 3)Stenosis of the shunt is significant after 6 months of age, if cardiac failure does not occur.

P645

Mid-term outcome after double switch operation

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Purpose: To evaluate mid-term outcome after double switch operation (DSO) for patients with atrioventricular discordance and ventriculoarterial discordance (AVD). Methods: Thirty-seven patients with AVD had undergone a DSO at our institute, and 33 hospital survivors were in my patients (male: female=22:8) and were reviewed. Age at DSO was 0 to 13.5 (5 +/- 2) years, and follow-up period was 12 to 151 (79 +/- 41) months. Results: Late death was observed in 2 patients. That was unexpected in both patients and they had had episodes of supraventricular tachycardia. One-year, 5-year and 10-year actuarial survival rate was 100%, 100% and 93%, respectively. Re-operations were needed in 5 patients (17%), including re-right ventricular outflow tract (RVOT) reconstruction in 2 patients, pacemaker implantation (PMI) in 1, thrombectomy of left atrium in 1, relief for pulmonary venous obstruction in 1. Catheter intervention was needed in 9 patients (30%), including percutaneous transluminal angioplasty and/or stent implantation for RVOT obstruction in 4 patients, for systemic venous obstruction in 5. Rhythm disturbances were observed in 6 patients (20%), supraventricular tachycardia in 5 patients and complete atrioventricular block in 1 who had needed PMI. Four patients needed antiarrhythmic agents. Eighteen patients performed a cardiopulmonary exercise test (interval from DSO to exercise test was 11 to 126 (68 +/- 37) months.) And peak oxygen uptake was 19.4 to 35.3 (26.3 +/- 4.1) ml/kg/min, which was 38 to 68 (52 +/- 8)% of predicted normal value. Conclusion: After DSO, in addition to impaired exercise capacity, some patients have serious post-operative problems of arrhythmia and/or obstruction of RVOT, systemic or pulmonary venous return. Therefore, careful observation is necessary to follow these patients after DSO.

P646

Neuropsychological and neurodevelopmental outcome of children with hypoplastic left heart syndrome following the three-stage Norwood procedure: the British experience

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Hypoplastic left heart syndrome (HLHS) occurs in 1:5000 live births, and 200 per annum in the UK. Due to insufficient number of donors in the UK, heart transplantation is currently not a first-line treatment option. The alternative is the staged Norwood Procedure, however most children in the UK are managed with terminal supportive care. Improved survival rates following palliation has led to an interest in understanding the impact of surgery on the neurological outcome of these children. Method: 11 children with HLHS post Fontan repair were assessed. A formal neurological examination was performed, including the Functional Independence Measure (WeeFIM). Cognitive functioning was assessed using Wechsler age appropriate Intelligence tests and behaviour using the Child Behaviour Checklist (Achenbach'91). Siblings were used as controls and underwent the same neurological and psychometric testing. Results: 11 children with HLHS, mean age 5.4 years (range 3.75 -7.25). 2 children had very mild hemiplegia. Only minor abnormalities were otherwise found. 4 had mild dysphasia, 4 difficulties with short-term memory and 1 lateral nystagmus. 10 siblings were assessed, mean age 6.6 years (range 4.5–9.0). 1 had lateral nystagmus (sib of above), remainder were normal. Differences in full scale IQ between HLHS and siblings were compared using an independent T test (equal variance not assumed). A non-significant trend was observed (t19= -2.03, p=0.057). The mean IQ for siblings was 101.1(SD=20.49) whereas the HLHS patients showed a mean IQ of 85.8 (SD=11.73). The mean difference between groups was -15.28(95%Confidence=7.28). Conclusion: There was no significant difference between the IQ of HLHS children post Fontan and siblings. Physical and functional prognosis following palliation in HLHS is good.

P647

Frequency distribution of congenital heart defects in Nicaragua

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Background: The incidence and frequency of congenital heart defects are believed to be geographically constant. Our observations in Central America indicate that this may not be correct. Methods: We reviewed the diagnoses of 188 patients, ages 1 mo to 20 yrs, with congenital heart defects seen at Rosales Hospital (HEODRA) in Leon, Nicaragua and compared the frequency distribution with published data for 1985–87 from Boston Children's Hospital (BCH). The diagnoses were determined by physical exam and 2-D echo and confirmed at surgery in 45 patients. Results: In comparison with BCH data, we found a higher frequency of VSD (especially conal septal defects), PS, PA/IVS, and PDA, and a lower frequency of left heart obstructive lesions including AS, coarctation and HLHS among Nicaraguan children. Common TGA was less frequent in Nicaraguan patients, although other conotruncal anomalies were not. We tested whether incomplete case finding of defects such as TGA and HLHS, that are lethal in neonates, might have magnified the

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

difference in frequency distribution. Recalculation of the frequency distribution of the BCH reference data to exclude these defects did not alter our findings. Conclusion Even after accounting for incomplete case finding, substantial differences in the frequency distribution of congenital heart defects were observed between Nicaragua and the US. Further investigation of possible genetic causes might yield insights into the development of congenital heart defects.

P648

The incidence of congenital dysplasia of aortic valves in Japanese neonates detected by echocardiography

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The incidence of congenital dysplasia of aortic valves in Japanese neonates detected by echocardiography Hatano.T ,Ikoma.M ,Nagano.Y. Nagoya The relative incidence of various types of congenital heart disease is different among the indigenous racial groups.As for the dysplasia of aortic valves,it has not been confirmed that Japanese neonates have significantly reduced incidence of bicuspid aortic valves compared to white ones.Incidence of 2% of the population We examined the 10560 neonates in our hospital to clarify the incidence of congenital heart disease,dysplasia aortic valves From September 1, 1988 to March 31, 2000, both 8012 well being neonates 1–2 day after birth and 396 sick in Neonatal Intensive Care Unit were examined by echocardiography All 11968 neonates were observed from definitive and multiple views including short-axis and longitudinal planes of aortic valves Our survey revealed cardiac malformations in 262 neonates, and 32 of them (1 per 1000 live births)had dysplastic aortic valves The anatomy of dysplasia contains 10 bicuspids(5 males and 5 females),a male unicuspid and a male quadricuspid aortic valves None of them has received medical and surgical treatment.As a result, we ascertained the lower incidence of dysplastic aortic valves in Japanese than white infants

P649

Noncompaction of the ventricular myocardium (NCVM) in pediatrics: Incidence, diagnosis, and outcome

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NCVM is an extremely rare disorder characterized by numerous prominent apical trabeculations and deep intertrabecular recesses To evaluate incidence, diagnostic methods and clinical course of NVVM in pediatrics, all consecutive patients at our institution were examined between 9/99 through 6/00. 9 of 2984 patients aged 10 days to 15 years were detected mostly neonates. Echocardiography was diagnostic in all cases and superior to magnetic resonance imaging (6/9) or cardiac catheterization (4/9) There was associated CHD in 6 patients, dysrrhythmias in 2, and a positive family history in 2. 7 patients developed heart failure with restrictive hemodynamics in three, 4 of them severely.Arrhythmia was found in 2 patients, thrombi and embolisation in 1 each. One patient died. NCVM in pediatrics is more frequent than suspected. Because of ominous prognosis early diagnosis is important Familiarity with the echocardiograph ic pictures is mandatory Long term follow-up studies are needed

P650

Surgical results for low birth weight infants from a multicenter consortium

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Introduction: Reports of surgeries for low birth weight or premature infants from different centers have suggested varying results The low birth weight infants from 1994–1998 from Pediatric Cardiac Care Consortium (PCCC) are investigated in this study Methods:We investigated 1491 procedures from 1395 patients with weights ≤ 2.5 kg and 11530 procedures from 10973 patients with weights 2.51–4.5 kg from PCCC PCCC consists of 45 cardiac centers that submit data from heart catheterizations, surgeries and autopsies to a central registry at the University of Minnesota for the purpose of improving care for children with heart disease Results Mortality for weight less than 2.5 kg was 30% and for greater than 2.5 kg was 21%. Common procedures and mortality for ≤ 2.5 kg are as follows: aortopulmonary shunts 405 [77 (deaths)], pulmonary artery band 105 (28), hypoplastic LV surgery 79 (58), total anomalous pulmonary venous connection surgery 62 (26), arterial switch operation 54 (15), pulmonary valvotomy 50 (13), VSD repair 48 (9), pacemaker generator 42 (14), aortic valvotomy 21 (10), truncus arteriosus conduits 21 (14), hemitruncus 11 (8). Mortality for weight is shown. (see table) Conclusions Low birth weight substantially increases mortality for infant cardiac procedures. Risk appears to commonly change up to 4.5 kg Mortality for several complex procedures such as hypoplastic LV surgery and truncus conduit placement is higher than for other open or closed procedures

Fetal Cardiology

P651

Outcome of structural heart disease diagnosed in utero: a case series

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Objectives To review outcome of fetuses with structural heart disease detected by echocardiography from 1983 to 1999 in one ped iatric cardiologic center Methods-A total of 99 fetuses with different types of cardiac defects, diagnosed at a median gestational age of 28.4 weeks (range 16 to 41) were included. Results-Of 99 fetuses with in- utero -diagnosed cardiac anomalies, 6 (6 %) showed normal cardiac status postnatally.35 % of fetuses with heart disease diagnosed before 24 weeks of gestation were terminated Of 93 fetuses 7 (8%) with a heart defect died in utero at a median gestational age of 32 weeks. Chromosomal abnormality was found in 28% of cases with a mortality of 73 %. Fetuses with normal chromosomes had extracardiac anomalies in 40% of cases. 46 % of them died. Intrauterine heart failure was detected in 27% of fetuses and was associated with univentricular heart (UVH) and intracardiac tumors, in 36% and 67%, respectively. 12 fetuses (13%) were found to have associated arrhythmia. 4 of these died. Of 76 live births (median gestational age 38 weeks, birth weight 2878 gm), a total of 37 (49%) neonates died. 24 neonates (32%) underwent cardiac surgery or invasive procedure; 6 (25 %) died after the procedure. Neonatal mortality was highest in fetuses with hypoplastic left heart syndrom (HLHS), ventricular septal defect (VSD) and UVH (87%, 64% and 50%, respectively) In long-term follow -up (median 3.8 years), 34 children of 76 live births (45 %) were alive, 95% of them were without symptoms Conclusions Our data indicate that despite planned delivery in a single tertiary pediatric cardiologic center the prognosis for fetuses with in- utero -diagnosed heart defect was poor. Poor outcome was largely attributable to associated extracardiac malformations and chromosomal abnormalities

P652

Fetal cardiac imaging by paediatric cardiologists provides improved detail over obstetric screening of congenital heart disease

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Fetal echocardiography is optimized by a team strategy of imaging by obstetrician-sonographer and obstetric scanners (O) and fetal paediatric cardiologists (FPC). We retrospectively examined 1037 studies (1995–1999), and identified 249 cases of major congenital heart disease The O diagnosis was compared to the FPC diagnosis and postnatal diagnosis. The rate of complete accurate diagnosis for O and FPC diagnosis were 51% (17% false pos 41% false neg) and 95% (2% false pos 5% false neg) respectively. Major differences in diagnosis or detail were found in 79 patients after FPC was completed and in 35/79 (149%) this was judged to have potential significant impact on management and pregnancy counselling. The complementary roles of O and FPC remain important. FPC can contribute with additional detail in some cases which may significantly impact on counselling and planning

P653

Etiology and outcome of fetuses with functional heart disease not associated with cardiac arrhythmias or structural defects

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The aim of this study was to assess the etiology and outcome of fetuses with functional heart disease detected by echocardiography A total of 63 fetuses (median gestation of 28.9 weeks) of 39 mothers were included 16 fetuses were twins; 5 of them had normal finding. The exclusion criteria were hydrops (N=13) pericardial effusion (PE, N=9), mitral valve regurgitation (TR, N=8), hypertrophic cardiomyopathy (HCM, N=7) and dilated cardiomyopathy (DCM, N=6). All cases with cardiac arrhythmias or structural defects were excluded. The etiology of functional heart disease was twin pregnancy in 16, maternal diabetes in 4, fetal anemia in 4, extracardiac malformation in 4,

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

antenatal infection in 3, reasons in 3, in utero indomethacin administration in 2, endocardial fibroelastosis in 2, congenital chylothorax in 2, hypoplastic lungs in 1, adenomatoid malformation in 1, maternal anaphylaxis in 1, idiopathic arterial calcification in 1, arteriovenous malformation in 1, pregnancy induced hypertension in 1 and unknown etiology in 2 fetuses. Antenatal managements were performed in 16 of 18 fetuses (88%): 2 abortions, 9 digoxin administrations, 2 pleural cenesis, 1 pericardial puncture, 1 blood transfusion and 1 amniotic fluid centesis. Altogether 13 deaths occurred (72 %): 2 stillbirths and 11 postnatally. Fetal hydrops, PE, IHB, HCM and DCM were associated with mortalities of 82 %, 33 %, 25 %, 0 % and 33 %, respectively. In the follow up of 3,5 years, 85 % of children are free of symptoms. These findings indicate that a functional heart disease in utero is associated with varying etiology and high mortality. The mortality was highest in fetuses with hydrops, pericardial effusion and dilated cardiomyopathy.

P054

Fetal dilated cardiomyopathy secondary to hemangioendotheliomas of the liver

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Introduction: The most common multifocal vascular lesion that involve the newborn are histologically forms of hemangiomas which in the liver are called hemangioendotheliomas. In the prenatal period they may present with dilated cardiomyopathy caused by high-output failure from volume overload. Case report: A 26 year-old gravida was indicated for the fetal echocardiographic by presenting increase of the heart area in the obstetric ultrassonography performed with 20 weeks of gestational age. Moderate right cardiomegaly was observed with no anatomical alterations. The myocardial contractility and the flow across the heart valves, the foramen ovale and ductus arteriosus were normal. Progression of the cardiomegaly was evidenced and with 30 weeks also documented increase of the left ventricle. With 29 weeks of age gestacional the patient was attended with premature contractions, cardiotocography and Doppler velocimetry indicating fetal hypoxia. An emergency cesarean section was performed. The male newborn weighing 1090g, Apgar score of 4/7, developed moderate respiratory distress. The echocardiogram showed moderate pulmonary hypertension. In the fifth day of life a clinical picture of congestive heart failure was evidenced without significant improvement with the associated clinical treatment. The presence of progressive hepatomegaly multiple hemangiomas of skin, ascites of the hepatic function, anemia and digestive hemorrhage were observed in the second week of life. Despite intensive care he died in the 23 day of life. At autopsy the liver contained numerous nodules and histologically the diagnosis of hemangioendotheliomas. Hemangioendotheliomas of the liver may present with fetal dilated cardiomyopathy. We pointed out the importance of fetal echocardiography in his diagnostic suspicion.

P055

A prenatally diagnosed case of complex pulmonary atresia with ventricular septal defect: echocardiographic and pathologic findings

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We describe a patient who had complex pulmonary atresia with ventricular septal defect (VSD), both of which were diagnosed and followed by prenatal echocardiography in the second trimester. The postmortem morphologic findings on this heart defect are also discussed. The patient's mother was 24 years of age, primigravida. In 18th week of gestation, a VSD was detected on routine prenatal ultrasonography. Fetal echocardiography was then performed, and this revealed a defective interventricular septum, dilation of the left ventricle (LV), narrowing of the right ventricle (RV), and a perimembranous outlet VSD. The aorta was overriding the VSD. Serial echocardiographic evaluations were performed at gestational weeks 22 and 24. These showed that the interventricular stability had deteriorated further. Also, the right ventricle diameter was unchanged but it was evident that the chamber was hypoplastic. Over the three exams, the RV/LV diameter gradually decreased from 0.95 to 0.7, to 0.54. We also detected left-to-right shunting through the VSD, and tricuspid insufficiency with a flow rate of 1.75 m/s. Postmortem morphologic examination of the heart revealed dextrocardia, left atrial isomerism, ambiguous atrio-ventricular connection, and a dilated LV with a double-outlet and opening to a hypoplastic right ventricle through a small VSD. The pulmonary artery, which had dysplastic valves that formed the roof of LV, was connected to a large ductus arteriosus. Autopsy also revealed a large atrial septal defect, abnormal pulmonary and systemic venous outflow tracts, and pericardial effusion. Fetal echocardiography is valuable in that it allows the examiner to follow transformations in interventricular connections and other changes in the abnormal heart over time. The ability to gather this information prenatally enables ethical decisions to be made when the malformations dictate no change of surgical treatments.

P056

Transplacental pharmacokinetics: treatment of fetal tachycardia with sotalol and/or digoxin

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Background: To reach an optimal therapeutic regimen in maternal-fetal pharmacotherapy of fetal tachycardia (FT), transplacental pharmacokinetics will be evaluated. Methods: Four patients with FT were treated with sotalol or sotalol and digoxin. Drug levels were determined in maternal blood (ML), amniotic fluid (AL), umbilical cord (UC) and neonatal blood (NL). Therapeutic range: sotalol: 0.6–2.5 mg/l and digoxin: 1.0–2.0 µg/l. Results: see table.gif. Gif image 9 EB AF = atrial flutter, SVT = supraventricular tachycardia, SR = sinus rhythm, FH = fetal hydrops, PT = pharmacotherapy, DS = closes, Eff = effect of treatment, R = relapse of, P = persistance of, ML-R = ML at relapse, ML-P = ML at birth. Doses of sotalol are in mg/kg/day and of digoxin in µg/day, sotalol levels are in mg/l and digoxin levels in µg/l. Conclusion: Amniotic fluid drug concentrations were 3 to 4 times higher than placental levels and maternal plasma levels were higher than fetal plasma levels. Therapeutic drug monitoring might be useful in choosing adequate pharmacotherapy, although sotalol and digoxin therapy weren't always effective within the therapeutical range. Additional studies are needed to refine our understanding of transplacental pharmacokinetics.

P057

Effects of respiratory stimulant dosapram hydrochloride on oxygen-induced constriction of fetal rabbit ductus arteriosus

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Dosapram hydrochloride, a respiratory stimulant, is used to treat idiopathic apnea of prematurity. The side effects reported are minimal, and what is more, there is no information about the effects on patent ductus arteriosus which is a very common congenital cardiovascular defect in premature infants. This study was carried out to investigate possible direct actions of this agent on oxygen-induced constriction of ductus arteriosus. The vessel was isolated from late-gestational fetal Japanese White rabbits and studied in vitro. The tension recordings were performed. Preparations were equilibrated at low oxygen tension (30–40 mmHg), dosapram hydrochloride (1–30 microM) had no effect on contractions induced by potassium, histamine or indomethacin. At the concentration of 30 microM, this agent increased ductal movement tension by 10% in the absence of vaso-constrictors. Preparations were equilibrated at high oxygen tension (350–400 mmHg), dosapram hydrochloride (1–30 microM) had no effect on contractions induced by high oxygen tension, potassium, histamine or indomethacin. Dosapram hydrochloride also did not inhibit ductal sensitivity to histamine, potassium or indomethacin. We concluded that physiological and therapeutic concentrations of dosapram hydrochloride (1–10 microM) does not inhibit sensitivity of the ductus arteriosus to certain vaso-constrictors and can not inhibit the maximum response to increased oxygen tension.

P058

Perinatal adaptation of the fetal great arteries

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Introduction: Fetal circulation is unique in that two ventricles function in parallel. At a moment of birth, the fetal circulation must immediately adapt to extrauterine life. After the first breath, pulmonary flow increases dramatically and is separated from systemic circulation. The size of the great artery usually correlates with the size of the mass and amount of flow through it.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

The purposes of this study were to investigate whether the perinatal hemodynamic changes influence on the size of aorta and the pulmonary artery. Methods A prospective study was performed The study group consisted of 50 full-term pregnant women who were planed to undergo C/S delivery Fetal echocardiography was performed one day before birth ad repeated 4 5day after birth The aortic and the pulmonic artery diameter were measured and correlated with birth weight, and other physical parameters. Results. In full-term fetuses, pulmonary arteries were bigger than aorta Aortic diameter correlated positively with birth weight In contrast, pulmonary artery diameter did not show any correlation with birth weight or other physical parameters but abdominal circumference. After birth, pulmonary artery becames smaller and aorta get bigger than before birth (p<0.01) The sum of the pulmonary artery and aortic diameter did not change After birth the size of two great arteries correlated positively with body weight Conclusion Assuming that the size of artery is related to the size of the area supplied, our study confirmed that the aorta and the pulmonary artery reflect perinatal circulatory changes In the fetus, pulmonary artery is bigger than aorta, which proves the right ventricular dominance After birth, diameter ratio of two great arteries approached to equity and the sizes of two great arteries correlated with body weight,and these findings supports that two circulations are separated and handle the same amount of blood.

P659

Maternal hypertension and altered fractal correlation behavior of fetal heart rate

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The purpose of the present study is to investigate whether temporal, spectral, irregular and fractal correlation behaviors of fetal heart rate dynamics is altered by pregnancy induced hypertension (PIH) of mother. Sixty fetuses who aged over 30 weeks and were not associated intrauterine growth restriction and whose mothers had PIH (PIH group) were studied Three hundred gestational age-matched normal control fetuses (control group) were also included We selected 5000 points of their fetal heart rate and calculated the power spectrum, approximate entropy, short-(_1 ≤ 30bpm) and long-(_2 > 30bpm) term fractal scaling exponent The power spectrum, approximate entropy, 1 and 2 reflect, periodicity, irregularity, short-term fractal correlation and long-term fractal correlation respectively. There were no significant differences in the mean (143bpm±3.4 vs 141.9bpm±3.1), variance (43.9bpm 2.1 2.2 vs 47.8bpm2±5.9), low- (131.0msec2±2.6 vs 138.5msec 2±12.2) and high (23.7msec2±1.0 vs 23.7msec2±2.1) frequency power and approximate entropy (1.716±0.011 vs 0.722±0.025) of the fetal heart rate between the PIH and the control group However, in the PIH group, _1 was significantly lower than in the control group (1.386±0.035 vs 1.481±0.036, p<0.0001), and _2 was significantly higher than in the control group (0.926±0.022 vs 0.860± 0.012, p<0.0001). It can be concluded that maternal PIH significantly alters not the periodic and irregular behavior of fetal heart rate but the short- and long-term fractal correlations behavior

P660

Fetal echocardiography as a prenatal screening test for congenital heart disease in Hong Kong

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Background: Fetal echocardiography is becoming more widely used to detect congenital heart defects (CHD) antenatally. The performance of fetal echocardiography, however, is not to be taken lightly because the diagnosis of severe CHD may result in termination of the pregnancy. Methods: 126 consecutive fetal echocardiograms done on 119 pregnant women in the Prince of Wales Hospital in the past four years were reviewed. The reasons for referral were maternal heart or medical problems, family history of CHD, abnormal 4 chamber view or other fetal anomalies. Fetal echocardiography was performed using standard method. All live birth babies had a complete physical examination after birth and an echocardiography was performed to confirm the prenatal findings if necessary. All aborted fetuses had a detailed autopsy by the pathologist. Results: Two third of the cases had fetal echocardiography before the 24th week of gestation. Nine(25%) patients were found to have abnormal findings. 27 of them were referred because of an abnormal 4 chamber view detected by obstetricians. The abnormalities included ventricular septal defect(3), atrioventricular septal defect(2), transposition of great artery(1), Fallot's tetralogy(4), coarctation of aorta(1), dysplastic tricuspid valve(1), total anomalous pulmonary venous drainage(1), hypoplastic left heart syndrome(2), double outlet right ventricle(3), dextrocardia(1) and complex cyanotic heart(1). 15 mothers chose to terminate the pregnancy because of severe CHD. One patient who had an antenatal diagnosis of double outlet right ventricle was found to have normal connections of the great vessels but grossly dilated RV and persistent pulmonary hypertension. The calculated false positive rate of fetal echocardiography is 2.3%, there was no false negative result. Conclusion: Although the false positive rate of fetal echocardiography is low, it warrants extra caution in the interpretation and management of all abnormal results.

P661

Prenatal diagnosis of congenital heart diseases and arrhythmias – own results

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Fetal echocardiography is a modern diagnostic method enabling an early detection of the structural anomalies and arrhythmias in the fetal heart. The aim of the study is to present our own results of the diagnostics of fetal's congenital heart diseases and arrhythmias in a group of patients with a high cardiological prenatal risk. 1575 prenatal echocardiographic studies were performed in 1226 fetuses. The group of 121 fetuses with structural anomalies of the heart and 104 fetuses with arrhythmias were analyzed. Within the group with congenital heart diseases the most common anomaly was CAVC – 30 cases (24.7%), which in 11 cases coexisted with complete heart block, next most common abnormality was VSD – 19 fetuses (15.7%), HLHS (12.3%), SA (6.6%). Another anomalies were: SP (5.7%), AtrAP (3.3%), ToF (5.7%), DORV (2.5%), CoA (4.9%), TAC (1.6%), TGA (1.6%), IAA corr I DAV III (0.8%), dextrocolon of LA (0.8%). The remaining group was made up of cases with complicated congenital heart defects. Among the group of 104 fetuses with arrhythmia the most common were supraventricular extrasystoles: beats – 61 fetuses (51.8%), then supraventricular tachycardia – 47 cases (16.3%), complete heart block – 22 cases (21.1%) and 1 case of sinus bradycardia. CONCLUSIONS: 1. Fetal echocardiography enables early detection of structural anomalies and arrhythmia in the fetal heart. 2. Early detection of these anomalies enables intrauterine therapy and labour in a medical centre, where early cardiological diagnostics and potential cardiosurgical treatment are possible.

P662

The natural history of the left heart obstructive disease in the mid-trimester fetus

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It has been suggested that hypoplastic left heart syndrome (HLHS) or severe mitral and/or aortic stenosis (MS/AS) progresses in mid-trimester. We reviewed serial prenatal echocardiograms of 14 fetuses with MS/AS and 5 with AS during 2nd and 3rd trimesters. We measured valve diameters of tricuspid (TVD), mitral (MVD), pulmonary (PVD), and aortic valves (AVD). We calculated the ratios of TVD/MVD and PVD/AVD, and compared them with our own standard. As a result, AVD was smaller than normal at the initial recording and further decreased as compared with the normal value with gestational days in all cases. There were 2 groups in the changes of MVD: it stayed lower limit of the normal in 3 cases, whereas it decreased with time and ended much lower than the normal in 14 HLHS fetuses. The serial changes of the TVD/MVD ratio showed 2 types, it was very high already at the 20th week and stayed as it was in one group (likely to be typical HLHS), and it gradually increased from the upper limit of normal in the others. We conclude that the left heart obstructive disease is progressive and late mid-pathophysiology will result in HLHS from, and this new opinion of MVD plays a critical role in the final morphology.

P663

Prenatal therapy for fetal heart block

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Prenatal therapy for fetal heart block has been established here – ximinami has been suggested to be effective, but only small numbers of cases have been reported until now. We will report five cases of fetal heart block treated in utero with beta stimulant, ritodrine. Four cases were without cardiac defects. Maternal SS-A antibodies were positive in three cases, and negative in one

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

case. One case was a left isomerism with Single Atrium. All cases needed pace maker implantation soon after birth. Their heart rates before therapy were from 50 to 60 per minute. The doses of salbutrine were from 50 to 100ug/minute. There were no side effects in mothers or fetuses. Fetal heart rates increased by 10–33% in four cases including the case of left isomerism, but in one case with positive SS-A antibody, heart rate didn't increase at all. We compared their cardiac functions before and after fetal therapies. Left and right ventricle shortening fractions, stroke volumes, combined cardiac outputs and renal blood flows increased after therapy, but CTAR/Cardio Thoracic Area Ratio) decreased by only 1–4%. We conclude that prenatal beta-stimulant therapy is effective not only in the cases of anatomically normal hearts but also in the case of left isomerism and that it may be effective not only for fetal heart rate but also for fetal cardiac function. We need more cases to evaluate effectiveness and side effects of fetal beta stimulant therapy for fetal heart block.

P664

Ebstein's anomaly during fetal life: diagnosis and perinatal outcome

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Ebstein's anomaly is the most common congenital disease of the tricuspid valve, corresponding to 0.5% of congenital heart diseases. Mortality rate is expressed in he in the 85% range in the neonatal period. The purpose of this report is to review the experience of a tertiary center of Fetal Cardiology in the diagnosis and management of Ebstein's disease. Material and Methods: Among 9500 fetal echocardiographic examinations from january 1987 until may 2000, 15 fetuses with Ebstein's anomaly were identified. Complete data of 12 of the patients were available for review. Results: Mean gestational age at diagnosis was 31.4 weeks. No risk factors for cardiac diseases were present in 82%. 2 cases occurred in pregnancies at risk (lithium ingestion in one and maternal diabetes in the other). Fetal heart failure was present in the first examination in 5 fetuses. Atrial tachyarrhythmias were present in 2 patients, and were reverted to sinus rhythm with maternal digitalis. Fetal hydrops were observed in 2 cases. There were 4 deaths in utero" and 3 in the neonatal period. All fetuses who died were in heart failure and the 3 neonates who died were in heart failure and the 3 neonates who did not survive had functional pulmonary atresia. Immediate neonatal surgery was performed in 5 babies, with 1 survival. The remaining patients are alive and well. Conclusion: Ebstein's anomaly is a severe anomaly when detected during fetal life, and early prenatal diagnosis may contribute to optimize perinatal management.

P665

Successful percutaneous valvotomie in a fetus with pulmonary stenosis intact septum.

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Pulmonary atresia with intact ventricular septum (PAIVS) associated with RV hypoplasia was diagnosed in a 27-week fetus. Doppler examination revealed holosystolic tricuspid regurgitation (TR), suprasystemic RV pressures and signs of heart failure by umbilical venous pulsations. Due to the bad prognosis, invasive intervensional therapy was anticipated. An ultrasound guided puncture of the hypoplastic RV was performed using a 18-gauge needle. The needle tip was directed into the RVOT and then advanced through the atresic valve into the main PA. A 2.5 F coronary balloon catheter (4mm inflated balloon diameter/ 1,5 cm length) was then inserted over a guide wire, placed across the pulmonary annulus and inflated. Immediately after the procedure turbulent (1.5 m/s) antegrade flow across the PV and holodiastolic regurgitation could be documented, the velocity of the TR jet had decreased to 1.8 m/s. RV filling had improved from a monophasic to biphasic filling pattern but the venous Doppler was unchanged. four weeks later the velocity across the PV had increased again to 3.4 m/s. TR had disappeared. Percutaneous valvuloplasty of an atresic PV in the fetus is feasible. It resulted in decompression of the RV, improved RV filling and disappearance of TR. Short time follow-up showed progressive re stenosis of the PV, no significant RV growth and no signs of coronary artery fistulae.

P666

Evolution of cardiac tumors (CT) in utero and after birth

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This study was undertaken to analyse the characteristics and evolution of fetuses with CT. Material and methods: Between 1986 and 2000, CT were diagnosed by echocardiography in 10 fetuses, at 21–36 wg and were followed-up for 5m–12 yrs (median 1.2 yrs). Results: 8 fetuses presented one or more intracardiac noduli suggestive of rhabdomyomatosis (R.): small, moderate in 7 and in one a large mass in the interventricular septum, in proximity of mural and aortic valves. Fetus n. 9, of mother with tuberous sclerosis (TS), showed at 31 wg a voluminous pericardial mass and smaller intracardiac nodula. The case 10 presented at 31 wg. with pericardial effusion and intramural masses in both ventricles. Outcome: 2/4 cases diagnosed before 24 wg. opted for termination of pregnancy; autopsy confirmed R.. CT grew progressively in utero, without true obstruction. After birth, cases 1–9 showed no relevant hemodynamic problems. The case 10 presented repeated ventricular tachycardias (VT) and surgical approach was attempted, at biopsy fibroma was diagnosed. This child died at 5m. as VT; 6/7 survived infants had TS: one child needed neurosurgery and one case 8 has bilateral polycystic kidneys and hypertension at 10 yrs. Cardiac masses regressed in all cases at follow-up. Conclusions: our data show a variable impact of CT found in utero and a frequent pericardial regression in R.

P667

Fetal echocardiography. Indications and results

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Introduction: The indication for performing a fetal echocardiography is responsible for different rates of incidence of congenital heart disease(CHD). The objective of this study is to show the results of a reference center in fetal echocardiography and to correlate the indications with the found alterations. Method: Retrospective analysis of the registered data of the fetal echocardiograms was performed including the period between november 1996 and july 2000. Results: 672 fetal echocardiograms were performed being in 608 the first study (591 pregnancies, 17 twin); and 64 control exams. In 208 exams (208/608)fetal heart abnormality was evidenced (34.2%) being CHD in 22(5.3%), functional heart alterations in 26 (4.2%), hypertrofic myocardiopathy of the left ventricle in 3(1.4%) and dextrocardia in 4 (1.9%). Arrhythmias were identified in 51 fetus (24.6%) and ventricular echogenic focus in 95 (45.6%). In the 22 fetus with CHD,the indications were: suspicion of a congenital heart abnormality seen on a routine obstetric ultrasonography in 16 (72.7%), presence of extracardiac anomalies in 4 (18.2%), fetal hydrops in 2 (9.1%), chromossomal abnormality in 2 (9.1%), arterial hypertension, presence of echogenic focus, arrhythmia and previous chromossomal abnormality in 1 of each respectively. Fifty one arrhythmias were confirmed among 84 indications by rhythm abnormality (60.7%).21 CHD and/or functional abnormality among 58 indications for extracardiac anomalies (36.2%), 19 CHD among 33 indications for suspicion of heart abnormality on the routine obstetric ultrasound (57.5%); and 6 CHD among 8 indications for nonimmune hydrops (75%).Conclusion: Up to 90% of CHD occurs in considered normal" obstetric patients. The fetal echocardiography is an important method in the diagnosis of fetal heart abnormalities if teamed an alliance with the experience of the obstetric ultrassonographist for the detection.

P668

The benefit of the Tissue Doppler Imaging in evaluation of the ventricular velocities and heart motions of the fetal heart

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The benefit of the Tissue Doppler Imaging in evaluation of ventricular myocardial velocities and heart motions of the fetal. The objective of this study was to evaluate myocardial velocities and heart motions of the fetal hearts by using Tissue Doppler Imaging(TDI). TDI was performed in 34 fetal hearts with gestational ages 20–35 wks. (mean 27+/-4.9 wks) to evaluate myocardial velocities and heart motions. The Toshiba, Power Vision, machine was used with an appropriate setting of colour-coded tissue velocities. The apical four chambers and apical or parasternal long axis views were the standard planes for measuring myocardial velocities and the evaluate the heart motions. The results showed the myocardial velocities of the posterior wall of the left ventricle during the early mid, late systolic phase were 1.61+/-0.71, 1.83+/-0.85, 0.93+/-0.45 cm/sec. and in the early mid, late diastolic phase were 1.22+/-0.81, 1.98+/-0.98, 1.17+/-0.67 cm/sec. respectively. The myocardial velocity of the anterior wall of the right ventricle during the early, mid, late systolic phase were 1.71+/-0.93, 1.40+/-1.03, 0.93+/-0.55 cm/sec and in the early,mid,late diastolic phase were 1.25+/-0.80, 1.69+/-1.22, 1.31+/-0.86 cm/sec respectively. The myocardial velocity of the interventricular septum could not be measured due to the abnormal septal motion

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

and the total heart movement. The fetal heart had anterior displacement during systole and posterior translation during diastole and also had counter-clockwise rotation during the systolic phase. Conclusion: the benefit of using TDI to evaluate myocardial velocities of the fetal heart is limited due to angle between the beam of the ultrasound and axis of measure and the total fetal heart motion. The myocardial velocities of the posterior wall of the left ventricle and the anterior wall of the right ventricle are not related to the gestational age.

P669

Outcome in 516 cases with congenital heart disease on fetal echocardiography: Multicenter study between 1998–1999

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Purpose: To report the incidence of each congenital heart disease (CHD) on fetal echocardiography and clinical impact on the outcome of diagnosed cases. Materials and Methods: Between 1998–1999, 516 consecutive cases of structural cardiac malformation have been diagnosed prenatally at 9 centers. The incidence of each cardiac malformation and their outcome according to the presence of associated factors was analyzed. Results: There were 365 cases of significant CHD, 104 cases of miscellaneous CHD, 47 cases of fetal arrhythmia. The five most common diagnosis in the fetus in order of frequency are ventricular septal defect (VSD)(n=113), heterotaxias (n=39), hypoplastic left heart syndrome (HLHS)(n=31), tetralogy of Fallot (TOF)(n=31), atrioventricular septal defect (AVSD) (n=28), and coarctation of aorta (n=20). These 5 frequent CHDs consisted of 70.7 % of significant CHD. The gestational age of the fetuses at diagnosis was 11–41 weeks. The overall rate of termination of pregnancy (TOP) in significant CHD was 33%. The TOP rate was 15% (n=17) in VSD, 50% (n=19) in heterotaxias, 39% (n=12) in HLHS, 69% (n=15) in TOF, 68% (n=19) in AVSD and 25 %(n=5) in coarctation of aorta. In 15 of 17 terminated VSD cases showed associated anomalies or chromosomal anomaly. The frequency of associated extracardiac or chromosomal anomaly was 17.9%(n=24) in VSD, 92% (n=36) in heterotaxias, 29% (n=9) in HLHS, 42% (n=13) in TOF, 54% (n=15) in AVSD and 35% (n=7) in coarctation of aorta. Five cases of d-transposition of great arteries with intact ventricular septum had been diagnosed prenatally, all of them had planned delivery and all survived arterial switch operation in neonatal period.

P670

Prenatal diagnosis of congenital heart disease affects pre-operative acidosis in the newborn patient

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Objectives: Congenital heart disease is the leading cause of death in the first year after birth. Prenatal diagnosis of the disease can optimize the pre-operative condition of the patient and lead to a better outcome. In this retrospective study we compared the occurrence of metabolic acidosis in patients with and without prenatal diagnosis of a congenital heart disease. Methods: Data of 498 patients who needed surgery for congenital heart disease within 31 days of life were analyzed retrospectively. Arterial blood gases at fixed time intervals and worst blood gas of 81 patients with and 327 patients without a prenatal diagnosis were compared, categorizing the patients on ductus-dependency, anticipated uni- or biventricular repair and left, right or no heart obstruction. Results: In the overall group significant differences were found in the worst arterial pH, (prenatal vs postnatal: 7.31 +/- 0.01 vs. 7.28 +/- 0.01, p=0.004) and in the worst pre-operative Base Excess (-1.93 +/- 0.45 mEq/l vs. -7.26 +/- 0.55 mEq/l, p=0.000) and worst lactate (3.14 +/- 0.17 mmol/l and 5.35 +/- 0.58 mmol/l, p=0.006), with acidosis more common among the prenatally diagnosed group. No significant differences were found in worst rates of pCO2 and pO2. In the group of patients with ductus dependent congenital heart disease the difference between pre- and postnatally diagnosed patients was more significant than in the group with non-ductus dependent lesions. Conclusions: Prenatal diagnosis of congenital heart disease minimizes metabolic acidosis in patients with congenital heart disease and will be associated with improved surgical results and prevention of cerebral damage among this fragile group of patients.

P671

What is the role of fetal 3D echocardiography?

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The development of fetal 3D echocardiography has for a long time been limited by the lack of a fetal ECG combined with frequent fetal movement. Recent technological developments have permitted the performance of fetal 3D echocardiography. The aim of our study was to evaluate the usefulness of 3D in the diagnosis of different fetal cardiac malformations. Six of 10 fetuses with a gestational age between 20 and 31 weeks presented with different congenital heart diseases detected using classical 2D echocardiography including: Valvular/annular stenosis of the aorta / probable interruption of the aortic arch 2 with transposition of the great vessels. A transabdominal scanning was performed on each pregnant woman using a KRETZ 5.0 D MT 3D echocardiography system. For every fetus, transverse and sagittal sections of the heart and great vessels were acquired to reconstruct 3D projections which was performed retrospectively. In normal hearts, the 3D study permitted rapid and easy acquisition of longitudinal and short axis views as well as to study the relation of the great vessels without changing the probe orientation. Moreover the 3D reconstruction allowed better assessment of the aortic arch. Of the 2 fetuses with transposition of great vessels, the 3D study revealed a subpulmonary VSD in one. The 3D study of the aortic arch in 3 fetuses with small left heart cavities permitted the diagnosis of coarctation with documentation of narrowing of the aortic isthmus. In contrast, a 3D study ruled out interruption of the aortic arch by perfectly reconstructing the aortic arch. Fetal real-time 3D echocardiography is a feasible and useful new technique. With future technological progress, a 3D study of the fetal heart will become routine particularly if 2D study is incomplete.

P672

Isolated non-compaction of the ventricular myocardium presenting as fetal complete heart block and hydrops: antenatal diagnosis and natural history

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Background: Non-compaction of the ventricular myocardium is a rare cardiomyopathy characterised by the persistence of numerous marked ventricular trabeculations and deep intertrabecular recesses with direct coronary supply by the ventricular cavities. Methods: Routine 20-weeks' ultrasonographic examination in a 20-year-old healthy primigravida detected fetal hydrops and bradycardia. Fetal echocardiography at 23 weeks revealed normal sites and connections but spongy or non-compacted myocardium with color flow entering the sinusoids. There was marked atrial and enlarged cardiothoracic ratio with complete heart block at 50bpm. Results: Immunological investigation and viral screen in the mother were negative. Amniocentesis, chorionic villous and fetal blood sampling performed for karyotyping, metabolic studies, fetal blood gas, blood film, virus investigations and infection screen were normal. At 34 weeks marked ascites, skin oedema and bilateral pleural effusions were noted. Ventricular rate remained at 50bpm. The pleural effusions and the ascites were aspirated and a Caesarean section scheduled. Postnatal echocardiography verified the presence of non-compacted ventricular myocardium. Cardiac function was very poor with a ventricular rate of 30bpm. The infant died at 14 hours of age and the echocardiographic diagnosis was confirmed at postmortem. Conclusion: This is the first case of non-compaction of the ventricular myocardium mentioned antenatally until the perinatal period. The severity of prognosis and the familial occurrence indicate the value of antenatal diagnosis, which is feasible using currently available ultrasonographic technology. The presence of non-compaction of the ventricular myocardium at a gestational age when the myocardium should already be compacted supports the theory of arrest in embryogenesis as the pathogenetic mechanism. Non-compaction of the ventricular myocardium, although rare can be added to the list of the aetiology of congenital complete heart block and fetal hydrops.

P673

Identical genome does not predict concordant cardiovascular structure and function

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BACKGROUND: Arterial stiffness is increased in infancy in the donor twin in Twin-to-Twin Transfusion Syndrome (TTTS). We hypothesised no cardiovascular differences in monochorionic diamniotic (MCDA) fetal twin pairs without TTTS. METHODS: We reviewed the fetal echocardiograms of 28 MCDA twin pairs with TTTS and 29 uncomplicated MCDA pairs examined in 1998–1999. One MCDA had fetal pulmonary valve stenosis (Z 4.4m/s),

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

This pair was excluded. ANOVA was used for analysis. RESULTS: Mean gestational age at scan was 23.7±3.6 weeks. Results are presented in the table. No differences were seen between the bigger(T1) and smaller twin(T2) in MCDA without TTTS. The four groups did not differ regarding fetal weight, heart rate, pulmonary and aortic valve diameters, left and right indexed ventricular output, aortic and pulmonary Doppler ejection and acceleration times. Ventricular hypertrophy was found in twelve recipients(R) and tricuspid regurgitation in seven. Abnormal myocardial function was present in four and mitral regurgitation in five. Six donors(D) had abnormal umbilical artery Dopplers and two tricuspid regurgitation. Four fetuses had congenital heart disease. Two had ventricular septal defect, one postnatal pulmonary valve stenosis (fetal velocity 0.62m/s) and one postnatal aortic coarctation. Two had single umbilical artery. CONCLUSION: Identical prenatal maternal hemodynamic differences results in identical fetal cardiovascular physiology but structural defects are not represented in MCDA. Circulatory imbalance may explain the antecedent arterial stiffness seen in infancy following TTTS.

P674

Clinical course of the fetal atrioventricular block in Japanese population: A multicenter experience

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The aim of this study is to elucidate the pre- and post-natal course of the fetal atrioventricular block (AVB) in Japanese population. Fifty-nine fetuses from 10 institutions were analyzed. Postnatal follow-up period was 0 day to 12 years (mean 2.55 years). Twenty-one had congenital heart defect (CHD) and 15 of them were left atrial isomerism. Thirty-seven had structurally normal heart, and 26 of them were positive maternal antinuclear antibodies. Remaining one case had cardiac tumor. Gestational age at diagnosis was 17 to 38 (median 25) weeks and all of the positive antibody fetuses were diagnosed after 20 weeks. Fetal hydrops (FH) were associated in 14 fetuses. Of the 59 fetuses, 34 survived, 4 terminated, 6 died in utero, 5 died in neonatal period, and 8 died after the neonatal period (2 unknown). Association of CHD (p = 0006) and presence of FH (p = .0001) were risk for death. Maternal administration of sympathomimetics successfully increased the fetal heart rate in 3 of 6 treated fetuses. Steroid was not effective in all 7. AVB was spontaneously resolved before birth in 2 fetuses both had no CHD or maternal antibody. Postnatally, all survivors with CHD had pacemaker implantation (PMI). Although neonatal mortality of the AVB patients with no CHD nor FH is only 7%, the number of PMI is continue to increase during follow-up. In addition, 2 cases died even after the PMI, and additional 5 cases have decreased cardiac function. In conclusion, CHD and FH were risks for death. Chance to have PMI was very high in the case with CHD, and the chance of PMI increase over time in the cases without CHD. The possible myocardial damage due to maternal antibody was continuing potential problem.

P675

Caveats in the recognition of abnormal ventricular topology in the human fetus

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Background: It is generally accepted that abnormal, left-handed ventricular topology can be accurately diagnosed postnatally, but the frequency, morphology and diagnosis in the fetus have not been examined in detail. Objectives: To determine predictors of ventricular architecture and assess accuracy of fetal diagnosis. Methods: Retrospective review of echocardiograms and cardiac specimens known to exhibit left-handed ventricular topology, examined as a tertiary centre for fetal echocardiography. Results: Differential insertion of the leaflets of the atrioventricular valves aids, but is not a prerequisite for, diagnosis. In its absence, the presence of the moderator band within the morphologically right ventricle is the best predictor of topology. Caveats to diagnosis involve anomalies of myocardial structure, such as marked ventricular hypoplasia, biventricular hypertrophy, anomalous muscular ridges and unusually positioned moderator bands. Frequency of recognition of left-handed topology improved with year of diagnosis in concert with improvements in imaging quality. Conclusions: It is feasible to diagnose left-handed ventricular topology in the fetus, but there may be caveats to diagnosis that are peculiar to the stage of development. Some of these may be overcome with improved ultrasonic imaging, but the possibility that a left-handed ventricular mass cannot be recognised should be considered when counselling parents following prenatal diagnosis.

P676

The benefit of the Tissue Doppler Imaging in evaluation of ventricular myocardial velocities and heart motions of the fetal heart

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The benefit of the Tissue Doppler Imaging in evaluation of ventricular myocardial velocities and heart motions of the fetal heart. Jaijaroensuk V. Bangkok, Thailand. The objective of this study was to evaluate myocardial velocities and heart motions of the fetal heart by using Tissue Doppler Imaging (TDI) technique. TDI was performed in 54 fetal hearts with gestational ages 20–35 wks (mean 27.1 +/-4.9 wks) to evaluate myocardial velocities and heart motions. The Toshiba, Power Vision, machine was used with an appropriate setting of colour coded tissue velocities. The apical four chambers and apical or parasternal long axis views were the standard planes for measuring myocardial velocities and the evaluate the heart motions. The results showed the myocardial velocities of the posterior wall of the left ventricle(PWLV) during the early, mid, late systolic phase were 1.61+/-0.71, 1.82+/-0.86, 0.93+/-0.45 cm/sec and in the early, mid, late diastolic phase were 1.32+/-0.81, 1.68+/-0.98, 1.17+/-0.67 cm/sec respectively. The myocardial velocity of the anterior wall of the right ventricle (AWRV) during the early, mid, late systolic phase were 1.31+/-0.95, 1.40+/-1.05, 0.90+/-0.55 cm/sec. and in the early, mid, late diastolic phase were 1.25+/-0.88, 1.69+/-1.23, 1.31+/-0.86 cm/sec respectively. The myocardial velocity of the interventricular septum could not be measured due to the abnormal septal motion and the total fetal heart movement. The fetal heart had axial displacement during systole and posterior translation during diastole and also had counter-clockwise rotation during the systolic phase. Conclusion: The benefit of using the TDI to evaluate myocardial velocities of the fetal heart is limited due to angle between the beam of the ultrasound and area of measure and the total fetal heart motions. The myocardial velocities of the PWLV and the AWRV are not related to the gestational age.

P677

Drug treatment of fetal tachycardia

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The pharmacological treatment of fetal tachycardia (FT) has been described in various publications. We present a meta-analysis reviewing the necessity for treatment of FT and regimen of drugs used in the last two decades and its mode of administration. Methods: Meta-analysis of the literature regarding FT in the last two decades. Results: The absence of reliable predictors of fetal hydrops (FH) has led many centers to initiate treatment as soon as the diagnosis of FT has been established, although a small minority advocate non-intervention. As primary form of pharmacological intervention oral maternal transplacental therapy is generally preferred. The mainstream drug in FT is digoxin, however, effectiveness remains a point of discussion. After digoxin sotalol seems to be the most promising agent, specifically in atrial flutter (AFl) and nonhydropic SVT. Flecainide is a very successful drug in the treatment of fetal SVT although concerns about possible pro-arrhythmic effect have limited its use. Amiodarone has been described favorably, but is frequently excluded due to its expensive side effects. In severely hydropic fetuses and/or therapy resistant FT, direct fetal therapy is sometimes initiated, to minimize the number of invasive procedures, an intramuscular or intraperitoneal injection that provides a more sustained release is to be preferred. Conclusion: Based on case data we conclude to propose a drug protocol of sotalol 160 mg 2x/day orally, increased to a maximum of 480 mg daily. Whenever sinus rhythm is not achieved, addition of digoxin 0.250 mg 3x/day is recommended. Only in SVT complicated by FH, either digoxin 1–2 mg IV in 24 hours and subsequently 0.5–1 mg/day IV or flecainide 200–400 mg/day orally is proposed. Initiating direct fetal therapy may follow failure of transplacental therapy.

P678

Fetal cardiac anomalies at the Institute Polish Mothers Memorial Hospital (1994–1999)

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5083 fetal sonographic + echocardiographic examinations were performed in our unit in 1994–1999. 1436 pregnant women with fetal malformations were evaluated. The medical records were completed by postnatal examination or by an autopsy report. Total 310 cases of fetal heart defects were detected. In 93

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

fetuses with ascites, fetal echocardiography allowed to diagnose congestive heart failure in 51 cases. There were 216 fetuses with premature contractions, 50 with supraventricular tachycardia and 16 with complete heart block. There were also other cardiac problems: heart tumors (9), ectopia cordis (12), myocardial infarct or myocarditis (15) and an aneurysm of interventricular septum (1) and LV diverticulum (1). In addition to cardiac problems in singleton pregnancies a specific cardiac problem in twins were such as transfusion-twin syndrome (11), conjoined twins with conjoined heart (6), and acardiac twins (4). Over the years in our referral center a significant increase of fetal CHD per year was observed and decreased number of fetuses with benign heart arrhythmias. In 1999, 82 fetuses with CHD were diagnosed at mean 30 weeks of gestation. In 1999 in Pediatric Cardiology Clinic were admitted 98 newborns with CHD. A common group of patients of this Clinic and our Department consisted of 15 newborns with CHD. 85% of neonates with CHD had prenatal diagnosis in our center in 1999. During prenatal life there are many cardiac problems which do not exist during the postnatal life. Polish obstetricians use prenatal cardiology but still pediatric cardiologists should be obliged to make this field better known for the society.

P079

Acute effects of smoking in the hemodynamics of fetal-maternal-placental unit

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Tobacco is the most frequently used drug during pregnancy. Maternal smoking causes an increase in vasoactive substances and a decrease in vasodilating substances in the umbilical cord, which may be related to acute and chronic perfusion changes in the fetal-maternal-placental unit. The sample was constituted by 21 pregnant women chronic smokers with a normal gestation, without risk factors for fetal disease. They were submitted to obstetric ultrasound evaluation and to fetal echocardiographic before and after smoking a cigarette with a standard concentration of nicotine of 0.5mg and 4mg of carbon monoxide. Mean gestational age was 30.20 weeks. The mean amount of daily used cigarettes was 9.67. Results obtained before and after maternal smoking showed an increase in maternal systolic blood pressure ($p=0.004$) and in diastolic blood pressure ($p=0.035$). There was an increase in maternal heart rate ($p<0.0001$) and in fetal heart rate ($p=0.044$). A decrease in S/D ratio in the left uterine artery ($p=0.039$) and in the right uterine artery ($p=0.014$) immediately after smoking was recorded. The S/D ratio in the fetal middle cerebral artery did not change ($p=0.078$), as well as in the ductus arteriosus ($p=0.154$), and in pulmonary artery ($p=0.958$). There was no significant change in the S/D ratio of the umbilical artery ($p=0.554$), in the left ventricular ejection fraction ($p=0.945$) and in the myocardial index of the septum primum ($p=0.836$). Exposition to smoking during pregnancy affects maternal and fetal physiologic variables without changes in fetal heart function. The observed decrease in uterine vascular resistance is probably related to a dose-dependent action of nicotine and others cigarette components.

P080

New electrode for in utero pacing for fetal congenital heart block

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Fetal complete heart block carries a poor prognosis when manifested with hydrops. Intrauterine pacing seems to be the next logical form of treatment. However, premature labor following hysterotomy remains a major obstacle. A new T-bar shaped electrode was developed, with the aim of pacing the fetus, with no need for intrauterine open surgical procedures. We describe a case where we believe is the first documentation of the voltage strength-duration curve for acute myocardial stimulation threshold of a human fetus that survived intrauterine pacemaker implantation. A 36-year-old woman was referred at 18 weeks' gestation with a fetus presenting with complete heart block (FHR = 47 bpm) and hydrops, associated with structural heart defects (left atrial isomerism and atrioventricular septal defect). In view of the poor prognosis, the patient consented to allow attempts at in utero pacing. In the 25th week of gestation, the new electrode was successfully implanted into the fetal myocardium through the tip of a modified spinal needle, under echocardiogram guidance. Stimulation resistance was 357 ohms and sensed fetal R wave of 8.4 mV. The voltage strength-duration curve remained relatively constant at pulse width > 0.6 msec. The new electrode was then connected to a Biotronik Axeos SR, single-chamber pulse generator, which was implanted subcutaneously in the maternal abdominal wall (pacing rate = 140 bpm). During the procedure, the fetus developed cardiac tamponade, managed with pericardiocentesis. The 1st POD echocardiogram revealed mild pericardial effusion. Fetal heart rate was stable, with low stimulation thresholds and no stimulation failures. However, the fetus died 36 hours after the procedure, probably due to cardiac tamponade. This case emphasizes that percutaneous fetal pacing with the new electrode in human fetus is a simple and reproducible method.

P081

Fetal atrial flutter: efficacy of Flecainide and assessment of drug levels in the fetal, maternal and funicular serum

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We report a case of a fetus of 30 weeks admitted with atrial flutter with an atrial rate around 440 bpm and 2:1 conduction complicated by hydrops. After excluding maternal ECG and echocardiographic abnormalities, the mother was treated with rapid digitalization (1.5 mg iv in 24 hrs, followed by 0.500 mg po b.i.d.) and metoprolol (100 mg t.i.d.). After four days, despite digoxin levels maintained in the therapeutic range, only a partial response in terms of rate control was seen, the hydrops was unchanged and the fetal contractility remained poor. Metoprolol was ceased and replaced with flecainide at an initial dose of 150 mg b.i.d. Serum flecainide levels were assessed 18 hrly. Therapeutic range was attained at the first sampling. At steady state concentrations the fetal heart rate remained around 170–180 bpm and a few improvement in myocardial contractile function and hydrops was noted. The fetus remained in a favorable hemodynamic state until delivery via elective cesarean section at 34 weeks. Maternal, neonatal and funicular assays of flecainide concentration taken during the delivery were consistently within the therapeutic range (810, 890 and 460 ng/ml respectively). A 1.98kg neonate was delivered with Apgars of 8 and 9 respectively. No signs of edema or pleural effusions were detected. ECG showed the persistence of atrial flutter with heart rate of 340 bpm and 3:1 conduction. Echocardiography demonstrated a preserved left ventricular systolic function with an ejection fraction of 50%. Anti-arrhythmic therapy with flecainide and digoxin was continued in the neonate but six months, despite restoration of the sinus rhythm in the third day of life. Conclusion: In this case of atrial flutter complicated by hydrops, flecainide was very effective in controlling heart rate with an absence of serious side effects, both fetal and maternal. Using regular assays the therapeutic range of flecainide can be accurately achieved and maintained which is helpful in assessing placental transfer particularly in the case of fetal hydrops.

P082

Prenatal diagnosis of pulmonary atresia with intact ventricular septum in utero and outcome

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We sought to determine the accuracy of prenatal diagnosis of pulmonary atresia with intact ventricular septum (PAIVS) including ventriculocoronary connections and to evaluate the outcome. From January 1991 to September 2000, 1,552 fetuses were evaluated. 8 had PAIVS among 868 fetuses diagnosed with congenital heart disease. The echocardiographic evaluation consisted of measuring the tricuspid and the mitral valve annulus, measuring the right ventricular dimensions, assessing the degree of tricuspid regurgitation, measuring the pulmonary trunk and branch pulmonary artery sizes, and assessing the ventriculocoronary connections. The mean age at diagnosis was 25.3 weeks (range 19–33). Three fetuses had ventriculocoronary connections and all of them had absence of the infundibular segment with a hypoplastic tricuspid valve without tricuspid regurgitation. One of these 3 had a right aortic arch, and 1 had tetralogy of Fallot with a ventral septal defect type malformation. The 5 fetuses with a triparticle right ventricle all had moderate tricuspid regurgitation, and 3 had right ventricular outflow tract obstruction. There was 1 twin pregnancy with both fetuses having PAIVS. Two pregnancies were terminated. 5 ultimately underwent surgery of which 2 died and 3 are alive. One fetus not yet born. It is possible to diagnose PAIVS including ventriculocoronary communications and associated anomalies. Prenatal detection of the associated abnormalities aids in family counseling and decisions on postnatal management.

P083

Heterotaxy syndrome: prenatal echocardiographic diagnosis and prognosis

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https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Our objective was to determine the accuracy of prenatal cardiac and non-cardiac diagnosis, prognosis and outcome of heterotaxy syndrome (HS). We reviewed our 9 year experience of prenatal diagnosis of HS. Twenty fetuses (12 left isomerism, 8 right isomerism) were identified with HS among 168 fetuses diagnosed with congenital heart disease. The diagnosis was confirmed by postnatal echo-cardiac catheterization, surgery, or autopsy. The mean gestational age at the time of diagnosis of the left isomerism was 28.6 weeks (range 17–36) and 26.5 weeks (range 21–32) for right isomerism. Of the 8 fetuses with right isomerism 5 had atrioventricular septal defect (AVSD) and D-malposition of the great arteries, 6 had totally anomalous pulmonary venous connections (all diagnosed postprenatally), 8 had atrio-caval juxtaposition, 7 had pulmonary stenosis or atresia, and 4 are alive following surgery. Two died without surgery, 1 pregnancy was terminated, and 1 died following surgery. Of the 12 fetuses with left isomerism, 8 had AVSD, 5 had bradycardia, 3 had complete atrioventricular block, and all 12 had interrupted inferior vena cava (IVC) with azygous continuation. There pregnancies were terminated and there was 1 intrauterine death. Four died after birth, 1 is in-utero, and 3 are alive. The characteristic findings in fetuses with right isomerism were atrio-caval juxtaposition, AVSD, double-outlet right ventricle, D-malposed great arteries, pulmonary stenosis or atresia, and anomalies of the pulmonary venous connection. The characteristic findings in fetuses with left isomerism were interrupted IVC with azygous continuation, AVSD, and bradycardia or complete atrioventricular block. It is possible to diagnose prenatally complex congenital heart disease including the pulmonary venous connections. HS carries a poor prognosis when diagnosed prenatally.

P684

Hypoplastic left heart syndrome: prenatal diagnosis and outcome

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The purpose of the study was to evaluate the impact of associated cardiac as well as non-cardiac diagnosis on overall outcome following prenatal diagnosis of hypoplastic left heart syndrome (HLHS). There were 2025 fetal echocardiograms performed on 1352 fetuses over nine years. There were 31 fetuses diagnosed with HLHS among 168 fetuses with congenital heart disease. Sixteen fetuses were diagnosed before 24 weeks of gestation and 15 after 24 weeks. The mean age at the time of diagnoses was 21.5 weeks (range 16 to 39.7). In utero diagnosed before 24 weeks, term parton of pregnancy was chosen. Parents of seven infants chose compassionate care, one each with trisomy 18, bilateral cerebral hemorrhage, left isomerism, two with Dandy-Walker malformation of the brain, and two without any associated malformation. There were 15 infants offered the Norwood palliation, 10 survived and seven died postoperatively. Of the seven who died, one had restrictive interatrial septum, one had moderate tricuspid regurgitation, and one had fetal obstructive pulmonary venous connection. The survival rate among infants offered surgery was 50%. The overall survival rate was 31%. The prognosis of prenatally diagnosed HLHS was poor. Parental counseling should include cardiac as well as the non-cardiac diagnoses.

P685

Atrioventricular plane nonconcordance with atrioventricular alignment discordance, superoinferior ventricles and double outlet right ventricle: fetal and neonatal echocardiographic findings

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Objectives: Visceroatrial situs, ventricular morphology, atrioventricular and ventriculoarterial alignments can reliably be assessed with fetal echocardiography. We herein report, to our knowledge, the first fetal echocardiographic diagnosis of a fetus with visceroatrial situs solitus, atrioventricular alignment discordance, superoinferior ventricles and double outlet ventricle. Case Report: A 22-year-old gravida 3 para 2 mother with a previous history of intrauterine fetal death, who had previously been followed-up at another center and who was referred to our unit because of the suspicion about her diagnosis, underwent fetal ultrasonographic evaluation at 29 weeks gestation. Ultrasonic examination of the fetus was carried out. Fetal echocardiographic evaluation demonstrated a left-sided stomach and levocardia. The aorta descended on the left side of the spine and a right-sided inferior vena cava drained to the right sided atrium. A four-chamber view of the heart was obtained, the right-sided morphologically left ventricle appeared prominent with a horizontally oriented interventricular septum resulting in superoinferior ventricles with the small right ventricle superior, leftward and anterior to the left ventricle. The great arteries were well developed and both were connected to the right ventricle with a posteriorly located pulmonary artery. Patent ductus arteriosus, large muscular ventricular septal defect and secundum atrial septal defect were also detected. A diagnosis of double outlet right ventricle with superoinferior ventricles was established. This arrangement was associated with ventricular inversion and L-looped ventricles. Neonatal echocardiographic evaluation confirmed the diagnosis. Accordingly there was four concordance (S, L, and A) with superoinferior ventricles and atrioventricular alignment discordance.

P686

The usefulness of color flow mapping on 3-vessel view of fetal heart

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Introduction: Three-vessel view of fetal heart is an important plane for screening congenital cardiac defects. The 3-vessel view is used to examine the number, the size, the arrangement, and the alignment of these vessels, ie superior vena cava, ascending aorta, and pulmonary artery. We applied color flow mapping on 3-vessel view in the fetuses with suspecting congenital cardiac defects. So, we analyzed the usefulness of color flow mapping on 3-vessel view for diagnosing significant outflow tract obstruction. Materials and Methods: From Dec. 1996 ~ Nov. 2000, we applied color flow mapping on 3-vessel view especially those fetuses with abnormal vessel number (non-visualization of pulmonary artery or ascending aorta) and abnormal vessel size (small pulmonary artery or ascending aorta). Results: In 3-vessel view of fetal heart, there were reversal of ductus arteriosus flow from descending aorta to main pulmonary artery in fetuses with pulmonary atresia or significantly severe pulmonary stenosis (N=12). There were reversal of aortic arch flow from descending aorta to ascending aorta in fetuses with severe coarctation of aorta and/or severe aortic stenosis/atresia (N=15). A small size of pulmonary artery or ascending aorta is an important finding of outflow tract obstruction, however, no evidence of flow reversal on distal arch or aortic arch indicates less severe outflow tract obstruction. Conclusion: Applying color flow mapping on 3-vessel view of fetal heart is a useful method for discriminating significant outflow tract obstruction in fetuses with congenital cardiac defects.

P687

Impact of early diagnosis of severe congenital heart disease

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Impact of early diagnosis of severe congenital heart disease. Marasini, M., Gurnhill, M., Inforzi, J., Villa, A., Contreras, A., Low, A., Makrowsky, J. Buenos Aires Argentina. Purpose: To study the impact of prenatal or early diagnosis of severe congenital heart disease (CHD) on mortality and costs. Methods: Between May 1998 and October 2000, 60 neonates required urgent interventional catheterism (IC) and/or surgery. Patients (pts) were divided into 2 groups. A) 49 pts born at our hospital, and B) 37 pts referred from other less complex centers. Results: Group A: 5/49 pts died, all after surgery (mortality 10%). Mean age at intervention: 9 days, mean hospital stay: 19 days and mean total cost: U$S 21,000. All had obstetric echography (OE) but only 27/49 were sent for fetal echocardiography: 15 with anomalies at the 4 chamber view on the OE (55%), 6 with evident CHD (22%), 3 with unclear CHD (11%), 2 arrhythmia (7%) and 1 intrauterine growth retardation (3%). 37/37 were referred from other centers. In the remaining 21, CHD were missed at OE but were detected early by one neonatologist. Group B: 9/37 pts died, 4 after surgery or IC (mortality 12.9%) and 5 before any intervention. Mean age at intervention: 25 days, mean hospital stay: 41 days and mean total cost: U$S 43,000. All had an OE but in none a CHD were suspected. Conclusions: 1) There was no difference in surgery mortality between the two groups, but 5 pts in group B died before any intervention could be carried out. 2) There was significant difference (p<0.05) in mean age at operation, hospital stay and mean total cost. 3) Training the obstetric echographer will enhance prenatal diagnosis which permits planning delivery in high complexity hospitals.

P688

Qualitative study of fetal echocardiography – decisions that follow

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Although antenatal diagnosis of CHD has been available since the 1970s, formalized Fetal Cardiology Centres are a more recent phenomenon. Within our multidisciplinary Fetal Cardiology Service, we conduct 400–450 fetal

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

echocardiograms annually with 60–80 positive diagnoses. While continually advancing the technological aspects of antenatal diagnosis of CHD, we are committed to ongoing research to understand the experiences of women and their partners who receive antenatal services. This qualitative study, employing in-depth interviews with 39 women and their partners before and after the birth of their baby with CHD, focused on how couples manage this experience. Constant comparative analysis of data revealed the agonising nature of decisions concerning further diagnostic testing and, in many cases, the option of pregnancy termination. Participants described the difficulties of making such crucial decisions in the time-pressured context of a progressing pregnancy. Some considered amniocentesis to detect chromosomal problems that would convince them to terminate the pregnancy. Others wanted this information to help with treatment decisions following birth. Others declared amniocentesis because pregnancy termination was not an option. Some participants expressed feeling vulnerable in relation to the influence that health care professionals had on their decision-making. Some felt that the professionals were too directive, especially related to termination, not a surprising finding perhaps, considering all couples conceived their pregnancies. Their insistence, however, that health care professionals explore the beliefs and values of those they are counseling regarding the antenatal diagnosis of CHD, warrants ongoing discussion amongst us as multidisciplinary colleagues of the perinatal ethical issues. Increasing cognizance and articulation of the influence of our potentially varying goals for antenatal diagnosis of CHD and various views of treatment for CHD on the experience of those we serve is necessary and timely.

P689

Fetal endocardial hyperechogenicity: a possible prenatal echocardiographic marker of maternal toxoplasmosis

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Introduction – Toxoplasmosis is a systemic infectious disease caused by Toxoplasma gondii. Acute infection during pregnancy determines well established fetal sequelae which occur mainly in central nervous system, heart and eyes. Routine prenatal echocardiographic observation of patients with acute toxoplasmosis raised the suspicion that fetal endocardial hyperechogenic foci could be a prevalent finding. Objective – To confirm the hypothesis that focal or diffuse endocardial hiperechogenicity occurs more frequently in fetuses of mothers with acute toxoplasmosis than in normal fetuses without risk factors for congenital heart disease. Design of the study: case control study with prevalent cases. Methods – Sixty consecutive fetuses cases from mothers with acute gestational toxoplasmose, detected by high IgM titles were examined by means of fetal echocardiography and compared to 353 normal consecutive fetuses from a low risk populational screening program (controls), in order to look for endocardial echogenic foci. Results – Among the cases, 57 (95%) presented focal or diffuse endocardial hiperechogenicity, while only 16 (5%) of the control fetuses showed endocardial foci. Perinatal follow up (mean=4,6 months) in 15 cases showed complete disappearance of the findings in 53%. Conclusion – Fetal endocardial hiperechogenicity (focal or diffuse) occurs more frequently in maternal toxoplasmosis than in normal pregnancies. This alteration, might represent an (inespecific) echocardiographic marker of endocardial inflammatory process, with a trend towards recovery in the first months of postnatal life.

P690

Development of Z-scores for cardiac dimensions from fetal echocardiography

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Background: Normalisation of cardiac dimensions to body size, using so-called Z-scores, is well-established in perinatal life. Such Z-scores have yet to be developed for fetal cardiac dimensions. The aim of this study was to produce formulae and nomograms allowing Z-scores to be calculated from fetal femur length (FL) and biparietal diameter (BPD), relative to cardiac dimension measured by echocardiography. Methods: Seventeen cardiac dimensions were measured using 2D echocardiography in 103 pregnancies with gestational ages ranging from 15–39 weeks. Regression equations were derived relating all 17 cardiac dimensions to FL and BPD. Z score formulae and nomograms were developed. Results: Relations between cardiac dimensions and FL or BPD were best described following natural log transformation: ln(cardiac dimension)= m.ln(FL or BPD) + c. For example, for ascending aorta diameter, the strongest relationship was with FL (R^2=0.92 for the natural log-natural log model). For calculation of the Z-score for ascending aorta diameter relative to FL, the following formulae were derived. Z-score = (ln(actual) – ln(predicted)) /0.13 where ln(predicted)= 0.84ln(FL) – 2.07. The scatterplot and Z-score nomogram for this example are shown on the attached graphs. Conclusion: This study now allows computation of Z-scores in fetal life for 17 cardiac dimensions from either FL or BPD. Whereas previous studies of normal data have allowed qualitative assessment of where abnormal cardiac dimensions lie with regard to the normal range, this study allows quantitative analysis (via Z-scores) of where such dimensions lie relative to the mean. This will permit mathematical assessment of serial growth of fetal cardiac structures in normal, and congenitally abnormal, hearts.

P691

Foramen ovale flow in fetuses with heart defects

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Size and flow across the Fo was assessed in 51 fetuses with CHD: 4 – PA&IVS, 4 – AT; 5 – Ebstein anomaly, 4 – TOF, 2 – TOF&absent PV, 10 – HLHS; 8 – CoA, 6 – DORV. In all fetuses with PA&IVS, AT and TOF Fo was big (Fo/IAS ratio = 0,48) with redundant septum primum. Those neonates needed Rashkind procedure after delivery. In all fetuses with HLHS Fo was restrictive (Fo/IAS ratio = 0.16) with left to right shunt. In 8 fetuses with CoA and 5 with DORV and hypoplastic LV Fo was small with right to left flow of increased velocity. In contrary to the previous papers we did not detect left to right or bi-directional flow across Fo in fetuses with CoA. We concluded that: 1. Redundant septum primum in fetuses might suggest restriction of the Fo; in the opposite 2. It is likely that small size of Fo is one of factors which determines development of the left heart. 3. Careful examination of the Fo flow and size in fetuses with CHD might be helpful in perinatal management of these infants.

P692

Genetic problems in perinatal cardiology

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We analyzed perinatal results and outcome of 170 fetuses with structural heart defects. Karyotype was checked in 72%. There was trisomy 18 in 15, trisomy 21 in 6, trisomy 13 in 3, other in 3. The most common defect among those fetuses was AVSD (9) and inlet VSD (8). There was one rare case of trisomy 13 in the fetus with HLHS. Atrio-ventricular valves in fetuses with Edwards syndrome had thick leaflets and looked quite different from normal AV valves. In 7 hypoechoic fetuses with simple heart defects [mild VSD, PS, suspected ASD] karyotype was normal but in neonates rare genetic syndromes were diagnosed based on clinical appearance and molecular tests. In 20 fetuses with conotruncal defects, inlet VSD, CoA FISH test for microdeletion of chromosome 22 was performed, which was positive in 1. Basing on the result of fetal echocardiography in combination with karyotype all families had careful genetic and cardiac counselling concerning the further course of pregnancy and fetal outcome. Just 4 pregnancies were terminated. In fetal chromosomal abnormalities, if parents did not decide to terminate pregnancy, perinatal care was organized in the best place for the patients. One neonate with Edwards syndrome was referred for the palliative home care. We concluded that: 1. One third of fetuses with CHD had chromosomal or genetic abnormalities. 2. Rate of pregnancy termination was low due to late diagnosis and ethical background of the families. 3. Thick dysplastic AV leaflets in hypoplastic fetuses with AVSD could indicate chromosomal abnormality. 4. Fetal hypertrophy with mild cardiac defects and normal karyotype could be a sign of other genetical syndrome.

P693

Fetal cardiac dysrhythmia

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In additional 2% of pregnancies exhibit a fetal cardiac dysrhythmia. We performed fetal echocardiography for evaluation of cardiac rhythm. After 2-dimensional echocardiography study of cardiac structure was performed M-mode and Doppler echocardiograms were analyzed for measurement of cardiac rate. Arrhythmias were diagnosed in 13 patients. We analyzed age at diagnosis, hemodynamic disturbances, and outcome. Mean gestation age was 33.2 weeks (23–38). Two patients with isolated ectopic beats which resolved during pregnancy, 3 patients had frequent sinus pauses, 2 patients had mild

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

sinus bradycardia and 5 with supraventricular tachycardia (SVT) with mean heart rate of 224 b/m (180–280). One of 5 patients had hydrops in which parents chose termination of pregnancy. 2 of 5 patients had dilated cardiomyopathy and 2 of 5 patients had normal heart. We used fetal SVT protocol. Three of 5 patients prenatally control was achieved cardioversion and haemodynamic improvement. One of 5 patients no change was noted and fetal death occurred. Supraventricular tachycardia may represent a serious problems for the fetus, but conservative approach must be advocated.

P694

Intrauterine growth retardation and its relationship to fetal cardiac and peripheral velocity waveforms

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Maximum flow velocity waveforms were studied at the cardiac level (ascending aorta, pulmonary artery and ductus arteriosus) and at the peripheral level (fetal internal carotid artery, descending aorta, umbilical artery, and maternal uteroplacental artery) in 12 patients with intrauterine growth retardation and 12 normal control subjects matched for gestational age and maternal parity. Gestational age ranged from 26 to 36 weeks (median, 30 weeks). All flow velocity waveforms were obtained with a sector scanner combined with a pulsed and continuous Doppler system with a carrier frequency of 2.5 and 3.0 MHz. Normal pregnancy was characterized by low fetal and placental vascular resistance's. The peak systolic velocity in the ascending aorta was significantly higher compared with the pulmonary artery. In patients with intrauterine growth retardation, reduced end-diastolic flow velocities were documented in fetal descending aorta, umbilical artery, and maternal uteroplacental artery, reflecting raised umbilical placental and uteroplacental vascular resistance's. Raised end-diastolic flow velocities were observed at the cerebral level, reflecting reduced cerebral vascular resistance ("brain-sparing" effect). Reduced peak systolic flow velocities demonstrated at the cardiac level may be secondary to reduced volume flow, increased valve or vessel size, or mixed shunting. The retrospective nature of the study did not allow differentiation between these variables.

P695

Fetal heart failure: causes and outcome

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Fetal heart failure (FHF) is being recognized more often since cardiovascular ultrasonography became a generalised technic. Although hydrops letalis from cardiac etiology is not always obvious, careful valvular and myocardial dysfunction detection with conventional 2D echo-Doppler (e-D) evaluation may give further information to access a better diagnosis and treatment. To identify cardiac causes of FHF, its management and outcome, e-D was performed in 2027 pregnant women referred to our centre. Isolated abnormal Doppler filling of vessels and ventricles was excluded. We identified 113 heart defects (HD) and myocardium (Myo) abnormalities and 75 rhythm or conduction (R/C) problems. Cardiac cavities enlargement and circumferencial pericardial effusion was present in 27 fetus at first evaluation. Mild to severe anasarca developped in 22/27 (81%) during follow-up. Isolated R/C (7/27) and Myo (10/27) were the major causes for FHF. Mild to severe tricuspid valve regurgitation (TRI) were present in 25/27, mitral (MR) in 4/27 and in 2/27 there was pulmonary absent valve syndrome (PRI). Cutaneo-Mucosa (C) HD also appeared associated to FHF in 7/27 - with 1 fallot co-existing with 1 third degree atrioventricular block (AVB). One severe MR from unguarded orifice, 1 nonfibrein TR, 1 isolated pulmonary valve dysgenesis and finally an Galeno arterio-venous fistula were also find. Therapy with digoxin was prescribed to mother in 5 dilated myocardiophaty and in 4 supraventricular tachycardia (SVT) complemented with Verapamil in one - endocardiectsia. Adenosine administration in a 22 gestacional age fetus with severe anasarca was also used (but the fetus died one week later). There was 6 in-utero deaths related to Myo, 1 to SVT and 3 to CHD. Congenital MR caused death immediately after surgical delivery and a Fallot/AVB, died at 2 days of life, despite energic measures including temporary pacemaker. All the others had programmed successfully delivery and postnatal management. In conclusion, although a high fetal mortality is still present in FHF we should perhaps, be more interventionist in some situations.

P696

Ductus arteriosus and foramen ovale restriction – diagnosis and etiologic aspects

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INTRODUCTION: There are few reports having studied functional abnormalities of the fetal heart such restrictive foramen ovale and restrictive ductus arteriosus. The purpose of this study was to describe the echocardiographic features in 21 fetuses without congenital heart disease and to correlate those findings with possible prenatal etiology. METHODS: Between 1983 to 2000, we reviewed our experience with 5400 high risk pregnancies referred for fetal echocardiography. In 21 fetuses we identified features of right-sided heart failure due to constriction of the ductus arteriosus (N=17) or restriction of the foramen ovale (N=4). Fetal and neonatal echocardiography excluded a structural heart defect specially coarctation of the aorta. We carefully investigated maternal ingestion of drugs which could explain those findings. RESULTS: Constriction of the ductus arteriosus was clearly identified in 14 fetuses and restrictive foramen ovale in 4. There were a further 3 fetuses in whom this diagnosis was retrospectively suspected because it was the only explanation for the right-sided heart failure in 1 case and for the 2 other cases in which prenatal echocardiograms had been posted as pulmonary atresia. The possible causes to explain the constriction of the ductus arteriosus were: maternal use of sympathomimetic drugs for nasal decongestion (n=2), anti-inflammatory agents (n=8), phenobarbital (n=1) and aspirin (n=2). In 4 cases we could not find any explanation (spontaneous?). Restriction of the foramen ovale could be associated with drug abuse (crack, n=4) and abortion attempt with prostaglandin E2 (n=1). In the remaining two cases we could not find any risk factors. Follow-up showed normalization of the heart between one week to four months. Only one fetus died. CONCLUSIONS: Although a combination of echocardiographic features can correctly identify those functional abnormalities of the fetal heart, further multicenter studies are necessary to establish the real correlation and risk caused by those drugs.

P697

Echocardiographic features of hydropic and non-hydropic fetuses with pleural effusions

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Fetal pleural effusions may lead to hydrops and are associated with mortality as high as 50%. A close monitoring of the fetus is essential to guide fetal therapy. To define the pathophysiology of fetal pleural effusions and their relation to hydrops, we reviewed 58 echocardiograms from 33 fetuses diagnosed with pleural effusions. Measurements included diameters of the RV, LV, inferior vena cava, aortic and pulmonary valve. Doppler velocities were measured above the aortic and pulmonary valves. From the standard 4-chamber view, the ratio of the effusion area over thorax area was calculated. Variables were converted into z values from regression equations based on normal data. Features of fetuses with and without hydrops were contrasted. Hydropic fetuses had higher effusion ratios than non-hydropic fetuses. Compared to normals, study subjects had lower dimensions of RV, LV and atrio-ventricular valves. In contrast, the inferior vena cava was dilated, and this finding was more pronounced in the hydropic group. Conclusion: Fetuses with pleural effusions have abnormal echocardiographic findings that can correlate with the presence or absence of hydrops. Prospective longitudinal data are needed to determine the implications of echocardiograms in the management of fetal pleural effusions.

P698

Diagnosis and treatment of fetal SVT

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To investigate the effect of transplacental therapy. ATL Ultra 9 Doppler Colour Ultrasound and FECG-D were used. Of 680 high risk fetuses (20–42 weeks), 23 cases (3.75%) were found to have supraventricular tachycardia (SVT). Digoxin was administrated intravenously to pregnant women at the initial dose of 0.8mg to 1.0mg. After three to four hours, 0.25mg was administrated intravenously or orally. If the effect is not satisfied, verapamil was used in combination. Eighteen pure SVT cases recovered with digoxin transplacental therapy. Three cases complicated with congestive Heart failure recored in conjunction with verapamil treatment. One of them complicated with CHD

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

clinical even by the combined treatment. Two cases were not given drug and immediately after birth. The conclusion is that fetal ultrasound examination is an essential method to diagnose fetal cardiac abnormality. Transplacental digoxin is therapy of choice for fetal SVT.

General Pediatric Cardiology, Prognosis/Natural History

P699

Aortic stiffness and blood-pressure difference between upper arm and thigh after coarctation repair

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There are no normal values for the stiffness of the aorta descendens (Aod) or the blood-pressure (B.P.) difference (diff) between the upper arm and thigh. Nevertheless the B.P. diff is often used to estimate the severity of a re-coarctation. We measured 4-times the B.P. at the right upper arm (UA) and thigh (T) in 50 patients (bodyweight=52.4 (17–17,8) kg) without significant re-coarctation (MRI and/or echocardiography) and 50 healthy controls (bodyweight=42,2 (+/-15,1) kg). Discarding the first B.P., we calculated the mean of the other 3 values, the measured the diameter of the aorta ascendens (Aasc) and the Aod 10-times. We calculated the stiffness-index b from the mean of the diameters and the B.P. We compared both groups using a t-test (p<0.05 significant; standard-deviation, (+/-std)). b-Aod b Aod Diff sys Diff dias Diff.MAD Coa 2.7 (0.8) 3.7 (1) 3.2 (0.1) 9.3 (8.2) 5.5 (9.1). CoA 4.1 (2.2) 3.7 (1.1) -2,3 (12,5) 4,6 (10,7) 4,7 (12,3). p=0,05 0.86 <0,05 <0,03 0.53. The stiffness of the Aod was lower than of the Aod (Windkesse) in controls independent of age. After repair of coarctation the stiffness of the Aod increased with age and was higher than of the Aod. The stiffness of the Aod showed in both groups a similar age dependence. The B.P.-diff showed a considerable scatter in both groups making it an unreliable tool to estimate the severity of a re-coarctation. The decreased function of the Windkessel in patients

P700

Cardiac arrhythmias in children with Marfan's syndrome

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Cardiovascular problems are crucial in the long term prognosis of Marfan's syndrome (MFS). Cardiac deterioration results mainly from aortic crisis, aneurysm formation or valve insufficiency. To date only a few reports focused on the problem of cardiac arrhythmias (ar) in patients (pts) with MFS. Method: We compared the frequency of ar in 24 h Holter recordings of 24 pts (10m, 14f, mean age 15.4 yr.) with a control group of 44 healthy volunteers (29m, 15f, mean age 15.9 yr.). Results: Ar were detected in 58% of the pts vs 15.9% of the control group. Holter recordings detected premature atrial beats in 29% premature ventricular beats in 8% and combined premature beats in 25% of the pts. Atrial tachycardia was present in 2;8% and ventricular tachycardia in 1 pt. In the control group premature atrial beats were found only in 4.6% and premature ventricular beats in 11.4%. Tachycardias were not detected. The hazard ratio for ar for MFS pts compared with the controls to develop ar was 3.6 for all kinds of rhythm abnormalities, 11.0 for atrial arrhythmias (both p=0.05) and 2.4 for ventricular arrhythmias (n.s.). Pts with mitral valve prolaps had a hr of 2.60 and those with mitral insufficiency had a hr of 2.07 for arrhythmias compared with the normal group (p=0.06). Conclusion: More than half of the pts with MFS show cardiac arrhythmias, mostly atrial premature beats. Ventricular dysrhythmias are rare in MFS. Pts with natural pathology have a markedly high risk of ca. We recommend that repeated Holter recordings should be obtained in the follow-up of pts with MFS. Further studies are needed to determine the clinical importance of the presented data.

P701

The natural history of ventricular septal defects in children

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This study was undertaken to identify predictors of outcome and quantitate the probability of spontaneous closure in children with isolated ventricular septal defects. We studied 252 children (129 male, 123 female) aged one day, 194 months (median 12 months) who had a diagnosis of an isolated VSD between May 1992 and May 1998 in Ankara University, Paediatric Cardiology Department. Patients were followed 1 to 8 years. Two-dimensional echocardiography in multiple views identified the site of the ventricular septal defect in all patients. Of these 252 patients, 129 were classified as perimembranous, 85 as muscular, 28 as inlet and 10 as outlet types. Cardiac catheterization was performed in 79 (31.3%) patients. The defects closed spontaneously in 71 (28.2%) cases most of them in the first 2 years of their life. Muscular defects were more likely to close spontaneously than other types. However, spontaneous closure rate of apical muscular defects were relatively infrequent. Aortic regurgitation developed in 20 patients with VSD and majority of them had outlet VSD. Ventricular septal aneurysm formation was present in 26 patients with perimembranous VSD. It was confirmed that a substantial proportion of patients with ventricular septal aneurysm formation is associated with left ventricular-to-right atrial shunt (7 of 26). The development of subaortic ridge was demonstrated in 9 patients with perimembranous VSD. Seven patients were complicated with infective endocarditis and 6 of them were undergone to surgical closure. In conclusion, VSDs are generally well tolerated and muscular defects were more likely to close spontaneously than membranous defects. Patients with VSD should be followed carefully for the development of secondary morphologic abnormalities such as aortic insufficiency, subaortic ridge, ventricular septal aneurysm and left ventricular to right atrial shunt.

P702

Screening for congenital heart disease (chd) in low-risk new-borns

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Routine echocardiography in high-risk groups of neonates is now widely practised. Screening all neonates may lead to earlier diagnosis, better treatment and higher levels of parent satisfaction. Aims: To assess the effectiveness and cost of an echocardiographic screening programme for CHD. Subjects and Methods: From 01/11/94–28/02/95 there were 2688 deliveries in The Royal Maternity Hospital Belfast. After randomisation and exclusion of high-risk infants 2665 new-borns underwent echocardiographic assessment, while 440 received the usual level of clinical assessment. Cases of CHD detected before hospital discharge were documented 'screened' and control infants diagnosed with significant CHD during 1995 were compared in terms of cost of management. The annual cost of screening was estimated and the rate of accurate diagnosis recorded. Results: 91 infants were identified with significant CHD in the screened group before discharge compared with 27 controls. There were 19 additional late diagnoses in controls and none in screened infants. During 1995 14 cases were 'picked-up' (before discharge) in screened infants and 5 in controls. The cost of subsequent management was £3,399/patient in the screened group and £7478/patient in controls. The mean time to complete diagnosis in the sub group was 2 days for studied cases compared to 11.0 in controls. The annual cost of screening for 1 infants was estimated as £37/child for the first year. Avoidance of referrals with innocent murmurs would cost at least £20/child in year. Sampling of parental support for screening showed almost universal approval. Conclusions: The addition of echocardiography to neonatal clinical examination greatly enhances detection of cases of CHD at a very early stage. Although screening is expensive, parents support it and once established, costs fall and benefits are long lasting. Screening should also reduce the cost of unnecessary outpatient referrals.

P703

62-year-old woman with unoperated single ventricle

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Background: Due to a poor spontaneous prognosis, nowadays most patients with univentricular (UV) physiology are considered for cavopulmonary connection early in life. But how to deal with patients in quite a good condition presenting for the first time in older life? Case Report: A 62-year-old woman was presented with double inlet left ventricle, L-transposition (rudimental right ventricular outflow chamber) and pulmonary artery stenosis. She was mildly cyanotic throughout life, but only a slightly reduced exercise capacity for decades and gave birth to two children. Now she was presented for increasing heart failure and atrial fibrillation. Echo, MRI and angiography confirmed the diagnosis. Catheterization showed normal systemic flow, normal pulmonary resistance and a Qp/Qs of 2.9. After optimizing medical treatment she was discharged. Conclusion: A small number of patients with UV-physiology can survive into the 5th decade without surgical treatment. If circulation is well

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

balanced, functional capacity and quality of life can be good, even in the long-term. Before the decision for palliative surgery (including all types of cavo-pulmonary or systemico pulmonary shunts) can be made, outcome and complications of these operations have to be considered carefully. The selected unoperated adult with CV-circulation maybe more likely to profit from conservative management and close follow up rather than from surgery.

P704

An 'epidemic' of anomalous origin of left coronary artery from the pulmonary artery at the red cross children's hospital, Cape Town, South Africa

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We have recently encountered a surprising number of patients diagnosed as having anomalous connection of the left coronary on the pulmonary artery (ALCAPA), a rare congenital heart defect. This lesion is of special importance as it represents a fully treatable cause of left ventricular dysfunction. From January 1 1999 to 30 November 2000, we have seen 11 patients with ALCAPA as compared to 3 patients from 1985 to the end of 1997. The mean age at presentation to our institution was 4.6 months. The mean age at diagnosis of the coronary abnormality was 3 years 3 months. Most children presented with a history of dyspnoea, coughing or living with feeding. The average length of time taken to make the diagnosis was 3 years and 3 months. All patients had abnormal ECG's. The most common finding was ST segment change rather than q waves in I and avL. One patient was diagnosed at autopsy. The rest all had confirmation of the abnormality at angiography. 6 patients have been repaired using the Takeuchi technique (baffle in the pulmonary artery), 1 patient has had ligation of the anomalous artery and one had had coronary reimplantation. 1 patient is awaiting surgery and 1 has defaulted. Improvement in LV function and clinical status has occurred in all patients undergoing surgery. This apparent increase in the incidence of ALCAPA may be nothing more than a statistical

P705

Prevalence of organised thrombosis in peripheral pulmonary arteries from patients with decreased pulmonary flow and "single ventricle" physiology: possible implications on the indication of Fontan-type procedures

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Low pulmonary arterial pressure is a basic requirement for candidates to total cavopulmonary connections. In the presence of decreased pulmonary flow, however, this is not a usual concern, since the resistance is low. However, the pattern of blood flow, as well as the increased hematocrit in such patients, brings the possibility of in situ thrombosis. We investigated the rate of organized thrombi in peripheral pulmonary arteries of possible candidates to total cavopulmonary connections. Methods: From the files of the Pathology department, we found twelve necropsy cases of patients older than 2 years with univentricular atrioventricular connections and decreased pulmonary flow (mean age = 152.8, median = 126 months). In one available microscopical slides of lung, we evaluated the number of arteries presenting eccentric intimal fibrosis and rounded-like lesions, both interpreted as organized thrombi, and the relative luminal area occupied by them. Results: Eight cases (66.7%) showed peripheral pulmonary arteries with organized thrombi. Among them, the percentage of compromised arteries varied from 9.5 to 38.9% (mean = 22.7%). The mean area occupied by thrombi relative to the total luminal area varied from 0.25 to 9.59. An occlusion index" was determined, representing the whole arterial tree (% of arteries with thrombi X mean occluded area) and varied from 4.2 to 11.4%. There was no association between age and presence of thrombi. The hematocrit values could be retrieved from the clinical files in 10 cases, and varied from 39 to 77.4% (mean values 54.6% and 47.7% among cases with and without thrombosis, respectively). Conclusion: Although the prevalence of organised thrombosis in peripheral pulmonary arteries was high among these patients, the percentage of compromised vessels was low. However, it could potentially impair the early and long-term results of the Fontan-type procedures.

P706

Cardiovascular findings and clinical course in williams syndrome (WS)

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We investigated the incidence of cardiac malformations in a series of 45 patients with (WS). The mean age and the follow up period were 3.5 ± 3.7 and 5.5 ± 5.5 years respectively. Twenty seven were male. Two girls were identical twins. Thirty eight patients (84 %) had cardiovascular anomalies, often combined. The most frequent malformations were supravalvar aortic stenosis (SVAS) in 25 (66%), 4 (11%) diffuse, myxomatous mitral valve (MV) in 12 (32%), 8 with mitral regurgitation (MR) (moderate or severe in 2); and pulmonary artery stenosis (PAS) in 11 (29%). Less frequent anomalies were pulmonary valve stenosis (PS) in 4 (11%), small ventricular septal defect (VSD) 2 (5%); coarctation of the aorta (CAo) 2 (5%) (one abdominal); patent ductus arteriosus, hypertrophic obstructive cardiomyopathy (HOCM), and fibromuscular subaortic stenosis (FSS) in one patient each. One child had Wolff-Parkinson-White pattern. Surgery or catheter intervention was performed in 42% of these cases. SVAS repair was carried out in 13 (one late reoperation), MV repair or replacement in 4, 2 surgical and one balloon valvuloplasty for PS, one balloon angioplasty for PAS, balloon angioplasty for CAo, by-pass and balloon dilation of abdominal CAo, and FSS resection were each. There were 3 deaths, 2 early after surgery and one late sudden death. All 3 patients had severe SVAS associated with moderate or severe MR. The rest of the patients are alive and well. In conclusion, the most frequent cardiovascular anomaly in WS was SVAS, and, differing from other series, was followed by myxomatous MV, and closely by PAS. Among the others, there was 1 case of HOCM, an association therefore not reported. WS patients at greater risk were those with left ventricular pressure and volume overload as seen in associated SVAS and MR.

P707

Aortic root size in childhood

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The most appropriate measure aortic root dimensions during childhood growth, and the best method of assessment of normality remains controversial. Furthermore, published data from authoritative sources for normal values of aortic root growth appear to differ in comparison with our observations. Patients with a variety of conditions including Marfan and Turner syndromes, as well as many other patients having some features of connective tissue disorders, although insufficient for a complete syndromal diagnosis, require accurate measurement for diagnostic and management purposes. Due to the paucity of adequate established reference data for our population, we have compiled reference data from 150 normal patients, aged 3 months to 18 years. This data has been collected from normal patients attending cardiology or oncology clinics. Cardiac assessment and 2 dimensional echocardiographic measurements have been performed by the same cardiologist, with careful attention to a standardized measurement technique. Results: Aortic root size measured over our paediatric population compares closely to a smaller series from a similar population, but varies considerably from commonly used reference data. Aortic root size correlates well with both height and body surface area. Confirmation of a previous observation that the annulus/sinus ratio is reasonably constant in relation to height. Conclusions: Sufficient data is now available to establish reliable nomograms of aortic root size in childhood in our population. Aortic root size and annulus to sinus ratio versus height provide readily available parameters for instantaneous and longitudinal assessment without need for BSA calculation.

P708

Clinical course of subpulmonic ventricular septal defect

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To assess the clinical course of subpulmonic ventricular septal defect(VSD), 1543 children of isolated VSD diagnosed during January 1995 to November 2000 were studied. The types of VSD were differentiated into perimembranous, subpulmonic, inlet, muscular and multiple VSD. The prevalence was 74.1%, 17%, 3.2%, 3.9% and 1.8% respectively. 263 cases of subpulmonic VSD, 131 male, 132 female, aged range from 30 days to 20 years(mean 4.5 years)were reviewed retrospectively. The initial echocardiogram showed aortic valve deformity in 92 cases and aortic regurgitation in 30 cases. The earliest age of the patient that having aortic valve prolapse was only 1 month old baby and AR was found at 2 months old in the same baby. At the mean follow up period of 1.4 years, 32 more cases of aortic valve deformity and AR developed. The overall prevalence of aortic valve deformity and AR were 39.5% and 23.6%. The mean age of the patient with aortic deformity and AR were 5.8 and 6.8 years respectively. 30 cases with large VSD and heart failure had surgical closure, with one dead. 3 cases developed Eisenmenger complex at

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

the initial presentation 2 cases died from heart failure and subsequent pneumonia before surgery. No one in our series had spontaneous closure of the VSD. Surgical closure and aortic valve repaired had been performed in 8 cases and Ross's operation had been done in 3cases. All had a good result. Conclusion: The prevalence of subpulmonic VSD and aortic valve lesion is high among Thai children. Closed follow up with echocardiogram and early surgery are necessary.

P709

Aorto-left ventricular tunnel: diagnosis, management and follow-up

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Aorto-left ventricular tunnel (AOLVT) is an extremely rare congenital cardiac defect. Our aim was to present the experiences with treatment and information on late results of surgery especially with reoperations for progressive aortic insufficiency (AI), or recurrence of AOLVT. In our Institution 5 pts with AOLVT were seen between 1983-1999 (age at diagnosis: 1 day - 6 yrs, mean 2.4 years). Three pts presented at neonates in high output failure due to severe AI. Echocardiography was diagnostic. We lost one newborn at the age of 3 days without surgery because of severe neurological complications, but the other two pts had successful emergency surgery. Follow-up period was 2.5 and 8 years. Both are in good condition with mild AI. The other 2 pts were referred because of heart murmur at their age of 4 and 6 yrs in 1985. Both needed reoperations after 10 and 11 years because of progressive AI due to the aortic root dilatation in one and the reopened orifice of AOLVT in the other. At reoperation aortic valve replacement was performed using a 25mm CarboMedics valve and in the other case the reopened orifice of AOLVT was successfully closed, monitoring by TEE. Both patients are symptomless after reoperation. Conclusions: AOLVT presents in two forms: as critically ill early infant age and in childhood with severe AI. 2 Echocardiography is the most important means of diagnosis and in the follow-up for AOLVT. 3 Early corrective surgery in AOLVT is the treatment of choice. 4. All pts should be re-examined regularly, because approximately 20% can be expected to require reoperations in the late follow-up.

P710

Management and outcome of heterotaxy syndrome with univentricular morphology

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The aim of this study is to clarify the outcome of the current management of heterotaxy syndrome with univentricular morphology, and analyze causes of death. Fifty-five consecutive patients with heterotaxy syndrome and univentricular morphology were included in this study (1988 - 1998, median follow up 3.2years). Other associated cardiac anomalies were pulmonic stenosis or atresia in 46 (83.5%), major aorto-pulmonary collateral arteries in 4(7.3%), extracardiac total anomalous pulmonary venous return (EX-TAPVR) in 17(30.9%). Nine of 17 with EX-TAPVR had severe pulmonary venous obstruction. Aortic arch anomalies were also found in 5(9.1%) including two cases of hypoplastic left heart syndrome. Thirty-nine(71%) patients underwent surgical intervention. Eleven(20%) patients had Fontan operation with two perioperative death. Three of them had late conversion to extracardiac conduit type TCPC, and two had take-down to bidirectional Glenn procedure with one death in each group. Two patients with hypoplastic left heart syndrome underwent Norwood operation were waiting for future Fontan. There were 28 deaths in total, and death occurred prior to Fontan in 24 of them. Causes of death prior to Fontan were as follows: 1) pulmonary venous obstruction in 7(29.3%), 2)ventricular failure in 7(25%), 3) sudden death with probable diagnosis of severe infection in 4(14.3%), and 4)other causes in two. Three of seven deaths due to ventricular failure were not associated with more than moderate atrioventricular valve regurgitation. Overall actuarial survival rate was 72.3% at 1 year, 68.6% at 5 years. Survival rate was significantly lower in patients with EX-TAPVR, comparing to patients without EX-TAPVR (35.3% vs 89.2 at 1 year and 22.1% vs 60.8% at 5 years). In conclusions, extracardiac total anomalous pulmonary venous return is still a complicating factor for long term survival. Meticulous management of ventricular failure regardless of atrioventricular valve regurgitation and prevention of severe infection were mandatory in following-up patients with heterotaxy syndrome and univentricular morphology.

P711

The level of parental knowledge, daily stress and adjustment in congenital heart disease in Korea.

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To investigate the correlation between the stress, coping and the level of knowledge of parents in congenital heart disease(CHD), 130 patents were recruited from Samsung Medical Center, Seoul, Korea between the period of June 2000 to August 2000. The level of stress, coping and the knowledge of the parents were access using questionnaire and the tools developed or modified by the authors, and the data was analyzed using the SAS program. The observations made from the study were as follows. 1. The average score of stress was 2.29 out of 5 points (=45.7%). The level of parental stress experienced by the subjects was shown to be moderate, and it was lower than that with the other obstacles such as cerebral palsy, cleft lip and cleft palate reported in Korea. 2. The average score for knowledge was 13 out of 20 points (=65.3%). Although parents are quite well aware of the immediate necessities for the diagnosis and treatment of heart disease for their children, they know little regarding the areas of lifestyle and long-term prognosis after treatment. 3. The average index of coping was 3.02 out of 4 points (=75.5%). The factors which affected coping were age, the condition of the treatment, the subject's age, and the state of perceived knowledge of the subjects: those who knew the name of the diagnosis, those who replied they know about congenital heart disease, and those who replied they have heard of endocarditis. 4. The correlation between the subjects' stress and coping, (r= -.27) and coping and knowledge (r=.34) was statistically significant. However, there was no significant correlation statistically between stress and knowledge. This study has shown that parental knowledge is an important aspect in coping of the patients. Moreover the patients' age demonstrated a significant variable on the level of stress, coping and knowledge. Further studies regarding guidance towards patients', which is the primary factor of stress are encouraged, in addition to finding emotional stability.

P712

Aneurysm of the ventricular membranous septum: Serial echocardiographic studies

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The natural incidence and natural history of aneurysm of the ventricular membranous septum(AVMS) has not been documented. We reviewed the records of 139 consecutive patients(pts) with a perimembranous ventricular septal defect(VSD), in whom the first echocardiographic studies(ECHO) were performed at age less than 3 months. Pts were divided into 3 groups. Group I consisted of 75 pts who had spontaneous closure. Group II consisted of 43 pts who still have a VSD at a mean age of 85 months. Group III consisted of 41 pts who underwent surgical closure. Pts were separated into 2 groups: 45 pts with congestive heart failure(CHF) and 74 without CHF. Color Doppler was performed to certify spontaneous closure. VSD cross-sectional area measured at the time of surgical closure was indexed to body surface area: VSD area index (VSDAI). In 70 of 139 pts(44%), AVMS was found (31% in group I, 60% group II, 59% in group III). Median age at AVMS formation was 5 months [range 1 day to 72 months]. No difference in age at AVMS formation could be found between 3 groups. As the first ECHO, 21 pts(13%) had an AVMS, 5 pts(7%) with CHF and 16(20%) without CHF. In 6 pts with an AVMS who underwent surgical closure, average age at closure was 39 months (range 9 to 99 months) and average VSDAI was 0.54 (range 0.16 to 0.94 cm2/m2). AverageVSDAI in 8 pts who underwent surgical closure at age less than 6 months was 3.6 (range 1.5 to 6.0 cm2/m2). We conclude that the patients of an AVMS is more favorable in the natural history of perimembranous VSD.

P713

A clinical pathway in open heart surgery of simple congenital heart disease in Korea

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To standardize the hospital management of simple congenital heart disease(CHD), we developed the 'clinical pathway (CP)' for the patients and analyzed the influences of the CP on quality improvement of outcomes in 60 simple CHD pts who had undergone an operation between June 1, 1998 and

October 31, 1998. The control group included 48 pts who had an operation for the same disease between the corresponding period of 1997. Method: Two types of CP were prepared according to the state and place of residence of patients. The pts were managed under the CP protocols in all processes of the subjects' education, examination, hospital stay, care, discharge from the hospital. In each process, the already determined evaluation items were assessed. After discharge, the performance of all the processes, hospitalization period, medical cost, treatment effects, complications and degree of satisfaction were assessed. Result: The hospitalization period prior to the operation (2.2 days vs. 4.1 days), the I.C.U stay (1.5 days vs. 2.7 days), ward stay after operation (4.5 days vs. 5.9 days) and total hospital stay (8.0 days vs. 12.5 days) were significantly shortened in the group to which the CP was applied, compared to that of the control group. The extubation time, the frequency of Lab during the hospitalization decreased in the CP group compared with the control group. Medical cost was also significantly lower in the control group. There were no cases of death, readmission or notable complication. A survey on the patients' guardians showed that they were satisfied with more than 85 percent of the questionnaire items. Accordingly, in the case of the simple CHD, the CP could reduce the hospital stay and medical cost as well as contribute to enhance the patients' satisfaction.

P714

Metabolic stress testing after repair of tetralogy of Fallot

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Forty-four patients status post complete repair of tetralogy of Fallot underwent transthoracic echocardiography and metabolic stress testing. Patients were ages 5 to 34 years and underwent repair in different surgical eras. Patients were divided into three groups based on age alone. Group 1 consisted of 20 patients < 18 years old, Group 2, 15 patients age 19 to 30 years and Group 3, 13 patients age > 30 years. There were no statistical differences between the groups when comparing right ventricular size, estimated right ventricular function, left ventricular function, or percent fractional shortening. The mean peak VO_2 (measured in ml/kg/min) in Group 1 patients was 37.1, in Group 2 was 27.5 and in Group 3 was 23.3. This data suggest that peak VO_2 measured during maximal stress testing in patients after repair of tetralogy of Fallot decreases as patients age. This decrease in peak VO_2 that occurs with age is consistent with findings in the normal population. Measured peak VO_2 after repair of tetralogy of Fallot is lower than reported age matched controls. The measured decrease in exercise ability in our patients is not related independent of ventricular function or type of surgical repair. This data will be helpful in evaluating patients who are being considered for right ventricular outflow tract reconstruction.

P715

Is prolonged use of prophylactic benzathine penicillin responsible for the development of ampicillin resistant streptococcal strains in oral mucosa?

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The most effective method for secondary prevention of acute rheumatic fever is intramuscular benzathine penicillin G. However, prolonged use of prophylactic penicillin may result emergence of penicillin resistant strains of streptococcus viridans in oral mucosa. The drugs used in infective endocarditis prophylaxis of patients with rheumatic heart disease (RHD) for dental, oral and upper respiratory tract procedures are oral amoxicillin or parenteral ampicillin. If there were penicillin resistant strains of streptococcus viridans in oral flora, amoxicillin and ampicillin would be ineffective for infective endocarditis (IE) prophylaxis. Thirty patients with RHD (15 female, 15 male) aged between 9 to 19 years (12.8±2.6 years) who have been received benzathine penicillin prophylaxis for 4 to 108 months (36.5±32.4 months) were enrolled the study. In vitro susceptibility of streptococcus viridans which were isolated from gingiva against ampicillin, clindamycin, cefazolin, erythromycin, clarithromycin, rifampin, and gentamicin was investigated by agar dilution method. 110 strains of streptococcus viridans were isolated. None of the isolated strains were resistant to ampicillin. Although previous studies demonstrate that oral penicillin prophylaxis results emergence of penicillin resistant streptococcal strains in oral flora the relationship was not clear cut for intramuscular benzathine penicillin prophylaxis. Our study showed that the prolonged use of intramuscular benzathine penicillin does not cause ampicillin resistance for streptococcus viridans. In conclusion, we suggest that ampicillin and amoxicillin can be used for IE prophylaxis in patients with RHD on benzathine prophylaxis.

P716

Relationship between relative lymphocyte concentration and heart failure severity in pediatric patients with heart disease

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Neurohormonal factors play important roles in the development of congestive heart failure(CHF). Increased cortisol secretion in response to the stress with CHF may decrease relative lymphocyte concentration (%Lc), and thus %Lc may be related to the severity of CHF. To test this, we investigated the relationship between %Lc and the severity of CHF in pediatric patients with heart disease (n=193). The severity of CHF was graded from 0 to 3 according to the clinical signs and symptoms of CHF. %Lc was corrected as %Lc / age-related average value of %Lc (c%Lc), since the normal range of %Lc varies with age during childhood. c%Lc was significantly lower in patients with severe CHF (grade 3: needed respiratory management) compared to other patients (p<0.0001). This relationship was observed in a subgroup of patients with cyanosis (n=40, p<0.05) or pulmonary high flow (n=45, p<0.05). c%Lc of patients in grade 3 tended to rise after resolution of CHF. In conclusion, c%Lc is decreased with severe CHF in pediatric patients, and may be an inexpensive and useful marker for the development of CHF.

P717

Oral anticoagulation in children, evaluation of a protocol-driven, nurse-run outpatient clinic

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Background and Methods: Oral anticoagulation in children is necessary following insertion of mechanical valves and some two-ventricular procedures. We recently changed from a junior doctor to nurse-run clinic. The nurse obtained finger-prick INR samples and prescribed warfarin according to a newly-devised anticoagulation protocol. This featured desired international normalised ratio (INR) ranges for different indications and dose recommendations depending on INR measurement. Readings < 1.8 or >6.0 and those +/- 1.5 out-of-range were referred for senior medical advice. Patient records were examined to assess the efficacy of the system. Results: In the 30 months commencing January 1996, 35 patients aged 4 months to 16.5 years were managed in the clinic (23 patient medication years). There were 991 INR tests with a mean of 16 days between samples. 50% of results were in target; of those out-of-range, 37% were above target and 63% below. 356 (52%) of all INR results led to a change in warfarin dose (average change 0.8 mg/day). 65% of dose alterations (233/356) deviated from protocol guidelines. Reasons for deviation included inappropriate protocol (65%) including inadequate dose range (e.g. large dose increments), no provision for higher doses or lack of flexibility, rapid movement towards new target range (25%), other factor (e.g. surgery, 4%), or unclear reason (6%). Concomitant drug administration in 7 patients had discernible effects on the INR (antibiotics=3, anticonvulsant=1, other=3). 4 patients were admitted a total of 7 times (mean stay 5 days), 6 due to INR >6 and 1 due to INR<1.8. There were no thrombosis or haemorrhagic complications. Conclusions: Anticoagulation control in children is a clinical challenge. INR control with acceptable error rates and low morbidity can be safely managed by a specialty nurse with appropriate guidance. Further development of the anticoagulation protocol is required.

P718

Idiopathic infantile arterial calcification: two cases

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Idiopathic infantile arterial calcification (IIAC) is a rare and usually fatal disease. In 80% of the known patients death occurred in the first six months of life. The etiology is not determined. IIAC is characterized by calcifications of the internal elastic lamina of the large and medium sized arteries along with proliferation of fibrous tissue within the intima. First case: A male three-week-old-newborn was admitted to the hospital with upper respiratory tract infection. His mother had had one stillbirth and one child who died at the age of 10 weeks. This child presented shortly after admission with signs of cardiac failure and electrocardiographic evidence of myocardial ischemia. His clinical condition deteriorated and he died in cardiac arrest. Second case: A eight months-old-girl with clinical signs of severe mitral valve insufficiency had several stenoses of the left coronary artery with complete occlusion of the LCX on cardiac catheterization. A mechanical valve was implanted and she did very well over the next year without cardiovascular problems. At the age of almost five

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

years she suffered a cardiac arrest with ventricular flutter and was successfully resuscitated at home. On admission she presented with pulmonary edema, low cardiac output and deteriorated ventricular function on echo. Stabilization was achieved in the intensive care unit. The following days she deteriorated neurologically and died in cardiac failure and pulmonary edema after unsuccessful resuscitation. As in most cases in the literature the diagnosis of IIAC was established by autopsy. Some patients who were diagnosed with IIAC were treated with diphosphonate successfully. Genetic counselling for the families is necessary as an autosomal recessive inheritance is discussed.

P719

Accessory orifices of the atrioventricular valve in atrioventricular septal defects: echomorphologic correlates

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Accessory orifice (valvular atrioventricular (AV) valve represents a rare form of congenital heart disease but can have significant effects on the patient. We examined the anatomical substrates in heart specimens and in patients to identify features that may help in diagnosis. We studied 32 specimens with atrioventricular septal defects (AVSD) that had accessory valvar orifices and compared our findings with echocardiograms from 13 patients. Sizes and location of the accessory orifices were analysed and the associated anomalies described. At echocardiography, we sought to establish any evidence of valvar stenosis or regurgitation. Accessory orifices were found in association with all variants of AVSDs. Three anatomic variants were identified: central bridging (36%), peripheral bridging (29%) and fenestration (36%). Only one valvar bridging was identifiable at echocardiography. The left AV valve was the most involved both at anatomical (67%) and echocardiographic (86%) observation. Left ventricular inflow obstruction was found in 9 patients and mild regurgitation in 3. In patients with AVSD, parasternal and subcostal short axis sections obtained the best diagnostic sensitivity. The absence of the usual trifoliate appearance of the left AV valve was the hallmark of the lesion. As the lesion represents an additional risk factor for surgery its recognition at echocardiography prior to any planned operation is extremely important.

P720

Coronary patterns, myocardial perfusion and coronary flow reserve assessed by positron emission tomography in patients after Fontan like operations

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Ventricular dysfunction frequently observed in patients after Fontan like operations (FLO) is a serious complication that might contribute to poor long-term results. Ischaemic heart disease will have debilitating consequences on a Fontan heart. Aim: Assessment of myocardial perfusion and coronary pattern correlated to clinical and haemodynamic parameters. Methods: 10 patients (15.8 years SD 5.1) after FLO had transoesophageal echocardiography and cardiac catheterization 9.3 years SD 4.2 after surgery. Myocardial perfusion was assessed by NH_3 PET at rest and after maximal vasodilatation; the results were compared with 10 healthy adults (28.1 years SD 6.3). Results: Ventricular function was normal in 4, and reduced in 6 patients. Angiographically 5 had a spiral course of the distal coronary arteries. Compared to normals myocardial blood flow (MBF) at rest was significantly higher in the FLO (0.94 SD 0.25 vs. 0.77 SD 0.17 ml/g/min; p<0.026), and coronary flow reserve (CFR) was significantly reduced (2.5 SD 0.68 vs. 4.1 SD 1.01; p<0.0035), especially in those with impaired ventricular function, coronary vascular resistance after maximal vasodilatation was significantly elevated in the FLO (44.0 SD 18.3 vs. 28.3 SD 8.45 mmHg/ml/g/min; p<0.037). Conclusion: Abnormal coronaries, altered MBF, and impaired CFR are common findings in FLO; increased CFR and reduced ventricular function are significantly correlated, seem to progress with time, and may be risk factors for the long-term outcome.

P721

Aortic valve prolapse under the age of 1 year in supracristal ventricular septal defect

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The aortic valve prolapse (AoVP) usually begins as early as 7 years of age in supracristal ventricular septal defect (VSD). However, recent echocardiographic study demonstrates that aortic valve involvement can develop during younger age. In this study, we investigated the incidence and characteristics of aortic valve involvement under 1 year of age by using two dimensional and color Doppler echocardiography. From 1983 to 2000, 140 patients with supracristal VSD underwent diagnostic evaluation at our institute (VSD and AoVP 38; VSD, AoVP, and aortic regurgitation (AR) 10; VSD without aortic valve involvement 52). In those 140 patients, 5 patients (3.5%) showed AoVP under the age of 1 year (mean age 4.6 months after birth). AR was recognized in 2 cases (2 months, 11 months, and 23 days after birth, respectively), which was not audible in either case. Two patients had a mild degree of failure to thrive. Four patients were male and 1 was female, indicating male predominance. In the youngest case, both AR and AoVP developed at 23 days after birth, and AoVP was recognized during both systole and diastole. In the case, her elder sister also showed supracristal VSD and AoVP. In all cases, only patch closure of VSD was carried out, and AR and/or AoVP improved after the operation. In conclusion, 3.5% of patients with supracristal VSD developed aortic valve involvement under the age of 1 year. In a patient with recurrence of supracristal VSD in siblings, aortic involvement might occur during quite early neonate period, and regular evaluation with echocardiography is mandatory. Even in such cases, early operation of VSD closure is effective, and their

P722

Psychosomatic development in long term follow up in children after arterial switch operation

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The aim of the study was to assess the impact of risk factors appearing in perioperative period on the psychosomatic development in children 5–8 years after neonatal ASO. Between 1983 and 2000, ASO was performed on 300 pts with TGA (94% in neonate period). 41 children in age 5–8 years entered in this study (28-TGA+IVS, 11-TGA+VSD, 2-TGA+AAA). All patients were in I NYHA clas. None of them required medication. Somatic development was estimated by BMI. Psychical development was examined using interview with parents, Terman-Merill and Bender-Koppitz test, WISC-R, Stone, Wechsler Intelligence Scale of Children, family and tree drawing. Intelligence (IQ), harmony of the development, visual - motor coordination were assessed. Normal or higher level of IQ was confirmed in 35 children (85%). In 6 pts IQ was slightly lower than normal. Visual-motor coordination was quite normal in 24 children (59%). Dysharmonic mental development was observed in 15 pts (46%). In all patients except one somatic growth after ASO were in normal range. Results of our study were correlated with following features: Apgar score, body weight, preoperative value in modified NYHA scale, age at operation, aortic cross clamping time, postoperative period. Shapiro-Wilk, Mann-Whitney, F-Snedecor, Fisher chi 2 and logistic regression) were used. Significance (p below 0.05) correlation was found between harmony of psychical development and preoperative NYHA (modified NYHA scale). No correlation was found among another features.

P723

Longitudinal follow-up of children undergoing surgery for tetralogy of Fallot in Montenegro

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In this study we analyse a group of 18 patients after a complete correction of Fallot's tetralogy. The aim of the study was to examine the results of the complete correction and to estimate the life quality of children who were operated on. At the time of operation the children's age ranged from 6 months to 11 years. Neuroponderal development was evaluated before and after the surgical treatment. We especially emphasized the importance of echocardiographic postoperative follow-up. Recanalization of 'patch' was found in 2 cases. Eight patients still had enlargement of right ventricle. Patients who were operated on at the age of 10 and 11 have larger enlargement of right ventricle. Only 4 patients had a significant pulmonary and mucuspid regurgitation that was found with the Doppler method. Pressure gradient ranged from 35–40 mmHg. Ten patients had pulmonary stenosis but with a very small gradient between right ventricle and pulmonary artery. The duration of postoperative follow-up was from 6 months to 13 years (mean 6.2 years). According to Binet-Simon's tests we evaluated that IQ with majority of children, ranged from 95–106. That corresponds to slightly lower values compared to healthy population. Social maturity (SQ), according to Doll's scale, ranged from 85 to 122, which corresponds to the values of healthy population. We conclude that our hypothesis a true: early diagnosis and operation of congenital heart disease give better operation results which directly enables better quality of life.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P724

Digitized ECG recordings in the premature infant < 32 weeks of gestational age

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Background: QT interval prolongation is associated with severe ventricular arrhythmias such as torsades de pointes and sudden death. Frequently used drugs such as cisapride, and macrolides are known to prolong ventricular repolarization. Normal values of QTc interval in prematures younger than 32 weeks are unknown. Methods: We prospectively studied 104 healthy premature infants of gestational age ≤ 32 weeks during the first 10 days of life. The 12-lead digitized ECG was recorded at a sampling rate of 500Hz, stored and transfered to a PC. A dedicated algorithm allowed to quantify ECG parameters. Results are expressed as mean ± SD. Results: Gestational age was 30,3 ± 1.6 weeks, and birth weight 1351 ± 341gr. The Apgar score at 5 minutes was ≤ 7 in 9 cases (9%). Corticosteroids were administered prenatally in 63 cases (60%) and caffeine postnatally in 92 cases (88%). A central catheter was inserted in 78 cases (75%). The ECG was recorded on day 4.4 ± 2. 9. The mean ECG parameters were the following: HR: 141 ± 17 bpm, PR interval 93 ± 21 ms, QT interval 290 ± 32 ms and QTc interval 443 ± 33 ms. QTc interval was > 440ms in 52 cases (50%). QTc duration was not correlated with gestational age but was inversely correlated with post-natal age with QTc longer before day 5 (448 ± 33 ms, n = 64 versus 430 ± 29ms, n = 40, p=0.005). No adverse cardiac events were reported during a 1 year follow up. Conclusion: A QTc interval > 440ms was found in half of the premature infants ≤ 32 weeks. This QTc prolongation was not correlated with gestational age but with post-natal age.

P725

It is not mandatory to give Aspirin to patients with modified Blalock-Taussig shunts

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Traditionally, patients having undergone pall ative modified systemic-pulmonary shunts have been treated with anti-platelets doses of Aspirin. Based upon our previous unpublished experience in another institution, and despite of controversial publications, we continue to discharge such patients without any anti-coagulation, with the exception of Norwood procedures, due to the well known risks of thrombose of the aortic segment of the ascending aorta. Between July 1998 and November 2000, we performed 35 modified Blalock-Taussig shunts in 33 patients aged between 1 day and 7 years (median 12 days), with weights between 1.6 and 29 kg (median 2.7kg). Indications for surgery were hypoplastic left heart syndrome in 8 patients who had a Norwood procedure (24.2%). Fallots tetralogy in 6 patients (18.3%), pulmonary atresia + ventricular septal defect (VSD) in 6 patients (18.3%), transposition of the great arteries with VSD and pulmonar stenosis in 3 patients (9%), tricuspid atresia in 4 patients (12.2%), obstructive rabdomiomia in 1 case (3%), and complex cardiac malformations in 1 patient (3%). The size of the shunt varied from 3 to 6 mm. Early mortality was 12% (4 patients) at total. 3 Norwood procedures and only 1 in 25 patients with shunt alone (4%). The 4 surviving patients with a Norwood procedure were discharged on 5mg/kg/day of Aspirin. All the remaining 29 patients have been followed with no anti-platelet therapy for a range of 1 to 26 months (median 9 months), until indication for further surgical therapy. So far, we have not had any complication secondary to partial or total shunt obstruction. Hence, we reckon that in the view of potential risks of anti-platelet therapy, there are no significant benefits in giving such treatment to these patients.

P726

A case of isolation of the left innominate artery from right aortic arch with chromosome 22q11.2 deletion

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It has reported that 22q11.2 deletion syndrome is often associated with various anomalies of derivatives of aortic arch system. We experienced a case of chromosome 22q11.2 deletion with right aortic arch and isolation of the left innominate artery. The patient was 11-month-old boy who had been diagnosed as ventricular septal defect (VSD) and sub-and supravalvular pulmonary stenosis (PS) during neonatal period. He had a simple partial seizure with spike focus on left parietal-temporal area at 7months old. Hypoperfusion of the left temporal lobe was recognized on brain SPECT while no abnormality was found on brain CT and MRI studies. On angiogram the left innominate artery (LIA) was isolated from the aorta. The right common carotid artery (RCCA) and right vertebral artery arcegradely supplied the right semisphere of the cerebrum and then perfused into the left-sided cerebrum via the circle of Willis. The RCCA also partially drained into the left common carotid artery (LCCA) through the thyrocervical collateral arteries. The LCCA and left vertebral artery filled arcegradely and retrogradly to the left subclavian artery. Accordingly, the closure of VSD and release of PS were successfully performed without any deterioration of the brain function postoperatively, although the communication between LIA and aortic arch was not established due to a small diameter of LIA (2.5mm). We consider that reconstruction of the LIA is required for preventing subclavian steal syndrome is required in future.

P727

The impact of improved echocardiographic detection of infective endocarditis on management in the pediatric age group

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All patients evaluated in our echocardiography laboratory for infective endocarditis (IE) from 5/1994 to 12/1999 were reviewed to evaluate the rate of detection, echocardiographic presentations & their impact on management of IE. Results: 90 patients were identified. 46 had positive echocardiographic findings (sensitivity 51%). 26 pts with indwelling lines were excluded. Twenty patients were analyzed. [median age 6.5y, range 0.14–27]. Patients were divided into two groups. Group A: no previous documentation of heart disease (n=4). Group B: pts with CHD (n=16). Group B included VSD (n=5) (31%), aortic valve (AoV) stenosis (n=3) (19%). Pulmonary stenosis (n=3) (38%), residual PDA after device closure (n=1) & miscellaneous (n=1). Four pts had a procedure <1 month prior to diagnosis (3 cardiac surgery & 1 cardiac catheterization). Small valvular rupture (SVR) was noted in 4 pts (2 ruptured into the LV, 1 into the RV & 1 into the RA). Anatomic involvement: Mitral valve (MV) (n=6), AoV (n=6) (including the 4 with SVR), MV & AoV (n=3), TV (n=1), MPA (n=3) & Blalock Taussig shunt (n=1). Organisms isolated by blood culture included streptococcus mitis (n=4), staphylococcus aureus (n=3), streptococcus pneumoniae (SP) (n=2), enterococcus (n=2), staphylococcus epidermidis (n=1) & streptococcus sanguis (n=1). Seven patients had culture negative endocarditis (35%). Twelve pts required surgical intervention & 8 were managed medically. (5% mortality, n=1). Only the left sided valves were involved in Group A (MV n=3 & AoV n=1). The only two cases of SP bacteremia had SVR. Early diagnosis of IE prior to clinical suspicion allowed early treatment in 3 patients. Conclusion: Improved echocardiographic detection of infective endocarditis allows early diagnosis, which may decrease morbidity. Clinical suspicion of Small valvular rupture should be higher with streptococcus pneumoniae bacteremia.

P728

Cardiorespiratory capacity in young patients after corrective surgery for tetralogy of Fallot

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Impaired RV-function following repair of tetralogy of Fallot (TOF) may lead to reduced cardiorespiratory capacity. Exercise testing with continuous O2-uptake measurement allows an objective assessment of the cardiorespiratory capacity. We used exercise testing to study cardiorespiratory capacity in patients after TOF repair. We measured maximal oxygen uptake (VO2 max), carbon dioxide output (VCO2), anaerobic threshold (AT), heart rate (HR), and blood pressure (BP) during treadmill testing using a modified Bruce-protocol. Consecutive TOF patients after a mean follow-up of 9.8±3.4 years (N=38, 22 male and 16 female, mean age 12.2 years, range 6–18) were subdivided by type of surgical intervention (sub- or transannular patch). The results were compared with those of an age-matched control group (N=47). Mean exercise time was shorter in the TOF group (12.06±2.4 vs. 14.34±2.3 min., p<0.001) and both VO2 max as well as VCO2 were lower (29.7±7.0 vs. 41.9±8.5 ml/kg/min, p<0.01, 1.23±0.6 vs. 1.74±0.8 l/min, p<0.01, respectively). AT was reduced in the TOF group (17.4±5.1 vs. 23.6±2.3 ml/kg/min, p<0.01). There was no difference in increase of heart rate or blood pressure. Between the TOF subgroups there were no differences in the parameters

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

except for exercise time (10.4±1.5 vs 12.5±2.3 min, p=0.01). Conclusion. Exercise capacity is reduced after repair of TGA. The type of corrective surgery in our study group did not influence cardiorespiratory capacity during mid-term follow-up.

P729

Health status and quality of life in children with Transposition of the Great Arteries

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The transition from the atrial to the arterial switch operation for treatment of transposition of the great arteries (TGA) was based on optimizing long-term outcomes and quality of life (QOL). We sought to assess health status and QOL in children after surgery for TGA during the time of transition in management strategy. Children previously enrolled as neonates in a prospective study of the Congenital Heart Surgeon's Society between 1985 and 1989 were eligible. Presumed survivors were sent a medical follow-up questionnaire and the Child Health Questionnaire for completion. Of 704 presumed survivors, 300 children, mean age 12±1 years, completed the questionnaires (44%), including 215 males (70%). Diagnoses included simple TGA in 202 (66%), TGA with VSD in 64 (22%) and TGA with VSD and PS in 20 (7%). The operative repair was by arterial switch in 189 (62%), atrial switch in 105 (34%; Senning, n 58, Mustard 47), and Rastelli in 12 (4%). Overall, QOL scores were significantly higher than normal control population, except for self-esteem (e.g. physical functioning= 93.3 vs 88.8 out of 100, p<0.01). Children after an arterial switch scored significantly higher than after an atrial repair in the areas of physical functioning (95.7 vs 91.2, p=0.01), bodily pain, general health perceptions, mental health and self-esteem, with no differences between the Senning and Mustard patients. Multivariable analysis confirmed the independent relationship of type of operation to QOL scores. Additional variables, including medical status, physical, behavioral and learning problems, and developmental milestones as assessed at follow-up were appropriately related to high and low scores on QOL scales addressing similar issues. QOL in children appears to be better with arterial than atrial switch. Factors predictive of better QOL need to be further defined.

P730

Time course of procalcitonin serum levels after cardiac surgery in children compared to other markers of inflammation

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Cardiac surgery induces a systemic inflammatory response syndrome (SIRS) causing an increase of different markers of inflammation. Therefore, differentiation between bacterial infection and SIRS in the postoperative period is difficult. In contrast with classic inflammatory indicators Procalcitonin (PCT) is considered to undergo only minor increases after surgery. Methods. Plasma serum levels of PCT, C-reactive protein (CRP), interleukin 6 (IL-6) and interleukin 8 (IL-8) were measured in 64 patients with congenital heart disease (mean age 5.3 years) who underwent cardiac surgery. Blood sampling was performed preoperatively, on the hours 1, 4, 12 and the days 1 to 6 after the operation. Results: Maximum PCT levels were found at the first postoperative day (mean 5.2 +- 1.1 ng/ml). CRP showed its peak two days later (mean 84.6 +- 7.3 mg/l). Highest mean values of IL-6 and IL-8 were 254.92 +- 44.2 pg/ml respectively 51.3 +- 12.5 pg/ml 12 hours after surgery. The difference in time where the maximum serum levels were observed was statistically significant (p < 0.001). Conclusion: PCT exhibits an increase in all patients similar to a more delayed change of CRP levels. As expected IL-6 and IL-8 peaked earlier. Thus, PCT provides – apart from its different kinetic – no advantage in the discrimination of SIRS from bacterial infection in this group of patients.

P731

Psychosocial competence and intellectual skills in adolescents with congenital heart disease

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An increasing number of patients with congenital heart disease are reaching adolescence and adulthood due to corrective surgery. They are mainly in good hemodynamic state and good physical condition. We tried to assess also the intellectual skills and psychosocial state. Questionnaires like Meyer-Probst Encephalopathy-Q, Youth-Self-Report (YSR), Child behaviour checklist (CBCL) were distributed to the parents and patients and a Hamburg-Wechsler-Test (HAWIK) and Test of visuelles of attention (TOVAC) were performed. There was a control group (I, n=20) consisting of patients with innocent murmurs or mild CHD without any necessity for treatment. In Group II (n=21) were pts after corrective surgery for acyanotic CHD and in group III (n=20) those after corrective surgery for cyanotic lesions. Groups were comparable for the socio-economic state of the family and age and sex distribution (mean age 11,8 years). HAWIK: There was no difference in the overall intellectual skills and the verbal part. Significant differences could be detected in the mathematical part (II worse than I p<0.01), in activity skills II and III both worse than I (p<0.01) as they were in the visual part (p < 0.008). No significance was found in the CBCL and TOVAC, although in all test group I was best and III better than II. To explain the higher results of group III compared to group II we suspect a more intensive training of the pts in group III as they are more taken care of for their cardiac problems too. Problems in social competence and logical thinking are also influenced by the family background. Overall we found our adolescents very well adapted to their situation. Further studies for the long-term problems – job, pregnancy – are to be carried out.

P732

Purulent pericarditis in childhood

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The patients with purulent pericarditis admitted between 1993 and 2000 to university hospital were analyzed retrospectively for their etiology, management and prognosis. Total number of patients was 22; 15 boys and 7 girls with a mean age 5.6±4.7 years (range 6 months to 13 years). Most of children presented with fever, tachypnea and chest pain. Cardiac tamponade was not seen in any children. The preceding or concurrent infections were septicemia (n=6, four of them were septic arthritis), infective endocarditis (n=2), pneumonia (n=2). 11 patients had no evidence of other focal infection. All patients were treated on parenteral antibiotics. Subxiphoidal pericardial drainage was performed in 14 cases for diagnosis and treatment, than all of them underwent surgical drainage. Five patients received only medical treatment (antibiotics). Three patients underwent surgical drainage without subxiphoidal pericardial drainage. Staphylococcus aureus was the most frequent causative organism of purulent pericarditis (n=9). Other organisms were Streptococcus pneumoniae (n=2), Escherichia coli (n=1), Haemophilus influenzae (n=1), Mycobacterium tuberculosis (n=1), Legionspira (n=1). No microbiologic agent was found in seven patients. Seven children died because of sepsis, the remaining 15 made a complete recovery and non of them had constrictive pericarditis approximately in five years. In conclusion, purulent pericarditis is common disease in our country. There are so many microbiologic agents of purulent pericarditis and its mortality is high.

P733

Outcome of balloon dilatation of native and recurrent coarctation of the aorta at a European Tertiary Care Centre

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All 75 balloon dilatation procedures performed on 67 patients with both native and recurrent coarctation, at a European Tertiary care centre between 1990 and 1998 were analyzed retrospectively. There were 35 patients with native coarctation. Mean age was 9 years (range, 2 months to 45 years). The mean peak to peak (PTP) systolic pressure gradient decreased from 27.8 ± 15.4 mmHg to 9.9 ± 9.7mmHg (p < 0.05). A PTP gradient of < 20mmHg was achieved in 20/35 patients (80%). Five patients have developed a recoarctation. One has undergone surgery, 1 patient received a stent, two are being followed up conservatively and another died during surgery for a second stage Norwood operation. A PTP gradient of > 20 mmHg was achieved in 5 patients. Three parents underwent surgical correction, and 2 being followed up conservatively. There were 32 patients with recurrent coarctation. Mean age was 12 years (range, 1 month to 42 years). The mean PTP systolic pressure gradient decreased from 35.2 ± 21.0mmHg to 15± 12.1 mmHg (p < 0.05). A PTP gradient of < 20mmHg was achieved in 26/32 patients (81%). Five of these patients required repeat balloon dilatation procedures after recurrence of their coarctation gradients. A PTP gradient of > 20 mmHg was obtained in 6 patients immediately following balloon dilatation. Only one patient

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

required surgery; one patient underwent a stent implantation and another patient required repeat angioplasty for a residual stenosis. The remaining 3 patients are normotensive and being followed up consecutively. Complications of the procedure are discussed. Details of stent implantations and repeat procedures are outlined. In conclusion, satisfactory immediate and medium term results for balloon dilatation of both native and reconstructed are achievable with few complications in selected patients.

P734
The analysis of nonlinear dynamics of the heart rate variability pre- and post-operation in congenital heart diseases
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Objective To investigate the clinical significance of the nonlinear dynamics method in analyzing the heart rate variability (HRV) of congenital heart diseases (CHD). Methods By using the nonlinear dynamics method and the power spectrum method, the authors analyzed the HRV signals pre- and post-operation in 2 groups of CHD patients: ventricular septal defect (VSD) and Tetralogy of Fallot (TOF). Results 1. There were no significant differences of all indices between two groups pre-operation ($P>0.05$). 2. The indices of the power spectrum and nonlinear dynamics pre-operation were significantly different from those post-operation ($P<0.05$, $P<0.01$). 3. After operations the indices of nonlinear dynamics in TOF group were significantly lower than those in VSD group ($P<0.05$) but power spectrum indices showed no difference. 4. Before operations the Poincare plot of HRV in two groups of patients displayed the pattern of comet. After operations, the Poincare plot of VSD patients showed the torpedo pattern at first and then gradually changed to the comet-like pattern at the 10th day after operations. The Poincare plot of TOF patients showed the complex patterns after operations, and remained in this pattern at the 30th day after operations. Conclusion Nonlinear dynamics method seems to be a more sensitive method than the traditional method and might provide more information about unexpected incidents of heart diseases.

P735
Major coronary artery anomalies in childhood
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Coronary anomalies are rare in children. Over a period of 10 years, we reviewed the data of children with serious coronary artery disease. Twenty-seven patients were selected (10 m–17.1 age at diagnosis 1m–14y). A heart murmur was found in 85% and signs of heart failure in 41%. Cardiomegaly or pulmonary congestion was found in 85% and only 26% showed a normal EKG. Echocardiography detected coronary dilatations in 70%, decreased LV function in 55%, mitral incompetence in 41% and anomalies of the mitral papillary muscles in 30%. In 12 patients, abnormal origin of the left coronary artery from the pulmonary artery (ALCAPA), was diagnosed, coronary fistula in 10, coronary stenosis in 4 and coronary thrombosis in 1. Treatment consisted of re-implantation in the ALCAPA group, 1 died. In the coronary fistula group, all except one were succesfully treated surgically or by intervention. In the 4 patients with coronary stenosis, 2 were succesfully treated surgically, and 1 died. The child with coronary thrombosis could be treated medically. Conclusion: severe coronary lesions are rare in childhood, with varying clinical presentation. However echocardiography can give a decisive clue to diagnosis. Succesful therapy is available for most of the cases.

P736
Musculoskeletal deformities in children after thoracotomy for congenital heart disease
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The standard surgical approach for closed heart procedures in small infants and children are done via a posterolateral thoracotomy incision, which results in the division of the latissimus dorsi and serratus anterior muscles. We have followed the evolution of 42 children (22 boys and 20 girls) operated with a left and right sided posterolateral thoracotomy in the forth intercostal space for congenital heart disease between 1983 and 2000. Additional median sternotomy was done in 9 (21%) patients. Mean age during operation was 10 ± 3.4 yrs (range 1 month–13 yrs). These patients (pts) were evaluated at mean age of 10 ± 5.1 (range 1–22) yrs. The evaluation was performed an average of 6 (range 1–13) yrs after thoracotomy. Thirty-nine (92%) of the pts had significant musculoskeletal deformities: 1) A scoliosis of 10 degrees or more was observed in 14 pts (33%), in 11 pts scoliosis was clinically and radiologically detected while in 3 it was diagnosed radiologically. Severe (50%) of these pts had aortic coarctation, 2) 15(35%) and 6(14%) pts had prominent elevation of the left and right shoulder, respectively. 3) Twenty-five (60%), 5 (12%) and 3 (7%) pts had left, right and bilateral.

P737
Rheumatic Fever in Western Australia – a 15yr experience from the tertiary children's hospital
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Introduction: Princess Margaret Hospital (PMH) is the only tertiary paediatric hospital in Western Australia (WA). The purpose of this study is to describe the clinical experience with rheumatic fever (RF) at PMH from Jan 1986 – June 2000. Methodology: A retrospective clinical review of medical records was conducted. Patients were identified using the PMH coding system. Those included had to meet the revised Jones criteria. Results. Forty six patients were identified with 44 being indigenous and 37 female. Age range, 18 months to 17 years. There was no evidence of a reduction in the number of admissions/year during the study period. Major presenting clinical criteria: Arthritis – 14, carditis – 14 and chorea – 24. Eight patients had more than one major criteria. A previous history of RF was recorded in 25% of first admissions. Forty-five patients had echocardiography with 32 found to have trivial to mild mitral regurgitation (MR) and only 12 of these had a radiate murmur detected. Patients with moderate carditis accounted for the longest mean stay of 31 days. Four patients required 6 surgical procedures including two mitral valve replacements. Compliance with 3M benzathine in the Metropolitan area demonstrated only 47% continue to have regular injections. Outcome: No deaths were recorded acutely or on follow-up. There was a recurrence rate of 25%. No patient reliably receiving prophylaxis had a recurrence. Of the patients presenting with significant carditis, 36% eventually required surgery. Conclusion: Acute RF and chronic rheumatic heart disease continues to present to PMH and cause significant morbidity amongst indigenous children in WA. Clinical follow up and penicillin compliance are major challenge for health professionals. Echocardiography is very valuable to aid the diagnosis of RF by demonstrating sub-clinical val

P738
Positive blood cultures in children with structural heart disease: do they have infective endocarditis?
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Patients with structural heart disease (SHD) and bacteraemia are generally considered at risk for developing infective endocarditis (IE). However, the diagnosis is often uncertain, and it is unclear if and for how long they should be treated. We retrospectively reviewed 58 such patients over a 5-year period. Medical records, echocardiography and laboratory results were reviewed. Patients were classified according to the Duke criteria for IE as having definite, possible or rejected diagnosis of IE. To ascertain whether any patients with a diagnosis of rejected IE subsequently developed the disease, follow-up evaluations in the 12 months after discharge were also reviewed. The most common organisms cultured were coagulase negative Staphylococcus species (39.2%), and Staphylococcus aureus (14.8%). Up to 40% of the positive cultures were considered to be possible contaminants. None of the patients had definite IE according to Duke criteria. Eleven patients (22%) were defined as having possible IE, and the diagnosis was rejected in the remainder (78%). Of those that were rejected, 88% had a firm alternate diagnosis, and in 82% the clinical manifestations resolved in under 4 days of antimicrobial therapy. None of the patients went on to develop IE. Positive blood cultures in children with SHD do not necessarily imply IE. The diagnosis should be considered in all such patients, but the decision to treat as IE is guided by careful consideration of clinical and laboratory data. Brief courses of antibiotics may be adequate in some patients provided they are reviewed closely. The Duke clinical criteria for IE appear to be applicable to children.

P739
Cardiac troponin I following congenital heart disease surgery on bypass – prognostic relevance and normal ranges
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https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Serum cardiac troponin I (CTNI) is an established marker of myocardial damage in adults but not children. Previous studies with small numbers have shown an association with more complex surgery and increased post-operative morbidity. 137 children undergoing surgery on cardiopulmonary bypass were prospectively enrolled with measurement of CTNI (Dade Behring Dimension heterogeneous immunoassay) at 0, 4, 12, 24 and 48hrs post admission. This data is combined with measurements on 80 patients retrospectively. CTNI was compared with ICU variables. Surgical procedures on 217 patients included patch closure VSD (n=44) repair tetralogy of Fallot (n=19), suture closure ASD (n=16), conduit replacement (n=13) and arterial switch (n=12). Median patients age 1.2yrs (range 7 days to 16yrs). Mean bypass time was 113±67mins and x-clamp 53±35mins. Patient mortality was 1.8%. On univariate analysis CTNI at all times individually correlated with an order of strongest correlation: bypass time, dobutamine requirement, dopamine requirement, x-clamp time, dialysis requirement, nitric oxide (NO) requirement, and ventilation time. There was a strong linear correlation between bypass time (p=0.007), adrenaline requirement (p=0.01), NO requirement (p=0.02), and mortality (p=0.03) with CTNI over time. CTNI was significantly different for the most common procedures: patch VSD, repair tetralogy of Fallot, suture closure ASD, conduit replacement and arterial switch with the difference greatest at the immediate and four hour samples and becoming less significant with the later samples. Within the arterial switch group there was very high correlation with CTNI at all times with x-clamp time (r=0.79–0.96), bypass time (r=0.5–0.9), ventilation time (r=0.70–0.82), dobutamine requirement (r=0.68–0.85) and dopamine requirement (r=0.69–0.85). Weaker correlation was seen with some of the variables in the other surgical groups. CTNI is a useful marker following cardiac surgery but may be most predictive in patients following arterial switch.

P740

Intrapulmonary shunting through bronchial circulation after surgery for congenital disease

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Two unusual cases of shunting through the bronchial circulation are reported, both ameliorated by catheter intervention therapy. In one case of simple transposition of the great arteries following arterial switch operation, dilatation of the left ventricle was investigated. The only possible cause detected was an angiographically large shunt through a dilated bronchial arterial circulation arising bilaterally from the thyrocervical trunks and from the thoracic descending aorta and draining via the pulmonary veins. Coil embolization of these bronchial vessels resulted in acute reduction of bronchial flow and reduction in left ventricular dimensions. In a second and unique case of left atrial isomerism with total left-sided pulmonary venous obstruction resulting from drainage of the left pulmonary veins to a blind-ending left atrium (absent left atrioventricular connection and intact atrial septum with no egress from the atrium), a dilated plexus of veins had developed along the left bronchial tree, coalescing into a venous trunk which joined the hemi azygos vein. This provided venous egress of the arterial supply to the lung which in this case and, typically for cases of unilateral pulmonary venous obstruction, was a systemic arterial supply from bronchial and acquired systemic arteries, with associated retrograde flow in the left pulmonary artery. Following atrial septectomy and total cavopulmonary shunt (Kawashima operation) corrected flow in this dilated primitive left bronchial venous system resulted in a significant right to left shunt from the hemiazygos vein to the pulmonary venous atria via communications with the pulmonary veins. Embolization of this dilated primitive bronchial venous trunk resulted in improved systemic arterial saturations. Following surgery for congenital heart disease, shunts which are deleterious may develop through the bronchial arteries or veins and these may be amenable to catheter interventional therapy.

P741

Pseudoaneurysm of right subclavian artery following modified Blalock–Taussig shunt: 2 cases report

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Infective endocarditis (IE) is still an important problem in patients with congenital heart disease. It is difficult to be diagnosed especially in patients with palliative shunt. One of the serious complications of IE is mycotic aneurysm, which can lead to morbidity and mortality due to aneurysmal rupture. We had 2 cases of huge pseudoaneurysm of right subclavian artery developed after IE. The first case having tricuspid atresia, pulmonary atresia, with right modified Blalock-Taussig shunt had pseudoaneurysm of right subclavian artery, diameter 10x12 cm, demonstrated by computerized tomography (Fig. 1a, 1b). This patient developed respiratory failure and required ventilatory support due to right lung atelectasis from mass effect on right main bronchus. She died in PICU. The second patient, a case of Tetralogy of Fallot with right modified Blalock-Taussig shunt, had pseudoaneurysm of right subclavian artery, diameter 4x6 cm, demonstrated by echocardiography (Fig. 2) with mass effect on right main bronchus. The patient underwent surgery for resection of pseudoaneurysm and died during surgery. We propose that the mechanism of pseudoaneurysm formation in these 2 cases is secondary to ruptured mycotic aneurysm at the subclavian artery site of modified Blalock-Taussig shunt.

P742

Spectrum of congenital coronary artery anomalies: an institutional experience

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Coronary artery abnormalities are often a part of complex congenital malformations of the heart or may occur as isolated defects. In either case they may be benign or have major clinical and surgical consequences. In this study we have analyzed the spectrum of congenital coronary malformations detected in pediatric patients referred to a tertiary care cardiology center. This retrospective study included 3804 new patients evaluated in the OPD between January 1999 to July 2000. Surgical, echocardiographic, catheterization and pathology data were included in this study. Coronary artery abnormalities were detected in 76(2%) of the patients. 12(3/70) had anomalies of origin and distribution and 64(84%) had coronary artery fistulae. The abnormal coronary was an isolated lesion in 28%(16) cases and was an associated finding in the setting of congenital heart disease in 75%(60) cases. Four really important lesions included ectopic origin from the pulmonary artery(ALCAPA) in 10.5%, ectopic origin of the right coronary artery from the ascending aorta or left sinus in 5.3%, single coronary artery in 10.5% and dual supply to the left coronary system in 2.6%. Other abnormalities included variations in course and distribution in 59.2%, coronary artery fistulae in 7.9%, myocardial bridges in 1.2% and atresia in 2.6%. Associated malformations were tetralogy of Fallot (42%), transposition of the great arteries (22.4%), atrial defects (7.2%), double outlet right ventricle (6.4%), truncus arteriosus (3.2%), patent ductus arteriosus (1.6%), supravalvar aortic stenosis (3.2%) and cardiomyopathy (3.2%). There was a significant association of coronary artery abnormalities with conotruncal malformations (67%, P<0.01). 65% case were identified preoperatively by echocardiography. In conclusion, coronary artery abnormalities can have important surgical and clinical implications and thus require accurate recognition. Directed echocardiography now enables non-invasive diagnosis in most cases.

P743

The effect of non-ionic contrast medium on renal function in children with organic cardiac lesions

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To assess the effect of non-ionic contrast medium on renal function in children with organic cardiac lesions, we studied the change of several renal function parameters before and after the routine cardiac angiography in 63 patients with organic cardiac lesions. Serum creatinine(s-Cre), serum BUN(s-BUN), urine N-N-acetyl-beta-D-glucosaminidase(u-NAG), urine beta2-microglobulin(u-BMG), urine alpha 1 microglobulin(u-AMG) values were evaluated before and 12 hours after angiography using three non-ionic contrast mediums, iohexol 350mg/ml(n=10), iopromide 300mg/ml (n=31), and iopamidol 370mg/ml (n=22). There was no significant change in s-BUN and s-Cre. However, u-NAG/Cre, u-AMG/Cre and u-BMG/Cre, as the parameters for the tubular function, significantly increased 12 hours after angiography. These parameter values returned to the pre-angiographic values 2 weeks later. There was no significant difference in change pattern of these parameters among three mediums. In neonates and infants(n=21), renal tubular function parameters significantly increased more than in the elder-age children(u-NAG/Cre 15.3+/-8.5 to 48.3+/-38.9, u-BMG/Cre 535.2+/-834.5 to 5252+/-10286, u-AMG/Cre 12.6+/-40.5 to 27.4+/-48.4, respectively). In 11 patients with CHF, renal tubular function parameters significantly increased more than in patient without CHF (u-NAG/Cre 19.4+/-9.1 to 51.5+/-39.5, u-BMG/Cre 80.7+/-112.1 to 6910+/-13916, u-AMG/Cre 22.4+/-57.0 to 53.9+/-64.5, respectively). This study

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

suggest that the non-ionic contrast medium have any pathologic effect to the renal tubular epithelium, especially more significantly in children during neonatal and infantile period, or with CHF.

P744
Noncompaction with uncut aorta anomalies
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Noncompaction of the ventricular myocardium represents an arrest in the normal process of myocardial compaction, resulting in the persistence of multiple prominent ventricular trabeculations and deep intertrabecular recesses. A neonat neonatal 18-month-old girl with tetralogy of Fallot (TOF) was referred to our hospital for surgical treatment. Her family history was unremarkable. Her brain CT showed multiple infarcts. The echocardiography showed a VSD, overriding of aorta, infundibular and valvular pulmonary stenosis, hypoplastic of pulmonary artery and depressed left ventricular systolic function. At the cardiac catheterization, during the left ventricle angiography, a thrombi moving from the left ventricle to ascending and to abdominal aorta has been spotted. Catheterization was stopped and she was transferred to intensive care unit. The observation revealed no neurological symptom. Her ECHO examination was tedious, and trombi with 0x1.5cm dimensions was seen in the left ventricle apex. There were excessive prominent trabeculations in the left ventricle apex and middle portion of the left ventricle posterior wall, and in the right ventricular apex. Echocardiographic study showed no trombi on the 4th of therapy. Beta blocking and anticoagulant therapy was started. However she died at the 3rd week of therapy because of a cyanotic spell. Her anteriangiograms showed TOF, and right arcus aorta with anomalous origin of left subclavian artery from the descending aorta, and honeycombed appearance of the apex and one half of the posterior wall of left ventricle, and the apex of right ventricle. Prophylactic anticoagulant therapy for the prevention of the embolic episodes should be encouraged and echocardiographic screening must be done in the first degree relatives to identify non-symptoms in the asymptomatic phase.

P745
Repair of complete atrioventricular canal defect
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In the period January 1995 to November 2000, 49 consecutive patients underwent repair of isolated complete atrioventricular canal (AVC) at the Red Cross Children's Hospital heart unit with a two patch technique and routine mitral valve cleft closure. A retrospective review of these 49 patients was done. Age at the time of repair ranged from 3 months to 185 months (mean age 24 months). 15 patients were under 12 months of age. Previous palliative pulmonary artery banding had been done in 24 (49%), and 33 (68%) had Down's syndrome. Rastelli classification was A (29 patients), B (5), C (12), and unknown (3). The mitral valve 'cleft' was routinely closed. A posterior suture annuloplasty was required in 13 patients. Patch augmentation of the posterior leaflet was done in 2. Right AV valve needed repair in 11 patients (suture annuloplasty 3, chordal shortening 1, commissure sutured 7). The hospital mortality was 8% (4 of 49) and there were no late deaths. Before surgery 14% (7) of the patients had severe left AV valve incompetence, 30% (15) had moderate left AV valve incompetence, 35% (17) had mild left AV valve incompetence, and 20% (10) had no left AV valve incompetence. 46 (93.8%) had a satisfactory repair of the valve (mild or no incompetence), and 2 patients required repeat operation for significant residual mitral regurgitation. A further 2 patients required revision of the VSD patch. There were no incidences of heart block. The remodeling of the atrioventricular valves in patients with complete atrioventricular canal defect is a crucial part of surgical repair. Variability in valve morphology is an important factor. Improved understanding of the structural and functional variability of the atrioventricular valve in this lesion has resulted in greatly improved success in repairing the defect.

P746
Doppler and color doppler flow mapping of the patent ductus arteriosus in preterm newborns and clinical findings
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To evaluate the relation between echodopplercardiographic and clinical findings in preterm newborns (PTNB) with Persistent Ductus Arteriosus (PDA), 61 PTN, with mean gestacional age 30+/-2w and mean birthweight 1,2+/-0,2Kg, were prospectively evaluated since the 2nd or 3rd day of life, using serial echodopplercardiographies with CFM examinations in order to determine the PDA diameter at rest, the presence of holodiastolic retrograde flow in the descending aorta (RFAo), the direction of shunting through the PDA. A clinical evaluation to detect the presence of clinical signs of PDA was accomplished concomitantly. The PTNB with PDA were divided into 2 groups: Group A (spontaneous closure of PDA until the 7th day life) and Group B (without spontaneous closure of PDA). Statistical analysis between the two groups was accomplished through the student t test, Mann Whitney and the Pearson's Qui square test. Significance was set as p<0.05. Twenty one PTNB showed PDA(34,4%), being 7 from the Group A and 14 from the Group B. The average diameter of the PDA in Group A was 1,4+/-0,6 mm and Group B 2,1 +/-0,6 mm (p=0,001). RFAo occurred in 14,3% of Group A and in 78% of Group B (p=0,01), and the presence of clinical signs of PDA occurred in 14,3% of Group A and 71,4% of Group B (p=0,013). The direction of shunting through the PDA was left to right in all PTNB, and no DA reopened after its closing. Pharmacological treatment of PDA was accomplished in 10 PTN of Group B, having been successful in 50%. In one a surgical treatment was done and the last 3 which didn't show clinical signs of PDA, died in the 4th day of life with pulmonary hemorrhage. Routine CFM and echocardiographic examinations in PTNB in the first 3 days of life, could detect early the cases in which there is an indication of pharmacologic treatment, even before the presence of clinical signs.

P747
Cardiac manifestations of mucopolysaccharidosis in childhood
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To assess the cardiac change of the mucopolysaccharidosis(MPS), we evaluated the 20 children(age:2-17year-old, male:female=28:2). Results: 1) Cardiac involvement was present in all patients. 2) The main echocardiographic findings were the abnormalities of mitral valve(MV), aortic valve(AV), and ventricular wall hypertrophy. 3) Mitral valve abnormalities were detected in 25 patients(83%). MV thickening without mitral stenosis or mitral regurgitation in 9 patients. MV thickening with mitral regurgitation in 15 patients. MV thickening with mitral stenosis and mitral regurgitation in 1 patient. 4) The AV abnormalities were noted in 16 patients(53%). AV thickening without aortic stenosis or aortic regurgitation in 6 patients, AV thickening with aortic stenosis in 5 patients, AV thickening with aortic regurgitation in 2 patients, AV thickening with aortic stenosis and aortic regurgitation in 2 patients. 5) The tricuspid valve thickening was detected in 1 patient. 6) Left ventricular wall thickening was detected in 8 patients(13%) and interventricular septal thickening in 6 patients(20%). 7) As the type of MPS was mainly type II(21/30)(type I: 3, type III: 3, unknown: 3), it was not possible to see the differences of cardiac manifestations between each type of MPS. Conclusions: Although none of our children with MPS did not have cardiac symptoms, all patients showed various cardiac manifestations. Therefore, cardiac evaluation should be performed regularly in all MPS patients, also we recommend infective endocarditis prophylaxis in MPS patients.

P748
The impact of managed care penetration on utilization of echocardiography among children referred for the evaluation of a heart murmur: A multi-center study
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Background: Managed care plans aggressively seek to contain costs, but few data are available regarding the impact of managed care on utilization of expensive technologies on children undergoing subspecialty evaluation. Methods: To assess the impact of managed care on utilization of echocardiography, records were abstracted for 1,077 children up to 2 years of age newly referred to a pediatric cardiologist for evaluation of a heart murmur at one of four academic tertiary care medical centers. Results: Multivariate analysis revealed that the fraction of managed care patients within an individual provider's case load was a strong predictor of whether a patient would undergo echocardiography (RR=0.85 [0.79-0.91] p<.001 per 10% increase in managed care case load). These results were adjusted for patient age, the overall utilization of echocardiography for the final cardiac diagnosis, the provider's years of experience and whether the provider was a nurse. The reduced utilization of echocardiography was most pronounced among patients with cardiac conditions associated with moderate overall utilization.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

suggesting that differences in utilization are more marked in a clinical setting where the indications for echocardiography are not clearly established. Of note, the individual patient's insurance type was not a significant predictor of echocardiography utilization. Conclusions: Increased managed care penetration was associated with reduced utilization of echocardiography among pediatric cardiologists suggesting that financial incentives do influence utilization of costly resources in settings in which indications are not clearly established.

P749

Quantifying and minimizing radiation exposure during pediatric cardiac catheterization

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Pediatric cardiac catheterization (CC) frequently requires prolonged fluoroscopy time (FT). We evaluated two new technologies: 1) a collimator-mounted dosimeter (DOS) measuring radiation exposure to air, and noninvasive semi-conductor dosimeters (MOSFET), measuring skin dose during CC. 44 patients (pts) (mean 20yr, 28M/16F) underwent CC with PA/lateral fluoro. FT was 1.4–42.2min. High mA 60 fps fluoroscopy (HMF) vs low mA pulsed fluoroscopy (LPF) settings were based on physician preference. The following correlations ($p<0.01$) were identified: 1) FT to DOS and FT to interscapular MOSFET (predicting radiation exposure and skin dose, respectively, based solely on FT and fluoroscopy output settings); 2) using an inverse square relationship, DOS to MOSFET; 3) LPF vs HMF DOS and MOSFET doses. Subsequently 12 pts (2yr–18yr; 5M/7F) were studied during general anesthesia radiofrequency ablation in the RAO/LAO projections with 4 thoracic MOSFETs and a retro-cardiac esophageal MOSFET. FT was 5.4–150.4min. Correlation ($p<0.05$) was noted between 1) FT and the esophageal MOSFET dose, and 2) esophageal MOSFET dose vs left scapular plus left clavicular MOSFET radiation doses. Thoracic MOSFETs outside the direct fluoroscopic field recorded minimal, if any, radiation exposure. Conclusion: 1) DOS or MOSFET can objectively assess CC radiation exposure; 2) minimal (if any) radiation dose is recorded at distant sites not directly in the fluoroscopic field. 3) MOSFETs placed at interscapular, left axillary, and right and left scapular sites can be used during CC to measure radiation exposure during combination of PA/lateral and RAO/LAO projections commonly used in pediatric CC; 4) strategies to reduce radiation should be employed during CC.

P750

Early outcome factors of Jatene's operation for transposition of the great arteries. Study of 120 neonates

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The purpose of this retrospective study was to analyze the perioperative factors involved with cardiac output, acute miocardial infarction or immediate mortality after Jatene's operation. From september 1984 to june 1996, 120 neonates with transposition of the great arteries (TGA) were operated at our institution. Ninety one (75.83%) had simple TGA, twenty-seven (22.50%) had TGA with ventricular septal defect and two (1.66%) had double-outlet right ventricle. Their average age at the time of the operation was 12.58 + 9.17 days (1–43). The average weight at the time of the operation was 3248.91+366.64g (2480 IR–4470.00). As Gittenberger-de Groot's classification, the usual coronary artery pattern was found in 88 neonates (74.57%), circumflex originating at right coronary artery in 18 (15.25%); single left coronary artery in three (2.54%), inverted coronary artery in one (0.84%), inverted circumflex coronary with right coronary artery in two (1.69%), intramural left or left coronary artery between great arteries in five (4.23%); intramural left anterior descending in one (0.84%). Atrial septostomy was performed in 86 (72.88 %). Forty-one of them (34.74%) developed low cardiac output, 10 (8.40%) acute myocardial infarction, and 31 (25.80%) died. By logistic regression the results showed that acute myocardial infarction ($p<0.01$) and aorta cross clamping time ($p<0.01$) were the risk factors contributing to low cardiac output. Likewise failure to perform preoperative atrial septostomy ($p<0.01$) and the cardiopulmonary bypass time ($p<0.01$) were the risk factors to acute myocardial infarction. The cardiopulmonary bypass time in single, inverted or intramural coronaries arteries, taken as a whole, was superior to cardiopulmonary bypass time when the more common patterns were found. Low cardiac output ($p<0.01$), acute myocardial infarction ($p<0.01$) and sepsis ($p<0.01$) were the risk factors to immediate mortality.

P751

Mitral valve disease associated with shone's-like complex: anatomic predictors of severe stenosis

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To assess the incidence and identify anatomic variants predictive of severe mitral stenosis (MS) in Shone's complex, echo studies from all pts with mitral anomalies and LV outflow and/or aortic (Ao) arch obstruction admitted to Children's Hospital of Wisconsin from 1991–97 were reviewed (n=24). All pts had a mitral annular hypoplasia (mean diameter 8.5 +/- 1.2 mm) with restrictive leaflet motion and abnormal papillary muscle architecture at presentation. Five pts (group 1) required surgery for MS at mean age of 3.8 years, 19 pts have not (group 2). All group 1 pts had a supramitral ring (SMR) intimately adherent to the leaflets with a mean Doppler gradient of 12.8 +/- 3.6 mmHg at the time of surgery. None of the group 2 patients had SMR, but all had trivial-moderate MS during infancy (mean gradient 5.8 +/- 2.7 mmHg; range 3–12 mmHg). Coarctation (22/24 pts) and hypoplastic Ao valve (22/24) were common in both groups. Long segment subaortic stenosis (subAS) with discontinuity between the Ao and mitral valves was consistently identified in all group 1 patients with an average Ao-mitral separation of 11.2 +/- 1.4 mm. All group 2 pts maintained Ao-mitral continuity and only rarely had discrete subAS (6/19 pts), none had tunnel subAS. At F/U, three of the group 1 pts have trivial MS (mean gradient 3.2 mmHg) after surgery, the other 2 had severe MS post-op and died following mitral valve replacement in one and conversion to single ventricle physiology in the other. There was no Doppler evidence of MS in 14/19 group 2 pts at a mean F/U interval of 5.1 +/- 2.5 yrs, the other 5 pts had trivial MS. We conclude that surgical intervention for MS with Shone's-like complex is generally associated with SMR during early childhood. It appears that aortic-mitral discontinuity and tunnel subAS occur with SMR, and may also predict severe MS. Mitral annular hypoplasia alone does not predict MS even with parachute-like changes, and valve function frequently normalizes during intermediate F/U.

P752

Effect of steroids on cardiac function of patients with Duchenne muscular dystrophy

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Long term steroids (S) have been proven efficacious in maintaining muscular strength in boys with Duchenne muscular dystrophy (DMD). Significant side effects such obesity, short stature, cataracts, osteoporosis are encountered. The effect of such treatment on cardiac function, a major determinant in the outcome of such patients, has never been reported. A total of 78 pts with DMD were studied. Echocardiographic parameters were compared in 26 boys on S (Deflazacort) for a mean period of 18.8± 3.8 months to 42 boys not treated with S. At time of evaluation pts receiving S were younger 9.9 ± 2.9 years compared to 13.6 ± 3.8. Systolic blood pressure (BP) expressed as Z score was significantly higher in pts receiving S: 1.22 ± 1.36 vs 0.23 ± 1.09, p = 0.001. Shortening fraction (SF) and ejection fraction were higher in pts receiving S: 31.0 ± 5% vs 27.1 ± 6%, 68 ± 7% vs 60 ± 9%, p<0.05. Left ventricle (LV) was dilated in the 2 groups. Z score 2.01 ± 2.45 in pts receiving S vs 1.97 ± 1.69 in pts not treated, p = ns. Dilated cardiomyopathy defined as a SF < 25% and/or LVS 95th percentile for height was present in 19/26 (24%) pts receiving S vs 22/42 (52%) in the others, p=ns. In conclusion, pts with DMD treated with S have a higher BP, their cardiac function appears better, however they are younger. Longer follow-up is needed to evaluate the impact of S on cardiac function in patients with DMD. The issue of a higher BP on a myocardium prone to dilated cardiomyopathy should not be neglected.

P753

Cardiac abnormalities in human immunodeficiency virus (HIV) + infected children with perinatal transmission

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To determine the prevalence of cardiac abnormalities in HIV-infected children with perinatal transmission, 100 children born to HIV-infected mothers, 51 males and 49 females, ages ranging from two months to twelve years, were prospectively studied. Serial clinical examinations, electrocardiograms (ECG), and echodopplercardiograms (ECHO) were performed during a five-year follow-up. The patients were divided into two groups: 1) control – 52 seroe-

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

veners (non-infected); 2) infected group – 48 children. The latter were either asymptomatic (N category – 14.6%) or symptomatic (A, B, C categories – 85.4%). The seroreverters were all clinically normal children with no significant pathological findings in the ECG nor in the ECHO. The infected group displayed the following findings: 1) clinical – fatigue or dyspnea (45.8%), tachycardia (52%), pathological murmur (16.7%), pericarditis with cardiac tamponade (4.2%), and heart failure (27%); 2) ECG: sinus tachycardia (52%), right ventricular conduction delay (18.7%), T wave abnormalities (35.4%), LV overload (12.5%), RV overload (4.2%); 3) ECHO: LV dysfunction (12.5%), LV dilation (12.5%), RV dysfunction (2%), large pericardial effusion (4.2%), mitral regurgitation (12.5%), tricuspid regurgitation (8.3%), and pulmonary hypertension (4.2%). Most of them belonged to the C category (severely symptomatic). We conclude that more than half of the children born to HIV-infected mothers seroreverted. Cardiac involvement was usually a late manifestation of clinically advanced HIV-infected children. The most frequent abnormal cardiac findings were sinus tachycardia, heart failure and T wave abnormalities. Left ventricular dysfunction was not uncommon. Cardiac tamponade was a serious complication of pericarditis.

P754

Electrocardiographic ST-segment depression and functional status in children with Fontan circulation

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Effort limitation found in patients with Fontan circulation could be related to ischaemic myocardial dysfunction. We have previously observed that Fontan patients have electrocardiographic ST-depression during exercise test and 24-hour ambulatory electrocardiographic (24 h ECG) monitoring. The purpose of this study was to investigate if the ST-depressions were correlated to functional status. STUDY GROUP: Forty-five children in two groups were examined. Group A consisted of 15 patients with Fontan circulation and group B consisted of 30 children with a structurally normal heart, matched for length and weight to group A. METHODS: Clinical and echocardiographic examinations were performed in all patients and controls. The clinical state was defined as NYHA functional classes I to IV. All 45 children then underwent 24-h ECG. An analysis of ST-changes was performed using a PC-based Holter System. ST segment-depression of more than 0.20 mV at 51+60 was regarded as significant. Patients in group A were examined 1–8 times (> 5 months between examinations), resulting in 56 recordings. The children in group B were examined once. RESULTS: Twelve of 15 patients analysed had significant 24-h ECG ST-depression in 9 of these patients on more than occasion. Three of these 12 patients were on digoxin at the moment of registration. None of the 30 matched healthy children showed 24-h ECG ST depressions. Comparing 24-h ECG ST-depressions in patients in NYHA I with patients in NYHA II–III showed a significant difference in deep ($p=0.0023$), duration ($p=0.0161$) and number of episodes/24h of ST-depressions ($p=0.0001$). CONCLUSION: ST-depression in daily activity seems to be a common finding in children with Fontan circulation. Repeated ST analysis on ambulatory electrocardiogram shows that ST depressions seem to be correlated to functional status and may be a useful tool in clinical praxis.

P755

Misdiagnosis of aortopulmonary window

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Aim: Aortopulmonary (AP) window usually presents with continuous systolic-diastolic murmur and signs of congestive heart failure. Diagnosis is said to be established by echocardiography. As we recently observed two patients, in whom the AP window was missed, we reviewed all our cases with this rare anomaly in order to analyse the features for a correct diagnosis. Results: Between 1981 and 2000 we saw 15 patients with AP window in our hospital. Only 10 of them were correctly diagnosed before surgical repair. Two patients underwent cardiac surgery (one PDA closure and one corrections of an interrupted aortic arch (IAA)), without having recognized the AP window. In three further patients, the defect was discovered as associated anomaly during corrective surgery of VSD, VSD plus IAA and IAA. Echocardiography had missed the diagnosis in two cases, cardiac catheterization was incomplete in two other patients and PDA closure was done in another hospital after clinical diagnosis. Conclusion: AP window can be found using two-dimensional echocardiography with pulsed and coloured doppler. Nevertheless there is a certain risk of missing the defect, especially when other anomalies are present. Therefore cardiac catheterization and cine-angiography are of major importance to detect AP window in these situations.

P756

The effectiveness of balloon angioplasty for the treatment of aortic coarctation

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The treatment of aortic coarctation is either by surgery or interventional catheterization. The advantages of angioplasty over surgery are: less invasive, shorter hospital stay and less expensive. The aim of this study was to assess the effectiveness of angioplasty without stenting. From June 1998 to July 2000 we treated 18 patients. Twelve were males and six female. Their age ranged from 0.10 to 55 years (mean of 27.5 years). Patients with angioplasty without stenting. All patients had an aortogram in left lateral view performed before and after the procedure. Thus it an observational study. Measurements were taken on 3 aortic segments (pre-coarctation, coarctation and post-coarctation) by two independents observers pre and post-procedure. The balloon/aortic isthmus ratio used was 1.0. The coarcted minimum lumen diameter pre-dilatation ranged from 1.78 a 6.32 mm and pos-dilatation ranged from 2.81 a 11.52mm. The equipment used was a cardiovascular measurement system-medirad, imaging system, version 1.0, 1995. There was no complication related to the procedure itself in our serie, like aneurysm or arterial injury. In conclusion our experience suggests that balloon dilation (angioplasty) is a feasible and safe procedure. In our study the mean increasing in diameter of the coarcted segment was statistically significant ($p<0.001$) by paired t-test. The mean, standard error and 95% confidence interval for the mean difference were respectively: 2.107 mm; 0.569 mm; [1.581; 3.813]. The minimum and maximum increasing were, 0.22mm and 7.88mm respectively.

P757

Nutritional management of chylothorax after cardiac surgery in children

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Chylothorax is the presence of triglyceride-rich lymphatic fluid in the pleural space resulting from a leak of the thoracic duct or one of its major divisions. The main causes of chylothorax are tumors, trauma and others. Usually postoperative chylothorax occurs after traumatic cardiothoracic procedure. Early nutritional therapy is important to avoid surgical intervention. This report describes 6 children (4 M, mean age 2.5y, range 15 days-10 years) who developed chylothorax following cardiac surgery and analyze the efficiency of nutritional therapy. Patients received medical treatment and enteral hyperprotein diet containing isolated proteins, medium-chain triglycerides (MCT) and no long-chain fat or alternatively elemental diets. In either case, vitamins and mineral supplements were added. All patients well increased clinical treatment. Four children had linfopenia and pericardial drainage and 5 had bronchopneumonia. The range of weight lost was 10%. There was no need for surgical intervention. The mean chylous drainage volume was 27 ml/kg/day for a mean period of 32.5 days. Seven patients received enteral nutrition and partial parenteral nutrition for 7 days. Only 1 patient received total parenteral nutrition for 2.5 weeks. In conclusion, all children well succeeded medical treatment with drainage of chylothorax and early nutritional therapy. The characters of enteral diets should always be hyperprotein, with MCT, mineral and vitamin supplements.

P758

Effect of prenatal diagnosis on one year neurologic and developmental outcomes in D-TGA

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By decreasing infant perioperative morbidity, prenatal diagnosis could improve neurodevelopmental outcomes in infants with critical congenital heart disease (CHD). Our purpose was to explore the impact of prenatal diagnosis on outcomes at age one year in children with CHD. We analyzed a database of children enrolled in perspective studies on surgical support techniques from 1988 to 1998. Selection criteria included a diagnosis of D-TGA, primary arterial switch procedure in early infancy, birth weight > 2.0 kg, no associated cardiovascular anomalies requiring additional procedures and no extracardiac congenital anomalies. Of 264 patients with D-TGA at enrollment, 15 had a prenatal diagnosis and 249 did not. Those with prenatal diagnosis, compared to those without, did not differ significantly in birth weight, gestational age, Apgar scores, preoperative intubation, preoperative acidosis, minutes of circu-

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

latory arrest, or total support time. At age one year, 213 patients returned for testing by neurologic examination and the Psychomotor Development Index (PDI) and Mental Development Index (MDI) of the Bayley Scales (1969 version). Although the differences were not statistically significant, children with prenatal diagnosis, compared to those without, had fewer possible or definite abnormalities on neurologic examinations (18% vs. 30%, P = .10) and fewer scores < 80 on the PDI (0% vs. 8%, P=0.02). Mean scores in those with and without prenatal diagnosis for the PDI (97.1 ± 12.5 vs. 96.5 ± 16.1) and MDI (108.4 ± 19.6 vs. 106.2 ± 14.4) were similar. Although these data suggest that prenatal diagnosis may reduce the incidence of neurologic abnormalities and severe impairment of motor function at age one year, our power to detect differences was limited by the small sample of patients with prenatal diagnosis. Further studies should include a greater number of children with prenatal diagnosis and a variety of congenital heart lesions.

P759

Giant ascending aortic aneurism in children

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Aortic aneurysm is rare in the pediatric age group. However, it is most commonly found in patients with Marfan syndrome. When aortic diameter is larger than 4 cm, the incidence of sudden death is high. Objective: To show the diagnosis and evolution of an aortic aneurysm in a child referred to Pequeno Principe Hospital. Case report: a 10-year-old boy was referred with a month's history of chest pain, heart murmur and faint started at school. He had normal height and weight, and was mentally normal. His neck examination showed visibly pulsating mass. The arterial heart sound was muffled, there were systolic and diastolic murmurs on the right external border, rate and rhythm were regular. Peripheral pulses were equal in four extremities and were bounding. An electrocardiogram showed a normal sinus rhythm without ectopy. A chest radiograph showed an abnormal dilation of the aorta. An echocardiogram revealed a grossly dilated aortic root and an aneurysm flap extending till the transverse aortic arch and severe aortic and mitral valve regurgitation. An angioresonance showed an aortic aneurysm, with 80 mm in transverse and 130 mm in area. The patient underwent a resection of the aneurysm, implantation of the aortic valve and ascending aorta. The diagnosis of Marfan syndrome was suggested by aortic biopsy. The postoperative course was uneventful. On the 10th day he was discharged in good condition, with mild mitral regurgitation. A one year follow-up, showed him to be asymptomatic. In a review by El Habbal, 4.3% of Marfan patients had serious complications by the age of 20, and with a negative family history of Marfan syndrome had more tendency for serious cardiovascular complications early in life. Conclusion: Early surgery in aneurysm is the best choice for a good follow up.

P760

Chest pain in children

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Chest pain is an alarming complaint in pediatric patients and often leads to a frightened family admitting to a pediatrician or pediatric cardiologist. Chest pain in children is rarely due to cardiac disease and usually the etiology is obscure. One hundred eighty five patients, 99 girls (53.5 %) and 86 boys (46.5 %) with chest pain were evaluated prospectively in our hospital. The girls were between 5 and 16 years old (mean 14) and the boys were between 6 and 15 years (mean 9). Chest pain was present between 6 months to 2 years in 137 (74 %) patients. Localization was on the left precordium at 149 (80.5 %) patients. The reason of chest pain was idiopathic in 123 (66.4 %) patients. Psychiatric, cardiac, gastrointestinal, musculoskeletal and pulmonary causes were present in 25 (13.5 %), 20 (10.8 %), 7 (3.7 %) 5 (2.7 %) and 3 (1.6 %) patients, respectively. One patient with lymphadenosis and one with chalcosis mellitus had chest pain.

P761

Discrepancies in parent and child health reports for children with heart disease

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Purpose: We sought to compare child versus parent reports of quality of life for children attending a general cardiology clinic, using the Child Health Questionnaire (CHQ-PF50, CHQ-CF87). Methods: Children ages 10–18 years without additional chronic illnesses and their parents completed the CHQ instrument. After data collection, children with similar conditions were grouped together. Five groups were analyzed: innocent murmur/trivial structural heart disease, supraventricular tachycardia (SVT) and 3 with structural heart disease (no minor, major interventions). Within group means were compared for children and parents separately. Results: The 5 groups included 127 patients ages 10 to 18.9 years (median = 14.7 yr). Among the 5, no differences were found for parent or child reports for physical functioning, behavior, mental health, family cohesion, and family activities. Differences or trends were found among cardiac groups for the following subscales: role/social limitations-physical (p= .05, parent) and more bodily pain/discomfort (p=.08, child), self esteem (p=.04, parent), and general health perceptions (p= .004, parent). The greatest differences were in parent reports of general health perceptions. However, interesting differences were noted between child and parent reports, especially for SVT and major cardiac interventions. In comparison to those with innocent murmurs/trivial structural heart disease, children with SVT reported more role/social limitations (difference between means=8.9) and bodily pain/discomfort (16.7) than parents indicated (4.7, 10.4 respectively). In contrast, children with major cardiac interventions reported less role/social limitations-physical (7.7), higher self-esteem (2.7), and higher general health perceptions (7.0) than parents (-5.3, 10.6, 21.4 respectively). Conclusion: Children with heart disease and their parents generally report similar health related quality of life. However, children with major interventions report fewer limitations than parents, and children with SVT report more discomfort.

P762

Hemodynamic response to isoproterenol infusion in children with valvular aortic stenosis

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The role of dynamic exercise testing in the assessment of children with aortic stenosis (AS) is well established, however it is inconvenient for small children and hemodynamic data acquisition during treadmill testing is technically difficult. To assess the response to an equal cardiac workload, isoproterenol infusion was administered to 14 patients with valvular AS aged 3–11.5 (mean 6.5±4.9) years, in incremental doses starting at 0.02mg/kg/min in 2–3 stages, while hemodynamic parameters were monitored by echocardiography. Eleven of 14 patients aged >4 years underwent treadmill exercise testing: endurance time was normal in all. Symptom, ST-T changes developed in 25%, abnormal heart rate response to exercise was noted in 25% of patients with severe AS (transvalvular Doppler mean gradient >40 mmHg, peak gradient >75 mmHg), but in none of the patients with mild-moderate AS. Abnormal blood pressure responses were documented in 43% and 75% of patients with mild-moderate and severe AS respectively. During isoproterenol infusion, significant increases in transvalvular mean (p=0.003) and peak (p=0.006) gradients, heart rate (p=0.005), systolic blood pressure (p=0.04), and significant decreases in diastolic blood pressure (p=0.007), LV diastolic volume index (p<0.01) were observed, stroke volume index failed to increase and CI, ejection fraction, fractional shortening increased insignificantly. no complications occurred. Negative correlations between the baseline mean gradient and increase in systolic blood pressure (r=-0.60), and increase in mean gradient (r=-0.66) were documented. Failure to increase the mean gradient by >50% the baseline value in response to isoproterenol had 80% sensitivity and 100% specificity for predicting severe AS. Isoproterenol infusion is a safe and practical means of assessing the response to increased cardiac workload in AS, allowing echocardiographic evaluation of hemodynamic alterations, which is difficult to accomplish during dynamic exercise testing.

P763

Dexamethasone given to premature infants and cardiac diastolic function in early childhood

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This study examined if dexamethasone (DEX) given to premature infants with bronchopulmonary dysplasia (BPD) resulted in cardiac diastolic dysfunction in early childhood. A neonatal rat model postulated DEX induces myocardial hypertrophy with fibrosis that may cause long term diastolic dysfunction. This has never been examined in animals or humans. We compared 7 children aged 3–6 years born at 26 weeks gestation and given DEX for BPD with 7 age and gestation matched controls using echocardiography to assess systolic and diastolic function parameters. All DEX patients had hypertrophic cardiomyopathy (HCM) that had resolved. Results: DEX patients had the same normal E/A and IVRT (24.9 ± 2.8 and 54.6 ± 6.3 ms) as controls (22.1 ± 3.0 and 47.3 ± 5.5 ms). Peak A velocities were the same in

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

DEX patients as controls 459 ± 15 vs 494 ± 56 cm/s, p=0.10) resulting in unchanged E/A ratios (1.89 ± 0.57 vs 2.15 ± 0.43, p=0.22). Peak E velocity and E-wave deceleration times were not different. We found no significant differences in systolic function parameters (VCFc, wall stress, ejection fraction). LV mass was the same between the groups confirming resolution of HCM. Conclusion: This data is consistent with normal myocardial relaxation suggesting long-term diastolic dysfunction does not exist in children who received dexamethasone as premature infants with regression of HCM.

P764

Role of the Doppler mean systolic pressure gradient in the assessment of severity of valvular aortic stenosis in children

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To assess the usefulness of the Doppler transvalvular mean systolic pressure gradient (PG) in evaluating the severity of valvular aortic stenosis (AS) in children, 35 patients with valvular AS aged 45 days-14.5 years (mean 7.9±4.8) were prospectively analyzed. Symptoms, physical findings, ECG, chest x-ray, treadmill exercise test results (11 patients), echocardiographic measurements of LV end-diastolic diameter, ventricular septal and LV posterior wall thickness, LV mass, ejection fraction, fractional shortening were evaluated and graded so that a stenosis score reflecting the clinical severity of AS was obtained for each patient. The mean and peak-instantaneous PG values measured by 2-dimensional color flow Doppler echocardiography correlated significantly with stenosis scores (r=0.70 and 0.60, respectively). The meanPG values at 95% confidence level that corresponds to mild, moderate, severe AS classified both according to stenosis scores, and transvalvular Doppler peakPG (<50 mmHg, 50–75 mmHg and >75 mmHg) were <25 mmHg, 25–40 mmHg and >40 mmHg, respectively. The sensitivity and specificity of a Doppler meanPG>40 mmHg for predicting the presence of symptoms were 100% and 68%, LVH on ECG 87% and 100%, ST-T changes at rest or on exercise 100% and 80% respectively. A Doppler meanPG>40 mmHg was highly sensitive (100%) and specific (100%) for predicting the need for intervention. Although these findings need to be verified in larger patient groups, the range of Doppler meanPG values obtained in this study that corresponded to mild, moderate, severe AS may serve as a useful guideline in estimating the severity of AS in children.

P765

Clinical profile of 2,294 children referred with cardiac murmur

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Objective: The study is to analyze the clinical profile of patients with cardiac murmur referred to a tertiary center of Pediatric Cardiology. Method: this is a retrospective analysis of 2,294 children, obtained during January 1995 – December 1997. The children were divided into two groups: normal group – 1,292 children (mean age 59.5 months); abnormal group – 1002 patients, mean age 8 months, with cardiac anomalies. All patients of abnormal group were submitted to echocardiography. For analysis, all the children were divided into 5 subgroups: I) 0 to 30 days; II) 31 days to 2 years; III) 3 to 6 years; IV) 7 to 12 years and V) 13 to 16 years. Results: There was predominance of normal children, in subgroups II and IV, 893 (69.1%). In the abnormal group, children were more prevalent in subgroup I and II, 678 (67.7%). Weight and age showed significantly lower in the abnormal group. Ventricular septal defect was the more prevalent anomaly. Complex cardiopathies were more prevalent in subgroups I (22.8%) and II (12.0%). Associated anomalies were found in all abnormal subgroups, with frequency of 12% – to 27%. Isolated anomalies were more frequent in older children. Three hundred seventy six cases (37.5%) were submitted to surgery. Sixty two patients (6%) died. Conclusion: The great majority of children with cardiac murmur have normal hearts. Pathologic heart murmurs is more prevalente in children of less than 2 years of age.

P766

Distal pulmonary artery growth in patients with pulmonary atresia/ventricular septal defect and tetralogy of Fallot: Does proximal pulmonary artery diameter play a role?

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BACKGROUND: Aggressive augmentation of the proximal pulmonary arteries is thought to improve distal pulmonary artery growth in patients with pulmonary atresia/ventricular septal defect (PA/VSD) and tetralogy of Fallot (TOF). We hypothesized that distal pulmonary artery size remains significantly smaller in these patients (pts) regardless of proximal pulmonary artery diameter. METHODS: Twenty-six pts with PA/VSD or TOF were compared to 25 control pts catheterized prior to the Rastelli procedure. Pulmonary artery diameters were measured distally after the takeoff of the upper lobe branches and proximally, at the narrowest diameter between the bifurcation and the upper lobe branches. Pt data were obtained from catheterization reports. Unimpaired samples t-tests were used to compare means. Statistical significance was defined as p<0.05. RESULTS: There were no significant differences between pt age (8.5 +/- 5.4yrs vs 11.2 +/- 5.9, p=ns), weight (36.2 +/- 22kg vs 40.9 +/- 14.2, p=ns), or BSA (1.55 +/- 0.94m2 vs 1.68 +/- 0.33, p=ns) between experimental and control pts, respectively. Distal pulmonary artery diameters were significantly smaller (RPA 11.2 +/- 2.6mm vs 15.6 +/- 2.8, p<0.001; LPA 12.1 +/- 3.1mm vs 15.5 +/- 2.7, p<0.001), while proximal pulmonary artery diameters were similar (RPA 15.5 +/- 8.6mm vs 18.8 +/- 3.1, p=ns; LPA 16.4mm +/- 8.3 vs 18.5 +/- 3.3, p=ns) between the groups. Right ventricle to systemic blood pressure ratios were significantly higher in pts with PA/VSD and TOF (0.48 +/- 0.1 vs 0.28 +/- 0.07, p<0.001). CONCLUSIONS: Distal pulmonary artery size in adult pts with PA/VSD and TOF was significantly smaller compared to controls. Normal distal pulmonary artery growth did not occur despite normal proximal pulmonary artery diameters. Further studies are needed to determine the factors associated with distal pulmonary artery growth.

P767

Ebstein's anomaly (EB): factors associated to poor outcome

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Ebstein's anomaly (EB): factors associated to poor outcome. Mota, C.C., Righy, M.L., Coutinho. To investigate EB and identify factors of poor outcome (hospitalization and surgery), 27 consecutive patients were selected between 1991–1998. Their ages (mean=28±7.4yr) ranged from 1d to 22y and the follow-up mean was 6.1±4.9y. Besides a retrospective investigation of clinical data, a blind analysis of ECHO, CATH, surgery and autopsy, considering the associated cardiac anomalies (ACA), anatomic type of AV and VA connections, RA, RV and LV sizes; mobility, displacement, dysplasia, type of attachment and morphology of TV leaflets. The frequencies of inpatients, surgeries and deaths were of 84%, 76% and 58% respectively. SVT was found in 52%. In group 1 (severe ACA), the presence of ACA constituted an associate factor to surgery, but not to death. In group 2 (mild or without lesions) upsetted LV, TV dysplasia, reduced leaflets and dilatation of RA were associated with statistically significant increase of invasive interventions: desaturation was identified as a risk factor of hospitalisation among neonates. This study provides a method to identify the factors associated to poor outcome and describes the marked heterogeneity of presentation.

P768

Cost implications of closure of atrial septal defects

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Objective: We sought to evaluate the relative cost of surgical and device closure of atrial septal defect. Background: Device closure for atrial septal defects is becoming an alternative to surgical closure. Methods: We examined the hospital-generated cost data in 82 patients who underwent surgical repair and 35 patients who underwent device closure of an ASD or patent foramen ovale (PFO) during a prospective clinical trial of the device. Results: The cost of device closure of ASD was $7837 less on average than surgical closure when the cost of the occlusion device was excluded (device closure cost $7397 – $2822 surgical closure cost $15234 + $3851, p < 0.001). When adjusted for a 5% failure rate of device closure, the cost savings was $7076. Conclusions: Device closure of ASD results in substantial hospital-related cost savings which will be an important consideration once new devices are approved for clinical use.

P769

Annuloaortic ectasia in children – a call for these cases

Oporto, V.L., Silva, C.M.C. Gomes, L.F.C., Neto, R.P., Melo, P. Carvalho, A.C.C., Andrade, J.C., Catani, R., La Rosa, C.A., Maluf, M.A., Buffolo, E.
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https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Annuloaortic ectasia in children – a call for three cases. Oporto, VT, Silva, CMC, Gomes, LFG, Nero, RP, Brito, F, Carvalho, ACC, Andrade, JC, Catani, R, La Rosa, C A, Maluf, MA, Bulbula, E Universidade Federal de São Paulo / EPM – São Paulo -Brasil Aneurysm of ascending aorta is a rare entity on children. The annuloaortic ectasia in children tend to progress rapidly with the end results of high morbidity and mortality and almost always require surgical intervention during the first decade of life, although surgical treatment in these cases have been rarely reported in literature. Between June / 1997 and November / 2000 3 cases of annuloaortic ectasia were diagnosed. The age ranged from 9 months to 14 years and their weight from 8 to 36 kg. Four of them had Marfan Syndrome (MS) and 1 had a bicuspid aortic valve as risk factor. The patients with M S have surgical indication and are in waiting list for surgery. The patient with bicuspid aortic valve underwent surgery for replacement of the aortic root and aortic valve (Bentall & de Bonno surgery) which was successful. Although rare in children this disease carries a major risk for dissection and rupture of aortic root for that reason surgical intervention is mandatory.

P770

QTc dispersion in children with ischemic heart disease: a study of infants with anomalous left coronary artery originating from the pulmonary artery

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Introduction: Increased QTc dispersion has been associated with adult cardiac ischemia. Children with anomalous left coronary artery originating from the pulmonary artery (ALCA) may develop ischemia secondary to a steal phenomena from flow reversal, often with an infarction pattern on ECG. Methods: QTc dispersion was measured in 14 infants with angiography confirmed ALCA and 10 age matched controls. We compared our data with the previously reported ECG findings suggestive of ALCA. These include: Q waves in lead I, aVL, V3-V6, abrupt loss of R wave in the mid-precordial leads, and T wave inversion in I, aVL, V3-V6. Results: Patients with ALCA demonstrated a QTc dispersion significantly lower than that of age matched controls (12.8 +/- 2.9 msec vs. 15.6 +/- 1.9 msec, P< 0.004). The most sensitive criteria was QTc > 12.5 msec in 11/14 (78%) patients. A Q wave in aVL was seen in 7/14 (50%) of patients. A Q wave in aVL and/or QTc dispersion > 12.5 msec, identified all patients with ALCA. In normals, 9/10 had no Q wave in aVL in QTc dispersion values <12.5 msec. Conclusions: Infants with ALCA demonstrated significantly lower QTc dispersion than normals. With a Q wave in aVL, sensitivity and specificity are > 90%. Therefore, this may be a useful screen for a suspected coronary artery abnormality in infants. Additionally, this suggests a mechanism for ischemia unlike that in adults. This may be from different autonomic function, tissue substrates, or etiologies of ischemia.

P771

Electrocardiogram interpretation and management in a pediatric emergency department

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The primary objective is to compare ECG interpretation between pediatric emergency staff and a pediatric cardiologist. Secondary objectives include the determination of inter- and intra-rater reliability. This prospective study involved the evaluation of electrocardiograms ordered by emergency department(ED) physicians at the Alberta Children's Hospital between January and July 2000. Electrocardiograms were analyzed according to indication, rate, rhythm, QRS-axis, intervals, morphology, and normality. If abnormal, it was determined whether cardiology follow-up was warranted. Electrocardiograms were re-distributed to the ordering physician, a second ED physician, and a pediatric cardiologist. Inter and intra-rater reliability was assessed by kappa (K) statistic. One hundred and twenty ECG's were ordered by ED staff during the study period. At this time 77 of the inter rater, and 92 of the intra-rater, ECG's have been reviewed. Preliminary data reveals that 16.9% of the original ECG's were determined to be abnormal compared to 23.4% and 22.3% for the inter- and intra-rater comparisons. The measure of agreement was .344 and .553 for inter-rater and intra-rater comparisons respectively. Acutely in the emergency department it was felt that 10.8% of the ECG's warranted referral to a pediatric cardiologist compared to 16.9% and 16.3% of inter-rater and intra-rater comparisons. The measure of agreement was .515 and .465 respectively. All comparisons were significant at p< 01. Preliminary results indicate that significant inter rater and intra-rater variability exists in the evaluation of ECG's by ED physicians. Some variability can be explained by the lack of clinical correlation during the inter- and intra-rater comparisons. Future addition of the cardiologist's evaluations will assist in determining the clinical significance of this variability. In addition the cardiologist's evaluations will help to assess the accuracy of the ED physician's ECG evaluation, and the appropriateness of referral.

P772

The results of a 16-week exercise rehabilitation program in children with post-operative congenital heart disease

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Purpose: To evaluate the benefits of 16 weeks of exercise rehabilitation in children with post operative congenital heart disease. Methods: Thirty-four patients (TET=22, TGA=6; FONTAN=3) volunteered to participate in the program. Subjects were divided into Rehab (n=17: TET=10, TGA=4, FONTAN=3) and Control (n=17: TET=12, TGA=5) groups. All but one patient completed the program. Exercise tolerance, hemodynamics, and cardiac function were assessed before and after 16 weeks of individualized exercise prescription using semi-supine cycle ergometry and echocardiography-Doppler. Rehab patients participated in formal exercise programs twice per week for the first 7 weeks and 3-4 times per week for the remainder of the program. Control patients were asked to participate in normal daily activities. Results: There was no difference in the age, height, weight, or BSA of the two groups of patients. There were no differences in baseline measures of total work, CI (7.9 vs 7.53 l/min/m), SVI (54.3 vs 50.9 ml), VE (42.8 vs 43.1 L/min), VO2 (1.03 vs 1.25 L/min), or HR (169.9 vs 158.6 bpm) at maximal exercise between groups. Following 16 weeks of exercise rehabilitation, the Rehab group increased its VO2 by 12.4% (p>0.05) and total work by 28.4% (p<0.02). Comparative values in the Control group remained unchanged. All of the patients had abnormal segmental wall motion at rest and during exercise. Conclusions: Children with post-operative congenital heart disease can benefit from a 16-week supervised exercise rehabilitation program. While motivation, scenarios, and rewards may vary from traditional adult rehabilitation programs, the concepts of frequency, intensity, and duration remain constant and are important determinants of the overall success of a pediatric program.

P773

Coarctation of the aorta: Blood pressure response and hormone analysis assessed in a standard exercise test

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15 patients (mean age 22 ± 11 years) with repaired coarctation were studied to elucidate hypertension, recoarctation, BP response and hormone response to exercise. The methods included treadmill exercise testing, MRI, blood pressure (BP) measurements (arm and leg) and blood tests for hormones. Gas exchange parameters and hormones were obtained using a modified Bruce protocol. Results: Thirty-seven patients (57 %) had hypertension at rest. Eight patients had a resting arm-leg BP gradient of more than 35 mm Hg (recoarctation). Systolic BP at rest was 128.4 ± 15.7 mmHg and 145.2 ± 20.1 mmHg in patients repaired before and after age/ear of age, respectively (p<0.005). Maximal oxygen consumption was 36.2 ± 7.9 ml/kg/min and 41.2 ± 10.3 in the two groups (NS). Table 1 BP and heartrates after exercise in relation to arm-leg BP gradient at rest are presented. In conclusion, hypertension at rest is a significant problem after coarctation repair. Surgery in infancy seems to give less long term hypertension and recoarctation. The BP gradient seems to be related to catecholamines rather than to renal hormone response. We estimate that more than 50 % of our patients will need intervention with balloon dilatation, medical treatment or both. Life-long follow-up is warranted.

P774

Correlation between increase of exercise systolic blood pressure and selected parameters of obesity in children after the surgical treatment of aortic coarctation

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Following ambulatory blood pressure monitoring (ABPM) parameters like load (SBPL and DBPL) and normal load (SBPF and DBPF) for systolic and diastolic blood pressure may indicate a risk of arterial hypertension. The aim of the study was to asses the correlation between mentioned above ABPM parameters and increase of systolic blood pressure in treadmill stress test among children after the surgical repair of the aortic coarctation. 16 children

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

(11 boys and 5 girls) in a mean age 12.37 years (SD±4.02) participated in this study. Mean time since the surgical treatment was 7.44 years (SD±3.53). All of children were operated with good effect. In all of children ABPM with SBPL, DBPL, SBPF and DBPF analysis was performed. Moreover the treadmill stress test with appraisal of systolic blood pressure increase between the rest and peak of the exercise (dESBP) was performed. Increase of systolic blood pressure during the stress higher than 60 mmHg indicate exercise hypertension. The correlation, meant by of Pearson correlation index (R), between dESBP and mentioned above ABPM parameters was assessed. Mean value of dESBP in examined group was 43,13 (±21,93) mmHg. Exercise hypertension was diagnosed in 4 cases (25%). The R value for each parameter was presented in the table 1. Table 1. Conclusions: 1. In children after the surgical repair of aortic coarctation significant correlation between SBPL and exercise increase of systolic blood pressure was noted. 2. In the case of DBPL, SBPD and DBPD no such correlation was found.

Cardiac Nursing

P775

One year outcomes of a nurse-driven pediatric anticoagulation program

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One year outcomes of a nurse-driven pediatric anticoagulation program. To study the impact of a nurse-run warfarin program, a protocol-based approach was used. Data included 49 patients with a mean age of 8.6 years resulting in 647 lab encounters, and diagnoses of mechanical valve (n=18), s/p Fontan (n=17), Kawasaki/aneurysms (n=8), and other (n=6). The INR value was elevated in 34 determinations (5%), and low INR values were present in 46 encounters in patients with prosthetic heart valves (7%). Compliance with lab encounters was 94%, with only 8 encounters delayed >1 week. No patients were lost to follow-up. There were no hemorrhagic events and one patient developed a left ventricular thrombus despite consistent INR results within target range. Prior to this program, 8 MD's, 5 R.N's, and other support personnel were involved. With the established protocol, one R.N is running the program, with MD consultation as needed. A nurse run protocol-based anticoagulation program for children can improve patient care, compliance, resource utilization, and may result in reduced complications.

P776

Withdrawal symptoms after cardiac surgery

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Providing children with effective relief from pain and anxiety following cardiac surgery is a standard practice, however, withdrawal symptoms (WS) from this medication may delay the patients' recovery. Between 9/99 and 02/00, 18 patients under the age of 3 years underwent cardiac surgical treatment using cardiopulmonary bypass. All children received continuous Midazolam infusion with either Fentanyl (6pts) or Morphine. In 12 patients the chest was left opened for 24–72hours postoperatively. Clinically significant WS were present in 9 patients (50%). The cumulative dose of Midazolam was higher in the group of children with WS (mean 113mcg/kg, SEM 30 vs 71mcg/kg, SEM 36, p=0.04) and WS were associated with the longer duration of Midazolam treatment (in all 5pts treated longer than 3 days - 100%- and only in 4/13pts -31%- treated for less than 3 days). Despite the cumulative dose of Midazolam being higher in the children with opened chest (mean 119mcg/kg, SEM 32) when compared with the patients where the chest was closed at the time of operation (38mcg/kg, SEM 17, p=0.02), the incidence of WS was the same (50%) in both groups. WS were present in 7/8 patients given Fentanyl (87%) and only in 2/10 patients given Morphine (20%). No difference was found in the cumulative dose of Midazolam given to the patients on Fentanyl infusion (mean 101mcg/kg, SEM 39) when compared with the patients receiving Morphine (mean 85mcg/kg, SEM 24, p=0.2). Midazolam infusion given longer than 3 days following cardiac surgery was associated with high incidence of clinically significant WS in our Unit. The concomitant use of Fentanyl may exacerbate the postoperative complications.

P777

Perceptions of illness severity and distress among mothers of children undergoing cardiac surgery

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According to the research literature on children with serious physical illness, maternal perception of the child's illness is a stronger predictor of the child's emotional adjustment than the clinician's assessment. However, it has also been reported that mothers' perceptions of their child's illness are related more to their own levels of distress and to the marital relationship than to objective disease severity. In the present study, assessments were carried out on a group of 54 families of children requiring cardiac surgery, before and 12 months after surgery. Mothers and physicians made ratings of disease severity and mothers' levels of distress were assessed using the General Health Questionnaire. The adjustment of the children was assessed using previously validated instruments. Preoperatively, there was poor agreement between mothers' and physicians' ratings of illness severity. Levels of maternal distress correlated significantly with physicians' ratings of illness severity, but not with those of the mothers themselves. Neither mothers' nor physicians' ratings of illness severity correlated with the children's preoperative adjustment. Postoperatively, significant correlations were found between mothers' and clinicians' assessments of the children's physical state. Mothers' ratings, but not those of the clinicians, correlated with adjustment of the child at follow-up. Preoperative maternal distress was the strongest predictor of maternal distress at follow-up irrespective of the child's physical state and adjustment. The strongest predictor of adjustment of the children at follow-up was their preoperative adjustment, regardless of the medical variables. The results indicate the complexity of mothers' appraisals of their ill children, and the potential value for overall clinical management of a deeper understanding of mothers' beliefs and perceptions.

P778

An evidence-based nursing approach to meeting the needs of children with congenital heart disease and their families and associated professionals

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A recent shift in emphasis from hospital to home care for children with congenital heart disease has added to parental responsibilities in the home. This has increased the need for children's cardiac liaison nurses from specialist centres to build networks of communication between the family, health and other care providers. This exploratory study aimed to determine the needs of children, families and professionals and to inform the development of an innovative cardiac liaison nursing service. Postal survey of families (n=207) indicated that daily activities of 98 children (47.3%) were affected by their heart disease with 90 (42.2%) perceiving their child as different from healthy peers. Since diagnosis 171 (82.1%) reported difficulties. Ninety one respondents (43.2%) required more information and 26.7% felt their healthy children were affected. Paediatricians (n=47) and senior nurses (n=22) from referring hospitals (n=34) reported a high level of satisfaction although areas requiring attention were arrival time of discharge summaries, procedure for liaison at discharge and provision of a keyworker. Telephone interviews with children's cardiac liaison nurses (n=9) revealed that seven undertook home visiting. Interventions included: psycho-social support (n=6), teaching (n=5), bereavement support (n=4) and practical care (n=4). Six nurses ran nurse-led clinics. Difficulties experienced were lack of time and travelling on a large geographical area. Autonomy, flexibility and recognition of family global needs were valued aspects of the role. These findings support the need for systematic provision of new services. A pilot randomised controlled trial evaluating a nursing intervention package at home is currently underway. This will influence the expansion of an evidence-based nursing service, enable the development of a model for good practice, contribute to the modernisation of cardio-thoracic practice and influence policy in transitional care from hospital to home.

P779

Technology and practice: designing a comprehensive paediatric monitoring system

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https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

The Patient Monitoring Practices Project was established in January 2000 with a mandate to examine and address continuous electronic monitoring of vital physiological parameters, including ECG, heart rate, respiratory rate/apnea and oxygen saturation. The scope of the Monitoring Project included: (i) examination of current monitoring practices throughout the hospital, (ii) examination of how geographical and space issues affect staff's ability to monitor patients safely and effectively, (iii) determination of monitoring equipment needs, (iv) development of recommendations to address systemic issues identified, (v) included in the recommendations should be the development of new patient monitoring criteria, policies, guidelines and procedures to address the practice issues, and (vi) implementation of all recommendations. Thorough assessments were conducted in all inpatient and ambulatory areas, using a variety of information gathering mechanisms. Literature searches and extensive benchmarking surveys revealed only modest amounts of useful information that contributed to recommendation development. The Monitoring Project yielded a comprehensive report that included twenty-four recommendations regarding: comprehensive implementation of criteria/policies/guidelines, enabling electronic order sets reflected new practice guidelines, investing significantly in education programs to raise the knowledge and skill level of staff throughout the hospital regarding monitoring and data interpretation, purchasing monitors to ensure consistency of equipment across the inpatient units; overcoming the geographical issues through use of advanced alarm annunciation technology; and implementing a structured quality management process to ensure that monitoring practices are safe, effective and appropriate.

P780

The use of propofol for early extubation in children

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In the Critical Care Unit at the Hospital for Sick Children, Propofol is used in the post-operative management of children who have undergone cardiac surgery. Propofol as an anesthetic agent, with its swift onset of action and rapid clearance from the blood, expedites early extubation of these children in the immediate post operative period. Propofol's anti anxiety property decreases narcotic requirements without compromising the child's comfort. Early extubation promotes a shortened length of stay in the CCU and allows the child to resume his/her normal activities sooner, thereby reducing the trauma of the surgical experience. For specific surgical repairs, such as the Fontan and the Bidirectional Cavopulmonary shunt, spontaneous ventilation promotes/supports hemodynamic stability. This poster will present a profile of Propofol that includes pharmacokinetics as well as benefits and risks. An algorithm showing inclusionary and exclusionary criteria will be presented, as well as a consideration of pertinent nursing implications. In the Critical Care Unit at the Hospital For Sick Children, Propofol is used in the post-operative management of children who have undergone cardiac surgery. Propofol as an anesthetic agent, with its swift onset of action and rapid clearance from the blood, expedites early extubation of these children. In the immediate post-operative period, Propofol's anti-anxiety property decreases narcotic requirements without compromising the child's comfort. Early extubation promotes a shortened length of stay in the CCU and allows the child to resume his/her normal activities sooner, thereby reducing the trauma of the surgical experience. For specific surgical repairs, such as the Fontan and the Bidirectional Cavopulmonary shunt, spontaneous ventilation promotes/supports hemodynamic stability. This poster will present a profile of Propofol that includes pharmacokinetics as well as benefits and risks. An algorithm showing inclusionary and exclusionary criteria will be presented, as well as a consideration of pertinent nursing implications.

P781

Reducing early mortality after the norwood procedure: integrating research into nursing practice

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Purpose: Early survival after the Norwood procedure has improved significantly in recent history. This study examined nursing care strategies for post-op Norwood patients that have coincided with improved early survival. Methods: Review of a comprehensive single ventricle database was undertaken to explore trends in morbidity and mortality as well as changes in medical and nursing practice. Nursing education strategies implemented as a result of these practice changes were also reviewed. Results: Data from 160 consecutive Norwood procedure patients were reviewed. The series was divided into 2 groups: Group A, before July 1996 (n=36) and Group B, after July 1996 (n=64). Hospital survival in the more recent group was significantly better, 91% vs 53% (p<0.001). In Group B, no deaths occurred in the first 7 days post-Norwood compared to 12 deaths in the early post-op period in Group A (p<0.001). Several changes in practice independently demonstrated an impact on early survival: a change in the isotope delivery system eliminating the need for bolus changes in the early post-op period, continuous SVO2 monitoring allowing a management strategy targeting systemic oxygen delivery rather than a specific arterial saturation, and use of complete alpha blockade to reduce systemic vascular resistance (SVR). Nursing education practices were modified to reflect these changes emphasizing early intervention as the patient with a decreasing SVO2, widening arteriovenous oxygen content difference or evidence of increasing SVR. Conclusions: The use of continuous SVO2 monitoring in the care of the Norwood patient provides a real-time, objective assessment of cardiac output and the adequacy of systemic O2 delivery. Improved understanding of Norwood physiology by the bedside nurse has enhanced recognition of problems and reduced response time resulting in dramatically improved early survival.

P782

Exploring compliance in adolescents with familial hypercholesterolemia: Hamilton, ON

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Familial Hypercholesterolemia (FH) is an inherited condition. All the adolescents with FH are on a low cholesterol/low saturated fat diet and some may also be on a cholesterol lowering medication to prevent the development of early heart disease. Compliance is one biggest issue with the adolescents. They are normal healthy teenagers that experience no ill effects from not following their diet or taking their prescribed medication. Ten adolescents (13-18 years), from the Pediatric Lipid Clinic were asked to participate in a taped interview with a nurse researcher. The interview focused on three questions: 1. What makes it easy for them to follow their diet or take their medication? 2. What were the biggest challenges for following their diet or taking their medication? 3. What ideas did they have for other kids in the clinic to help them learn to follow their diet or take their medication? Results revealed that supportive parents who were role models and brought appropriate products into the home, helped the adolescent. The biggest challenge was going out with their peers. These adolescents felt they had control in lowering their cholesterol levels. Clinic appointments were important to help monitor how they were doing and educated them about new products and heart disease. These adolescents wanted an occasional group session with other adolescents in the clinic. It is from the adolescents who experience Familial Hypercholesterolemia a chronic condition, that we as nurses can learn new strategies to improve compliance for FH as well as other chronic diseases.

P783

Psychosocial needs of the child in hospital and his parents: The role of a liaison sister at the German Heart Center in Munich

Yvonne Hesthaler
Pediatric Nurse, Munchen, Germany

At the German Heart Center in Munich a new post was created 2 years ago. The merit for a nurse with plenty of experience in pediatric cardiology and with a special interest in caring for the child and his parents during the hospital stay for investigation or operation had developed over the years. The liaison sister is responsible for optimal information of the parents regarding the non medical side of routine procedures like cardiac catheterization, catheter intervention, operation and ward routine during intensive care. She contacts all patients (babies, children, adolescents and adults) daily, gets to know their families who visit or stay in hospital together with their child. She finds out about their needs regarding information, and refers them to other members of the psychosocial team. She helps if accomodation is needed and gives support to anxious parents during the hospital stay. Sufficient information before operation for the child and the parents is one of the main tasks. It takes time to find out how much information is tolerated at the moment. Parents and children are taken to the intensive care ward and the equipment is explained individually. A special group of interest are the adolescents and young adults who appreciate a daily visit and very often an informal chat in addition to the specific information needed. A further important task is to write to parents who have lost their child during the hospital stay and refer them support if they wish. Working as a liaison sister is an extremely rewarding task. While all parents appreciate information regarding procedures and operation, the need for further informal contacts are rather individual.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

It is up to the parents to decide whether or not they want any help, but on the whole it is much appreciated.

P784

Daily interdisciplinary patient care rounds: from concept to reality

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The Hospital for Sick Children (HSC) Toronto Canada, underwent strategic transformations and role redesign in the summer of 2997. Before introduction of interdisciplinary care (defined as collaborative discussion, planning, treatment, and evaluation across the continuum of care through mutual participations among health care team members and the family) was a common theme identified during this process. The development and framework for daily Interdisciplinary Team Rounds was recognized as a hospital-wide need and identified as an important standard to achieve. For the Cardiac Inpatient Unit (4D), this was a priority initiative for implementation. An interdisciplinary working team (which included representation from Nursing, Medicine, Professional Services and Facilities) was established. Clarity of purpose, efficiency, structure, membership, communication, family participation, and locus were identified as elements which would need to be addressed. It was also established that daily rounds would not be intended for teaching or training, rather they would be directly linked to the plan of care, using it as a guide for discussion and tracking progress of the patient/family. Daily inpatient patient care planning rounds were implemented on the Cardiac Unit at the end of August, 1999 and continue to date. A comprehensive evaluation of these Rounds is currently underway. This poster presentation will highlight details of the planning process (formulation of aims, identification of selected changes and desired outcomes, bench marking and development of measurement tools, description of tools, and analysis of results). Has the implementation of daily patient care planning rounds improved delivery of interdisciplinary care on the Cardiac Inpatient Unit? What have we learned as we implemented these rounds? As we act to hold the gains made what needs to be done?

P785

The development of nursing guidelines. will it improve the care of the neonate post cardiac surgery?

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HSC, Toronto, ON Canada

The United Nations Convention (1992) on the right of the child acknowledges that children are especially vulnerable and have a right to expect special consideration. Its underlying ethos, like the Children's Act (1989), is that in every situation the needs and voice of the child must be heard and respected. Neonates under going cardiac surgery are unable to voice their demands and expectations for care, therefore the CCU at the Hospital for Sick Children, Toronto, Ontario, Canada have developed a role within it's nursing structure called a cardiac resource nurse. The objective of this role is to promote and enhance the care of the paediatric cardiac patient. Since September 2000 the group has been working towards the development of guidelines to advance the care for specific neonatal cardiac defects. Our poster presentation will highlight the aforementioned guidelines through an illustration of a children's caricature.

P786

Can adolescents with CHD provide each other emotional support within a group setting?

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To form a support group for adolescents with CHD in order to provide them an opportunity to address various psychosocial needs common to the chronic illness. The need for support was identified for adolescents living with chronic illness. Issues they face often include struggles with: isolation, body image, desire to be like peers, fears about future, autonomy, control, and treatment adherence. Addressing these needs within the group format allows for opportunity to interact, express conflicting emotions, share information, discover commonalities, instill hope, and to learn from peers and facilitators. The environment would need to be comfortable, safe, and fun, all the while building on self esteem and acceptance issues. The age of the group to be targeted is 11 to 17 years. A team has been developed to organize and run the group consisting of an RN, Social Worker, Child Life Therapist, Psychologist and an adult with CHD. Goals and objectives were established. The group would meet one evening per month, which would include a fun activity along with an educational component. A mailing list was generated from the clinic database of those adolescents that fit our criteria. A mail out was sent to 75 youths and their parents, which included an invitation and letter describing the format of the group. The group is held have proven to be a comfortable and safe venue for youth to talk openly about the issues that surround adolescents with CHD. Attendance at this group has been encouraging and has shown the need for such a group to be supported. Thus far our return rate has been 100%.

P787

Don's say his name or he'll appear

Curry J A
Green Lane Hospital, Auckland, New Zealand

This poster display will show, by way of a timeline, how a partnership of care develops between a young family coping with a child with congenital heart disease (CHD) and near impossible to control Supraventricular Tachycardia (SVT), and the multidisciplinary hospital team. At 3 years of age this child is the youngest in New Zealand to have a radiofrequency ablation. He was diagnosed antenatally with complex CHD, congenitally corrected transposition of the great arteries, large ventricular septal defect, mild subpulmonary stenosis, severe tricuspid regurgitation. He was later diagnosed with Wolff Parkinson White Syndrome and recurrent SVT. The family reside 50km from Green Lane Hospital, New Zealand's only centre for paediatric cardiology. At age 10 months, he underwent 3 open heart surgeries, resulting in the insertion of a mechanical valve. Over the next 2 years the family coped with multiple admissions, sometimes daily, for potentially fatal SVT (rates > 200bpm), as well as managing medications, weekly blood tests and caring for two older siblings. I will show strategies/protocols put in place by the combined health team to ease the stress and reduce the length of these admissions. For example, education of parents in early recognition of SVT, direct admission to the paediatric cardiology ward following a phone call, parents supplied with topical local anaesthetic cream for application prior to transport, specific protocols and care plan in place for his cardioversion. He went forward for EP study and ablation on a moderately urgent basis as it was no longer possible to control his practically an exam SVT. Since the successful ablation he has had no SVT, has achieved normal milestones and leads a happy and active life.

P788

Impact of rheumatic fever

Oxford C Y
Green Lane Hospital, Auckland, New Zealand

Rheumatic Fever remains a significant disease among certain sectors of the New Zealand population. On average, 84 cases of Acute Rheumatic Fever (ARF) are reported annually, with 62% of these cases occurring in children aged 4–14 years. Of these, 84% are of Maori and Pacific Island descent. A national Rheumatic Fever Register was established in 1966. Currently the rate of Rheumatic Fever notifications is 2.5 per 100,000. This rate is high for a developed country and has not declined markedly since the mid-1980's. The annual notification rate for New Zealand's anglo-saxon population is 0.3 per 100,000, in contrast to the Maori (indigenous) population's rate of 6.8 per 100,000 and Pacific Island people's rate of 23 per 100,000. Despite the evidence of this disparate incidence, specific racial predilection has never been substantiated by research (Neutze,1995). Factors believed to contribute include overcrowding, poor housing, poor hygiene (Neutze,1990) and inadequate primary and secondary treatment programmes (Stollerman,1997). ARF and it's sequelae – chronic rheumatic heart disease, result in 450 hospital admissions and a significant utilization of the health budget annually. New Zealand has a small total population (< 4 million). The city of Auckland has the largest Pacific Island population in the world. Measures needed to reduce the incidence of this debilitating disease include identifying and training culturally specific resource people to enhance prevention strategies and be community role models.

P789

Late diagnosis of complex congenital heart disease-a case profile

Oxford C Y
Green Lane Hospital, Auckland, New Zealand

Congenital Heart Disease (CHD) is the most common of all congenital abnormalities. This case profile outlines the presentation of Sam, a twin, who at 1 week post-partum was admitted in a collapsed state to his local hospital. The product of a normal and well monitored pregnancy, Sam's relatively late presentation raises some concerns about the adequacy of ante partum and

post-partum care in centres where there is a lack of paediatric cardiac expertise. Sam, his twin Ben and their parents were required to relocate from their home town some 400 miles away for the duration of Sam's treatment. Ben was a boarder on the ward during Sam's in-hospital stay, cared for by his parents. The chance finding upon 5 weeks post-partum and just 1 day prior to Sam's planned discharge that Ben too has complex CHD is evidence that such conditions are not always found in their presentation. Maternal and child health care in New Zealand is first and primary care providers such as midwives and Plunket Nurses visit new mothers and babies in their homes. Early detection of CHD may be facilitated through appropriate training of these health care professionals, especially as many births are now happening at home or mothers and babies are returning home within hours of delivery. New Zealand is a relatively small country (population 4 million) and our unit is the country's only tertiary paediatric cardiac referral centre. Increased cardiac knowledge in primary carers throughout the country can be promoted through our expert nurses providing theoretical and clinical training opportunities such as seminars and facilitating clinical exposure to this patient group. Ultimately we are keen to develop an advanced nurse practitioner programme for this area of specialty practice.

P790

Evaluation of a cardiovascular-thoracic pediatric nurse practitioner program

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Children's Memorial Hospital, Chicago, IL, USA

Purpose: Limitations on residency programs, increased patient acuity, and the impact of managed care has prompted the utilization of nurse practitioners by specialty practices to assist with patient care management. This study examined the effectiveness of our cardiovascular-thoracic surgery pediatric nurse practitioner program which began in 1995 and evaluated the impact on family-centered care. The pediatric nurse practitioner (PNP) collaborative patient responsibilities include admissions, discharges, preoperative and postoperative evaluations, and minor procedures. Methods: After being pilot tested on a sample of 25 parents, physicians and nurses, 5 separate questionnaires containing both closed and open-ended questions evaluating the role of the PNP were sent via mass mailing to 799 parents, 55 staff nurses, 20 collaborating physicians, 45 specialists and 524 primary care physicians. The overall response rate was 25% (325 parents, 96% staff nurses, 75% collaborating physicians, 33% specialists, and 11% primary care physicians). Results: The PNP program was well accepted by the parents, nursing staff, and physician groups. Parental satisfaction regarding the care provided by the PNPs was achieved 95% of the time. Among staff nurses, 100% felt that the PNPs improved communication with the nursing staff and enhanced patient care. Greater than 90% of the collaborating physicians felt that the PNPs decreased physician work-load and effectively performed specific components of the role. Ninety-three percent of the specialists felt that the PNPs' request for consultation was appropriate, implemented recommendations effectively, and facilitated multidisciplinary collaboration. Seventy-three percent of primary care physicians felt that the PNPs provided timely and adequate communication about the surgery and hospital stay. Conclusion: The consensus from all groups was supportive of the PNP role. The PNP program met its objectives to enhance patient care and parental satisfaction.

P791

The adult congenital patient in the cardiovascular intensive care unit: unravelling the ties that bind them

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University Health Network, Toronto General Hospital, Toronto, ON, Canada

The adult with congenital heart disease is an emerging population. Their complexities challenge the expertise and experience of health care providers caring for this expanding patient population. The challenges posed by these patients during their early, postoperative period are frequently very daunting. Complex cyanotic lesions such as the Tetralogy of Fallot or Fontan patient do not always experience an uneventful postoperative course. In an effort to improve care and provide a better guide for nursing practice, we undertook a 3 year retrospective chart review. To clarify the adult congenital patient's early postoperative needs, we examined their major problems and challenges. These include: achieving early hemostasis with the endpoint of maintaining adequate preload, supporting moderate to severe ventricular dysfunction, managing complex arrhythmias and providing a full spectrum of respiratory support. From July 1997 to June 2000, 198 patients were reviewed. 58% of these patients had undergone a previous repair or palliative surgery. The presentation of data will include: total blood loss and replacement, incidence of reexploration for bleeding and/or tamponade, combinations of inotropic support used and length of time required, the incidence and timing of early postoperative echo, need for epicardial pacing, antiarrhythmic therapy, the initiation of afterload reduction and duration of ventilatory support. The ICU length of stay statistics will also be shared. The ability to anticipate and better define the postoperative problems of the adult congenital patient, allows for early identification of these problems and timely intervention of effective therapeutic modalities. Through the evaluation of the information and data that patient population provides, we gain an understanding of patient trends and specific needs. The lessons learned and impact on the nursing care provided, are invaluable.

P792

Inter-provincial cardiac services providing family-focused care

Lois Hawkins, Lee Legge, Cathy Morrison, Cory Robinson
Stollery Children's Health Centre, University Of Alberta Hospitals, Alberta, Edmonton, Canada

Two provincial pediatric cardiac centers have joined forces to better meet the needs of infants and children requiring heart surgery. In a country as large as Canada, with populations that are spread out and sparse, resources need to be combined to provide excellent medical care to specialty groups. Success has been the outcome for children and families from Winnipeg that are involved in just such an effort. In 1995, the Winnipeg Children's Hospital developed a multi-disciplinary family support program to assist families travelling out-of-province for pediatric heart surgery. Services included medical information, emotional support, education and preparation for the out-of-province experience, financial support, and other supports as needed by individual families. In 1997, the program was evaluated. Program services lessened many of the negative aspects of out-of-province travel. The highest-rated services were medical information, travel arrangements and financial assistance. Deficiencies included, gaps in continuity-of-care, follow-up in and beyond the recovery, hospital, and accommodation. Program services did not have as direct an impact on parental stress. The needs of parents coping with additional stresses were not completely met. These families, in particular, missed their home support network and the program's support services did not completely fill this gap. In 1998, a focus group of seven families was conducted. Two major themes emerged: the need for continuity throughout the cardiac surgery experience, and the interest in preoperative support and preparation. These findings led to program modifications. Educational material was developed and efforts were made to enhance collaboration between centers. A third program evaluation is currently underway. Findings will be presented, with further recommendations from the professional team and families.

P793

PICU/CVICU Newsletter

Rosita V. Maloy Arlene B. Angulo
Texas Children's Hospital, Houston, TX, USA

To improve and enhance communication with hospital and unit users, educational publications and programs resulting for effective, efficient patient care, staff development and opportunities for staff participation and contribution to the newsletter.

P794

Protein losing enteropathy in children post-Fontan: Is heparin or corticosteroid therapy most efficacious?

Miller Judi, J. A.
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Protein losing enteropathy (PLE) is a rare and life-threatening complication which can occur following the Fontan operation. Children diagnosed with PLE post Fontan present with a unique set of challenges. The exact pathophysiology of the disease is unknown, and the child and family are faced with serious debilitations, tenuous symptom management and a dismal prognosis. Current treatment of PLE post-Fontan remains associated with a very high mortality and morbidity rate, with a five year survival of 58% and a total mortality rate of 93%. Non-pharmacological treatments have included baffle fenestration of the Fontan circuit, Fontan takedown and transplant. Current pharmacological management is directed towards symptom control with diuretics, and attempts to alter the disease process involve both heparin and corticosteroid therapies. Outcomes for the pharmacological treatment of PLE remain variable. Until larger research trials are conducted, the use of either heparin or corticosteroid therapies will remain a process of trial and error, and ultimately be influenced by the individual child's response. Future research to

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

compare the effects of heparin and corticosteroid therapies would advance the treatment protocols for this vulnerable population and potentially optimize their outcomes. This poster will present an integrative methodological critique of the strengths and weaknesses of the relevant research, and a comparison of drug therapies involving the pharmacodynamics and pharmacokinetics of heparin versus corticosteroids.

P795
Standardazzarion of inotropic drugs
Arlow B. De La Cruz
Texas Children's Hospital, Houston, TX, USA

Standardazzarion of inotrope drugs in the operating room and intensive recovery developed by the multidisciplinary care teams thus providing stability to patient's vital signs, fluid volume status, minimizing errors and efficient dispensing and delivery of medications.

P796
Parent Power: The INR home testing project
Sparrow, Heather D
Green Lane Hospital, Auckland, New Zealand

A number of children, including an increasingly younger age group are being discharged home requiring anticoagulant therapy post cardiac surgery. An INR blood test accessed by venepuncture is commonly available at community laboratories but very young patients and older children with needle phobias prefer a finger-prick blood test to a venepuncture. The fingerprick INR test was only available at the central hospital and often involved long travelling times from rural communities for families able to access it. The introduction of finger-prick, home testing, INR machines onto the New Zealand market found informed parents of cardiac children asking 'Why can't we do this for our child'? This paper tracks the progress of the home testing INR machine project that began with this question from the parents and resulted in INR testing machines being used in homes nationally. The paper discusses the process and the management of issues such as parent selection and education, safe prescribing, risk factors and quality controls for the machines, the solutions and the testing process. The perils and the pitfalls we experienced in the process will also be described.

P797
A study of how professionals and mothers recognize the needs of children with congenital heart disease
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This study examined how professionals and mothers recognized the needs of children with congenital heart disease (CHD), in order to improve cooperation between professionals and mothers when supporting the children and their families. The type of needs recognized by professionals and mothers were compared, as were the needs recognized among professionals. A questionnaire was administered to 192 professionals (nurses, doctors, public health nurses, kindergarteners and teachers) whose work was related to children with CHD, and 588 mothers of children with CHD, in September 1999. 159 (82.8%) professionals and 330 (56.1%) mothers responded, of these responses, 156 and 292 were used for the analysis, respectively. The responses were analyzed using Needs Scale for Children with CHD, developed by Hirose in 1999. It consisted of 6 categories. The needs recognized by professionals showed significantly higher scores than those recognized by mothers in 5 categories, Play and Social/Cultural Activity, Habitude of Daily Living and Social Relationship, Psychological Domain, School Life and Knowledge of Disease. However, there was no significant difference between the needs recognized in the Medical and Physical Domain. Comparison among professionals was not significant in all 6 categories. It was suggested that professionals recognized the total needs of children with CHD, but mothers concentrated on the needs within the Medical and Physical Domain.

P798
The role of nitric oxide in heart failure
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Heart failure (HF) is a pervasive pathophysiologic condition that occurs in children with congenital heart disease. Regardless of the etiology, HF causes altered gene expression, functional and structural transformation of cardiac myocytes, and finally, attenuated cardiac function. The physiologic manifestations of HF induce compensatory mechanisms through stimulation of neurohumoral pathways that cause an activation of the renin-angiotensin-aldosterone and sympathetic nervous systems, and augment the release of vasoconstrictor substances. Although homeostasis is initially achieved in the acute stages of HF through activation of these processes, the effects eventually contribute to the pathophysiology and progression of HF. Enhanced vasoconstriction and reduced vasodilatory response to exercise are specific attributes of patients with chronic HF. Although compensatory mechanisms and neurohumoral effects have been associated with the abnormal vasomotor tone that is typical of the patient population, these findings indicate that additional factors may be involved in this atypical response. There is growing evidence to suggest that this effect may be exacerbated by an aberrant production of endothelium-derived vasoactive mediators. Specifically endothelium-derived nitric oxide has been identified as an attributable element in the pathogenesis of HF. Nitric oxide is a well-recognized mediator within the cardiovascular system. However, increased production of this free radical gas molecule, as is seen in patients with HF, may prove to be deleterious. This poster will review the abnormalities of the nitric oxide pathway that occurs in patients with HF.

P799
Patent ductus arteriosus in premature infants: implications for nursing care
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With advanced medical care, more severely premature infants are surviving. The presistence of a patent ductus arteriosus (PDA) continues to be a risk. To further explore this population of patients we reviewed all patients born between January 1, 1998 and January 1, 1999 who were admitted to DeVos Children's Hospital neonatal intensive care unit with a gestational age less than 38 weeks and who required intervention for a PDA. There were 97 patients who received indomethacin and 9 of these infants required surgical ligation of the ductus. The surgical and the non-surgical groups were compared regarding these variables: birth weight, gestational age, number of courses of indomethacin, incidence of necrotizing enterocolitis (NEC), use of high frequency ventilation (HFV), and hospital mortality. Results: Significant differences found between the surgical and non-surgical groups were: mean birth weights of 853.6 and 1263.68 grams ($p=0.038$), mean gestational age 25.33 and 28.36 weeks ($p=0.009$), and mean number of courses of indomethacin per patient were 2.00 and 1.14 ($p=0.012$), respectively (data were analyzed using T-test). The surgical and non-surgical group incidence of NEC was 2 (22%) and 9 (10%) ($p=0.024$ by ANOVA). HFV was 7 (78%) and 30 (34%) ($p=n$ by ANOVA), and hospital mortality was 2 (22%) and 7 (8%). Conclusions: If the infant has a lower gestational age and/or birth weight or requires more than one course of indomethacin, the nurse should be aware of the increased potential need for surgical ligation of the PDA. NEC may be an indication for surgical ligation of the PDA. The nurse must appropriately assess cardiovascular and respiratory status. Nursing interventions can be tailored to meet the needs of the patient and family.

P800
Operating room nurses in the pre-operative phase for cardiothoracic surgery
Pollman, J., Minden, M., Bradley, V., Kolvar, E., Carpenter, D., McDougall, J., Preston, M., Meglahn, S., Denny, F. Lofland, C.
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Preoperative visits for pediatric cardiac surgery patients were traditionally the responsibility of the same day surgery nursing staff. With an ever increasing caseload and a demographically diverse patient population from a wide geographic area, improvement in efficiency was mandatory. Twenty-four months ago, cardiac surgery operating room nurses assumed responsibility for same day preoperative visits. Guidelines for cancellation prior to invasive testing were established. Communication was increased between Cardiac Surgery, Cardiology, Anesthesiology, Clinical Laboratories and Blood Bank. In eighteen months, ten variables were identified to have the greatest effect on the efficient completion of the preoperative phase. Problem solving techniques were then applied to the most commonly occurring variables in order to reduce their frequency. The time of preoperative preparation declined from 4 hours (+/- 120 min) to 2 hours (+/- 30 minutes). Most cancellations were declared in the preoperative assessment phase thus preventing unnecessary invasive preoperative testing and the loss of expensive disposable supplies. Canceling cases in the pre-operative assessment phase resulted in a savings of approximately $2,000 in disposable supplies per cancellation compared to

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

cases cancelled the day of surgery. Utilization of cardiac surgery operating room nurses on the same day surgery preoperative assessment of cardiac surgical patients can improve efficiency, contribute significantly to cost containment, and better serve the patient.

P801

The influence of lactation support services on breastfeeding success among infants with congenital heart disease

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It is widely perceived that infants with congenital heart disease (CHD) are not able to breastfeed successfully, and must be supplemented. In spite of recent studies indicating that oxygen saturations were maintained at higher and less variable levels, and that breastfed infants with CHD gained weight more quickly and had shorter hospital stays, mothers of infants with CHD often are not encouraged to breastfeed. The purpose of this descriptive study is to determine if lactation support and education impact success at breastfeeding and breast pumping duration among this high-risk group of infants with CHD. This project asks mothers of infants with CHD to respond to a written survey when their infant is at least 6 months old, following cardiac surgery in the neonatal period. The survey addresses questions related to breastfeeding initiation, use of breast pumps and supplementation, and duration of breastfeeding/milk expression. Sources of breastfeeding support and education or perceived lack of support, and mother's level of satisfaction with her breastfeeding experience are evaluated. Data retrieved from this ongoing study were compared to data from a previous study at the same institution, conducted prior to the development of a formal lactation support program at the hospital. Breastfeeding duration rates at 3 months and 5 months post-cardiac surgery were compared. A significant increase ($p \leq 0.01$ chi square) in the number of mothers able to exclusively breastfeed, or continue to provide at least partial breastfeeding or breastmilk feeds to their infants has been demonstrated. Results suggest that given support and education necessary to initiate and maintain lactation, mothers can successfully breastfeed their infants with CHD, and provide breastmilk for durations suggested for the general population by the Healthy People 2000 initiative.

P802

Psychological and physiological effects of long term alprostadil therapy on infants with hypoplastic left heart syndrome

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As more families are choosing heart transplantation as a treatment option for Hypoplastic Left Heart Syndrome, increasing numbers of infants are on long term Alprostadil therapy. Treatment may be as long as 3–5 months and patients are managed either in the Critical Care Unit (CCU) or on the Cardiology ward. These duct-dependant infants are maintained on continuous intravenous Alprostadil infusions. Consideration must be given to a number of side effects including apnea, seizures and fever. Psychologically and physiologically infants need security and containment to enable them to maintain a state of wellbeing. Fever challenges care providers as there is a need to balance being able to meet the infants developmental care needs while risking a febrile response. It is imperative to rule out an infectious process by coordinating related testing. At the Hospital for Sick Children, creative solutions to these challenges have been met by using light blankets suspended off the infant with a floss cradle, light dimming, & nesting. Teaching is done with the family to identify, encourage and promote infant self-regulation behaviours. Understanding altered family dynamics and sustained interactivity theory are key elements in the care of the infant and family. Another reported side effect of long term Alprostadil therapy is a rare and temporary disorder called hypertrophic osteoarthropathy. Clinical manifestations of this self limiting symptom are soft tissue swelling, particularly hands and feet as well as cortical proliferation of long bones. This can be associated with significant pain and discomfort which requires the collaborative approach of the interdisciplinary health care team. This presentation will highlight three unique and complex physical and psychological effects of long term Alprostadil therapy on the patient and family.

P803

Using a family bed to enhance family centred care in the cardiac unit

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The Cardiac Program at The Hospital for Sick Children in Toronto, Canada provides medical and surgical treatments to a large multi-cultural population. Nurses in the program have a strong appreciation for cultural diversity, recognizing the importance of children having their loved ones present during their hospital stay. In many cultures, children are raised from a very young age sharing a bed with their parents and/or their siblings. Over the past several years there has been an increase in the number of requests for a 'family bed' received by nurses in the Cardiac Unit. They are sought to reduce parent anxiety by facilitating closer parent infant sleeping arrangements. Nurses have expressed concerns relating to patient safety and potential liability issues surrounding this practice. A literature review was done by members of the Cardiac Program's Nursing Practice Council to address the use of a 'family bed' in this practice context. Other units within the hospital and other Health Care Centers were also contacted to ascertain whether the use of the family bed has ever created a dilemma in their practice. A gap in nursing practice was then identified which led to the creation of practice guidelines. The family bed guidelines were developed to assist nurses to employ the proper knowledge, skill, and judgement around the decision to incorporate a family bed into the patient care setting. These guidelines have been given approval by The Hospital for Sick Children's Risk Management Team, and are being considered for implementation throughout the organization. This poster presentation will elucidate the current literature available related to this practice, the development of the Cardiac Program's guidelines for family bed use, the potential risks associated with the like, and the implications for nursing practice.

P804

Preparation of child and family for cardiovascular surgery at the Hospital for Sick Children

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Over 550 children and families are prepared for cardiovascular surgery at the Hospital for Sick Children each year. We recognized that these preparation days were often too lengthy, families were overwhelmed with new information, and unidentified issues existed in postponement of surgery. To prepare families while supporting them in a holistic manner, we examined both the roles of our interdisciplinary team members and the structure of the existing pre-operative day. An important goal was incorporating the family into the health care team, fulfilling the hospital mission of family centered care. We recognize parents are the most consistent caregivers, and that being an active part of the health care team enables them to move more easily from the hospital to home. To prepare families prior to their pre-op day, a triage phone assessment by the Surgical Nurse Coordinator was instituted. This enabled identification of issues that could delay surgery. A package including a follow-up letter, information on directed donation and use of blood products, pain management, is mailed to reinforce the initial information. To ratify a need for parent and sibling support, the Child Life Department developed a pamphlet that includes strategies parents can use to prepare the child and siblings for the hospital admission. Re-organization of the day allowed for more effective use of time (ie. earlier arrival time avoided traffic delays, and use of labs during less busy hours). While the pre-operative day has a certain structure to it, we recognize the need for flexibility to allow for the patient and family to obtain the greatest benefit.

P805

Developing a Competency Based Orientation in an Acute Care Pediatric Cardiac Unit

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Purpose. To ensure that orientation for nursing staff provides the tools and education needed to become a competent practitioner. The nursing orientation to the Cardiac Program has historically been to train individuals to carry out those duties that nurses perform the most. The orientation consisted of a general hospital nursing orientation and a unit based orientation. There was an unwritten belief into the orientation about who came with ten years experience received the same orientation as new graduate nurses. Competency Based Orientation provides a framework that allows each individual to identify and define their learning needs. The orientee becomes a shared responsibility among all members of the nursing team. The orientation also highlights adult learning principles and learning instead of teaching. Competencies were developed using four nursing education priorities. 1) Family needs that relate to high-risk patient care. 2) Frequent any nursing practice that is often

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

performed. 3) Fundamental: needs that are essential aspects of effective nursing practice, 4) Basic: educational needs for all hospital employee's. The tools used to measure and evaluate practice and the competencies were created before implementation (Donnelly and Hallett, 1998), provide a challenge to educators to ensure they are presenting what is needed to survive in the organization rather than extraneous material that is nice to know". A variety of learning options utilizing an array of learning styles are available to new staff members including: unit procedures, observation of expert staff, practice under supervision, videotapes, pre and post testing and independent study. Competency Based orientation provides the new orientee with a "real world" environment that will integrate them into the highly specialized role of pediatric cardiac nursing."

MAY 30 Time: 11:30–12:30

Session 5
Catheter Interventions

P306

Balloon dilation of aortic stenosis in infants less than 6 months of age

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42 patients aged 1 day–6 months underwent balloon dilation of the aortic valve from 1988–98. We examined outcomes and predictive parameters in the 3 subgroups Grp 1: 1–7 days(n=15), Grp 2: 8–30 days(n=10), and Grp 3: 1–6 months(n=16). Median follow-up was 5.2 years(s: 6m–10y). There were 12 deaths, with 12 occurring in the group undergoing dilation on the first 30 days. The overall survival at 10 years was 72%, with 42%, 65% and 92% for Grp 1, 2 and 3 respectively. For those surviving beyond one month the freedom from intervention rate was 70% and 21% at 5 and 10 years. Aortic valve annulus >25 mm/m2 (26/42) was the only univariate significant predictor for this basic parameter for survival with 10 year survival of 88% and freedom from reintervention of 35%. Multivariate analysis of a range of anatomic elements allowed 80% prediction of outcome. Balloon dilation of the aortic valve is effective in the first 6 months of life in patients with adequate left heart structures.

P307

Stent implantation for children under 6 years old

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National Cardiovascular Center, Osaka, Suita, Japan

Purpose: To evaluate the feasibility of stent implantation in small children under 6 years old. Subjects: We attempted stent implantation for 17 lesions in 12 patients under 6 years, specifically, the pulmonary artery (PA) 14 lesions, pulmonary vein (PV) 2, coarctation of aorta (CoA) 1. Age and body weight ranged from2 to 79 (median 38) months, 2.9 to 22.0 (11.0) kg. Methods: We implanted stents using a from loading technique (4 cases), conventional technique (4 cases), or combined technique (3 cases). We implanted a Palmaz P308 or P308 stent (Johnson & Johnson) in the PA using a 9F sheath, except for 3 lesions where a P128 or P207 was implanted as a larger vessel could not negotiate with the lesion. P104 or 154 stent was implanted for PV stenosis in a 4.5 kg baby, while a P 106 was implanted for CoA in a 2.9 kg baby. Repeat catheterization was performed after 3–15 (6) months in 8 patients with 12 lesions. Results: A P128 stent dislodged from the left PA and impacted in a benign portion of the right PA. Further procedures were abandoned in this case, otherwise stents were successfully deployed without complications. The minimum lesion diameter significantly increased from 0.8–6.5 (3.5±1.7, mean±SD) mm to 3.6–12.0 (6.7±2.0) mm (p<0.01). Although follow up the diameter, 1.5–12.0 (5.8±2.9) mm, was not significantly decreased, there was a variable degree late luminal loss. 0–59.5 (20.9±21.8) %. Late luminal loss requiring redilation occurred in one PA lesion (7.1 %) and 2 PV lesions (58 and 59 %). Conclusions: Stenting is a treatment option even in small children. Although redilatation might be occasionally necessary because of late luminal loss, particularly in PV stenosis.

P308

Pulmonary arteriovenous fistula - successful percutaneous transcatheter embolisation

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Pulmonary arteriovenous fistulae (PAVFs) are rare vascular malformations of the lung and may present with cyanosis and vascular malformations in other parts of the body leading to epistaxis, neurologic manifestations or cerebral abscesses. Purpose of the study: To assess the effectiveness of percutaneous transcatheter closure of PAVFs in our patients. Patients and methods: Transcatheter embolisation of PAVFs using spring coils was performed in three patients (2 males and one female). The age at presentation was between 8 months and 3 years. They presented between 1989–1999. All had severe cyanosis and clubbing at presentation. There was no skin or neurologic manifestations. Their saturations at the onset ranged between 60–72%. After pulmonary angiography and localisation of the fistula, multiple spring coils were used to occlude 6 fistulas in the 3 patients using a total of 32 coils. This was done in 3 settings (more than one setting for each patient). Results: complete occlusion of PAVFs was achieved in all the patients. There was no acute or long term complications in all 3 patients on follow up (1–5 years). Arterial saturation rose from mean 66.3% to 94%. The chest x-ray showed dramatic regression of the PAVFs shadows in all patients on follow up. Conclusion: transcatheter coil embolisation is a safe and effective method to treat PAVFs non surgically. Patients may require multiple settings to occlude these fistulas completely.

P309

Transcatheter closure of secundum atrial septal defect using amplatzer septal occluder : short-term outcome

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The study is to review the short-term outcome of transcatheter closure of secundum atrial septal defects (ASD) using Amplatzer Septal Occluder (ASO). From January 1997 to October 2000, 210 patients with secundum ASD underwent successful transcatheter closure using ASO. Nineteen patients with right to left shunts were excluded from the analysis. Total of 191 patients with left to right shunts were reviewed. The patients were assessed for possible complications and the presence of residual shunts using transthoracic echocardiogram at 24 hours, 3 months and at one year. Their median age was 10 years (range 2 to 69 years) and median weight was 23.9 kg (range 8.9 – 79 kg). Five patients had transcatheter closure of patent ductus arteriosus and 2 had balloon valvuloplasty for valvular pulmonary stenosis performed at the same sitting. The median ASO device size was 20 mm (range 4 to 36 mm). The mean procedure and fluoroscopy times were 90 minutes (range 30–210 min) and 25 minutes (range 5–111 min) respectively. Mean follow up was 20.8 +/- 12.4 months. Complete occlusion was obtained in 169 of 191 (88%) patients at 24 hours, 128 of 132 (96.2%) at 3 months and 100% at one year (n=102). There was one incidence of malposition/dislodgement of the device which was successfully retrieved via transcatheter approach and one embolized into the right ventricular outflow tract which was removed surgically the next day together with ASD closure. No haemodynamic disturbance occurred in either patient. There were no major complications eg. thromboembolism, endocarditis or device fracture noted on follow-up. In conclusion, transcatheter closure of ASD using ASO is safe and effective. However, a long-term follow-up is warranted before it is recommended as a standard procedure.

P310

Transcatheter closure of patent ductus arteriosus in adults

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From Nov 1995 to Sept 2000, a total of 216 patients with patent ductus arteriosus (PDA) underwent transcatheter closure by Cranston coil. Eighteen (8.3%) of them aged over 18 years were evaluated retrospectively. There were 7 males and 11 females with aged ranged from 18 to 77 years (38.4±19.8 years). Total of 20 procedures were performed on these 18 patients who were isolated PDA in 16, associated with other lesions in 2. Two patients underwent second time transcatheter closure for the residual ductal shunt within one month. The calculated Qp/Qs ratios by Fick's principle were 1.07–4.05 (1.70±0.90). Six patients had mild pulmonary hypertension. The average pulmonary vascular resistance were 1.87±1.04 Woods units. The types of PDA included A1 in 3, A2 in 10, C in 1 and E in 4. The narrowest diameter of ductus were 1.40 to 5.10 (3.29±1.89)mm. A total of 42 coils had been placed through transfemoral approach. Temporary occlusion of ductus by balloon had been performed on 5 patients. No patients had coil embolization in the pulmonary arteries or aorta. All patients had been follow-up studies regularly with auscultation, chest x-ray film and echocardiograms. The significant residual ductal shunt was detectable for 3 weeks in 2, but completely disappeared after re-insertion of 2 coils. All other 16 patients had a complete closure of PDA immediately after coil placement. Acute hemolysis with

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significant anemia occurred since second day after procedure in 2 who including 3 patients with anoxemia. The complication disappeared completely after the repeated coil placement. In conclusion, transcatheter closure of PDA with coil in adults is a safe & effective method. Acute hemolysis could be overcome with the further placement of coils to close the residual shunt.

P811

Initial and longterm results following catheter intervention for neonatal critical pulmonary valve stenosis/atresia: a single operators experience

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Isolated neonatal critical pulmonary valve stenosis(PS)/atresia(PA) is rare, presents with cyanosis & can be technically difficult to alleviate in the cath lab. Since 1989, 26 neonates (age:6+/-11 days, weight 3.4+/-0kg) presented with critical PS/PA requiring PGE1 and were considered for valvuloplasty(V). Initial echo demonstrated severe TR (80%), RV dilatation/reduced function(46%), hypertrophied small RV(42%) and suprasystemic RV pressure(100%). PA was suspected by echo in 20(85%) of which 5 were truly atretic at cath. All pts underwent successful V (balloon to annulus ratio 1.26+/-0.1) without mortality. Five pts with PA required valve perforation first followed by V. Post V, the RV/aortic pressure ratio (1.3+/-0.2 vs 0.44+/-0.2 p<0.0001) & pulmonary valve gradient(51+/-17 vs 6+/-5mmHg;p<.0001) decreased significantly. A 'stage assisted' approach, which we initially reported in 1994, was necessary & feasible in 16pts(62%) resulting in reduced fluoroscopy 37+/-28 vs 81+/-19minutes,p<.005) and fewer balloon catheters utilized (1.4+/-0.9 vs 3.5+/-1.7,p<.001). In 21pts(81%), PGE1 was successfully discontinued 7.9+/-3.8 days post V while 3pts required surgical PDA ligation and 2 required RV outflow tract augmentation & BT shunt placement. At latest follow-up(47+/-36mths) there has been 1 late unrelated death & 1 repeat V. Latest echo follow-up(43+/-36mths) demonstrates persistent gradients in 6(71+/-9mmHg), improved TR and 1 pt with persistent ASD. All pts are functional class I and 2 require a daily diuretic. Conclusion: Catheter intervention is the treatment of choice for critical PS/PA utilizing a variety of techniques. V with long term follow-up there is persistent gradient relief, significantly improved TR & resolution of right to left atrial level shunting.

P812

A "smart" stent for severe stenosis of the right pulmonary artery

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The SMART (Shape Memory Alloy Recoverable Technology) stent (Cordis-Johnson & Johnson manufacturer) is currently in use for vascular purposes, but so far as we are aware, not yet in the pulmonary arteries. We report the successful insertion of this self-expandible tubular stent in a 15 years-old 25 kg female patient with corrected pulmonary atresia and ventricular septal defect, who had developped a severe right pulmonary branch stenosis. She had a complicating severe kyphoscoliosis, right cardiac failure and functional disability (NYHA class III), with supra-systemic right ventricular (RV) pressures that were squeezing the left ventricle (LV). She had undergone a previous attempt of a Palmaz-Shatz stent insertion, which failed in the view of technical difficulties derived from the distortion of the cardiac and thoracic anatomy. The 10 mm x 40 mm SMART stent appeared to be easy accurate and secure to deploy very flexible on its advancement onto the stenotic area, and provided an excellent anatomic and functional result. Indeed, the RV pressure dropped to 50% systemic, hence significantly enhancing LV performance. On follow-up, the patient is much more active, in class I NYHA, thus having achieved a much better quality of life. Clinical studies are required in order to define eventual further indications for the use of this stent in this cohort of patients.

P813

Patent ductus arteriosus closure using the guanucro detachable coil

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Patent ductus arteriosus (PDA) represents the third most frequent cardiac malformation in Costa Rica. Most of them are adequate for coil embolization using Guanucro detachable coils. During the last three years, 65 patients with PDA had their defects closed via cardiac catheterization at the National Children's Hospital. San Jose. Age varied from 6 months to 120 months with a mean age of 42 months. Weight range was 7 to 30 kg with a mean of 14 kg. Complications included three coils that migrated to the pulmonary arteries, one of whom were recovered. Two patients (3.1%) presented with hemolysis that warranted a second coil 3 and 7 days later respectively. The most frequently used coil size was 5 mm in diameter and 5 loops. Ductal diameter varied from 1.6 to 4.2 mm, with a mean of 2.4 mm. Pulmonary artery pressure was between the normal range in 54 cases and mild to moderate degrees of pulmonary hypertension were found in 11 patients, 8 of them were children with Down syndrome. Seven patients required two coils. At one week follow up, 92% of our patients had the defect occluded. At one month follow up, 95% were completely closed. Associated problems in our patient population consisted of 2 cases with pulmonary stenosis that were treated during the same procedure, two cases had ventricular septal defects, one patient had an AV septal defect, one patient had peripheral pulmonary stenosis. Closure of certain patent ductus arteriosus can be safely performed using the detachable Guanucro coil. The 5 mm diameter and 5 loops coil was adequate for the majority of the cases.

P814

Transcatheter occlusion of patent ductus arteriosus using coronary platinum coils

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Background: Although transcatheter occlusion of patent ductus arteriosus (pda) using stainless steel coils has become popular, one of the disadvantages of stainless steel coil is incompatibility with magnetic field. On the other hand, commercially available coronado platinum coils (TPC) has proven to be safe in magnetic field. PURPOSE: To develop a method to occlude PDA using commercially available TPC. METHODS: Ten patients with PDA (4 boys and 6 girls; 1 to 7 years; 9 to 22 kg) were included. The minimal size of PDA ranged from 0.5 mm to 3.6 mm and Qp:Qs ranged from 1.1 to 2.6:1. We applied either anterograde or retrograde method using retrievable system. A 5 Fr multi-purpose catheter was advanced into descending aorta across PDA to deliver TPC. Either smaller end or larger end of TPC was grasped by 3 Fr biopsy-nip. The coil-biopsy-nip were advanced and pushed out slowly through 5 Fr multi-purpose catheter. Three to 5 1/2 loops of larger end of TPC was placed in the aortic ampulla and the remaining 1/2 or 1 loop of smaller end of coil was in the main pulmonary artery. If the coil position was suboptimal, the coil was redeployed. RESULTS: In 9 of 10 patients that had PDA <= 2.7 mm in diameter, we successfully occluded PDA using 1 to 5 TPC. In 8 of the remaining 9 patients, an echocardiogram confirmed complete occlusion until 3 months after procedures. In 1 patient, we gave up to occlude PDA using these coils because of malfunction of the biopsy-nip. CONCLUSION: It is feasible to occlude PDA <= 2.7 mm using commercially available TPC and this method can be alternative to that using stainless steel coils in closing small PDA.

P815

Transcatheter retrograde closure of muscular ventricular septal defects with the amplatzer ventricular septal defect occluder: case report

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Objectives: The aim of this study was to close muscular ventricular septal defect(MVSD) in a child with a Amplatzer ventricular septal defect occluder (AVSDO). Methods: A 5-year-old girl with MVSD underwent transcatheter closure using the AVSDO. The device is a modified self-centering and repositionable Amplatzer device that consists of two low profile disks made of Nitinol wire mesh with a 7-mm(d) connecting waist. A soft J-tipped 260-cm exchange 0.035 in guide wire from a retrograde femoral arterial approach was passed from the left ventricle to right ventricle, guided by right coronary catheter, and advanced into the pulmonary artery, where it was snared from a percutaneous femoral venous approach. The catheter was removed and a 6-F long sheath was advanced over the wire from the femoral artery to the right ventricle. Under fluoroscopic and transesophageal ultrasonic guidance, the first disk was deployed and pulled gently against the septum, which was both felt and observed by TEE. The sheath was pulled back and the second disk was deployed. The device was released when its position was optimal and interference with atrioventricular valve structures had been excluded by TEE with color flow Doppler. After release of the device, both color Doppler echocardiography and left ventriculography were performed to detect residual shunts. The patient was discharged on the third day after the procedure on

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

aspirin 3 mg/kg daily for six months.Results The location of the defect was midmuscular.The MVSD diameter was 3.7-mm.The selected device size(waist diameter) was 6-mm.Device placement was successful and complete occlusions occurred immediately. No complications were observed.Conclusions.This encouraging initial clinical success indicates that the AVSDO is a promising device for transcatheter closure of MVSDs in children and retrograde approach is as effective as anterograde approach.Further clinical trials and longer follow-up are needed before the widespread use of this technique can be recommended.

P816

Developing of transcatheter occluder for fenestrated Fontan: from basically to animal experiments.

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Fenestrated Fontan is effective procedure for borderline of Fontan candidate. We had closed fenestrations with Rashkind PDA occluder, Clamshell ASD occluder, Angel wings ASD occluder and coil. But transcatheter occluder to close fenestration is not available except coil in Japan now. Therefor we are developing transcatheter occluder for fenestrated Fontan with simple and less thrombotic structure. We report animal experiment with this occluder. [Method] This occluder is made with shape memory nitinol wire. This occluder is making left atrial side three hooks, right atrial side disc and one string for self-centering. The shape memory appears over 30 degree and keeps form stiffly. But occluder is changed to soft in cold water and easy to load inside of catheter. Occluders implanted to foramen ovale of pigs (20-25kg) with fluoroscopy and echo monitoring instead of fenestrated Fontan baffle. Retrieve was also tried by retrievable catheter. Chronic experience was done to know endothelialization to occluder. Fenestrated Gore-tex sheet with occluder implanted right atrial appendage in chest chip by surgery. [Result] Implantation was possible in three pigs under fluoroscopy and echo guide. Retrieve was also possible after implantation with retrieve catheter after implantation. Sacrifice of acute experiments showed occluder set foramen ovale of pig stiffly. Echo monitoring showed relationship between foramen ovale. Endothelialization to occluder was also good because Gore-tex sheath, and occluder covered by fibrous tissue and endothelium fully six month and one year after implantation and those dogs done not have any complication. Branch pulmonary arterial fracture is doing now.

P817

Severe aortic valve stenosis in early infancy; predicting factors for survival

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Purpose of the study: Retrospective multicenter study on the result of treatment of severe aortic valve stenosis in infants(age up to 3 months), stressed on possible differences for surgical valvotomy(SV) or balloonvalvuloplasty(BV) as initial treatment. Study period was 1991–1999 with a follow-up until 1-1-1999. Results. 44 patients were studied; 25 had SV(gr.I) and 19 had BV(gr.II) as first treatment. In gr I bodyweight was lower(p<0.01) and there was more protein-dependancy(p<0.05). Procedure-mortality: gr I 2/25 and gr. II 1/19. In-hospital mortality: gr I 3/25 and gr II 4/19. There were 25 reinterventions and 16 patients no reintervention (gr I 6 and gr II 10). Aortic insufficiency after the first intervention. Mild: in gr I 8 and 7 in gr.II. Severe in gr.I 5 and in gr II 2. Stepwise logistic regression: mitral Z-score(p<0.004), protein-dependancy(p<0.01)and apex-forming LV(p<0.2) are significantly related to mortality and BV is significant related to freedom of reintervention(p<0.01). Conclusion: For severe aortic valve stenosis in infancy BV is preferable over SV as initial treatment based on a lower reintervention-rate, not a higher mortality and not more aortic valve insufficiency. For a small LV (mitral and/or aortic Z-score <-2) a Norwood procedure is a better option.

P818

Transcatheter occlusion of residual patent ductus arteriosus after surgical ligation

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Purpose: To evaluate the immediate and short to moderate-term results of transcatheter occlusion of residual patent ductus arteriosus (PDA) after surgical ligation using three types of occluder. Methods: Between March 1999 to November 2000 among 270 patients with PDA who underwent transcatheter occlusion, 12 patients (4 male, 8 female) had residual PDA after surgical ligation. Patients ranged in age from 4 to 48 years (median 15.9 years) and in weight from 15 to 82 kg (median 45.6 kg). Of these 12 patients, three types of occluder were used. Rashkind device, detachable Cook PDA coil, and Amplatzer duct occluder (ADO) were used in 1, 1 and 10 patients, respectively. Physical examination, chest radiograph, and echocardiography were performed 24 hours after device placement in all patients. 10 patients completed a 1 to 18-month follow-up. Results: A total of 12 devices were successfully implanted in the 12 patients. There were no complications. Minimal ductus diameters ranged from 2.0 to 8.0 mm (median 3.0 mm). According to the angiographic classification described by Krichenko et al, 11 PDAs were type A, 1 PDA was type B. Angiography showed that 9 patients (75.0%) had complete immediate closure. 3 (25.0%) had a trace residual shunt at 10 minutes after implantation of the device. Echocardiography revealed complete closure in all patients 24 hours after the procedure. 10 patients were followed-up 1 to 18 months. There was no instance of the device migration and ductus recanalization. Conclusions. Transcatheter closure is a safe, easy and effective non-surgical method in the treatment of residual PDA after surgical ligation. It may be an alternative to second surgery. Minimal ductus diameter more than 2.0 mm is suitable for ADO occlusion, 2.0 mm or less for detachable coil.

P819

Transcatheter closure of right-to-left interatrial shunts with amplatzer septal occluder

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We report our results of using Amplatzer septal occluder to close right-to-left interatrial shunts in systemic oxygenation. Between April 1998 and March 2000, 8 patients at a median age of 9.1 (range 2.1 to 17.5) years and median weight 28.2 (range 7.5 to 57.6) kg underwent transcatheter closure of interatrial right-to-left shunts under general anesthesia with transesophageal echocardiographic guidance. The right-to-left interatrial shunts were associated with Fontan fenestrations (n=4), pulmonary atresia or critical pulmonary stenosis post-right ventricular outflow tract reconstruction (n=3) and balloon valvoplasty (n=1). Transcatheter occlusions were performed at a median interval of 2.3 (range 2.03 to 17.3) years after the aforementioned interventions. The procedural time ranged from 75 to 250 (142±55) minutes and fluoroscopic time from 13 to 42 (26.9±11) minutes. A single occluder, ranging from 6 to 24 mm in size, was placed in all but one patients who required 2 occluders (18 and 20 mm). There were no procedural failures or complications. The systemic arterial oxygen saturation rose from 79±9 % to 94±2% (p=0.0008), while the right atrial pressure remained unchanged (11±4 vs 13±4 mm Hg, p=0.60). Color Doppler imaging revealed no leak through all devices at a median of 0.6 (range 0 to 5) month after their placement, although trivial to moderate rough separate atrial communications were noted in 2 patients. Post-Fontan patients (n=4) received long-term warfarin, while the others (n=4) had 6 months of aspirin. One patient, however, developed episodes of presumed transient ischemic attack. Elevated jugular venous pressure with hepatomegaly was noted in 3 patients on follow-up. The Amplatzer septal occluder effectively eliminates right-to-left interatrial shunts and improved arterial oxygen saturation. Nonetheless, systemic venous congestion might worsen and warrant diuretic therapy.

P820

Transcatheter closure of patent ductus prestenosis (pda) in infants using the amplatzer duct occluder (ado)

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12 infants under 1 year of age (age: 1 to 11 months, median: 4.7 month; body weight: 2.6 to 8.7 kg, median: 4.4 kg) with moderate-to-large PDAs were considered for transcatheter closure with the ADO. All of them were presented with clinical symptoms of heart failure. In 6 patients pulmonary hypertension were present. The mean PDA diameter, measured angiographically, at the narrowest end was 2.9 ± 1.0 mm (range: 1.5 to 5 mm) and devices with its cylindrical portion 2 to 4 mm larger than the PDA were selected. A 6 or 7 F long sheath was used for transvenous delivery of the ADO. 10 out of 12 patients had successful device placement with complete PDA occlusion. No

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

obstruction of the pulmonary arteries or aorta and no late complications were observed (follow-up period: 4 months to 3 years). In 2 infants (2.6 and 4.4 kg bodyweight) the attempt of transcatheter closure was not successful and the procedure had to be abandoned. Procedure related difficulties occurred in 9 of 12 cases and led to relatively long procedure and fluoroscopy times (procedure time: 50 to 180 min, fluoroscopy time: 5,2 to 49 min). In infants with PDA the ADO offers an alternative to surgical treatment but further improvement of the implantation system is necessary before the procedure can be recommended as treatment of choice.

PB21

The changes of the heart haemodynamics after atrial septal defect closure by amplatzer septal device occluder.

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The aim of investigation was the assessment of size and shape early changes of the heart right and left ventricles (RV, LV) and heart haemodynamics after intravascular closure of atrial septal defects (ASD) by Amplatzer Septal Device Occluder. 5 children at the age of 5.33±1.97 years with ASD from 0.6 to 3.0 cm in diameter were operated. Heart echocardiography (EchoCG) was made before operation and on 1 day, 30 days and 90 days after operation. Heart catheterization and angiocardiography (ACG) in two projections were made before and after the occluder implantation. 27 ACG and 15 EchoCG parameters were studied. There was an increasing of indices of myocardium relaxation and Veragut. The decreasing of end diastolic index (EDI) of inflow and outflow sections (IS, OS) of RV is lower then decreasing of end-systolic index (ESI) that leads to a small decreasing of perfusion index and simultaneous increasing of banish fraction (BF) of IS, OS and whole RV. It is necessary to define that OS plays the main role of these RV changes because its EDI and ESI decrease more then three times while the analogous indices of IS change less then a quarter. After the short period of decreasing the mitral blood circulation increased to the end of the first month. Time of isovolemic relaxation (TIR) of lung vents (LV) have been decreased just after the operation and then practicaly remained the same. Diastolic diameters of LV and RV practicaly remained the same, also the systolic diameter of left atrium (LA), which after a small decreasing became like before operation. Thus, the most optimal for styding of the early changes of intracardiac haemodynamics after ASD occlusion are: indices of myocardium relaxation, Veregur, EDI, ESI of OS of RV, time of isovolemic relaxation of LV and systolic diameter of LA.

PB22

Transcatheter patch occlusion of cardiac defects: early clinical experience.

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Transcatheter patch (TP) occlusion of atrial septal defects (ASDs) in piglets has been found effective and safe. Polyurethane TPs require 48 hours to be endothelized in the septal wall, through fibrin infiltration and inflammatory reaction. Endothelialization occurs after 10 days. TP requires minimal rim (patch diameter 2 mm larger than defect diameter); it is retrievable and retractable in the introducing sheath (9–12 F). Requirements for a successful subsequent TP repair is full test occlusion by the stang balloon and no interference to critical structures. The following heart defects inappropriate for disk device repair, were occluded by TPs since December 1999: Fourteen secundum ASDs, 1 sinus venosus ASD, 1 membranous ventricular septal defect(VSD) and 1 patent ductus arteriosus(PDA). All but 3 TPs were supported by double balloons. The ASD size varied from 12–34 mm (med 27), the VSD was 12 mm and the PDA 21 mm. Patient age varied from 1.5–58 years (med 19). All patients had immediate full occlusion of their defects by the balloon/patch; however only 15/17 were successfully endothel by the released patch after 48 hours. One patient who suffered an anesthesia related respiratory arrest received continuous heparin, in another patient the balloon/patch was not in contact with the septum. In both patients the patches were retrieved through the introducing sheath. An additional case had significant residual shunt from premature leakage of the occluding balloon. In conclusion, TP application is easy, effective and safe in the occlusion of heart defects inappropriate for disk device repair, despite initial learning and technical problems. Advantages include wide application and safety margin, disadvantage is the need for 48 hours hospitalization. Larger clinical trials are justified.

PB23

Interventional treatment of acute lung bleeding during open heart surgery

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Acute lung bleeding in adult patients during open heart surgery is a severe complication with a considerable mortality. We report a case of a 71 years old woman with aortic valve disease who underwent valve replacement. On restarting pulmonary perfusion severe bleeding occurred from the endotracheal tube. Weaning from cardiopulmonary bypass appeared to be impossible because of air embolization into the left atrium, verified by transoesophageal echocardiography. Bronchoscopy showed severe bleeding from the right lung. After puncture of femoral vein a 6 French Wedge catheter was positioned into the right pulmonary artery. Selective pulmonary artery wedge-angiography of the right upper, middle and lower lobe artery was performed using a C-arm for selective demonstration of the pulmonary venous return. Catheterization was done under bronchoscopic guidance. Pulmonary venous return of the lower lobe showed mass of air bubbles in the right lower pulmonary vein and contrast medium entering the right bronchus. The wedge catheter was replaced by a 10 mm diameter Opta™ balloon catheter using a 0.35 inch exchange guidewire and the right lower pulmonary artery blocked by inflation of the balloon. Bleeding stopped immediately. The patient was weaned from the extracorporeal circulation and transferred to the intensive care unit with the balloon catheter in place and unilateral ventilation of the left lung. After 16 hours the balloon was deflated and removed under bronchoscopic guidance. No further bleeding or air embolization occurred and the patient was weaned from the respirator the next day. Conclusion: Acute lung bleeding can be treated by temporary occlusion of the feeding vessel. This considerate and quick procedure can be performed during open heart surgery. For intraprocedural imaging a mobile C-arm and a bronchoscope are needed."

PB24

Amplatzer occlusion systems in unusual positions: different

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Occlusion systems like the Rashkind devices have been used for several indications during the past years. New devices have superseded these systems, involving a broadened spectrum of therapeutic options. Between 2/99 and 6/00 six patients at the age of 0.8 years to 15.8 years, mean 7.3 years and a bodyweight between 8.6 Kg and 45.7 Kg, mean 22.7 kg were treated using 5 Amplatzer PDA occlusion systems and 1 Amplatzer ASD occlusion systems. In one patient a venous fistula between spleen and kidney was closed by an 6 mm ASD occluder. A large mayor aorto pulmonary collateral artery was occluded by an 8x6 PDA occlusion system and a post traumatic arterio-venous fistula between vertebral artery and superior caval vein was closed by 6x4 PDA occluder. The remaining three patients with complex cardiac malformations had venous fistulas with right to left shunt, two after Fontan procedure and one after Glenn anastomoses. In two patients a connection between left caval vein and coronary sinus and one hemiazygos vein were occluded by PDA systems. A 9 months old baby with supra (stenosed) pulmonary artery and fenestration after Fontan procedure was treated with a 4 mm and 5 mm Amplatzer ASD occlusion systems. One of them was implanted into the fenestration, the other one into the main pulmonary artery. The occlusions were successful without intra- and post-interventional complications.

PB25

Simultaneous stent and pacing system implantation.

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We present the case of 2,5 years old girl, who underwent the total correction of Fallot Tetralogy at the age of twenty month. The right pulmonary artery has been damaged and the atrio-ventricular disimpery occurred during the operation. The child was asymptomatic till the age of two and half. During hospitalization the complete AV block and severe right pulmonary artery was diagnosed. The patient required cardiac pacing and dilatation of pulmonary artery. Because transvenous pacing leads could disturb correct fixation of the stent we decided on perform implantation of cardiac pacing system and stent at the same time, during one procedure and anesthesia. They underwent without complications with good result. Actually in 2 years follow-up the

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

child feels good, she has no cardiac insufficiency and cardiac arrest. Probably the complex intervention procedure in children will be much more frequent in the future

PB26

Severe aortic coarctation in infants less than 3 months:successful palliation by transumbilical and transfemoral artery balloon angioplasty

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The optimal management of severe aortic coarctation in the young infant is controversial, although we and others have used balloon angioplasty effectively. We analyzed and compared the immediate and follow-up results of transumbilical(TU) and transfemoral(FA) balloon angioplasty. Successful palliation is defined as avoiding surgical intervention for six weeks or longer. During a 5.5 year period ending July 2000, 45 neonates and infants less than 3 months old underwent TU(N=22) or FA(N=23) balloon coarctation angioplasty. The age of the patients in the TU group varied between 1 and 21 days, whereas in the FA group it was 7 to 90 days. Associated defects were more common in the TU (16 of 22) than in the FA (10 of 23) group. Acute reduction (p<0.001) of peak gradient occurred in both the TU (34±12 vs 4±5 mmHg) and FA (45±17 vs 8±6 mmHg) groups along with an increase (p<0.01) in the diameter of the coarcted segment. Improvement in heart failure and/or hypertension occurred in both groups. Surgical repair of coarctation within six weeks of BA was undertaken in 1 of 22 in the TU and none of the FA patients. Blood loss requiring transfusion occurred in 2 patients in each group and pulse perfusion requiring warming of contralateral extremity and/or heparin occurred only in FA group (6 of 22). At 1.5 years mean follow-up (6 months to 4.5 years), 8(36%)in the TU group and 6(27%)in the FA group underwent repeat balloon angioplasty and 3(14%)in the TU group and 6(26%)in FA the group required surgical repair. Based on these data it is concluded that effective palliation was accomplished in both TU and FA groups. Balloon angioplasty is equally effective in both groups but the femoral artery complications are present only in the FA group

PB27

Coil closure of the small patent ductus arteriosus in infants and small children without arterial access

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Background and Objectives: For coil closure of the patent ductus arteriosus (PDA), arterial access is traditionally considered mandatory. Arterial access necessitates heparinization and can result in bleeding or pulse loss, especially in small infants. We describe a technique of coil occlusion of small PDAs using venous access alone. Methods and Results: Of the 248 patients who underwent PDA coil closure at our institution (July 1998 - July 2000), we attempted to coil occlude PDA in 36 patients (age: 4 months-6 years, median 11 months, wt: 4.5 - 13 kg, median wt: 8kg, size of duct at PA insertion 1.8- 3 mm) electively without arterial access. The duct was crossed via the pulmonary artery and a hand injection in the aorta, just beyond the duct ampulla was used to outline the duct anatomy. Doppler color flow imaging was used in the cath lab to confirm duct closure. For 8 patients (22%) arterial access was obtained because of accidental arterial puncture (4), unsatisfactory echo window (1), and embolization of coils to descending aorta requiring retrieval (2) and additional coil delivery via arterial route (1). Heparin was not administered in any of the patients except when arterial access was required. There were no procedural complications. Complete closure was achieved in 33 patients (92%) in the cath lab and a colour Doppler after 3 hours in the remaining three patients showed no residual flow. The mean fluoroscopic time was 8.5 ± 3.3 min (range: 2.2 - 9.5 min). Follow up data at 3 months was available in 27 patients (75 %). None had any residual flow. Conclusion: It is feasible to coil occlude carefully selected patients with small PDA using venous access alone. The potential advantages include avoidance of heparin use and arterial injury

PB28

Biopsiome-assisted simultaneous delivery of multiplecoils for occlusion of the large patent ductus arteriosus

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Background and Objectives: Coil occlusion of the large patent ductus arteriosus (PDA) is technically difficult and associated with frequent embolization. Occluder devices are useful but expensive alternatives. We describe a novel method that allows biopsiome-assisted delivery of multiple Gianturco coils simultaneously for occlusion of the large patent ductus arteriosus (PDA). Methods and Results: Seven patients (2.5-64 years, median 15 years) with large PDAs (4.9±1.0 mm, range: 3.5-8.4 mm), PA mean pressure 34±16 mm Hg, pulse pressure 67±19mm Hg) underwent biopsiome-assisted occlusion with multiple coils at our institution. Two or more coils were interwinded as one coil and held by a biopsiome (5 F) and pulled into a short introducer. The coils were then deployed in the PDA via a long sheath (7-11F) previously placed across the duct via the femoral vein. Additional coils were deployed from the arterial ends if residual flows were seen. The jaws of the biopsiome were opened once the position of the coils was deemed satisfactory. The procedure was uneventful in 5 patients (fluoroscopy time: 6-23 min) and prolonged in 2 patients (fluoroscopy time of 72 and 120 min) because of dislodgement of the coil mass and embolization of an additional coil. Successful coil deployment was feasible in all patients. Two patients had transient hemolysis and required repeat coil deployment for flow elimination. Final PA pressures declined to 22±9 mm Hg. The most recent color Doppler showed complete elimination of flows was achieved in all patients. LPA flows were unaffected in all but one patient who had a 8 mm Hg gradient. Conclusion: Biopsiome-assisted PDA occlusion using multiple coils delivered simultaneously may be a promising alternative to devices for transcatheter closure of large PDAs.

PB29

Neointimal proliferation of pulmonary artery after stent implantation in children with congenital heart defects

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Intravascular stents are well accepted in the management of vascular obstruction including peripheral pulmonary stenosis (PPS) in children with acquired congenital heart defects. However, it is not clear about neointimal proliferation as a late complication in pulmonary artery (PA). Patients and methods: Fifteen stents were placed in 9 patients with postoperative PPS at the age of 12.2 +/-5.5 years. Original diagnoses were TGA, TOF, PA/VSD, SV in 2 each and PA/IVS in 1. Six patients were after total correction and 3 were after palliative surgery with bidirectional cavopulmonary shunt. On pulmonary arteriography performed 18.5+/-10.6 months after stenting, a gap between stent and contrast medium in PA was measured in each stent and neointimal proliferation ratio (NPR) was obtained by the gap divided by adjacent stent internal diameter. Results: NPR was ranging from 5.8 to 33.6% (mean: 17.5+/-7.7 %) showed a positive correlation to the degree of the stent deformation, which obtained from maximum and minimum stent internal diameter ($p<0.001$, $r=0.76$). Moreover, there was a linear correlation between NPR and the dilatation ratio of pre- to post-stenting PA diameter ($p<0.001$, $r=0.73$). However, there was no relation between NPR and PA pressure, hemoconcentration, follow-up time or flow characteristics in PA. These results suggest us that neointimal proliferation in PA after stenting might be related to the deformation of stent and the forced dilatation of original PA.

PB30

Comparison of atrial septal defect closure using amplatzercm septal occluder with surgery

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Objectives and Methods: Our study reports the results of a comparison of closure of ASD surgically with transcatheter closure using the Amplatzer TM Septal Occluder in one patients between January 1999 to November 2000. Results: There were 120 patients who had ASD. All 79 patients in group I (surgery) had a successful operation with only two patients with a mild residual shunt. There were 41 patients enrolled for transcatheter closure of the ASD (Group II). The median age for group I was 25 (from 2.3 to 64) years old compared to 12.7 (from 2 to 70) years old in group II ($p = 0.179$). In group I, the mean ASD diameter measured was 25.4±9.7 mm compared to 23.4±5.4 mm in group II ($p = 0.001$). Devices were deployed in 35 patients with sizes from 10 to 30 mm (median = 24mm). Device was not successfully deployed in six patients. One patient had a device embolized into the right ventricle (surgically removed and closure of the ASD). Complications were found in 20 patients in group I and 6 patients in group II. Hospital stay in group I was longer than in group II ($p<0.001$). Average charge was US$ 2,650±580 in

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

group I compared with US$ 4,760±397 in group II (p < 0.001). Complete occlusion was found in 33out of 34 group II patients (97%) during the follow-up periods (18.6±7.6 months). Conclusion: The AmplatzerTM Septal Occluder is a new device for closure of different sizes ASD with excellent closure results. The benefit for each patient was demonstrated in lower morbidity and a shorter time spent in the hospital.

P831

Coronary arteriovenous fistula – nonsurgical treatment with transcatheter coil embolization.

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Coronary arteriovenous fistula (CAVF) was treated traditionally by surgery. Recently transcatheter closure has been reported. This study evaluates our results with nonsurgical treatment of CAVF using coil embolization. After hemodynamic and angiographic evaluation, a coronary angioplasty guide wire (GW) was inserted selectively into the CAVF. A floppy Teflon catheter was passed over it. The GW was replaced by a stiffer GW to insert a 5F Judkins right coronary catheter to the fistula as deep as possible. Multiple spring coils (average 7) were embolized through this catheter to achieve complete closure or filling of CAVF. Patients were followed up with color Doppler echocardiographic examination. Over the past 3 years we anticipated to close CAVF in 5 children – age 10 months to 11 years. CAVF originated from the right coronary artery in 3 and from the left in 2. Four drained to the right atrium through a dilated tortuous pathway with a narrow distal end. One drained to the right ventricular apex through a dilated tubular right coronary artery. Immediate complete closure was achieved in 3, while 1 required 2 procedures. On follow up with Doppler echo 6 months later, 4 (80%) had complete closure and 1 had tiny residual shunt. There was no immediate or late complication. Our experience suggests that trans catheter coil embolization is a simple, safe and effective nonsurgical treatment for CAVF.

P832

Middle and large PDA detachable coil closure.

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PDA detachable coil closure is a method of choice in treatment of small PDA in Europe. Coil closure if ductus > 3 mm is still discussed and controversial. The modification of technique is required. The rate of complication is higher than in treatment of small ducts. We present results of closure PDA > 3 mm in the group of 96 ducts treated in Paediatric Cardiology Department in Gdańsk in years 1990–2000. In this group 17 ducts > 3mm were found. We present 15 cases of PDA > than 3 mm treated with detachable coil closure. In 12 children modification technique of implantation was needed, and 12 PDA's were closed with more than 1 coil. Mean time of procedure was 36.8 min. One case of coil migration occurred. Residual flow and hemolysis was observed in 1 case; patient required surgical intervention. In 2 cases of PDA's bigger than 2 mm coil closure was impossible – The Amplatz Duct Occluder was successfully used. PDA coil closure is useful method of treatment patients with duct > 3 mm but needs modification of implantation technique.

P833

Nonsurgical closure of anomalous artery supplying right lung lobe.

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Anomalous artery supplying right lobe is a rare malformation. It may be a part of a complex abnormality, scimitar syndrome or pulmonary sequestration. Clinical manifestations are not prominent. The symptoms depend on the severity of pulmonary hypoplasia, the size of left-to-right shunt and pulmonary artery pressure. That abnormality may be asymptomatic or may caused the heart failure in neonates period. Surgical ligation or nonsurgical closure can be performed in patients with indication for treatment. We report two cases of nonsurgical closure of anomalous artery. The first is a 2-month old baby with scimitar syndrome, pulmonary hypertension and heart failure. The second is asymptomatic 9 year-old boy with scimitar syndrome and left ventricular enlargement. Nonsurgical treatment – coil embolization was performed. In both cases for complete occlusion of anomalous artery 11 coils had to be implanted. There were no complications during procedure. Immediate and follow-up results were good. During 2-years follow-up the pressure in pulmonary artery in 2-month-old boy were normalized. The size of left ventricle in second case decreased. Nonsurgical embolization of anomalous artery may be alternative method for treatment in some patients. This procedure is safety and less invasive than surgical ligation.

P834

PFO closure: has its time come too?

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PFO closure: has its time come too? The role of the patent foramen ovale (PFO) as a pathway for paradoxical embolism has been established in patients with cryptogenic stroke. In addition, size of the PFO, amount of right to left shunting and coexistence of an atrial septal aneurysm seem to influence severity and recurrence rate of ischemic stroke and transient ischemic attack (TIA). Antiplatelet drugs, warfarin, surgical and catheter closure have been used to prevent recurrent ischemic events. Between 6/1995 and 11/2000 202 patients (mean age 42.2 +/- 19.6 years) have undergone PFO closure at our unit. Diagnoses were ischemic stroke 106, TIA 81, peripheral arterial embolism 5; 54 had multiple events. The patients received 5 different devices: 13 Rashkind occluders, 18 Amplatzer septal and 82 Amplatzer PFO-occluders, 73 CardioSeal and 16 Starflex devices. Time of fluoroscopy was 8.3 +/- 4.5 minutes. Early complications included 2 device embolizations, 5 retroperitoneal hematomas and 2 cardiac perforations; no patient died. Eight patients had late atrial arrhythmias. On TEE 3 to 6 months after implantation we found 13 residua. Leaks which occured mainly with Rashkind and CardioSeal occluder. We followed 175 patients for 3 to 62 (24,6 +/- 14,2) months. Three patients had a TIA following the procedure. We now overlook 170 patients with 204 symptom-free patient years. Catheter closure of the PFO is a simple, effective and quick method which ensures a high closure rate, avoids life-long anticoagulation and has a low recurrence rate of neurologic events. It has become the most frequent catheter intervention in our unit. As there also seem to be connections of a PFO to diseases like migraine or transient global amnesia, the range of PFO-closures has come.

P835

Should management of coarctation be different in children and adults?

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Background: Whether indication for and outcome of balloon angioplasty for coarctation are the same for childhood, adolescence and adulthood has not been established. Results and follow up of this procedure are compared for different age groups. Methods. Balloon angioplasty for coarctation of the aorta was performed in 45 patients from 1990 to 2000. We classified according to age and native / recoarctation. Groups A (n=30) (mean 4.8 years) and B (n=14) (median 29 years) included native coarctations. Groups rCoA A (n=31) (mean 6.1 years) and rCoA B (mean 20 years) included recoarctations. Follow up included 2D-Doppler echocardiographic studies and angiography or MRI. Decreases in pressure gradients were compared using independent-samples T-test. Kaplan-Meier and Logrank analyses were performed to compare long-term outcome. Results. No mortality occurred. Immediate success was equal in groups A and B: 94% (30patients) and 94% (31 patients) in group rCoA A also. Dilatation was unsuccessfull in all 3 re-coarctations of group rCoA B. Residual pressure gradients decreases were 23,1 mmHg in group A, 51,4 mmHg in group B and 18 mmHg in group rCoA A. Independent samples T-test (2-tail, unequal variances) determined a significant difference (p<0,001) in decrease of pressure gradients between groups A and B. Hospital stay varied from 42–48 hours for all patients. Follow up ranged from 0.1- 9 years. The Kaplan-Meier curves of groups A and B are not significantly different. Aneurysm formation was encountered in one patient. Conclusion: Data presented and those reviewed from published reports show that balloon angioplasty for native coarctation in both selected children and adults is the first choice therapy, although immediate results of balloon angioplasty for native coarctation in adults group are lower. In recoarctation, we recommend balloon angioplasty only in the pediatric age group.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P836

Asd and pfo percutaneous closure in infants requiring liver transplant.

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Closure of any interatrial communication is mandatory before liver transplant (LT). Five patients (age 0.83±0.18 yrs, weight 7.66±1.22 Kg), underwent percutaneous closure of ASD (3) or PFO (2). Two Cardioseal (17 mm) and 3 Amplatzer (5.7-10 mm) devices were implanted under fluoroscopic monitoring. All devices were correctly positioned, with abolition of the shunt. Mean procedure and fluoroscopy time were 63±12 (55-90) min and 22±6 (13-30) min. No complications occurred. One patient died 1 month later after a second LT. At a mean follow-up of 2.39 yrs (mean 2.4±0.6) echocardiographic control demonstrated optimal result in the 4 survivors. Permanence closure of ASD and PFO before LT is feasible in a very low body weight with and carries an anesthesiologic risk lower than that of surgery. Our experience promotes percutaneous closure of ASD or PFO as the elective therapeutic approach in infants candidates to LT.

P837

Pulmonary balloon valvuloplasty in neonates: mid term follow-up.

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To assess the results of pulmonary balloon valvuloplasty performed during the first month of life, we reevaluated 26 pts at a mean follow-up of 4±1.4 yrs (range 2.2-10.5 y). Mean age and weight at dilatation were 9 days and 3.3 Kg. Right ventricular pressure was suprasystemic in 20, pulmonary circulation was duct dependent in 13 (50%). Valvulop atry was always effective, peak gradient diminishing from 66±20 to 19±8 mmHg. One complication occurred: femoral arteriovenous fistula, requiring surgical correction. Five patients (19%) required pulmonary redilation after 1, 3, 4, and 6 months. At follow-up all patients were asymptomatic, with normal body growth and physical activity. Maximal transvalvular Doppler gradient was 37±15mmHg, being >50 mmHg in 2 (12%) and more than 50 in 1 patient. Pulmonary incompetence (10 pts, 38%) was never more than moderate. Balloon dilatation of neonatal critical pulmonary valve stenosis is safe, effective, and offers durable results.

P838

Congenital muscular vsds closure

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Surgical closure of congenital muscular ventricular septal defects (MVSDs), situated in the low or apical part of the interventricular septum, is still associated with significant morbidity and mortality. The aim of this study is to report our experience in percutaneous closure of congenital MVSDs. Between August 1998 and November 2000, 7 patients (pts) aged 5 months to 23 years (mean 10.7±12.9 years) underwent transcatheter closure of a MVSD. The patients underwent right and left heart catheterization. The location of the defect was defined by angiographic views. A arterio-venous run-in was created (IJV-RFA). After release of the device both color Doppler echocardiography and left ventriculography were performed to detect residual shunts. All patients had a chest X-ray and a transthoracic color Doppler echocardiographic study at 24 hours after the procedure and at the follow up in the outpatient clinic. The MVSD diameters at the sizing balloon ranged from 4 to 8 mm. Pulmonary/systemic flow ratio (QpVQs) varied from 1.7 to 2.5 (mean 1.9±0.34). A VSD-Amplatzer occluder device was successfully delivered in all patients. Immediate complete closure or tiny residual leak was obtained in 6 pts. The device was surgically removed and the defect closed in 1 pt. There was no mortality. The mean F-up is 204±56 days. The device is in an appropriate position, not interfering with the adjacent cardiac structures with no evidence of residual shunt in all pts.

P839

Long-term results of balloon angioplasty for pulmonary artery stenosis after arterial switch operation

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To evaluate the long-term results of balloon angioplasty (BA) for pulmonary artery stenosis (PS) after arterial switch operation (ASO) in patients with transposition of the great arteries, 35 patients were recatheterized a median of 4.5 years (1.2-9.3 years) after BA. Fifty stenotic lesions were dilated. The median age at BA was 2.3 years (0.4-12 years). To adjust growth-related change in the size of the pulmonary artery, the stenotic diameter was expressed as a percent of normal (%N). The growth-adjusted stenotic diameter increased from 46±13 to 72±30%N, and the pressure gradient (PG) across the stenotic lesion decreased from 36±23 to 17±15 mmHg immediately after BA. The right ventricular-aortic systolic pressure (RV/Ao) ratio significantly decreased from 0.69±0.25 to 0.51±0.11 after BA. Compared with the immediate data after BA, there was no significant change in the growth-adjusted diameter of the stenotic lesion (72±30% N after BA vs. 69±27% N at follow-up, p>0.05), and the PG (17±17mmHg after BA vs. 21±21mmHg at follow-up, p>0.05). The RV/Ao ratio also did not change (0.51±0.11 after BA vs. 0.51±0.22 at follow-up, p>0.05). Restenosis, defined as the growth-adjusted stenotic diameter occurring at the level close to a pantaloon valve, occurred in 11%. Our long-term follow up data suggest that the stenotic lesions of the pulmonary artery treated by BA do not decrease in size with the growth.

P840

Transcatheter closure of muscular ventricular septal defects using the amplatzer ventricular septal defect occluder: an initial experience

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We report our initial experience in the transcatheter closure of muscular ventricular septal defects (VSD) using the Amplatzer VSD occluder (AVO). Fifteen muscular VSDs in 6 patients were successfully closed using the AVO. One patient had this procedure as the primary therapy. The VSD's were closed in conjunction with surgical interventions in the rest: Three patients had prior pulmonary artery banding. One patient had a persistent residual midmuscular VSD despite surgical closure. Two had surgical closure of a perimembranous and inlet VSD respectively in all but one patient the deployment of the device was assessed via the right internal jugular vein. The median age was 52.7 [range 5-136.] months and the mean weight was 18.2 +7.5 (range 6-29) kg. The mean Qp:Qs ratio was 1.82+1.28 (range 1-4.3). Six apical VSD, 3 mid muscular and one anterior muscular VSD were closed using the AVO. The median device size used was 8 (range 6-14) mm. The mean fluoroscopy time was 127.5 + 69.9 (range 65-235) minutes. In 2 patients we had recurrent kinking of the sheath due to the angle of the VSD. One patient subsequently had cardiac tamponade while attempting to reposition the sheath. In the other patient after attempting many different sheath sizes, we finally had to dilated the VSD prior to the occlusion. Complete occlusion was achieved in all but 2 VSDs at a median 14.5 (range 5 to 29) months follow up. One patient developed pulmonary vascular disease. We conclude that the Amplatzer VSD occluder is a safe and effective device for the transcatheter closure of muscular VSDs. However more clinical trials are warranted before it can be recommended for general use.

P841

Closure of multiple atrial septal defects (asds) using single amplatzer septal occluder.

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Objective: To study the feasibility and results of device closure of more than one atrial septal defects using a single Amplatzer septal occluder. We present our experience of 8 such cases. Material and method: Between May 1998 & Nov 2000, 76 patients underwent attempted device closure of ASD. Eight of these had more than one defect in the area of fossa ovalis. Their age ranged between 4-44 years (mean 20.1 years) and weights ranged between 15-72kg. (mean 64kg.). The procedure was carried out under general anesthesia and with the guidance of TEE. Routine left and right heart catheterization was performed. Under the TEE the largest of the ASD was crossed with a multipurpose catheter and was then used using a balloon occlusion catheter. Amplatzer septal occluder 2mm larger than the stretch diameter of the defect was deployed, such that the rims of the device would overlap the smaller defects, resulting in complete closure of all the defects with one device. Results: Two patients had cribriform defect, 2 had 3 defects and 4 had 2 defects in the area of fossa ovalis. The size of the largest defect ranged between 5-17mm (mean 11mm). The balloon stretch size ranged between 10-26mm (mean 20.5mm). The device size ranged between 12-36 mm. (mean 21mm). The device was successfully deployed in 6/8 (75%) patients. One patient underwent two procedures as the largest of the defect could not be crossed on

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

first sitting. There were 2 unsuccessful attempts. These patients have been operated successfully. Four of the 6 patients have completed 6 months follow-up and show complete closure on echocardiography. One patient continues to show small residual shunt at 3 months. Conclusion: Multiple ASDs can be closed using single Amplatzer device, provided they are close to each other.

P842

Percutaneous balloon angioplasty for a 5-month-old infant with neuroblastoma and renovascular hypertension

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A 1-month-old female infant presented with poor weight gain, ill humor, and diminished sucking ability. Severe hypertension (systolic blood pressure, 200 mmHg) was noted. An echocardiogram was performed showing 31% function in the left ventricle, and an abdominal ultrasound showed a mass around the right renal hilum. Laboratory tests showed elevated plasma renin activity and aldosterone levels, >20 ng/ml/hr and >1600 pg/ml, respectively. The urinary VMA level was 135 E g/mg Cr and HVA was 54 E g/mg Cr. Biopsy specimens of the mass revealed neuroblastoma. A renogram showed 96% function in the left kidney and only 4% function in the right kidney. A renal arteriogram confirmed that there were two left renal arteries feeding the upper and lower pole, respectively. They were constricted in the proximal region. The angiogram also showed a hypervascularity in the right adrenal gland suggestive of a mass, and a few collateral arteries fed to the right kidney. Despite chemotherapy and administration of an ACE inhibitor, marked left ventricular hypertrophy and renal insufficiency were found. To improve her symptoms, percutaneous balloon angioplasty for the two left renal arteries was performed, approached via the left femoral artery with a 5 Fr balloon catheter (Rady TEAUTH, 2.0 mm / 10 mm, Boston Scientific) over a 0.014 valve wire. After angioplasty, her blood pressure fell slightly, and the serum BUN and creatinine levels improved to within the normal limit. At that time, at 5 months of age, she weighed only 3100 g. To the best of our knowledge, this is the youngest reported case of renovascular hypertension treated by percutaneous balloon ang

P843

Transcatheter occlusion of the patent ductus arteriosus (pda): a comparative study of two widely used devices

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Objectives: To identify any difference in outcome using the Cook detachable coil and the Rashkind double umbrella in PDA occlusion. Design: Retrospective study of patients in whom PDA occlusion was attempted using the Cook detachable PDA coil over a 4-year period. Comparison of these results with occlusion using the Rashkind double umbrella in the same UK regional centre. Patients and methods: From May 1996 to May 2000, 71 children and 6 adults underwent attempted PDA occlusion with the Cook coil. Between 1989 and 1996 110 children and 9 adults had a similar procedure carried out using the Rashkind double umbrella. Results: The rate of immediate complete occlusion was 24% compared to 29.9% for the Rashkind device. The figure for complete occlusion after 24 hours with the PDA coil was 93% compared with 61.5% in the Rashkind group (p>0.1). The overall closure rate in the coil group was 72% versus 74.6% for umbrellas. Complications were rare. There were 4 device embolisations in umbrella patients versus 2 in coils. Haemolysis occurred in 1 patient receiving an umbrella and 2 in the coil group. Turbulence was noted within the left pulmonary artery or descending aorta in 4 patients in whom a coil had been deployed but was absent in the umbrella group. Conclusion: The outcome in terms of complete duct closure using the Cook coil is comparable with figures obtained using the Rashkind umbrella despite subtle differences in the initial occlusion rate. Both devices have a good safety profile in the short and medium term.

P844

Stent implantation for ductus venosus of asplenia syndrome with total anomalous pulmonary venous connectio

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We succeeded in stent implantation for ductus venosus (DV). Three days-old male neonate was admitted to our Hospital with severe cyanosis. He was diagnosed as asplenia, single RV, single atrium, PA, CAVC, PDA, intracardiac type of TAPVC and bilateral SVC. As pulmonary congestion was getting worse, we decided to implant the stent in the DV. We implanted the stents into the DV via umbilical vein. After the procedure, there was enough flow through the DV and no sign of pulmonary congestion. Operative mortality rate of the palliative repair of TAPVC during neonate is extremely high. For the strategy of complex congenital heart disease with TAPVC such as our case, stent implantation for DV early after birth is the most effective and less invasive, decisi

P845

Percutaneous closure of atrial septal defects (asds) with the amplatzer device

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Percutaneous closure of ASDs with the Amplatzer device has been employed safely and effectively. In this study we report our experience with this technique. Since 10/97, 46 attempted implantations were performed in 45 patients (pts) at a mean age of 20 ± 16 years. Single defects were present in 40 cases, multi-fenestrated septum in 3, and 2 separated defects (2 devices implanted) post surgical ASD and a PFO in 1 each. All pts/op were selected with ambulatorial TEE. The mean ASD diameter was 14 ± 5 mm (8.7-26.3). TEE was repeated in 32 pts 3 months after the procedure, and in those with persistent shunt at the 1-year old implantation was seen could in 1 pt due to obstruction and 6/pts mild. In the successful cases, the mean waisted diameter was 19 ± 4 mm (10.5-36), and the mean diameter of the connecting waist of the device was 20 ± 4 mm (10-36). Four pts had non-sustained episodes of SVT. One patient had femoral artery thrombosis due to an embolized fragment of a ruptured sizing balloon requiring surgery. Total occlusion occurred in 25/45 immediately after the procedure, and in 39/45 at a mean follow up of 12.2 months. Right ventricular end diastolic diameter decreased from 135 ± 25% of upper normal range for age and weight to 96 ± 10%. Three pts have residual shunts measured at < 1 mm, and 3 at 2-4 mm on Echo, all with normal RV dimensions. There was no late embolization, endocarditis or cardiac surgical hospitalizations. Percutaneous closure of ASDs with the Amplatzer device is a safe and effective procedure in well-selected patients. Residual shunts are uncommon and do not seem to cause any significant hemodynamic burden to the right ventricle.

P846

Balloon mitral valvuloplasty in patient younger than 18 years of age. Immediate and follow-up results.

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From 6/97 to 6/00, 47 patients (pts) younger than 18 years of age underwent anterograde transseptal balloon mitral valvuloplasty (BMV) for rheumatic mitral stenosis (MS). Thirty-six (76%) pts were female, 7 (15%) were pregnant and 30 (62%) were in functional class (FC) III or IV (NYHA) at the time of the procedure. All were in sinus rhythm and the echocardiographic score varied from 5 to 10 (mean 7.7 ± 1.2). Double balloon and Inoue techniques were employed in 32 (68.5%) and 14 pts (30.5%) respectively. Success was achieved in 44 pts (93.6%). Mitral valve area by planimetry increased from 0.92 ± 0.19 to 2.01 ± 0.50 cm2. Mean diastolic gradient and mean left atrial pressure decreased from 22.1 ± 5.9 to 3.2 ± 3.3 mmHg and from 28.7 ± 6.4 to 12.5 ± 6.2 mmHg respectively. Severe mitral regurgitation occurred in 2 pts (4.3%) and pericardial tamponade in 1 (2.1%). Follow up data was available in 36 pts a mean time of 681.9 ± 399.8 months. Twenty nine (91.6%) pts were in FC I or II. Restenosis occurred in 12 pts (33.3%), 10 requiring successful redilation procedure. There were 3 late deaths. BMV is a safe and effective procedure for patient younger than 18 years of age with rheumatic MS, being the treatment of choice at our institution. Significant late morbidity and mortality remain worrisome. Restenosis was common and occurred at a short time in this series probably due to the high prevalence of the disease in our country, and maybe a more malignant course. However, redilation was feasible with no additional risks.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P347

Interventional procedures in the treatment of neonates with congenital heart defects.

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Aim of the study. To assess the immediate results of endovascular surgery in neonates. Material and methods: Interventional procedures were performed in 2700 patients with congenital heart defects. The age of 789 (26,7%) patients ranged from 36 hours to 12 months. The patients with Rashkind procedure are excluded from the study. Twenty five (3.5%) patients were below 30 days of age. All patients were critically ill. Results: In 4 cases after ineffective Rashkind procedure restrictive atrial septal defects was dilated with balloons sized 6–12 mm. As a result SaO2 in these patients increased from 34,6±4,2% to 60,2±3,9%. In 12 patients with critical valvular stenosis of the pulmonary artery (VSPA) transluminal balloon valvuloplasty (TLBV) was carried out. As a result systolic pressure gradient (SPG) between pulmonary artery and the right ventricle decreased from 102,6±24,0 to 25,1±14,3 mm Hg, SaO2 increased from 74,0±4,4% to 89,1±6,5%. Balloon valvuloplasty of VSPA was performed with good effect in two patients with tetralogy of Fallot, whose state was very critical, with SaO2 below 40%. In 6 patients with critical valvular aortic stenosis (VAS) TLBV was carried out. As a result SPG between the left ventricle and the aorta decreased from 89,2±24,6 to 20,7±12,8 mm Hg, and left ventricular ejection fraction increased from 28,5±10,3 to 59,4±9,2%. In 1 patient with II type pulmonary artery atresia patent ductus arteriosus was dilated. SaO2 increased from 40% to 88%. Two patients (8,0%) died. The deaths were caused by ballistic lesion of the left atrial wall during ASD dilatation and bilateral pneumonia after successful TLBV of valvular aortic stenosis. Conclusion: Endovascular interventions are effective for the treatment of some congenital heart defects in critically ill infants during the first months of life.

P348

Use of occluders for the treatments of arteriovenous and venoarterial blood shunting.

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Purpose of investigation: to demonstrate the possibilities of occluders in the treatment of 22 patients with following defects: atrial septal defect (ASD), aorto-pulmonary septal defect (APSD), patent ductal arteriosus (PDA), communication between right pulmonary artery (PA) and left atrium (LA). Material and methods: five types of occluders have been used: Amplatzer Septal Occluder (ASO, AGA-Med, USA), Amplatzer Duct Occluder (ADO, AGA-Med, USA), Buttoned atrial device (BAD, Sideris, Greece), Buttoned ventricular device (BVD, Sideris, Greece), Patch Occluder (PO, Sideris, Greece). ASO was used in 13 cases: 12 with ASD, 1 with APSD (remalization); ADO – in 1 case (PDA). BAD was used in 5 patients: 2 with ASD, 3 with PDA. PO was used in 2 cases of ASD. BVD was used for the closure of RPA – LA communication. The size of ASD closed varied from 8 to 34 mm, PDA from 5 to 12 mm. APSD diameter was 4 mm. Results: ASDs were successfully closed with ASO in 10 patients, in 2 cases the defect could not be closed due to miscounting of their sizes. ASO was also successfully used for APSD closure, and ADO – for PDA closure. BAD, BVD and PO were successfully implanted in all the cases. Immediately after the procedure residual shunting was seen in 3 patients: 2 with ASD after BAD and PO implantation and 1 with RPA – LA communication (despite the shunting SaO2 increased from 68% to 92%). Follow-up results were studied in 11 patients with ASD and 2 patients with PDA: 8,2±2,4 months after the procedure. Only 1 patient had residual blood shunting after PO implantation. Conclusions: The use of special occluders is an effective procedure with definite indications.

P349

Selection of occlusion devices for pda: gianturco coil and/or amplatzer duct occluder?

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Objective: The objective was to first retrospectively review device selection and effectiveness of PDA occlusion with either the Gianturco coil or the Amplatzer Duct Occluder, then devise and implement a device selection strategy. Results: From 5/95 to 12/99 at the Children's Hospital of Wisconsin, 79 patients (pts) underwent percutaneous device occlusion of a PDA. Coil delivery was performed in 70 pts and Amplatzer duct occluder in 9 pts. Follow-up echocardiograms were performed in 64 coil pts. The median maximal ductal diameter of these 64 pts was 2.0 mm (range 0.5 – 3.5 mm). Forty-two pts had a PDA smaller or equal to the median of 2 mm (Group A) and 22 pts had a PDA larger than 2 mm (Group B). Complete occlusion as defined by no residual ductal shunting on follow-up echocardiogram was documented in 40 of 42 (95%) pts in Group A and in 18 of 22 (82%) pts in Group B ($p = 0.08$ Chi-Square, $p = 0.17$ Fisher's Exact Test). The median maximal ductal diameter of the 9 pts with an Amplatzer duct occluder was 2.3 mm (range 1.5 – 3.0 mm). All 9 have complete and effective occlusion as seen on follow-up echocardiogram. Beginning in 1/00, a device selection strategy was employed: coil occlusion was initially chosen for PDA < 2 mm and Amplatzer for PDA > 2 mm. A coil has been placed in 8 pts and an Amplatzer in 7 pts, all with complete occlusion to date. Conclusion: Coil occlusion of a PDA may be less effective in the patient with a larger ductus. The device strategy of coil for PDA less than or equal to 2 mm and Amplatzer > 2 mm has been effective.

P350

Interventional procedures for complex treatment of patients with pulmonary artery stenosis.

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Goals: The patients with congenital or iatrogenous stenosis of the pulmonary artery (PAA) enter the high-risk group for the radical or hemodynamic correction procedures. Depending on the anatomy and hemodynamics of the defect multi-stage methods of surgical treatment, including endovascular procedures are being elaborated. Material and methods: At different stages of the treatment for PAA we have performed 34 different interventional procedures to 65 patients. In 18 patients after the reconstruction of the right ventricular outflow tract transluminal balloon angioplasty (TBA) of the PA segments of the PA was performed. For pulmonary artery stenosis or hypoplasia. In 6 patients TBA of the stenosed PA was performed using the approach through the fenestrated pulmonary anastomosis. TBA of the stenosed Blalock-Taussig anastomosis was carried out in 11 patients, and in 2 of them a simultaneous TBA of the anastomosis and the PA were performed. In 2 patients we performed TBA of the stenosed aorto-pulmonary collaterals with stenting in one case. In 2 cases we performed catheter closing PDA. In 19 cases we have carried out the embolization of the large aorto-pulmonary collaterals with Gianturco coils. 36 collaterals were closed completely with 80 coils. In 7 patients from this group we have also performed the dilatation of the stenosed PA with stent implantation in two cases. Conclusion: In some cases interventional surgical procedures are effective for the treatment of patients with PAA. They allow improving the anatomy and the hemodynamics of the pulmonary circulation and preparing this group of patients for radical or hemodynamical correction of the defect under favorable circumstances.

P351

The use of stents in stenotic pulmonary arteries in patients with congenital heart defects.

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Purpose: to show the possibilities of stenting at eliminating pulmonary artery stenoses. Material and methods. Twenty-five stents were used to treat 23 stenosed segments of the pulmonary arteries in 18 patients. The patients' age varied from 3.5 to 27 years (mean 13.6±6.2 years). Results: After stenting the diameter of stenosis increased in average from 5.5±2.1 mm to 11,5±2.1 mm ($p<0.0006$), and systolic pressure gradient fell in average from 51,1±12,7 to 17,4±17,9 mm Hg ($p<0.0005$). The ratio of systolic pressures in the right ventricle and the aorta (RV/Ao) decreased from 0.79±0.07 to 0.48±0.06 ($p<0.0033$). Immediate good effect of stenting was seen after dilatation in 21(95,5%) out of 22 segments of stenosed PA. Serious complications were not seen. Three technical mistakes were noted: in 1 case the underplayed stent migrated into the lower lobe branch of the PA, in 1 case the stent was excessively implanted into the trunk of PA, in 1 case the stent's position within the stenosis was suboptimal. Optimal position of the stents in the place of stenosis was obtained in 22 (88.5%) of 25 stents. Long-term follow-up studies were carried out in 12 (66,7%) patients in whom 16 stents have been implanted in 6 to 38 months (mean, 15±11,6 months) after the procedure. In all of the cases repeated catheterization and angiography were performed. All the stents were patent: no cases of migration were seen. Only in 1 case (6,3%)

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

stent was restenosed due to neo-intimal hyperplasia. Conclusions: stenting is an effective, but technically rather complexated procedure. The rate of immediate success was 95.2%, with 6.6% of late restenoses.

P852
Balloon dilatation and stenting of brachiocephalic arteries in patients with supravalvular aortic stenosis
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Purpose of investigation: to demonstrate the possibilities and the effectiveness of endovascular methods in patients with supravalvular aortic stenosis associated with aortic arch branches pathology. Material and methods: by November 2000 we have performed 13 endovascular procedures in 11 patients with congenital pathology of brachiocephalic arteries. In 10 patients supravalvular aortic stenosis was associated with congenital pathology of the left common carotid artery; in 1 patient there were pathologies of the left common carotid artery and of the brachiocephalic trunk. Results: As the first stage to prevent cerebral hypoxia during surgical correction of supravalvular aortic stenosis with cardiopulmonary bypass, the patients were submitted to balloon angioplasty of the affected brachiocephalic arteries which allowed to perform surgical correction of supravalvular aortic stenosis without significant risk. After balloon angioplasty mean area of the stenosis has increased from 11.2±0.5 mm2 to 18.7±0.3 mm2. The first stage of treatment in one patient with congenital supravalvular aortic stenosis associated with congenital stenosis of the left common carotid artery and brachiocephalic trunk consisted of balloon angioplasty of the left common carotid artery and brachiocephalic trunk stenosis. It allowed performing surgical correction of supravalvular aortic stenosis as the second stage. However, there was a need for implantation of the Smart stent (Cordis) 3.5 years later due to recurrent lesion of the brachiocephalic trunk. Angiographic and hemodynamic results after endovascular procedures were good. No complications were encountered. Conclusions: Endovascular methods of treatment of the stenoses of proximal segments of the aortic arch branches can be a method of choice in complex treatment of patients with congenital supravalvular aortic stenosis.

P853
Our experience of 175 transcatheter closures of patent ductus arteriosus.
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Aim of the study: to assess the opportunities of transcatheter closure of patent ductus arteriosus (PDA). Materials and Methods: To October, 2000 175 pts with PDA underwent the attempt of transcatheter closure. Patients were from 5 months to 24 years old. Native and recanalized PDA closure was performed using Gianturco coils (COOK) in 169 pts, DuctOccludo (PFM) in 4 pts, Amplatzer Duct Occluder (AGA Med. Corp.) in one and Buttoned device Sideris in 2 pts (in one case the combination of coil and button device used). Ductus diameter varied from 1.1 up to 9.0 mm. Gianturco coils and DuctOccludo have been used at ductal diameter less than 4.0 mm. Coil diameter exceeded the ductal diameter by twice. In 3 cases with ductal diameter from 4.5 to 6.0 mm we implanted 2 coils simultaneously using 2 delivery catheters. In 9 patients with concomitant congenital heart diseases (aortic stenosis, coarctation of the aorta, pulmonary artery valvular stenosis (PAVS), PAVS and peripheral stenosis) we performed one-step endovascular procedures such as balloon angio- and valvuloplasties and PDA embolization. Results: Complete occlusion of the ductus was achieved in 167 pts (95.4%). In 5 cases we couldn't implant coils because of ductal kinking, incompatibility of the DuctOccluder and ductal forms and PDA diameter more than 6 mm. Coil migration to the pulmonary artery occurred in 6 cases, all the coils were removed with basket device. The complete closure was achieved in all the patients using Amplatzer Duct Occluder and Buttoned device without complications. Long-term results (from 6 month to 6 year) were learned in 130 patients. In 127 cases we noted complete PDA occlusion, in 3 patients with incomplete ductal closure we performed repeated embolization with coil implantation in 2 cases and Buttoned device in one. Conclusion: transcatheter closure of the PDA is an effective and non-traumatic method.

P854
Transcatheter closure of postoperative complex ventricular septal defects in children using the amplatzer ventricular septal defect occluder.
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Two patients who had previously undergone surgery for complex congenital heart disease had residual VSDs which were inaccessible at surgery. They underwent successful device closure using the Amplatzer VSD occluder (AVSDO). One 6 year old child had TOF, Pulmonary atresia and multiple VSDs and had previously undergone unifocalization with VSD closure. She had a posterior muscular VSD hidden behind the tricuspid valve which showed significant shunting and failure post-operatively. The VSD was successfully closed using a 12 mm AVSDO, with no residual shunt on follow up. A 8 month old child with TGA and a very large VSD undergoing to single ventricle underwent PA banding initially. At 11 months of age, as PA pressures were still high, she underwent septation of the ventricle with arterial switch operation. There were 2 significant residual VSDs apically, below the VSD patch and the child remained on CHF. The larger VSD was closed using a 10 mm AVSDO, with complete occlusion. The smaller VSD was partly covered by the device and shunt was minimal across it, on follow up. AVSDO is a promising device for transcatheter occlusion of complex, surgically remote VSDs associated with other complex congenital cardiac lesions.

P855
Transcatheter therapy for treatment of arterial thrombosis in pediatric patients
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Objective: To evaluate the safety and patency rates of transcatheter recanalization of arterial thromboses in pediatric patients. Patients: 9 patients (age: 4 days – 19 months) tetralogy of Fallot (n=2), valvar aortic stenosis (n=2), pulmonary atresia (n=2), transposition of the great arteries (n=1), congenital thrombosis of the descending aorta (n=1) and acquired protein c resistance with iliofemoral thrombosis of the external iliac artery (n=1). In 7 patients the femoral (n=6) or subclavian (n=1) arterial thrombosis was due to arterial lumbar catheterization. Methods: 5 patients had recanalization using an anterograde femoral venous approach with passage of the arterial side via a ventricular septal defect (n=3) or the ductus arteriosus (n=2). 4 patients were treated using retrograde arterial catheterization via the axillary artery (n=2) or the femoral artery (n=2). The occluded vessels were recanalized using 0.018-inch and 0.035-inch guide wires. PTCA catheters with diameters ranging from 2 mm to 4 mm were used in 8 patients. 1 patient was dilatated with a 5 mm PTA catheter. 1 to 15 dilatations with pressures ranging from 10 to 22 atm were performed. Results: There were no complications. Repeat angiography or duplex-sonography 1 – 6 months post-interventionally showed completely patent arteries without residual stenosis in 8 patients. 1 patient had partial recanalization. Conclusion: Transcatheter recanalization of arterial thrombosis was safely performed in pediatric patients. It may be an effective alternative to fibrinolytic therapy.

P856
Is the amplatzer duct occluder is really safe and effective in the occlusion of moderate to large sized ductus arteriosus, even in infancy ?
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The aim of the study was to evaluate the Amplatzer duct occluder in terms of efficacy and safety for the occlusion of patent ductus arteriosus. From 1998 to November 2000, 216 patients had undergone occlusion of ductus arteriosus (55 Rashkind, 51 Sideris, 81 coils, 29 Amplatzer). We focused here on these 29 patients (including 3 infants) who had occlusion with the Amplatzer duct occluder. The mean age was 24 +/- 20 months, minimum 3 months and the mean weight was 11 +/- 15 kg (range 3.9 to 74 kg). Duct diameter was 4.0 +/- 1.2 mm (1.8 to 6 mm). Transcatheter occlusion was performed under local anesthesia in 24/29; 5 patients. One patient required an arterio-venous loop for implantation. A transvenous approach was solely used with echocardiographic control in 7 remaining patients. Implantation succeeded in all but one patient. No complication occurred except one hemiplegia following implantation that resolved after temporary occlusion of the duct ampulla by a balloon catheter. During follow-up, the rate of residual shunting on echocardiography decreased with time : 21 % at one month, 12% at 3 months, and 4 % at 1 year. The Amplatzer duct occluder has several advantages : 1/ occlusion of large duct is feasible, 2/ a weight of 3–4 kg is not a contraindication for transcatheter occlusion, 3/ implantation can be achieved by a sole transvenous approach, 4/ rate of complete occlusion is high. To conclude the Amplatzer duct occluder is really a safe and effective device for transcatheter occlusion of moderate to large sized duct, even in infancy. We clearly recommend its use instead of coil in the occlusion of ductus arteriosus with a diameter above 2.5 to 3 mm.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P857

Interventional closure of defects in the oval fossa – experience with four different types of devices

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Nonsurgical closure on interatrial communication seems to be an attractive method. Several devices have been used but not all have been successful bring far away from being an ideal device. Patients and methods: In cooperation with our adult cardiologists we performed 76 implantations out of 112 selected patients aged between 3 and 74 years. We used five different devices: ASDOS, CardioSeal/Starflex, ASD and PFO occluder AGA, and Helex device (in Berlin). There were 49 ostiumsekundum type defects and 27 PFO's with a neurological indication for closure. Results: In 76 patients, out of 112, we implanted a device. The rate of success was linked to the experience, the device and the indications. Success rate: ASDOS 47/47, CardioSeal/Starflex 58/74, AGA 14/20. Time of fluoroscopy (in min): ASDOS 31, CardioSeal 14, AGA 9. There was significant residual shunting in one PDA, we failed to implant a second device, the child was operated. In one adult a Starflex device embolised in the abdominal aorta nearly asymptomatic. The device was explanted and a 20 mm AGA system was implanted. There was no life-threatening event. Conclusion: With experience the interventional closure is a safe and successful method. We use different devices depending on the configuration of the defect, e.g. multiperforated CardioSeal, defect diameter over 20mm AGA ASD-device. The future perhaps will bring us closer to the ideal device.

We thank Prof G. Haverich (Hannover) and Dr M. Schneider (Berlin).

P858

Treatment of pulmonary artery stenoses by repeated balloonangioplasty

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Introduction: Although balloonangioplasty (BAP) of pulmonary artery stenoses is the method of choice, restenosis is frequent. Stent implantation may be helpful to reduce the frequency of restenosis, however, it is not recommendable in infants and small children. Our study addresses the question, whether repeated angioplasty improves the initial results leading to long-lasting maintenance of right ventricular pressure reduction and vessel diameter increment. Patients and methods: Between January 1996 and March 2000 25 patients with 62 pulmonary artery stenoses (2 MPA, 29 LPA, 31 RPA) and mostly hypoplastic pulmonary vessels underwent repeated balloonangioplasty with up to four dilatations. At the time of the first intervention the mean age was 4.8 years (2 months to 25 years). The mean time interval between the procedures was 9.7 months. Results: The vessel diameters steadily increased (1. BAP = 4.6 mm, 2. BAP = 5.7 mm, 3. BAP = 6.1 mm, p < 0.001) and the pressure relation between RV and LV gradually declined (1. BAP = 74.1%, 2. BAP = 56.2%, 3. BAP 52.2%, p<0.03). Conclusion: Repeated balloonangioplasty within a fixed time schedule improves the initial results and may be advantageous particularly in infants and children.

P859

Percutaneous occlusion of patent ductus arteriosus (pda) with pfm's duct-occlud system – a personal experiences

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A retrospective analysis of the data were performed to asses the efficiency and safety of percutaneous occlusion of PDA with pfm's Duct-Occlud system using standard and reinforced (hourglass and cone-shaped) spiral coils. From June 1997 to November 2000 cardiac catheterization for intended percutaneous occlusion of PDA was performed in 45 pts. (mean age 6,57 years – range from 3 months to 52 years, mean weight 24,6 kg – range from 3,9 to 94 kg, mean minimal diameter of PDA 1,7 mm – range from 0,6 to 4,2 mm). There were more female pts. (57,77%). 3/45 pts. (in 66%) had residual shunt after surgery, others had native PDA. 23/45 pts. (51,11%) had long-tubular or long-conical, 20/45 pts (44,44%) had short-broad, 1/45 pt. (2,22%) had long-aneurysmal and 1/45 pt. (2,22%) had window type of PDA. Coil was successfully implanted – mostly retropulmonary (77,5%) – in 40/42 pts. (95,23%). 2/45 (4,44%) procedures were abandoned because stable positioning of coil could not be achieved in 1 pt. and because PDA was too large in another one (minimal PDA diameter was 4,2 mm). Occlusion was not attempted in 3/45 pts. (6,66%) because of pulmonary artery hypertension in 2 pts. and window type of PDA in another one. The fluoroscopy time ranged to average 16 min. A long-term closure with 1 coil was achieved in 36/40 pts. (90%) without complications except 1/40 (2,5%) early embolisation to the left pulmonary artery. 2/40 pts. (5%) needed second coil implantation. 18 months after first procedure 2/40 pts. (5%) have residual silent PDA (1 pt. 1 year and 1 pt. 6 months after procedure). They will probably need another coil. Conclusion: the pfm's Duct Occlud system is a save and effective coil type device designed for percutaneous occlusion of long tubular, long-conical and hourglass type PDA of small-to-moderate size. A retrospective analysis of the data were performed to asses the efficiency and safety of percutaneous occlusion of PDA with pfm's Duct-Occlud system using standard and reinforced (hourglass and cone-shaped) spiral coils.

P860

Mid-term results of combined angioplasty and valvuloplasty of the aorta and aortic stenosis in newborns and infants

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Coexistence of coarctation of the aorta (CoA) and aortic valve stenosis is well documented. Balloon dilatation of CoA and AVS as isolated lesions has been reported previously by several authors. However there is only few case reports combined intervention for both lesions during single catheterization procedure. We presented nine neonates and infants with both native CoA and AVS who underwent successful balloon dilatation in a single session between August 1996 and January 1999 in Baskent University in Ankara. Patients' age ranged between 28 days and 4 years (median 54 days). Left ventricular systolic function was decreased in four patients. Three patients had echocardiographic findings of endocardial fibroelastosis. Mean aortic valve gradient and coarctation gradient were 66.2±18.9, and 18±12.9 mmHg respectively before the procedure. Aortic and coarctation gradients were decreased 24.5±15 (p<0.0001) and 8.0±8.2 mmHg (p= 0.0021) respectively. Balloon coarctation angioplasty revealed recoarctation in three patients (33.3%) in median 41 months of follow-up. Aortic regurgitation was observed in one patient. Three patients died because of endocardial fibroelastosis and intractable heart failure. Two patients were operated on for subaortic stenosis (myectomy and Konno) with aortic valve replacement in each one patient). Survival rate was 66.6 %, event-free survival rate was 22.2% in 63 months follow-up. Nine nine children with coarctation and valvular aortic stenosis who underwent successful balloon dilatation both CoA and AVS during a single procedure are presented. Recoarctation is frequent in infants under three months. Mortality is high in infants with endocardial fibroelastosis. Reintervention or operation was high in these patients.

P861

Initial experiences with transcatheter applications using the starflex and cardioseal implants.

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Objective: to review our initial experience with transcatheter applications with the StarFlex and Cardioseal devices. Methods: A total of 21 pts (9 males, 12 females) aged 13.5±11.0 years (range 3.4–44.1 years) underwent transcatheter occlusion of communications at our institution between June 1999 and November 2000. Diagnoses: 12 pts had secundum atrial septal defect (ASD), 3 fenestrated Fontan, 1 Mustard baffle leak, 4 patent ductus arteriosus (PDA) and 1 a huge pulmonary arteriovenous malformation (PAVM). The procedures for the ASDs and atrial leaks were performed under general anesthesia and transesophageal echocardiography while the rest under conscious sedation. Results: The median stretched ASD diameter was 15.0 mm (range 5–25 mm), while 2 pts had multiple defects. All 4 postoperative atrial defects were 5 mm. The median stretched PDA diameter was 5.5mm (range 4 to 7 mm) while the PAVM measured 12 mm. We implanted 12 StarFlex (sizes 22 to 40 mm) and 9 Cardioseal (sizes 17 to 28 mm) devices. the PAVM pt had 2 devices implanted. The device to stretched diameter ratio was 2.1±0.5 (range 1.7–3.4). There were no complications and, during 7.1±4.6 months of follow-up, all patients were asymptomatic with improved right and left ventricular dimensions and no arm fractures. Only 2 pts had residual trivial shunt 6 months after device placement. The device was mildly protruding in the superior vena cava in 2 pts and in the left pulmonary artery in 5 pts without flow disturbances. Conclusions: The StarFlex and Cardioseal devices may be used for occlusion of various cardiac communications with good early outcome and hemodynamic improvement.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P862

Closure of atrial septal defects with the cod buttoned device: results of FDA approved us clinical trials

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The centering on demand (COD) buttoned device for transcatheter occlusion of atrial septal defects (ASD) is a rounded 4th generation device with an unperforated centering ring. We hypothesized that the COD mechanism would increase the effective occlusion rate and decrease the device/ASD ratio compared to previous generation devices. The COD device was implanted in 24 patients (pts) in a 9 month period ending November 2000 under an FDA approved clinical trial. Pt ages were 1.5y-79y (median 5.6) and weighed 12–100.8 Kg (median 22.1). ASD size by echo 5 to 35 mm (9.3 +/- 5.8 mm), and 8 on 22 (16.5 +/- 4.8 mm) balloon stretch. The QP:QS was 1.8 +/- 0.8. The device size was 35 +/- 8 mm. (median 35). Effective occlusion, defined as no (N=16, 67%) or trivial (N=8, 33%) residual shunt occurred in all 24 patients. The device/ASD ratio was 2.1 +/- 0.44. In one patient a clot noted in the left atrium attached to the occluder resolved with rTPA, no other complications occurred. In the short follow-up period (1–9 mo) there has been no re-interventions.

P863

Transcatheter coil occlusion of tube fenestration after the extracardiac Fontan operation: simple, safe and inexpensive

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Objective: To review our institutional experience of using detachable coils for occlusion of tube fenestration after extracardiac Fontan. Methods: This is a descriptive clinical study of all patients who have undergone extracardiac Fontan with tube fenestration since our introduction of this surgical modification. We retrospectively reviewed medical records, echocardiography, hemodynamic and angiographic data. Results: Between May 1999 and November 2000, 24/27 children (median age 3.9 yrs; range 2.4–8.7 yrs) supported extracardiac Fontan with tube fenestration (diameter 4–8 mm, surgically clipped to variable diameters). Five patients had confirmation of spontaneous fenestration closure at cardiac catheterization and 5 by transthoracic echocardiography with concurrent consistent improvement in resting oxygen saturation (SaO2). Eight children underwent fenestration occlusion by transcatheter placement of 12 detachable coils (diameter: of 5–6 mm) and 8 awair cardiac catheterization. The median (range) serial SaO2 increased from 92% (85–96) to 96.5% (94–99) ($p<0.004$), while the right atrial pressure rose from 10.5 mmHg (7–13) to 13 mmHg (7–15) ($p<0.004$) after coil implantation. Angiographically, 2 patients had complete occlusion of their shunt, 5 had trivial and 1 patient had a significant residual shunt. At a median follow-up of 3.5 (1–17) months, the resting SaO2 was 94.5% (90–99). There were no immediate complications, but coil embolization, thromboembolic events, or documented hemolysis. Conclusions: The use of tube fenestration during extracardiac Fontan allows for a simple, safe and inexpensive postoperative occlusion by means of transcatheter detachable coil implantation.

P864

Balloon angioplasty of stenotic right ventricle to pulmonary artery graft: does it work?

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Reconstruction of the right ventricle (RV) to pulmonary artery (PA) passage by various grafts often results in stenosis with or without insufficiency and often necessitates multiple re-interventions. An effective palliative transcatheter intervention is desirable to postpone or reduce the number of surgical re-interventions. Balloon angioplasty (BA) of the stenotic RV to PA grafts has been used but there are only few reports with various results and with small number patients involved. From April 1996 to April 2000, 10 BA procedures were performed in 9 patients (age 1.7 to 17 years, mean 8.2 years, 7 male and 2 female). Diagnoses included TOF (5), d-TGA (2), common arterial trunk (1), and pulmonary atresia with VSD (1). The sites of stenoses were at the proximal graft anastomosis (5), pulmonary valve (2), distal anastomosis (1) and diffused graft stenosis (2). Prior to BA, the RV pressures were 75.4 ± 15.5 mmHg and RV/LV pressures 0.7 ± 0.2. The RV to PA systolic pressure gradients were 41.2 ± 11.6 mmHg. All except 2 patient had single BA. Two patients had double BA. After BA, the RV pressures were 56.0 ± 14.9 mmHg (23% reduction, $p<0.005$). The RV to PA systolic pressure gradients were 26.4 ± 14.7 mmHg (31% reduction, $p<0.001$). Balloon rupture occurred in 2 patients with no hemodynamic consequence or vascular injury upon retrieval. No patient had worsened pulmonary regurgitation. These patients were followed for 17.5 ± 15.9 months by echocardiography. Four patients needed surgical intervention for graft replacement. Our results suggest that BA of stenotic RV to PA grafts significantly alleviates the graft stenosis but the improvement is limited and palliative. Further reductions in pressure gradient in those patients who do not respond to BA might be achieved with stent implantation but with the potential risk of open pulmonary regurgitation.

P865

Transcatheter closure of patent ductus arteriosus (PDA) with the Amplatzer ductal occluder (ADO) in children : immediate and medium term results

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To assess the immediate and medium term outcome of transcatheter closure of patent ductus arteriosus with the ADO device. Ninety patients underwent transcatheter closure of a PDA between January 1997 and November 2000. Fourteen patients (11 females and 3 males) had attempted placement of the device when there was failed coil occlusion or large tubular duct. The median age was 8.5 months (2 -132 months) and the median weight 8 kg (4–29kg). The mean ductal diameter was was 3.98 +/- 1.31 mm. Immediate and medium term results were evaluated by colour flow echocardiography at 24 hours, 3 months and 1 year. Thirteen patients had the device placed successfully. Complete closure was seen immediately by angiography in 5 patients, residual trace shunt (clinically silent) in 2 patients, mild shunt in 6 and moderate shunt in 1 patient, with similar findings at 24 hours. By 3 months, 10 (77%) patients had complete closure, 2 (15%) remained to have mild shunt and 1 patient had trace shunt. At one year follow up all patients (100%) had complete closure. One patient had an unsuccessful attempt during the early series due to the inavailability of an appropriate device size. One patient was complicated by a partially obstructed descending aorta flow due to an oversized protruding disc. The median hospitalisation stay was one day. The Amplatzer ductal occluder is an easy and effective technique for selective ductal anatomy with low rate of complications and short hospital stay.

P866

Analysis of myocardial perfusion after percutaneous transluminal coronary rotational ablation in Kawasaki Disease

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Background: Percutaneous transluminal coronary rotational ablation (PTCRA) has been performed on coronary artery stenosis with calcification after Kawasaki disease (KD). It is essential to identify the assessment of myocardial perfusion after PTCRA. Method: Three KD patients (age 17 and 21 years old) underwent PTCRA for localized stenosis with calcification. Myocardial perfusion imaging by technetium-99m tetrofosmin single photon emission computed tomography (SPECT) was obtained before and after PTCRA. Exercise stress SPECT was performed under bicycle ergometer stress and at rest on the same day. Result: The targeted lesion for PTCRA was the left anterior descending artery of two patients, and the circumflex coronary artery of one patient. After PTCRA, coronary artery angiography showed that the coronary stenotic lesion reduced in all patients. In case 1, the myocardial perfusion image improved after PTCRA. After 6 months from PTCRA, the myocardial perfusion image became worse and re-stenosis was observed at the segment of PTCRA in the coronary angiography. In case 2, the myocardial perfusion image improved at rest, although became worse under the stress comparison of previous test. The coronary artery lesion was showed coronary stenosis rate under 50% in the coronary angiography. In case 3, the myocardial perfusion image improved after PTCRA, and the stenosis lesion became good configuration on the coronary angiography. Conclusion: PTCRA was useful for re-vascularization in coronary artery stenosis with calcification after KD. And myocardial perfusion imaging is useful to evaluate effect of PTCRA and assess re-stenosis after PTCRA.

P867

Stent implantation for pulmonary artery lesions after surgical repair of congenital heart disease

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https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Stent implantation for pulmonary artery lesions after surgical repair of congenital heart disease Kobayashi T., Shimahara M., Kobayashi T., Suzuki M., Namuki T., Koike N., Inoue Y*. Gunma Children's Medical Center, Gunma University School Medicine*, Gunma, Japan Pulmonary artery stenosis or atresia after surgical repair of congenital heart disease is intractable. Conventional surgical or balloon dilation therapy for such lesions has been unsatisfactory in many cases. We treated patients with pulmonary artery stenosis or atresia after surgical repair of congenital heart disease by stent implantation and assessed the short-term efficacy of stent implantation. The 5 patients who underwent stent implantation were diagnosed as TOF with pulmonary artery stenosis (n=4) or TGA with pulmonary artery atresia (n=1). A balloon-expandable endovascular stent (Palmaz stent, Johnson & Johnson) was used in all cases. The patients ranged in age from 2.5 years to 13.8 years. Most (4 of 5) were older than 11 years. Their weight ranged from 9.1 to 47 kg. Percutaneous stent implantation was performed unilaterally in 2 cases and bilaterally in one case for the treatment of postoperative stenotic lesions. Intraoperative stent implantation was performed unilaterally in 2 cases for a postoperative occluded pulmonary artery in conjunction with surgical angioplasty. After stent implantation, the diameter of the stenotic lesions increased from 5.7 +- 2.0 mm to 9.0 +- 0.8 mm (mean increase, 169 +- 46.8%). The diameter of the occluded pulmonary artery increased to 6.2 mm or 6.5 mm after intraoperative stent implantation. There was no acute complication of stent implantation. Follow-up catheterization revealed focal intimal ingrowth within the implanted stent. In conclusion, percutaneous stent implantation is an effective treatment. For patients with an occluded pulmonary artery, intraoperative stent implantation in conjunction with surgical angioplasty is a useful method. Long term follow up is necessary because of the possibility of restenosis due to intimal ingrowth.

P868

Interventional occlusion of congenital vascular malformations with the detachable Cook coil system

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Introduction: Nonsurgical closure of pathologic vascular communications may be achieved by coil embolization. Different systems which are used in pediatric patients with congenital heart disease (e.g. PDA) allow controlled release of the embolization devices, however, they are too stiff for coil occlusion of small or tortuous vessels. Microcoils delivered through a small flexible microcatheter are advantageous, however, a simple release control mechanism has been missed so far. Methods and patients: A new detachable coil system (I.A. Cook) combines flexibility with a simple release control mechanism. The system consists of a wide range of coils with a variety of configurations, sizes and degrees of softness, which are premounted on a delivery wire. A 3F microcatheter serves as a delivery catheter. Five children aged 8 days to 10 years underwent heart catheterization for interventional occlusion of different congenital vascular malformations (2 coronary artery fistulae, 2 aortopulmonary collaterals, 1 hemangioma). The diameter of the target vessels varied between 3.8 mm to 5.6 mm, the length between 10 mm to 22 mm. Results: After introduction of a 4F guiding catheter up to four (018

P869

Interventional treatment in native and postsurgical aortic coarctation in 39 children.

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An 11-month-old boy with no previous medical problems was admitted to hospital in heart failure due to sudden sepsis (Staphylococcus aureus), endocarditis and ventricular arrhythmia, which were successfully treated. ECHO showed a huge mass in the right ventricular cavity indicating a cystic teratoma. Three weeks later, the boy was submitted to surgery. A cystic tumor (6 x 4.5cm) was excised from the right ventricular cavity. The tumor originated from the interventricular septum but it was also strongly adherent to the apex and partly to the right ventricular free wall. The papillary muscles of the tricuspid valve were partially involved in the tumor mass. The child recovered uneventfully and pathologic examination revealed a mature cystic teratoma. The operative procedure, pre- and perioperative ECHO studies, pathologic, gross and microscopic findings, four year follow up and a review of the literature are presented.

P870

Atrial septal defect -the difference of heart rate variability (HRV) in patients treated with surgery or Amplatzer Septal Occluder.

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The aim of this study was to compare the parameters of HRV between two groups of patients (pts) with secundum atrial septal defect (ASD) treated with transcatheter implantation of Amplatzer Septal Occluder (ASO) or surgically (Surg). Material: In 13 pts aged 10.9 (2.5- 26) y treated with ASO the size of ASD assessed with TTE was 6 – 19 (mean 11.7) mm. In 9 pts aged 10 (3 – 40) y treated surgically – the size of the ASD was 8–22 (mean 13,4). Methods: We examined five time-domain indices of HRV determined from 24 h Holter recordings in both groups before (I), 1 month (II) and 3 months (III) after closure of ASD. SDNN, SDANN index, SDNN index, rMSSD, pNN50. Results (table) *p<0,05 – comparison with initial value (ANOVA) Conclusion: HRV parameters were significantly reduced 1 month after surgery what suggest autonomic system disturbance, but they tended to normalisation after 3 months. 12 pts treated with ASO constant HRV improvement was noted after 1 and 3 months. Tab 1. Tab 2

P871

Percutaneous transluminal septal myocardium ablation (PTSMA) in patients with hypertrophic obstructive cardiomyopathy (HOCM)

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Introduction: PTSMA by alcohol-induced occlusion of septal branches with resulting reduction of LV outflow-tract obstruction (LVOTO) is a new treatment option in symptomatic adults with HOCM. Methods and patient: We report about the treatment of a 13 years old boy. The diagnose of HOCM has been established within the first year of life. The boy was treated with verapamil for 11 years, when sotalol was prescribed to alleviate his symptoms. Nevertheless he remains dyspnoeic and exercise tolerability was reduced. Doppler-echocardiography shows MI I° and SAM III° with LVOTO of 80 mmHg with 28 mm septal thickness. Results: The diagnosis was confirmed by heart catheterization. After placement of a temporary pacemaker lead the 2nd septa branch was determined as target vessel by pediatric balloon occlusion (1.5 mm over the wire balloon) followed by myocardial contrast echocardiography. The branch was then occluded by injection of 3.5 ml ethanol (96%). LVOTO was reduced from 63 mmHg to 38 mmHg at rest and from 98 mmHg to 77 mmHg post extrasystole. The creatine kinase peak was 670 U/l after 9 hours. Standard-ECG exhibited RBB, brady- or tachyarrhythmia were not observed during 24h-Holter-ECG. Follow-up: Three months later the echocardiographic examination shows a septal thickness of 22 mm and a pressure gradient of 50 mmHg.

P872

Percutaneous treatment of middle aortic syndrome – 4,5 years experience.

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The purpose of the study is to evaluate the results of percutaneous treatment of middle aortic syndrome. Seven patients (pts) aged 4–17 years (mean 13.4) with middle aortic syndrome underwent interventional treatment (5 pts stent implantation, 2 pts balloon angioplasty). Five pts underwent stent implantation, 1 pt additionally balloon angioplasty of coexisting renal artery stenosis. Angiography revealed 3.5–6.5 cm long segment stenosis (minimal diameter 2,5–5 mm) in thoracic and/or abdominal aorta. Pressure gradient ranged 40–90 mmHg (mean 63.2 mmHg). Three Palmaz 5014, three Palmaz 4014, one Palmaz 3008 stents were used and expanded with 7–10 mm balloons. Pressure gradient after implantation was 0–35 mmHg (mean 13,6 mmHg), angiography confirmed stents patency and proper position. Heparine (for 48 hours), aspirin and acenocumarol (for 3–6 months) were administered after procedure. In 1 pt thrombosis of the stent occured 6 days after implantation and was successfully treated with local infusion of rt-PA. Thrombolytic treatment was followed by balloon dilation and second stent implantation. In 36–54 months follow-up 1 pt had elective redilation of the stent (after 6 months), 2 underwent successful redilation due to neointimal hyperplasia

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

after 9–42 months. Spiral CT angiography confirmed good result and no aneurysm formation. In 2 pts (11 and 12 years old) with severe arterial hypertension due to complex form of middle aortic syndrome balloon angioplasties of aorta, renal and mesenteric arteries resulted in possibility of pharmacological control of hypertension. Stent implantation produced satisfactory early and long-term results. Significant neointimal hyperplasia causing arterial hypertension can develop and can be successfully treated with further balloon dilations. In complex forms of middle aortic syndrome balloon angioplasty can improve pharmacological pressure control but long-term result is worrisome and needs further evaluation.

P873

Balloon valvuloplasty and surgical valvotomy in neonates with critical aortic stenosis

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To assess the outcome of balloon valvuloplasty and surgical valvotomy in neonates with critical aortic stenosis between 1990–2000. A retrospective analysis was undertaken of all patients presenting to a tertiary centre and who required intervention. Twelve were subjected to balloon valvuloplasty and five to surgical valvotomy at a median age of 10 days (2–42 days) and 9 days (1–21 days) respectively. There was no difference in age, weight, aortic annulus or left ventricular dimensions in either group. There was one death in each group. However the death in the valvuloplasty group was not related to the procedure. Mild to moderate aortic regurgitation was seen after either procedure. Four patients in the valvuloplasty group developed femoral artery thrombosis which resolved with thrombolytic therapy and 2 had cardiac perforation treated conservatively. The mean Doppler gradient was reduced from 48 +/- 16 mmHg to 12 +/- 5 mmHg ($p < 0.01$) in the valvuloplasty group compared to 34 +/- 15 mmHg to 12 +/- 1 mmHg ($p < 0.01$) in the surgical group. The surgical and valvuloplasty groups had a mean hospitalisation stay of 20 +/- 18 days with a mean ICU stay of 4 +/- 4 day and 8 +/- 5 days and a mean ICU stay of 2 +/- 1 day respectively. None of the patients in the surgical group required re-intervention whereas 5 in the valvuloplasty group required further balloon valvuloplasty. The most rapid rise in gradient occurred within the first month after ballooning. Both procedures offered effective short and medium term palliation in infants with critical aortic stenosis. Balloon valvuloplasty had a higher re-intervention rate, but shorter hospitalization.

P874

Interventional cardiology in newborns – one centre experience.

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Between 1992–2000 (9 years) cardiac interventions were performed in 210 newborns. Balloon atrioseptostomy in 125 patients (pts); with different complex congenital heart defects. BAV was performed in 45pts with critical/severe AS. Pressure gradient decreased from mean 72mmHg to mean 16mmHg. Early death occurred in 11pts (24%), late death in 2pts (4%). Good result is stable in 33pts (72%) in mean follow-up 34 months. BPV was performed in 33pts – 24 with critical/severe PS, 9 with Fbrocin anomaly, 1 with complex TGA, 5 with ToF. In PS group gradient decreased from mean 77mmHg to mean 17mmHg. Saturation increased from mean 79%HbO2 to mean 92%HbO2. 1pt died 6 hours after valvuloplasty (RVOTO perforation); 6pts required repeat valvuloplasty. Good result is stable in 21pts during mean 25 months follow-up. In 2pts with dysplastic valve procedure was insufficient. 5pts with ToF had hypoplastic valve and one/both pulmonary arteries. Saturation increased in all (mean 20%HbO2). 1pt is after correction. 4 in follow-up. One required elective B-T shunt (discontinuation of the RVA). Other interventions were performed in limited number of cases. 1pt with septic renal failure underwent balloon angioplasty of CoA. Because of short follow-up results need further evaluation. In 2pts after arterial switch operation mechanical ventilation was discontinued after microcoils closure of aorto-pulmonary collaterals. In 1pt with critical ToF and PDA, right B-T shunt and Rocck procedure were performed. Due to lungs hyperperfusion after surgery, PDA was closed (coils) with good result. 1pt with ToF, PA, I PA stenosis underwent right B-T shunt and prematurely stent implantation to PDA. In one premature baby with sepsis, 7cm long distal part of common was stented from pulmonary arteries. Different interventional procedures can be performed in newborns as a final or palliative treatment.

P875

Transcatheter versus surgical closure of atrial septal defect in adults

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Transcatheter closure of secundum atrial septal defect (ASD) has been accepted as an alternative to conventional surgical approach. Adult population has important added risks compared to children which may affect the safety of the procedure. This study was undertaken to evaluate the efficacy and safety in 53 patients (41 female: 12 male), mean age 34.5 years who underwent successful transcatheter closure from January 1997 to October 2000 (group 1) compared to 156 patients (125 female, 31 male), mean age 28.6 years who underwent surgical closure, from June 1992 – December 1996 (group 2). In group 1, 38 (53%) were symptomatic of which 14 had palpitation, 10 had effort dyspnea, 2 angina pectoris and 2 had paradoxical embolic stroke. While in group 2, 34 (22%) had documented history of either effort dyspnoea or palpitation. Transesophageal echo (TEE) was used to assess the defect and surrounding structures prior to stretch balloon sizing of the defect. The size of the defect in the surgical group was assessed intraoperatively. In group 1, the calculated mean Qp/Qs was 3.1 + 1.4 and 17% of them had pulmonary vascular resistance more than 3.5 woods unit. 2 had SVTs and one coronary artery disease patients underwent successful radiofrequency catheter ablation and angioplasty respectively prior to ASD closure at the same sitting. The mean procedure time was 98 minutes; screening time was 19.6 minutes and one hospital day as compared to 6 hospital days in group 2. Major complication was not encountered in group 1 while in group 2: 4 had pericardial effusion, 1 had pleural effusion and 2 had surgical wound infection and required intervention. Clinical improvement was noted in all symptomatic patients regardless of the treatment group. Only 1 patient had residual shunt noted at 24 hours following transcatheter ASD closure but these sealed-off completely at 1 months follow-up and none noted in the surgical group. Transcatheter ASD closure in adult population is effective and safe. It offer better alternative to surgery especially to those planned for other interventional cardiac procedure.

P876

Usefulness of Amplatzer occluders in closure of asd and other undesirable shunts.

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Purpose: One center retrospective analysis of Amplatzer occluders applied for closure of ASDs, PDAs and veno-venous hepatic fistula. Method: Transcatheter ASD closure with Amplatzer Septal Occluder (ASO) was attempted in 80 pts aged 0.4 – 54 (mean 14.5) y. The size of ASD ranged from 4.0 to 40 (mean 11.5) mm in TTE, 4–36 (mean 12.8) mm in TEE, the stretch diameter – from 5 to 36 (mean 16.1) mm. In another 4 pts aged 0.6 – 42 (mean 12)y PDAs 2–4 mm in diameter were closed with Amplatzer Duct Occluder (ADO). Five years old boy with critical desaturation (45%) after unsuccessful Fontan operation with partial hepatic vein exclusion and huge 20 mm intrahepatic veno-venous fistula was treated with implantation of 8/6 mm ASO. Results: ASD was successfully implanted in 77/80 pts with ASD. In one pt embolization of ASO to aorta occured. In 2 pts we could not achieve correct position of ASO and the procedure was abandoned. The defect was single in 61 pts, double in 19 pts with aneurysm of IAS in 4 pts, with insufficient antero-superior rim in 20 pts. The size of implanted devices ranged from 5 to 38 (mean 18.6) mm. In one pt with two distant ASD two ASO were implanted. Complete closure of PDA was confirmed in all pts after 24 h and in all but 2 ASD pts after one year of follow-up. In examined patients after ASO implantation closure of hepatic fistula was achieved with rise of saturation up to 85%. Conclusion: Implantation of Amplatzer occluders became the treatment of choice in selected patients with ASD (including multiple), with PDA and major vascular fistulas.

P877

TRANSLUMINAL COIL EMBOLIZATION OF THE CONGENITAL CORONARY FISTULAE

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Aim of the study: to assess the opportunities of transluminal embolization of the coronary fistulae. Materials and Methods. In October, 2000, 14 pts underwent the attempt of transluminal embolization of the coronary fistulae using Cianoacrylic coils. Patients age varied from 11 month to 44 years old (mean

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

7.2±2.7 yrs) and weight 11mm 9 to 74 kg (mean 28±21 kg). Localization and fistulae diameter were determined after selective right and left coronary angiography. Six patients had the fistulae between the RCA and the RV see RA. 1 – between the RCA and the trunk of the PA. 3 – between the LAD and the RV, 4 – between the Cx and the RV see RA. Occlusive coils diameter was approximately at twice more than the fistulae diameter. the number of the implanted coils was determined by the fistulae size from 2 to 20 coils were implanted to every patient. Results. Complete coronary fistulae occlusion was achieved in all of the cases There were noted the following complications: coil migration in the PA in 2 patients (all the migrated coils were removed using a basket device), femoral artery thrombosis in 2 pts (unfortunately, one patient died after repeated ineffective thrombectomy was complicated by common iliac artery thrombosis and acute renal insufficiency resistant to treatment) and a wire-related perforation of the fistula in one case which led to immediate hemopericardium with complete fistulae thrombosis. Long term results were learned in 13 pts in 6 months to 16 years. All the patients led a regular life and had no complaints. A selective coronary angiography was carried out in 4 pts. Radionuclide test of myocardium was being performed in 2 pts under loading conditions. The complete occlusion of the fistulae was confirmed in all the cases. Conclusion: transluminal embolization of the coronary fistulae is a safe and effective method.

PB78

Cost-comparison of transcatheter and surgical closure of atrial septal defect in children

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Background - Transcatheter closure (TCC) has been recently introduced for treatment of atrial septal defect type II (ASD). The devices in use have been so far quite expensive. Aim - Cost of TCC were compared to the cost of surgical closure of ASD. Methods - We analyzed the cost of the procedures and hospital stay. Seven consecutive children underwent TCC. These patients were compared with 7 other children who had surgical closure of their ASD before introduction of TCC. The following parameters were compared - Age, weight, length of hospital stay, total hospitalization cost, residual shunting at discharge (echo). - The mean age and weight of both groups were comparable. The procedures were successful in all patients. None of the patients in both groups had any major complication. Mean length of hospital stay was 0.6 ± 0.8 days for TCC vs 8.4 ± 0.9 days for surgery, p<0.001. The mean total hospital cost of the TCC was 14,222 ± 1,441 SFr (=7,964 US$, rate 0.56) vs 37,632 ± 2,192 SFr (=21,068 US$) for surgery, p<0.001. There was no residual shunting with surgical closure, while 4/7 (57%) after TCC had complete closure at discharge. Further studies on long-term effectiveness of TCC are needed. The currently available clinical data demonstrate complete closure in over 90% of patients at one year follow-up. Conclusions - Despite the devices for TCC are still expensive, the procedure remains almost two thirds cheaper than surgical repair of atrial septal defect. The patients with TCC have significantly shorter hospital stay. TCC is a viable alternative to surgery.

PB79

Undilatable Pulmonary stenoses – successful treatment with the cutting balloon

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Introduction. Balloonangioplasty is the treatment of choice for central and peripheral pulmonary stenoses. However, about 10% may be non dilatable even with the use of high pressure balloons. Methods: The cutting balloon (Ex. Cardiologie) is a new therapeutic alternative to treat such refractory pulmonary stenoses. Three to four microsurgical blades (0.1 to 0.4 mm thickness) are mounted longitudinally on the surface of the cutting balloon. Inflation of the cutting balloon produces longitudinal, 0.17 mm deep incisions in the wall of the target vessel. We report about a 5 years old boy with pulmonary stenosis with multifocal blood supply. Results: After three steps of unifocalisation a severe pulmonary stenosis (1 mm) remains on the right side. With high pressure angioplasty (18 atm, Bypass speedy, Ex. Cordis) relief of the stenosis was not achievable. The angioplasty balloon was exchanged for a cutting balloon (4 mm). After incision of the vessel wall with the cutting balloon angioplasty was repeated with a 6 mm angioplasty balloon leading to complete relief of the stenosis and increase of vessel diameter from 1 mm to 3.5 mm. Follow-up: Follow-up catheterization three months later shows restenosis of the vessel, which could be easily inflated. However, proximal to the former now another stenosis was now evident and refractory to angioplasty. Its diameter was to large for the 4 mm cutting ballon and the stenosis remains undilatable. Conclusion: The cutting ballon allows dilation of otherwise refractory stenoses. Currently its use is limited due to the maximal size of 4 mm.

PB80

Femoral artery thrombosis following cardiac catheterisation in children under 10 kg: risk factors and management

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Background: Damage to the femoral vessels is a well-recognised complication of cardiac catheterisation. We examined the incidence and management of suspected femoral artery thrombosis (FAT) in children and infants <10 kg following cardiac catheterisation. Methods: Information from all consecutive catheterisation procedures between 1991-8 was derived retrospectively from computerised records. Patients >10 kg and those without arterial access were excluded. Linear regression analysis was used to assess potential risk factors for FAT. The diagnosis of FAT was made on clinical grounds (absent or reduced foot pulses or temperature difference between limbs 6 hours post-procedure). Results: 216 patients <10 kg (mean weight 6.7 kg) with arterial puncture were identified. 182 (70%) procedures were diagnostic; 64 (30%) were interventional. Mean procedure duration was 85 (SD +/-45) minutes. FAT occurred in 35 cases (13%). Procedure times were longer in the FAT group (104 mins vs 79 mins, p=0.06). FAT was more likely to occur following interventional than diagnostic catheterisation (25% vs 13%, p=0.05, but NS if time for procedure was taken into account). There were clear, but non-significant, trends towards increased FAT with larger sheath size, smaller patients, and when guidewire rather than cannulae were the first operator. However there was no clear relationship between risk of FAT and side of puncture (a surrogate for difficult access), prophylactic use of dextran 40, intra-procedure heparin dose, serum haemoglobin concentration or systemic oxygen saturation. FAT was managed with early streptokinase +/- heparin infusions and, although bleeding was common, only one transfusion was required. Pulses and limb perfusion were restored in all cases without long-term sequelae. Conclusions: FAT is common following cardiac catheterisation but, whilst the problem is readily treatable, the message is clear – don't waste time in the cath lab.

PB81

Pulmonary valve balloon dilatation in patients with critical pulmonic stenosis and congestive heart failure: is it an indication for transjugular approach?

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Background: Pulmonary valve balloon dilatation (PVBD) is conventionally performed by the femoral route. However in patients with tricuspid regurgitation and congestive heart failure (CHF), it may be impossible to cross the valve by this route and transjugular approach may be required. In view of the potential for complications with transjugular approach, it is generally reserved to only when the femoral approach fails. We attempted to find if any pre-catheterization parameter could predict the failure of femoral approach, so as to use transjugular approach decisively. Methods: Over the past 8 years, 15 patients underwent transjugular PVBD after a failed transfemoral attempt. These were compared with a control group of 10 randomly chosen patients with CHF who had a successful transfemoral PVBD. We analysed data for differences in clinical, echocardiographic and hemodynamic parameters between these two groups. Results: One infant in the study group sustained a tear of the internal jugular vein and underwent surgical valvotomy. There was no difference in severity of echocardiographic grade of tricuspid regurgitation in two groups. The mean right atrial pressures and the right ventricular systolic pressures were also not different in the two groups (14.8 and 108.1 mmHg in the study group versus 13.5 and 115.6 mmHg in controls respectively). The only predictors of failure of the femoral approach were a large cardiothoracic ratio on chest x-ray (0.84 in the study group versus 0.73 in controls, p<0.02) and lower cardiac index (1.96 versus 2.8 l/m2; p<0.005) suggesting that the study patients had more severe disease. A cardiothoracic ratio of 0.8 or more was more often found in the study group (8 versus 4). Conclusion: PVBD should perhaps be first attempted by the transfemoral approach even in patients with CHF. Patients who fail the transfemoral approach are likely to have more severe disease. A cardiothoracic ratio of 0.8 or more identifies such a subset of patients.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P082

Complementary use of detachable coils and amplatzer duct occluders for closure of patent ductus arteriosus (intermediate term results)

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Purpose. Several different devices were evaluated for the percutaneous of closure of patent ductus arteriosus (PDA), and important drawbacks were found in all of them. To overcome these drawbacks, both detachable Cook PDA coils and Amplatzer duct occluders (ADO) were used for the percutaneous closure of PDA. Methods&patients: Between September 1996 and September 2000 a total of 124 patients underwent transcatheter occlusion of PDA at a median age of 4,5 years (range 0,5–29 years) and at a median weight of 19,5 kg (range 6–69). Three patients were adults. Results:Detachable Cook PDA coils were used in 76 patients (multiple coils in 5), with a median PDA diameter of 1,7 mm (range 1,1–2,2 mm) and ADO were used in 48 patients with a median PDA diameter of 3,9 mm (range 1,9–10,5 mm) (p<0.0001). Devices were successfully implanted in all 124 patients. During the follow up period of 3–48months (median 28months) complete closure was achieved in 73 of 76 patients in coils group (96%) and in 46 of 46 in ADO group (100%)(p <0.01). There was no death, arterial or venous complications, thromboembolism, haemolysis or other morbidity. Conclusion: According to our experience, the complementary use of detachable Cook patent ductus arteriosus coils for smaller (< 2 mm PDA) and Amplatzer duct occluders for the larger PDA (>2mm) can be recommended as a treatment of choice with excellent results. However, further long term follow up studies are needed to support our recommendations.

P083

Echocardiographic considerations in patients selected for transcatheter closure of atrial septal defect.

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The aim of the study was to analyse the role of TTE and TEE in selecting patients and monitoring the procedure of ASD closure with Amplatzer device. 16 children aged 4 – 19 years, mean 10,1 years were initially selected for the procedure based on the TTE. The defects were located in central or anterior- superior part of the septum, the diameter ranged from 5 to 21 mm, median 11,8 mm, with rims> 5 mm. Qp:Qs> 1.5, there was double defect in 1 child. After the TEE assessment of morphology and diameter of the defect at the time of implantation 1 child was excluded from the procedure because of too small inferior rim. In 15 children the diameter of ASD ranged from 7 to 20 mm, median 12,7 mm and correlated well (r =0,9) with balloon stretched diameter which ranged from 10 to 23 mm, median 14,9 mm. The procedure was performed under TEE guidance. In 1 child the device was withdrawn because it was unusable. Finally, device closure was performed in 14 patients. The diameter of Amplatzer device was from 11 to 24 mm, median 16,4 mm. After the procedure the position of the device, atrio- ventricular valves function and systemic and pulmonary venous return were estimated by TEE. Follow-up examination at 1,3,12 months after implantation included clinical examination, ecg and TTE. In 13 children, closure of ASD was complete, in 1 a trivial haemodynamically insignificant residual shunt remained. No procedural complications were encountered. Conclusions: 1. TTE is useful for initial selection of patients with ASD for transcatheter closure. 2. TEE is essential for definitive qualification and monitoring the procedure.

P084

A successful percutaneous transluminal renal angioplasty (ptra) in 2-year-old boy with renovascular hypertension

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PTRA is well accepted in the managements of renovascular hypertension. However, the application for small children was rare and its results were not satisfied. We report our successful experience of 2-year-old boy with severe renal artery stenosis. The patient was followed as multiple cardiac tumors complicated with tuberous sclerosis. His cardiac function was depressed from neonatal period. At 16 months of age, he suffered with acute encephalopathy and developed renal and heart failure. He has also severe hypertension resistant to any multiple medications. At 21 months of age, an aortography was performed and severe right renal artery stenosis (0.5mm) and no left renal artery were demonstrated. At this moment, surgical and interventional treatment seemed to be difficult because of severe stenosis in one-side functioned kidney and small child with 6.6kg body weight. At 25 months of age, his cardiac and renal functions were deteriorated with respiratory infection. Despite of intensive care, a blood pressure and fluid management was not controlled. At 29 months of age, PTRA was performed to save his life. Catheter was introduced through a 6F sheath inserted from right external carotid artery. The initial balloon catheters using 2.0 and 2.5mm in diameter was not successful. Therefore a cutting balloon with 2.5mm diameter was tried and an adequate expansion was obtained, following safety stent implantation (2.5x12mm). After PTRA, a blood pressure decreased immediately and was well controlled with antihypertensive medications. Plasma renin activity and serum creatinine dramatically decreased from 293 to 41.5 ng/kg/h and from 2.1 to 0.1 mg/dl, respectively. PTRA was successfully performed in 2-year-old infant with renovascular hypertension resistant to multiple medications.

P085

Results of percutaneous closure of patent ductus arteriosus with detachable coils

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The aim of the study was to assess safety and efficacy of transcatheter closure of PDA. Between June 1996 and October 2000 65 patients aged from 9 months to 17 years, median 7,17 years, underwent percutaneous closure of PDA. 8 patients had residual PDA after surgical ligation. All children were asymptomatic. The median PDA diameter as in narrowest segment assessed in TTE was 2.54 mm (range 1 to 4.5) and on angiography 1,76 mm (range 0,8 to 3,5). By angiographic classification of PDA: 46 patients had type A, 21 type E, 8 type D, 8 type I, 3 type B and 3 type C. In 80 patients, one coil was implanted, while in 4 patients two coils and in 1 three coils were placed. Residual shunt was detected in 7 patients and disappeared within 3 months to 1,5 year in all except 1. In 3 children reimplantation of PDA was required and in 1 reintervention was required. No intraprocedural or short-term and late follow-up complications were encountered. No obstruction of left pulmonary artery or descending aorta was diagnosed. Conclusions: 1. Transcatheter closure is an effective therapy for patients with PDA diameter up to 3,5 mm. 2. Complete closure in most small-sized PDA is achieved with one implanted coil. 3. Residual shunts after coil embolization close spontaneously in the majority. 4. Coil occlusion of PDA is a safe procedure, no complications were found.

P086

Local rt-PA treatment in thrombosis complicating interventional procedures or cardiac surgery

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The purpose of the study was to evaluate effectiveness of local, angiographically and/or echocardiographically guided rt-PA treatment in patients (pts) with thrombosis complicating cardiac surgery or interventional procedures. Eleven pts aged 7 days-17 years (mean 7 years) were analyzed. 7 pts after surgery (2 BT shunts, 2 arterial switch operations, 2 Fontan operations, 1 ASD with PAPVD correction) developed pulmonary artery-, SVC-, IVC-thrombosis or intraventricular thrombus. In 4 pts thrombosis complicated interventional procedures (1 pt – femoral artery thrombosis after PDA coil closure, 1 pt with middle-cava syndrome – thrombosis of stent implanted to aorta, 1 pt – thrombus on IAS during ASD sizing, 1 pt – circumflex artery thrombosis after coil occlusion of coronary artery fistula). Signs of thrombosis lasted 0–42 (mean 10 days). Local thrombolysis with catheter placed at site of thrombosis was started with rt-PA bolus (0,02–0,5 mg/kg), in 9 pts was followed by local rt-PA infusion (0,01–0,1 mg/kg/h) for 20 hours – 4 days (mean 1,7 day), according to immediate result. In 9 pts thrombolysis alone or combined with interventional procedures (1 pt with SVC thrombosis – its post-thrombolytic patency allowed stent implantation, 1 pt with aortic stent thrombosis – second stent implantation followed successful thrombolysis, 1 pt after achieved patency of circumflex artery – balloon angioplasty and coil reposition were performed) resulted in vessel patency restoration and intracardiac thrombus resolution. Results are stable during 3–58 (mean 30) months) follow-up. 1 pt after Fontan operation with IVC and pulmonary thrombosis died before repis. In 1 pt with SVC thrombosis and massive venous collaterals thrombolysis was unsuccessful. Local rt-PA treatment is effective in different thromboembolic complications after cardiac

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

surgery or interventional procedures also in long-standing thrombus. Angiographic and echocardiografic guidance allows treatment modification.

P887

Balloon occlusion tests in patients with complex cardiac anomalies

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Purpose: Diagnostic value of temporary balloon occlusion tests (BOT) in pts with complex heart defects. Methods and material: BOTs were performed in 21 catheterized pts with mean age of 5,3 (0,4- 12,1) y. Pulmonary pressure (MPAP) and saturation (S) were estimated before and after BOT of RVOT (8 pts), B-T shunt (6 pts), Waterston shunt (3 pts), azygos vein (2 pts), IVC (1pt) and large hepatic fistula (1pt). In 2 pts ASDs were occluded to assess RV or LV function. Results: In 15 pts after BOT of RVOT or B-T shunt, MPAP decreased from 23,5 to 15,7 mmHg and successful first or second stage of Fontan operation (FO) was performed. No significant changes in MPAP in 3 pts after Waterston shunt and 1 after RVOT occlusion was demonstrated so they were disqualified for FO. BOTs of azygos veins in 2 pts after Glenn procedure resulted in an increase of MPAP and S an closure with coils were performed. In 1 pt after FO and increase of S during BOT, IVC-RUPA conduit was closed with ADO and PLE disappeared. In patient FO hedge membrane fistula was closed with ASO with permanent increase of S. BOT of ASD in pt with hypoplastic RV showed increase of CVP so surgery was contraindicated, but ventilator dependent infant with ASD and mild LV hypoplasia: LA pressure decreased after BOT and successful surgical closure of ASD was performed. Conclusions: Temporary balloon occlusion test imitating potential results of permanent closure of any vascular connection can be very helpful to predict definitive treatment. It has special importance in borderline indications for various types of veno-pulmonary connections.

P888

The Amplatzer TM septal occluder for transcatheter ASD occlusion. A comparison with the buttoned device in 158 patients.

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To report the experience with the Amplatzer-TM Septal Occluder (ASO) and compare results with those of the buttoned device for transcatheter ASD closure. Methods: Transcatheter ASD closure was performed in 94 patients (pts) with ASO (group 1, 1998-2000) and in 64 pts with the buttoned device (group 2, 1992--1997). Age of pts was 31+/-20 (3.4-90) years in group 1 versus (vs) 17+14 years in group 2, weight 57+/-21 kg in group 1 vs 44+/-21 in group 2 and stretched diameter was 22+/-5 mm in group 1 vs 19+/-6.5 mm in group 2 (p<0.04). Implantation succeeded in 95 % of pts in group 1 and 75 % in group2 (p<0.05). The Fluoroscopy time was 8 +/- 7 minutes in group 1 and 24 +/-14 minutes in group 2 (p<0.05). Immediate surgery was required in 1 patient in group 1 (embolization) and 2 pts in group 2 (1 embolization, 1 residual shunt). One death occurred after implantation in group 1 unrelated to the device. Atrial arrhythmia occurred in 3 pts in group 1 and 2 pts in group 2. During follow-up, no patient was operated upon in group 1, but 8 pts in group 2 because of shunting (n = 7) and atrial perforation (n = 1)(p>0.05). At latest follow-up, the rate of complete occlusion was 95 % in group 1 vs 69 % in group 2 (p<0.05). In conclusion our experience with ASO compares favorably with the buttoned device. Implantation failure rate of successful implantation is higher and rate of complete occlusion is better with ASO than Buttoned device. ASO is clearly for the present time the treatment of choice for transcatheter occlusion of ASD.

P889

Percutaneous closure of the blalock taussig shunt: Experience and results in 13 patients.

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Introduction: Blalock Taussig shunt (BT) has been essential to increase pulmonary blood flow. When not needed, closure can be performed percutaneously. Eventually, a BT is left open during the post operative period to assure adequate flow. We present our experience in closure of BT shunts in the catheterization laboratory. 13 patients were referred for closure, as an emergency (6) or as elective procedure (7). Material and Methods: Mean age: 8,7 ± 4.5 y and weight 25,4 ± 10.9 Kg. 12 had complex congenital heart diseases (CHD) and one simple CHD. 8 cases (previous Glenn). One patient had 2 BT. Previous balloon occlusion assessed the result in 5. There were 14 BT in 13 patients. Right 4, left 10, double 1. 57 coils were placed. 4.3 coils per BT. Coil/BT diameter ratio: 1.4. Technique: Arterial access in 11, venous in 2. Catheter size: 4F: 1, 5F: 10 and 6F: 3. Gianturco coils in 3 patients and Jackson detachable in 10, since 1996. Associated techniques: Pulmonary valvuloplasty 1, Stent implantation 1, simultaneous balloon occlusion of PDA 3. Previous balloon test occlusion: 5. Important to secure first loop and carefully coil up the BT. Additional coils were implanted until total occlusion. Results: Total occlusion 13 BT (92%) and 1 failure. Complications: Coil migration: 3 (successful snaring 2, 1 coil surgical retrieval). Conclusion: Percutaneous closure of BT's is simple and effective. Can be performed in the post operative period, being our elective technique. Balloon test occlusion can assess the indication. Detachable Jackson coil system seems the ideal tool for this purpose.

P890

Transcatheter closure of large venous collaterals using Amplatzer duct occluder

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Objective: To assess the feasibility and effectiveness of the Amplatzer duct occluder for transcatheter occlusion of large venous collaterals. Background: Development of systemic venous collaterals is common after cavopulmonary shunt operations. They lead to systemic desaturation, hypoxia and effort intolerance. Management of such large venous collaterals is challenging and conventionally involves surgical ligation. Patients and Methods: Large venous collaterals were encountered in two Fontan patients and in one cavopulmonary shunt patients with marked cyanosis and effort intolerance. These vessels represented a) A dilated azygous vein draining into the pulmonary venous atrium, b) A dilated left superior venacava draining into the pulmonary venous atrium via coronary sinus, c) A large paravertebral collateral arising from the innominate vein draining into the inferior venacava. Transcatheter closure of these 8mm, 5.2mm and 7mm collaterals was accomplished using a 14/12-mm 8/6-mm and 10/12-mm Amplatzer duct occluder respectively. Femoral venous approach was used in first patient while right internal jugular approach was used in the other two patients. Result: Successful transcatheter closure of large (> 5 mm) venous collaterals using Amplatzer duct occluder with improvement in the systemic saturation [mean pre 82% to mean post 93%, p = 0.0041]. Conclusion: Occlusion of large systemic venous collaterals can be accomplished safely and effectively using Amplatzer duct occluder. Amplatzer duct occluder is a self-centering, self-expanding device, which causes occlusion by thrombosis. Advantages of this technique include delivery through a small sheath, ease of delivery and repositioning and retrievability. This approach should be considered as an alternative to surgical ligation procedures for management of large venous collaterals.

P891

Transcatheter pda occlusion with the Amplatzer device

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Sixty nine patients (28 males,41 females) underwent percutaneous occlusion of PDA with the Amplatzer device in the last 3 years at our institute. Their ages ranged from 10 months to 50 years (mean age 12.2+/- 11.8 years). the procedure was performed from the transvenous route using a 5 to 8 F sheath. In six patients the PDA had to be crossed from the aortic end. The PDA measured 3-9 mm on the angiogram (4.3+/-1.8mm). The PDA was type A in 49, type B in 4 type C in 1 and type E in 5 patients. Two patients had severe PAH. Multiple angiographic views were required in 16/61 patients as the PDA was not profiled in the conventional lateral view. Device deployment was successful in all patients-in two patients a larger device than the size deployed initially was required. Device sizes used were 6-4 in 13, 8-6 in 29, 10-8 in 14, 12-10 in 4 and 14-12 in 1 patient. Immediate total occlusion was seen in 30, trivial to small residula flow in 18 and significant flow in 13 patients. The mean fluoroscopy time was 14.1 +/- 8.8 minutes (range 8.3 to 31 minutes). Doppler echocardiography at 24 hours showed persistent trivial flow in 2/61 patients. No gradients were seen in left pulmonary artery or descending thoracic aorta. One infant had loss of femoral pulse after the procedure. There were no late complications or reopening of duct on follow-up till date. Transcatheter closure of PDA with Amplatzer device is a safe and effective technique for majority of patients.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P892

Left atrial mechanical function following atrial septal defect closure with the Amplatzer septal occluder

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Background Although transcatheter closure with the use of occluder devices has been increasingly used in the management of atrial septal defect (ASD), there are no studies examining the impact of such an intervention on left atrial (LA) mechanical function. This study determined LA volume and function in a group of ASD patients (pts) treated with the Amplatzer septal occluder (ASO) 12 to 48 months (24 ± 11.5 months) after the intervention and compared the findings with those of normal controls. Methods: Eighteen pts with ASD (age 8.9 ± 3.5 years) successfully treated with ASO (device size 16 to 34 mm, mean = 19.8 ± 5.7 mm) and 15 normal controls (age 9.0 ± 3.3 years) with similar sex distribution were studied. LA volumes were determined at mitral valve (MV) opening (maximal, Vmax), at onset of atrial systole (P wave of ECG, Vp), and at MV closure (maximal, Vmin) from the apical 2- and 4- chamber views, using the biplane area- length method. LA passive emptying function was assessed with the LA passive emptying volume (PAEV) = Vmax - Vp and the LA passive emptying function (PAEF) = PAEV / Vmax. LA systolic function was assessed with the LA active emptying volume (ACTEV) = Vp - Vmin and the LA active emptying function (ACTEF)= ACTEV / Vp. Transmitral flow was assessed with pulsed Doppler. * Results: Conclusion: Implantation of the ASO in patients with ASD is associated with a slight decrease in LA size without significant changes in LA mechanical function or in the transmi

P893

Transcatheter Closure of 'swiss Cheese' Ventricular Septal Defects Using Amplatzer VSD Occluder.

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Objective: To assess the feasibility and effectiveness of Amplatzer VSD occluder for transcatheter closure of 'swiss cheese' septal defects. Background: Despite significant improvements in the diagnostic, interventional and surgical techniques management of 'swiss cheese' septal defects remains controversial. Residual VSDs and ventricular dysfunction contribute to significant postoperative morbidity and mortality. Patients: Transcatheter closure of a large anterior trabecular VSD and a large apical muscular VSD was accomplished using two Amplatzer VSD occluders in a 18 month old infant (weight 7.2) with a large perimembranous VSD and 'swiss cheese' trabecular septum and palliation with a pulmonary artery band. Interventions: Standard right and left heart catheter sation and angiography. Right internal jugular venous approach for balloon sizing and closure of apical VSD with a 4 mm Amplatzer VSD occluder through a 5 French delivery sheath. Right femoral arterial approach to cross anterior trabecular VSD from the left ventricle, guide wire snared from right ventricle and retrieved through right femoral vein. Balloon sizing and closure of the defect with a 12mm Amplatzer VSD occluder through a 7 French sheath via right femoral vein. Continuous TOE monitoring during balloon sizing, device deployment and release. Small residual shunt on angiography post occlusion. Surgical closure of large perimembranous VSD with pulmonary artery debanding two months later. Conclusion: Transcatheter closure of 'swiss cheese' VSDs using Amplatzer VSD occluders is feasible, safe and effective. The technique can be used for closure of muscular septal defects situated in different parts of the trabecular septum. A collaborative approach with preoperative transcatheter closure of muscular VSDs followed by surgical closure of perimembranous inlet or outlet VSDs may be the optimal approach for patients with multiple VSDs and 'swiss-cheese' trabecular septum.

P894

Transcatheter Closure of Residual Ventricle to Pulmonary Artery Communication after Fontan Operation

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Objective: To describe the use of transcatheter interventional procedures for closure of residual ventricilo-pulmonary arterial shunts in the setting of a Fontan circulation. Background: Left-to-right shunt through a residual ventricle to pulmonary artery communication is a rare complication following Fontan operation. It can result in persistent pleural effusions, ventricular volume overload and ventricular failure. Conventional management involves surgical division of such communications. Patients: Two Fontan patients with significant left to right shunt through a residual ventricle to pulmonary artery communication. Interventions: A seven year old boy developed persistent pleural effusion and ventricular failure two weeks post Fontan operation. At catheter mean Fontan pressure was 17 mmHg. Angiography documented a large left-to-right shunt from the left ventricle to the main pulmonary artery (Qp/Qs of 1.8:1). This ventricule-pulmonary communication which measured 4 mm was successfully occluded using a 17-mm Rashkind double umbrella device from the right internal jugular vein through an 11 French long sheath. This resulted in cessation of pleural drainage and ventricular function improvement. An 18-year-old female underwent cardiac catheterisation for reduced exercise tolerance and ventricular failure six years post Fontan operation. Pressure in the Fontan circuit was 15 mmHg. Angiography revealed anterograde flow from the pulmonary artery via the recanalised pulmonary arterial trunk. Calculated Qp/Qs was of 1.5:1. Balloon sizing revealed minimal diameter of 4 mm. This communication was successfully occluded using an 8/6-mm Amplatzer duct occluder from the right femoral vein. Conclusion: Transcatheter closure of residual ventricle to pulmonary artery communication in setting of a Fontan circulation is feasible. This technique is safe and can be effectively used for management of significant left to right shunt in this scenario as an alternative to surgical approach.

P895

ASD closure with Amplatzer occluder in children: selection of candidates.

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Secundum atrial septal defects (ASD) amenable to transcatheter closure are those with well-defined borders and clearly separated from both Vena Cavae, Coronary Sinus and Tricuspic valve. Objective: To establish the sensibility and specificity of transesophageal echocardiography (TEE) to select the candidates for ASD closure with Amplatzer. Material: Between January '98-March '00, 47 consecutive patients(pts) aged 5 to 16 years (ys), X: 8 ys with ASD were evaluated by TEE. The major diameters of the defect as well as the length of its borders were measured. Results: 22 of 47pts had a defect smaller than 20mm. X: 20 ± 3.3 (9–26mm), the appropriate borders and were referred for Amplatzer closure. The defect was 3.5mm larger when measured with an inflated balloon in catheterization (X:22.5 ± 5.3mm, 12–34mm). The ASD was successfully closed in 21of 24 pts(87.5%). The size of defect was underestimated in 3pts subsequently referred to surgery. In all 25pts sent to surgery the defect was larger than 20mm as confirmed by the surgeon. Lack of appropriate borders was noted in 25% of these. Conclusion: TEE showed a sensibility of 95.4% and a specificity of 95% to adequately select the candidates for Amplatzer closure in children. There is a 10–20% TEE underestimation of the size of the defect measured with an inflated balloon.

P896

Transcatheter closure of Atrial septal defects – experience with two different systems

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Secundum type atrial septal defects (ASD's) are undergoing transcatheter closure for more than a decade. This is done not only in childhood but with an increasing number in adults. We report our experience in a mixed population. > From 1997 to 2000 we treated 74 pts age 2 - 77y, weight 10–91kg. Procedure was done in the same setting under general anesthesia and guidance of T.E.E. and systemic anticoagulation and heparinisation of the delivery sheath. We implanted 80 devices. In one pt two devices were implanted for two defects in the same session. There were 39 pts treated with 20 CardioSeal-devices (CS) and 50pts with Amplatzer-septal-occluders (ASO). Qp:Qs was 1.8±0.6:1, it was larger in the ASO group, as was the age, but this was not significant. Duration of the procedure and radiation time were not different (4.5 – 27 min, mean 10.2). Size and location was also different in the ASO group: there were larger defects included, more located to the aortic rim. This is due to the technical specifications of the different devices. Successfull implantation was possible in all pts. There was no early or late embolisation. We encountered only one serious complication. In a pt with insufficient systemic anticoagulation a cerebral infarction with hemiparesis occurred within the first 12 hours after the procedure. Symptoms resolved quickly and are almost gone two years after the implantation. On follow up with TTE and TEE there are two trivial residual leaks. CS-devices are mainly selected in defects up 20 mm and smaller pts, whereas ASO-devices can be implanted also in defects exceeding even 30 mm. We summarize ASDs are to address with

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

transcatheter closure when the appropriate device for the pt and defect is selected in all ages.

P897

Transcatheter closure of the patent ductus arteriosus. are results dependent of devices and learning curve ?

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Objectives: To assess our results with different devices for percutaneous treatment of 148 patients with Patent Ductus Arteriosus (PDA) over a period of 9 years. Population and Methods: Age of patients ranged from 3.5 months to 63 years (median, 3,6 years) and weights from 5.0 to 76 (median, 15) Kg. Four patients had residual shunts after surgery. Rashkind devices (umbrellas) were used in the first 5 years and coils (detachable or Gianturco) were used in the last 4,5 years. Results: Five patients with pulmonary hypertension were considered unoperable. Of the remaining 143 patients, two had a PDA too large and were operated. Total closure was achieved at a single catheterization in 115 patients with implantation of a Rashkind device (31), one or more coils (82), a Rashkind device plus coils (1) and a detachable balloon (1). A second procedure was required in 25 patients for total closure, and this was obtained with a second umbrella (5), implantation of coils after a previous umbrella (10) and coils for residual flow after a first coil (4). Long term leaks after a single procedure reduced from 36 % (Rashkind devices) to 4 % (with detachable coils). Total occlusion was obtained in 138/143 (97%, CI 95 %—100%). In the whole series there are 5 patients with residual flow (two after recent coil implantation and one without implantation). Procedure time reduced progressively from 132,8 + 49,4 (mean + S.D.) minutes (min.) in the first year to 51.3 + 20.4 min. in the last year ($p < 0.01$). Fluoroscopy time reduced from 19.5 + 9.7 min to 8.1 + 3.7 min. ($p < 0.01$) in the same period. Patients treated with detachable coils also had procedure and radiation times significantly shorter. Complications: One umbrella and one coil embolized to the pulmonary artery and were retrieved percutaneously. Two coils embolized to the femoral artery; one required arteriotomy. Conclusions: Detachable coils provide better results than Rashkind devices, with the additional advantage of being cheaper. Percutaneous closure of the PDA is safe and efficient. Procedure duration and radiation time decreased significantly after the initial learning period.

P898

Experience with the interventional closure of atrial septal defects – a retrospective single center experience in 659 patients over a period of 10 years.

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Development and results of the interventional techniques to close intracardial communications were evaluated retrospectively for the period 1990 to 2000 with respect to size of defects and occluder, residual shunt, complications and fluoroscopy time. Methods: Starting with the Rashkind Occluder (RO) (n = 21) in 1990, the Sideris Device (BT-D) (n = 112) was used between 1992 and 1997. Since 1997 the Amplatzer Occluder (AO) (n = 508) and CardioSeal (CS) (n = 15) were exclusively implanted. Results: With the RO (PFO n = 6, ASD n = 6, fenestrated Fontan (FF) n = 12) only small defects with a median diameter 9 mm (2,5–9 mm) could be closed. With the introduction of the BT-D defects with a median diameter of 12 mm (3–27 mm) in PFO (n = 26) and ASD (n = 85) were treated successfully. Since 1997 it became possible to close defects with median diameters of 12 mm (4 - 26 mm) in PFO (n = 251), ASD (n = 353), and FF (n = 4) using the AO. CS (n = 15) was only used for PFO. Complete closure rate for RO / BT-D / AO was 81/72/94.3 %, the explantation rate 0/12 +/0 9%, and the median fluoroscopy time 25/17.5/ 7.5 min. With the introduction of the AO not only larger defects but also those with an unusual anatomy such as multiperforated lesion (n = 4) (3 of them with 2 Amplatzer Occluder), septal aneurysms (n = 69, and defects with a right-to left shunt (n = 15) could be closed. Conclusion: Experience and the technical developments during the last 10 years have made the interventional closure of intraatrial communications to a routine and safe method allowing to close larger and more complex defects.

P899

Closure of atrial septal defects larger than 24 mm diameter with the Amplatzer Septal Occluder (ASO)

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Background: Since the last years, interventional closure of ASD closure was limited to small to moderate defects. Introduction of ASO extended this limit. Aim: To study effectiveness and safety of large (>24 mm) ASD closure with ASO. Patients and Methods: Out of 493 patients, 67 patients (median age 35 yrs [6–68.5 yrs] median weight 63 kgs [15.5–92 kgs]) had defects larger than 24 mm stretched diameter (median 26 mm (24–34 mm)). Mean (SD) Qp/Qs was 2.8(1.2). Procedure was performed under fluoroscopy and TEE control. Balloon-sizing of the defect was performed in the usual way. Deployment of the ASO sometimes needed special manoeuvres: opening of the left atrial disc towards the right-upper pulmonary vein, or towards the mitral valve, or opening of central waist and partially the right atrial disc before a full contact of the left atrial disc onto the interatrial septum. Results: Mean (SD) fluoroscopy time was 15(11) min. Mean (SD) procedure time was 59.5 (36.5) min. Devices used were as follows: 24 mm (23 pts), 26 mm (15 pts), 28 mm (11 pts), 30 mm (6 pts); 32 mm (8 pts); 34 mm (4 pts). Complication rate was 4 %: (a) one patient had embolization of the ASO. He underwent surgery with no complication. (b) one patient had a moderate pericardial effusion with fever; (c) one pt had transient (<1 h) atrial fibrillation. At discharge only 5 pts (7 %) showed a tiny residual leak. Median follow-up was 8 months (1–20 months). At 6 months follow-up only one pt showed a trivial residual shunt. Conclusions: Transcatheter closure of large ASD with ASO is safe and effective in experienced hands. Sometimes, particular manoeuvres during device deployement are needed to achieve an adequate device position.

P900

Late dislocation of an ASD-Occluder after interventional closure of a defect with insufficient anterior rim

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Early dislocation of ASD-devices is a well-known complication. However, late dislocations has so far not been described. We report a late dislocation 6 months after implantation of an ASD device. A 3 year old girl presented with a 12 - 14 mm ASD of secundum type with pulmonary pressure of 22/10(15) mmHg and a left to right shunt of 60%. Balloon sizing revealed a defect size of 15 - 16 mm. Transesophageal echocardiography (TEE) showed an adequate left atrial (LA) size and sufficient rim around the ASD except in the area of the so called anterior rim beyond the aortic root. A 33 mm NMT Medical starflex self centering system was implanted under general anesthesia. The system was positioned without problems; one arm of the device was placed directly to the aortic root on the LA side. TEE confirmed correct placement of the occluder. Repeat transthoracic echocardiography up to 4 months after intervention showed a correct position of the device without a residual shunt. After 6 months a small shunt was visible with a bulky appearance of the occluder. TEE revealed a rotation of the device with dislocation of part the left disc into the right atrium, thus creating a shunt. Because of risk of rhombus formation and embolization surgery was performed via limited anterolateral thoracotomy. Intraoperatively rotation of the completely endothelized device was confirmed. Further follow-up was uneventful. Stable positioning of devices with arms like the Starflex device is problematic particularly in big ASDs with an insufficient anterior rim. Late dislocation is a potential risk in mid-term follow up. Round shaped devices and devices with more than 4 arms theoretically provide more stability in such cases.

P901

Implantation of Cheatham Platinum (CP) stents for reconstitution in childhood

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Reconstitution after surgical repair in infancy is seen in more than 10%. In some cases with moderate or no hypoplastic aortic arch dilation and stent placement is an alternative approach to surgery. The new CP stent (NuMED Canada, plin Germany) is specially designed for use in congenital heart disease and is expandable in a range of 8 to 25 mm with a maximum shortening of <20%. We implanted the CP stent in 3 children (age 4, 7 and 16 years, body weight 11, 25 and 80 kg) after surgery for coarctation in infancy (patch insertion, resection and end-to-end anastomosis and subclavian flap repair). Two children had arterial hypertension proximal to the stenosis, the youngest patient has a dilatative cardiomyopathy and is listed for heart transplantation. The pressure gradients in the cath-lab were 25, 40 and 65 mmHg in general anaesthesia. For all patients an S-zig design was chosen and the stent

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

lengths were 28, 28 and 56 mm. In all cases the new balloon-in-balloon (BIB) technique (NuMED Canada, plus Germany) for implantation was used. The stents were placed without any complication in all 3 patients. No residual gradient was seen in the two smaller children, a residual gradient of 8 mmHg remained in the bigger patient due to aortic arch hypoplasia and a tight ratio of the proximal stent. Echocardiographic follow up for 3 – 6 months showed no significant gradient (< 20 mmHg) in 2 patients. In the last patient a gradient of 35 mmHg was registered and a redilation is scheduled. We conclude, that the implantation of a CP stent with a BIB is suitable and safe for children in a wide range of body weight. The immediate results are satisfying, long-term data have to be collected.

P902

Can large atrial septal defects larger than 25 mm effectively and safely be closed by intervention?

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Aim of the study was to judge whether an Amplatzer Septal Occluder (ASO) can be used as a safe therapy instead of surgery for closure of even large atrial septal defects larger than 25 mm in diameter. Method: We report our experiences in 54 patients (pts.) out of a cohort of 506 patients after successful ASO implantation within a period of time of 3 years (yrs.). Results: Median defect diameter was 28 mm (25 – 38), median age was 40.8 yrs. (10.1 to 77.7 yrs.). Body weight ranged from 36.9 to 120.0 kg (median 71 kg). Due to an inevitable reduction of the stent size with increasing distances of the discs fixed at the thicker part of the atrial septum in larger defects we implanted devices 2 – 4 mm larger than the measured stretched diameter. Fluoroscopy times ranged from 7.0 to 29.4 minutes, with a median of 9.8 minutes. Follow-up studies were obtained after 48 hours, and one, six, and twelve months, then yearly. The median period of follow-up is 0.98 yrs. (0.1 – 3.0). Complete occlusion rate after 3 months was 91.4 % increasing from 42.6% immediately after implantation. A trivial hemodynamically insignificant residual shunt remained in 8.6 % of the pts. 5 pts. showed transient atrial tachyarrhythmias within the first three months after implantation and 7 remained in their preexistent chronic atrial fibrillation. Conclusion: The excellent results in the short and medium term make Amplatzer device implantation to a recommendable safe and effective alternative to surgery even in selected cases with large defects larger than 25 mm and make it to our treatment of choice due to the less invasiveness of the method. Final judgement, however, is only possible after long-term follow-up.

P903

Cardiac interventions in hypoxic children with stented/blocked systemic to pulmonary shunts.

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Five patients (Pt) with previous stenting to pulmonary shunts underwent cardiac catheterization (CC) to evaluate shunt function and/or hemodynamics. The diagnosis included: (Pt1) pulmonary atresia (PA/IS), ventricular septal defect (age 20 days). (Pt2) tricuspid atresia, (PA/IVS 15 days); (Pt3) d-transposition of great arteries with preoperative banding and shunt (6 months). (Pt4) mitral atresia, hypoplastic left ventricle (12 years). (Pt5) tetralogy of Fallot (1.5 yrs). The CC was undertaken as an emergency procedure in 3 (Pt1, 2 stabilized with prostaglandin infusion, and 3). The CC findings related to the shunt included stenosed distal end of shunt (BoS): 3 (Pt1, 2, 4); narrowed subclavian artery 1 (Pt1); stenosed confluence of pulmonary artery 1 (Pt1), complete occlusion of proximal BoS 2 (Pt3-cause:5 chronic). Interventions were done in all pts: balloon angioplasty (BA) of the shunt 3(distal BoS 3) with % increase in diameter (PTD) Pt 1 = 50%, Pt2 60%, Pt3 200%; BA of the subclavian artery 1 (Pt1 PTD= 37%). BA of the confluence of PA 1 (Pt1, no change). BA of pulmonary valve and right ventricular outflow tract 1(Pt5); catheter directed thrombolytic therapy 1(Pt3). Intervention resulted in improved saturation in all but one patient (Pt1) with confluence PA stenosis who required a contralateral shunt. One patient required blood transfusion related to the CC. No CC related complications were noted. Surgical intervention to improve saturations was deferred in 4/5 pts. Mean(+/-SD) saturation at discharge were 81+/-8%. At mean follow up of 5.8+/-2.8 months, patients have continued to do well. In conclusion: interventions directed at improving pulmonary blood flow/oxygenation in pts with stenosed/blocked systemic to pulmonary shunts can avoid surgery. Palliative surgery can be deferred in most such pts.

P904

Possible predictors of clinical outcome in neonates with critical aortic stenosis

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Background and Objectives: Balloon dilatation (BD) has become the initial treatment for neonates with valvar aortic stenosis (VAS). However neonatal critical aortic stenosis sometimes can better benefit from surgical Norwood procedure. Exact criteria in borderline cases are not yet clearly established. The present criteria reflect more of left heart size dimensions, than clinical or haemodynamic parameters. Therefore, above the conventional parameters, morphologic and haemodynamic clinically relevant parameters were evaluated. PATIENTS AND METHODS: Between November 1993 and November 2000 19 patients (12 pts <6 days, 6 pts 7–30 days) underwent BD. Ball-to-annulus ratio = 1. Aortic, mitral valve and ascending aortic and diastolic left ventricular diameter measured on 2D echocardiography, ductal patency and shunt direction on color-flow map were evaluated. Comparison was made with respect of the clinical outcome. RESULTS: Eleven patients survived. One patient died BD procedure related. Seven patients deaths during the observation period were not procedure related. Among the 11 survivors, one patient required surgical reintervention. All survivors had mitral left-right shunt direction at ductal level. Among the 8 deaths, all 5 pts with mitral right-left shunt at ductal level died. Both survivors and nonsurvivors had left heart dimensions within the normal percentiles. Conclusions: 1. The direction of the ductal flow was better related to the clinical outcome, compared with left heart structure dimensions. 2. Critical VAS pts with right-left ductal flow probably better benefit from the Norwood procedure.

P905

Mid-term follow-up data after ballon dilatation for valvar aortic stenosis in neonates and infants

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Objectives: Ballon dilatation (BD) has become the treatment of choice in neonates and infants with valvar aortic stenosis (VAS). Immediate results are well studied, mid-term data are sparse. Therefore we report our mid-term follow-up data. PATIENTS: Prevalence patients: 19 neonates, 34 infant underwent BD were followed and categorized into 3 age groups: A: < 6days, B: 7–30days, C: 1–12 months. Methods: Early (<30days) and late (>30 days) mortality, reintervention rate (RR), early Doppler gradient (EDG) after BD at hospital discharge and late Doppler gradient (LDG) at maximum follow-up (MFU) severe PAI (Pascotis' insufficiency (vAI) at MFU were correspondingly studied. Data are expressed in percent or as mean value ± SD. Statistical analysis was performed by using ANOVA. $p < 0.05$ was considered significant. RESULTS: see attached table. Conclusions: Mid-term results of aortic balloonvalvuloplasty in neonates and infants seems to bear a low complication rate in the respect of late mortality, reintervention rate, EDG, progression, AI progression.

P906

Spontaneous closure of major aorto-pulmonary collateral arteries (MAPCAS) after creating the duct in an adult

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In patients with reduced antegrade pulmonary artery perfusion development of MAPCAs is frequently observed. Increasing recirculation may lead to cardiac insufficiency but these vessels contribute essentially to blood oxygenation. We report a case of a 29 year old patient with double outlet right ventricle, pulmonary artery stenosis, duct dependent left sided pulmonary perfusion and left sided MAPCAs. Spontaneous occlusion of a left sided, modified Blalock Taussig shunt occurred early after operation at the age of 25. During the following years his clinical condition aggravated due to decreasing saturations and increasing hematocrit of 74%. The patient refused further surgical treatment but decided to be treated interventionally. Angiogram revealed a duct dependent left pulmonary artery with severe stenosis and kinking. A 9 mm x 40 mm self-expanding stent (Bard Angiostent, Memorherm) was implanted via the left carotid artery. The stent did not fully expand due to the kinking of the duct. In order to avoid pulmonary edema or rupture of the ductus arteriosus, the stent was not balloon dilated. The oxygen

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

saturation increased to 90 % and the patient was discharged on the following day. After three months elective recatheterisation was performed. At this time the hematocrit was decreased to 36%. The stent showed slightly more expanded without intimal proliferation. None of the pre-existing MAPCAs could be demonstrated. The patient reported significant increase of everyday workload. Conclusion: This case report demonstrates spontaneous self closure of MAPCAs after optimizing antegrade central pulmonary perfusion in an adult. Improved clinical condition was confirmed by reduced hemiatocrit as well as improved oxygenation. Also the duct was enlarged, the overall recirculation has been reduced.

P907

Outcomes of percutaneous radiofrequency-assisted valvotomy and balloon dilatation in infants with pulmonary atresia and intact ventricular septum

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Background: Primary surgical management of neonates with PA-IVS has been challenged with reports of catheter-assisted valvotomy and subsequent balloon dilatation of the pulmonary valve. As experience is limited, risk factors, safety, efficacy, and long-term outcomes are not known. This study analyses the outcomes of percutaneous (RF, valvotomy and balloon dilatation performed over an 8 years period at a single institution. Methods: The divisional database was searched for patients diagnosed between January 1992 and August 2000. Primary and follow-up echocardiograms, catheterization reports, angiograms, surgical reports and clinical charts were reviewed. Results: A total of 52 children were diagnosed with membranous atresia of the pulmonary valve with intact ventricular septum, of these, 50 underwent attempted catheter-assisted valvotomy and balloon dilatation of the pulmonary valve. There were 16 (32%) boys, the median weight of the patients was 3.25 kg (mean 3.4±0.95). In 11 (22%) patients right ventricule-coronary connections without a RV-dependent coronary circulation was present. Perforation was successful in 27 children. Complications included: loss of pulse (n=4), rhythm disturbances (n=7), perforation to the outside of the heart (n=5), MPA aneurysm (n=1), severe tricuspid regurgitation (n=1), and death in the catheterization laboratory (n=1). A Blalock-Taussig (BT) shunt was required in 14 (48%) patients between 1 and 24 days after the intervention. In 1 patient, a BT shunt was created first and the RF-perforation attempted at day 10. In 5 patients, RVOT repair was performed between day 0 and 47 after the intervention. Four patients died (BT shunt occlusion, sepsis and arrhythmia), 15 patients are on a biventricular track, 5 patients are at an end-stage for 1 ½ ventricular repair, 4 patients are in an intermediate stage, 1 patient was lost to follow-up and in 1 patient a Fontan was completed. Conclusions: Primary treatment with catheter-assisted valvotomy in PA-IVS is an efficient alternative to surgery and allows sparing and/or delaying cardiopulmonary bypass procedures.

P908

Stent implantation to stenotic bioprosthetic valves in the pulmonary position

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We report our experience with percutaneous stent implantation to relieve obstruction in bioprosthetic valves in the pulmonary position. A database search identified 9 patients (6 male) who underwent stent implantation across stenotic bioprosthetic pulmonary valves between July 1996 and July 1999 at the Hospital for Sick Children, Toronto. Catheter intervention was initiated if echocardiography revealed Doppler estimates of right ventricular pressure (RVp) >2/3 of systemic arterial pressure (or systolic septal flattening with an estimated gradient of >50 mmHg across the valve prosthesis). Catheterization was performed under general anesthesia at an age (mean ± SD) of 9.3 ± 3.5 yrs and weight of 32.0 ± 17.1 kg, 5.9 ± 1.8 yrs after surgical insertion of a bioprosthetic valve in the pulmonary position. 7 had Tetralogy of Fallot, 1 had congenital pulmonary stenosis/insufficiency, and 1 had a Rastelli operation. All had systolic septal flattening and RV dilatation with moderate/severe pulmonary insufficiency pre-intervention. Fluoroscopy times were 33.1 ± 5.5 min. Seven patients received single P4014, and 2 received single P308 stents (Palmaz, Johnson & Johnson) without significant complications. The RVp decreased acutely from 83 ± 16% systemic to 41 ± 10% (p <0.001, n=9), and the transvalvar gradient decreased from 49.7 ± 8.5 mmHg to 11.0 ± 5.9 mmHg (p <0.001, n=8). During the follow-up period (10.9 ± 8.1 months, n=8), 1 patient had an unsuccessful attempt at re-dilating the stent (RVp 60% systemic) and underwent uneventful surgical pulmonary valve replacement. None of the remaining patients had echocardiographic evidence of systolic septal flattening, and RV dimensions did not change significantly. Stent implantation is a safe and effective means of providing palliative relief of obstructed bioprosthetic valves in the pulmonary position, and may delay the need for pulmonary valve replacement.

P909

Randomized comparison of 2 transcatheter closure methods of patent arterial duct.

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A trial of arterial duct occlusion with a Rashkind double umbrella (DU) or wire coil (WC) was undertaken for pediatric patients weighing >10 kg with ductal shape < or =3 mm in diameter. Forty patients were randomized (2 were excluded due to a ductal diameter >3 mm on aortography) to either DU (n = 20) or WC (n = 18) groups. The groups did not differ significantly with respect to baseline characteristics. Data were compared as an intention-to-treat analysis by group of randomization. Cross-over occurred only in the DU group, where in 4 patients (20%) the duct diameter was < or =1 mm and could not be entered for umbrella placement. All remaining DU group patients had duct diameters > or =1.5 mm (p <0.0001). There were no embolizations or secondary implants in the DU group, but in the WC group there was 1 early and 1 late embolization, with 6 patients (33%) having 2 coils or more. Mean procedure and fluoroscopy times did not significantly differ. There was angiographic duct closure in 4/13 (31%) of the DU group and 4/13 (22%) of the WC group (p = 0.69). Combined with an echocardiogram, closure in 11/17 (65%) DU patients and 13/18 (72%) WC patients (p = 0.64) was documented prior to hospital discharge. One WC group patient received thrombolytic therapy for a femoral artery thrombus. Follow-up at a median of 6.5 months (range, 1.2 to 37 months) showed closure by Doppler echocardiography in 15/19 (79%) DU patients vs. 14/18 (78%) WC patients (p = 1.0). With a tendency towards similar procedural characteristics and outcomes, the higher cost of the double umbrella favors the use of coils for closure of the small arterial duct.

P910

The role of transesophageal echocardiography in transcatheter closure of secundum atrial septal defects by the Amplatzer Septal Occluder

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Aim: to define the role of transesophageal echocardiography (TEE) in closure of atrial septal defects by an Amplatzer septal occluder (ASO). Patients and methods: 200 patients with ASD in indium were examined by TEE between September 1995 and June 2000. Results: 28 patients (18%) with partial or totally deficiency of the posterior or inferior-anterior or inferior-posterior rim were not suitable for transcatheter closure. 54 patients (27%) had centrally positioned ASD, 92 (46%) had an ASD with sufficient superior-anterior rim, 9 patients had multiple ASD's and 8 presented with septal aneurysm associated with single defect and 4 with multiperforated aneurysm. Two patients were excluded during basic catheterization and 170 underwent implantation of ASO. ASO was correctly positioned in 144 cases at the first attempt. In others we demonstrated with TEE: unstable position of the left atrial disk (12 cases), opening of both atrial disks in the left atrium (5 cases); deployment of the device in patients with multiple ASDs through the smaller defect (3 cases) and in 1 case the device was too small and had to be replaced by a larger one. Conclusion: Morphological variations of the ASD are common. TEE is crucial for recognition of ASD morphology, measurements of rims and dimensions of ASD that are vital for proper patients selection. TEE allows precise guiding and positioning of the ASO, which is essential for safe and effective ASD closure.

P911

Echo-guided closure of atrial septal defects – is fluoroscopy still necessary?

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A method of echo-guided closure of atrial septal defects (ASD) using the Amplatzer septal occluder (ASO) without the use of fluoroscopy was

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

described by Ewert et al in 1999. However, it is still not known whether the device is safe enough or suited for routine use. All children intended to have an interventional ASD closure after April 1th, 2000 were enrolled in a prospective protocol. Catheterization included standard pressure registration and oxymetric shunt calculation. Correct catheter position were confirmed by transthoracic echo. Balloon sizing and ASD implantation were performed under transesophageal echo-guidance. So far, 18 patients (pts.), age 3.25 to 17 (median 7.2) years, body weight 12 to 60 (median 21) kg, have been enrolled. Total procedure time ranged from 43 to 130 (median 74) minutes. ASD was excluded in 1 and judged too large for intervention after balloon sizing (>38 mm) in 2 pts. ASD closure without the need of fluoroscopy was completed in 11 pts. (device size 10 to 22 (median 17) mm). In 4 pts (device size 28 to 34 mm) fluoroscopy had to be used because of problems with configuration or positioning of the device. Fluoroscopy time was 2 to 10 (median 4.0) minutes. Successful ASD closure was possible in 2 of them (both 30 mm). In the remaining 2 pts the device was withdrawn because no complete occlusion (28 mm) resp no stable position (34 mm) was obtained. Fluoroscopy is not necessary for hemodynamic assessment and balloon sizing of ASD in children. Echo-guided closure of smaller and medium sized defects with the ASO is suited for routine use. Larger defects require the combined use of fluoroscopy and transesophageal echocardiography. Extremely large defects remain surgeon's property.

P912

Mid term results of stent implantation in congenital and acquired cardiovascular disease

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Objective: Pre- and postoperative coarctation of peripheral pulmonary arteries and other variable vessel stenosis associated with congenital heart disease can be effectively widened by stent implantation. We describe indications, effectivity and complications in mid term follow up. Patients: From 1994–5/2000 in our institution 119 Pts aged 1 day to 22 years were treated by stent implantation. 44 Pts had no previous surgery or transcatheter intervention, 75 were postoperatively. Stenotic sites were: Mustard Baffle (n=6), pulmonary artery branch stenosis (n=34), coarctation of the aorta (n=12), pulmonary venous obstruction (n=2), stenoses of anastomoses (n=7), aortopulmonary fistel (n=1), Fontan tunnel (n=1), systemic veins (n=2), aortopulmonary collateral pathways (n=4), ductus arteriosus (n=44), renal artery (n=2), peripheral arteries (n=1), right ventricular outflow tract (n=3). Methods: Most of the stenoses were augmented with balloon expandable stents (Jo-Med, Palmaz, NIR), some we used a self-expanding covered stent for sealing a tunnel leakage (Endotex). In 8 pts stent implantation was performed intraoperatively. In 80% of the interventions general anaesthesia could be avoided. Results: In 58 pts a single approach was successful, in 16 pts redilatation of the stents was necessary, in 17 pts a second stent was placed. Surgical removal of the stents was performed in 35 pts, mostly ductus stents during consecutive corrective or palliative surgery. Dislocation of stents was the most frequent complication, hemodynamic deterioration with low cardiac output occured in 8 pts. Conclusion: Stent implantation is effective in mid term widening vessel stenoses of variable locations. The indication has to be weighed against the risk of surgery. In-stent-stenosis due to intima proliferation may occur and require reintervention, the rate of complications after the initial learning curve is low.

P913

Trans-catheter (Amplatzer) versus surgical closure of asd: a prospective comparison of results and cost.

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Objective: To compare effectiveness, complications and cost of Amplatzer and surgical ASD closure. Methods: 43 consecutive patients were prospectively followed after ASD closure (27 Amplatzer, 19 surgical). Parameters assessed were procedural success, complications, regression of right ventricular dilation (RVED) dimension and CTR, at 6 months post procedure, and cost (derived from institutional accountancy data). Results: Amplatzer closure was successful in 24/27 (89%) of patients. Surgical closure was successful in all 19 cases (including 3 failed Amplatzer procedures). There were no deaths. Only 1 (3.7%) complication (device embolisation) occurred in the Amplatzer group but 9 surgical patients (47%) had complications; 4 pericardial effusions (both requiring readmission, 1 requiring drainage), 3 small pneumothoraces, 1 small pleural effusion and 1 SVT (p <0.005). There was no significant difference in regression of RV dilatation at 6 months (median tricuspid ring diastolic annular decrease: Amplatzer 17.5%, range 0–45.8, surgical 15.1%, range 5–57.9). Median CTR decrease: Amplatzer 7.5% range 0–26, Surgical 7.5%, range 0–31). Hospital stay was significantly shorter with transcatheter closure (median 1 day vs 6 days for surgery) but transcatheter closure was more expensive overall than surgery (respective costs £5529 +/– 2974, vs £3476 +/– 2100). Conclusions: Amplatzer ASD closure has a lower chance of success with a single procedure than surgery but is associated with significantly fewer early complications. Resolution of RV dilatation is similar for both techniques. Hospital stay was shortest for the Amplatzer group but cost, on average, was higher than for surgery.

P914

Long-term outcome of pulmonary artery stent implant: A single center experience over 10 years

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We reviewed all patients who underwent pulmonary artery (PA) stent implant from 9/1989 to 2/2000 to determine the efficacy and long-term outcome of PA stents. Patients were divided into four groups: Tetralogy of Fallot/pulmonary atresia (TOF/PA), congenital branch pulmonary stenosis (CBPS) status post arterial switch (ASO) and status post Fontan operation (Fontan). Results: 612 stents were implanted in 200 patients: 430 stents in 207 TOF/PA patients, 93 stents in 46 CBPS patients, 45 stents in 16 ASO patients and 43 stents in 31 Fontan patients. Mean age 12.2 years (range 0.1–47.8). Mean weight 30kg (range 4.9–96). Mean follow-up interval 5.2 years (range 0.2–10.2). In the TOF/PA group mean systolic gradient decreased from 40 to 9 mmHg*, mean vessel diameter increased from 5.6 to 11.4 mm*, and the right ventricle:systemic arterial pressure ratio (RV:SA) decreased from 0.63 to 0.45*. CBPS group mean systolic gradient decreased from 45.5 to 8.5 mmHg*, mean vessel diameter increased from 4 to 9 mm* and RV:SA decreased from 0.59 to 0.44*. ASO group mean systolic gradient decreased from 43.9 to 7.8 mmHg*, vessel diameter increased from 7.6 to 13.3mm*, and RV:SA ratio decreased from 0.67 to 0.39*. Fontan group mean vessel diameter increased from 6.1 to 12.7 mm*. Complications included: pulmonary edema (n=5, 1.6%), stent migration (n=7, 2.3%), hemoptysis (n=2, 1.0%) and death (n=1, 0.3%). Cumulative survival was 0.98 at 10 years for TOF/PA group, 0.92 for CBPS group, 1.0 for Fontan group and 0.94 for ASO group * (p<0.01). Conclusion: Pulmonary artery stent implant is safe and effective therapy with low complication rate and excellent long term survival up to ten years.

P915

Transcatheter perforation of atretic pulmonary valves

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Opening the atretic pulmonary valve in patients with pulmonary atresia and intact ventricular septum (PA/IVS) provides antegrade pulmonary flow while decompressing and promoting growth of the hypertensive right ventricle (RV). Surgical relief of RV outflow tract obstruction continues to be widely used to achieve this objective because of the lack of an easy to use transcatheter technique. We used the new Nykanen radiofrequency (RF) perforation system in 5 patients (3 neonates and 2 older patients – 22.15 and 30 months) with pulmonary atresia and intact ventricular septum. The neonates were duct dependent and on a prostaglandin infusion and the 2 older patients had surgical systemic-pulmonary shunts. The atretic pulmonary valve was easily perforated with a brief application of 5–W of RF energy. In the neonates the coaxial catheter could be passed directly over the RF wire to allow introduction of a .014wire. In the 2 older patients the coaxial catheter would not track and the perforated valve was re-crossed with a .014

P916

Multicentre experience comparing two systems for coil closure of persistent ductus arteriosus

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Two systems for interventional closure of persistent ductus arteriosus (PDA) with detachable coils have been used in three pediatric heart centers. This study aims to compare the results of both devices. Patients: Between Jan 1996

and Dec 1999 95 children (mean age 4 5 (5 7) yrs, mean weight 17.2 (12 9) kgs) underwent coil closure as initial treatment for PDA. Coil size selection was based on PDA diameter. A Cook(r) detachable coil was used in 58 pts (group B), a PFM(r) detachable coil was used in 37 pts (Group B). Results: Mean age, weight, Qp/Qs and PA/AO pressure ratio were comparable in both groups. PDA diameter was slightly larger in group B (p<0.05). In most patients only one coil was used initially. Early embolization occurred in 3/58 pts in group A and in 4/37 pts in group B. Hemolysis requiring reintervention occurred in only 1 pt in group A. Late systemic embolization occurred 1 pt in group B. Residual shunting on echo shortly after implantation was comparable in both groups. During follow up a 2nd coil for significant residual shunting was needed in 2/58 pts (A) and 5/37 pts (B). Long term follow up revealed a trend towards earlier spontaneous closure of residual shunts in group A. Conclusion: Both detachable coil systems show comparable effectiveness with a low complication rate.

P917

The change of configuration of the Amplatz Septal Occluder in undersized defects

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Purpose: To describe the change of configuration of the Amplatzer Septal Occluder (ASO) when placed into small defects. Method: Three ASO devices (24mm, 16mm and 10mm) were placed sequentially into smaller size defects at 2mm decrements. The configuration of the device was recorded on high resolution video. The change in configuration was observed for each device. The change in the disk diameter and the interdisk space were measured and the trend of change documented. Results: As the devices were placed into smaller holes, the diameter of both the disks decreased as the thickness of each disk increased. The most obvious change was the increase in the interdisk space with placement in smaller defects. (see Figure 2) Conclusion: Very often, an oversized ASO is used for closure of atrial septal defects. This study confirms the change in the configuration of the device in small deficiencies.

P918

Balloon valvuloplasty for infant aortic stenosis; acute results and subsequent need for the Ross procedure.

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Balloon valvuloplasty (BAV) is commonly employed as the initial therapy for infants with critical aortic stenosis (AS). There is little information regarding need for subsequent surgical intervention after BAV, however. We retrospectively reviewed our experience with BAV in infants under 6 months of age and their subsequent need for surgical intervention within the next 36 months. Variables examined were change in aortic valve gradient, development of AI, acute and short term morbidity and need for surgical intervention. Twenty infants with a mean age of 1.9 +/- 1.6 months underwent BAV at our institution between 1-97 and 11-00. 7 were approached from the carotid artery. The peak gradient pre-BAV was 73.5 +/- 28.8 mmHg and was reduced to 20.0 +/- 17.4 mmHg ($P < 0.0001$), a 72% reduction. Nine patients (45%) had CHF before BAV. Function improved in all 9 pts. No patient had AI pre-BAV. 14 (70%) developed AI post-BAV. Three (15%) had moderate to severe (3-4+). 5 (25%) had mild (2+) and 6 (30%) had trivial (1+) AI. The mean balloon annulus ratio was 0.96 +/- 0.07, range 0.84 to 1.2. All patients with AI had balloon annulus ratios < 1. Six (30%) required surgical intervention 15 h +/- 11 months after BAV, range 6 to 36 months. Five of the 6 required a Ross procedure, 4 for severe AI, 1 for combination AS/AI. No patient with trivial AI progressed and only one patient with 2- AI worsened to 3-4+ AI. There were no post-BAV deaths but one patient died following the Ross procedure (18%). We conclude that BAV is an effective therapy for infant AS. Surgical intervention was needed mainly for those pts with 3-4+ AI immediately post-BAV.

P919

Transcatheter closure of symptomatic atrial septal defect (ASD) in infancy.

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To assess feasibility of trans-catheter ASD closure in symptomatic infants. Between December 1996 - September 2000, eight infants (age 0.2 - 1.8, mean 8 months) (3 - 10.7kg, mean 6.1kg) underwent attempted Amplatzer septal occluder (ASO) closure for ASD. Two neonates (3.3-3.9kg), one with cranial PVS and one with pulmonary atresia intact septum, post successful wire perforation and ballooning of PV with persisting cyanosis and unsuccessful trial of balloon ASD occlusion underwent 3, 8mm ASO closure. Subsequent late (FT) 14 minutes for one and the other under echo guidance only. Four infants underwent closure with 11 - 15mm ASO (FT 13 - 37, mean 24.5 minutes). One other infant had FT 94 minutes with multiple attempts at occluding two large ASDs, finally, 14mm ASO successfully implanted. Subsequently noted cerebral vascular occlusion, gradual recovery. Another infant 6kg had FT 38 minutes, balloon sized 18mm ASD. Multiple attempts at implanting sequentially 14mm, 16mm, 15mm ASO failed. There was haemodynamic instability and subsequent embolic brain phenomena noted; gradual recovery occurred. Subsequent surgery revealed multiple fenestrated ASDs, one large (15X 14mm) with no superior rim and four smaller ASDs. Follow up period, (2 - 48, mean 23 months). One patient two months post implant developed septicemia and endocarditis vegetation on device necessitating surgical removal. Other patients remain well with no residual ASDs. Trans-catheter ASO closure is feasible in symptomatic infants, more demanding in the large multiple ASDs and more vigilance required for technical skills and haemodynamic monitoring.

P920

Surgical treatment for ASD ostium secundum in the device era

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Between 1993 and 1999, 284 ostium secundum atrial septal defects (ASD) were closed in our centre, with the ASD being the primary indication for intervention. This was done surgically (N=196) or, since 1998, with the Amplatzer septal occluder (N=88). This report evaluates the evolution of the means of closure and the results of the respective techniques. The median age of surgical patients (45% male) was 3 years (6 months - 24 years). 29% had associated minor cardiac anomalies and 10% chromosomal anomalies. The median age of device patients (46% male) was 5 years (6 days - 60 years). 2% had associated minor cardiac anomalies. There were no early or late deaths in either group. In terms of morbidity 3 surgical patients had post-operative pericardial effusion requiring drainage, and one developed mediastinitis. Of device patients there were 100 attempted device implantations in 95 patients. Two patients suffered embolic cerebrovascular accident during the procedure, and 11 had initial failure of the implantation. Of these, 5 had unsuitable anatomy at catheterisation (multiple defects 2, inadequate rim 1, oversized defect 2, associated IVC 1) and subsequently had surgery. Four patients had later successful device closure. Two patients await further management. One device was surgically removed eight weeks after insertion for endocarditis. 31 device patients demonstrated early residual shunting (data for 83/88). One of these patients had a small residual shunt at 12 months follow-up (data for 16/31). The relative numbers of device to surgical closures for fossa ovalis ASD were 14:22 in 1997, 54:25 in 1998 and 24:11 in 1999. We conclude that device closure of ASD is associated with significantly greater morbidity than surgery. Surgery is indicated in younger patients and those with anatomical contraindications to device closure.

P921

Novel transseptal puncture technique for transcatheter closure of complex patent foramen ovale

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Introduction: Transcatheter closure of patent foramen ovale (PFO) may be difficult or impossible in patients with significant septal overlap (SO) between septum primum (S1) and septum secundum (S2). We compare results using the original atrial septal defect" approach and a novel transseptal puncture delivery technique. Methods: 23 patients, age 16 - 72 years, with SO ranging from 8 - 16 mm (mean = 12.0 +/- 2.4) underwent transcatheter closure of PFO between July 1997 and November 2000. In Group 1 (N = 9) the delivery sheath was passed through the PFO to the left atrium (LA). In Group 2 (N = 14) a Brockenbrough transseptal puncture was performed just below the site of SO to pass the delivery catheter to LA. S1 and S2 were sandwiched" together between the superior aspects of the device occluders, preventing further flow. Results: There was no difference between groups in either age or SO. In Group 1 there were technical difficulties with device delivery in 5/9 (55%): failure to implant (2), failure of LA occluder to appose the septal surface due to folded S1 tissue under the device (2), and partial collapse of LA occluder (1) at PFO channel. Right to left shunting, by contrast transesophageal echocardiography (CTEE) increased significantly

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

after successful" device placement in one additional patient. There was complete acute closure by CTEE in 3/7 (42.8%) implanted devices in Group 2, there were technical difficulties in 0/14 pts: all devices were implanted, were flush with the LA septal surface and were fully opened on both sides. Complete acute closure occurred in 9/14 (64%). Conclusions. The new transseptal puncture simplifies transcatheter closure of PFO by eliminating long SO as a risk factor for procedural failure and poor outcome.

P922

Balloon pulmonary valvuloplasty to the small pulmonary valve in tetralogy of Fallot.

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The purpose of this study is to assess the role of balloon pulmonary valvuloplasty (BPV) in patients with tetralogy of Fallot (TOF), particularly with small pulmonary valve. Our hypothesis is that the pulmonary valve grows following BPV which results in preventing from transannular patch or corrective surgery. From June 1993 to July 1998, 22 patients (15 males and 7 females) of TOF with small pulmonary valve underwent cardiac catheterization, which was followed by corrective surgery. The age at cardiac catheterization ranged 8 mo to 62 mo (mean 25+/-15 mo). Six patients (Group 1) underwent corrective surgery with transannular patch and 16 patients (Group 2) did not require transannular patch. The diameter of pulmonary valve was significantly smaller in Group 1 (61.0+/-9.4 % of normal vs. 75.9+/-15.7 % of normal in Group 2: p<0.05). Particularly in 16 patients required repeated cardiac catheterization, 5 patients (Group 3) underwent catheterization only, and catheterization as well as BPV were performed in remaining 5 patients (Group 4). In Group 3, the increase in diameter of pulmonary valve was not significant (73.5+/-15.7 % of normal to 79.0+/-17.1 % of normal). However, in Group 4 the diameter significantly increased from 50.6+/-21.4 % of normal to 65.1+/-18.3 % normal (p<0.05). In conclusion, small pulmonary valve of TOF grows after BPV, which triggers the role in avoiding transannular patch repair.

P923

Interventional catheterization after glenn or fontan procedures.

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Introduction: Univentricular heart patients may need several operations before definitive result". BT shunt or PA banding often disrupt pulmonary arterial anatomy. Further surgery on those vessels is difficult. New vessel formation may increase cyanosis. Such vessels can be treated in the cathlab, avoiding surgery. We present our experience in therapeutical interventions in Fontan-type (F/G) operated cases. Material and Method: 35 patients (pts) with a previous (F/G) operation were catheterized because of clinical deterioration, in one or more sessions. Ages ranged from 3.5 to 30 years, mean 9.3. Stenosis of one or both pulmonary arteries were present in 17pts. Balloon angioplasty (BAP) in 8pts. Stent implantation in 9. Abnormal vascular connections closed in 12. Coil occlusion of the azygos vein: 5pts. Bronchial embolization for hemoptysis: 3 (coils or mako spirals). Coil occlusion of residual BT: 2. Intrahepatic veno-venous fistula closed with an Amplatzer vein occluder, and 1 pt with unnecessary IVC-RPA conduit, had it closed with an Amplatzer Duct Occluder. Unnecessary IVC-RPA conduit closed with an Amplatzer Duct Occluder. In six pts stents were placed: atrio-pulmonary connection-2, Superior and Inferior Vena Cava-3, iliac vein opening-1. Results: Pulmonary branch stenosis ballooning gradient diminished from 8.6 to 3.1mmHg and diameter of stenosis increased from 2,88 to 6,14mm, mean values. In stented patients, same values changed from 9.1 to 0.28mmHg and 5,2 to 10mm respectively. Only temporary improvement was observed in 4/9 pts, treated with BAP. All abnormal connections were completely closed. Significant saturation rise in 2 pts treated with Amplatzer Occluders and in 3/5 pts with azygos embolization. Conclusions: Follow-up catheterization in patients after (F/G) procedures, with clinical deterioration, may demonstrate complex lesions, amenable to treatment in the cathlab, avoiding the added risks of further surgical procedures.

P924

Extended application of the retrievable spiral coil in the treatment of patent ductus arteriosus with various modifications of the closing configuration.

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The transcatheter closure of ductus arteriosus using embolization coil has been acknowledged as a standard treatment in small sized ductus, but the limitations in the application to larger ductus have been encountered. To assess the mid-term results and efficacy of retrievable spiral coil in transcatheter closure of ductus arteriosus and to evaluate the effect of extended application of the device to a larger ductus with configurational change, we investigated 100 patients (median age: 20 months, male: female: 67/33 cases). The obtained results were: 1. The closure rates (CR) on 12 months follow-up were up to 94%(82 out of 87) in small to moderate sized (<4mm) ductus and 77%(10 out of 13) in larger ductus (>4mm). 2. The narrowest diameter of ductus was larger in patients with residual shunt (4.3±0.6mm) than without residual shunt (2.7±1.0mm, p=0.028). 3. We applied original shaped devices in 55 patients (CR=95%) and attempted some modifications with the closing configuration such as: wedge shape, 21cases (CR=100%), cross bar shape, 18cases (CR=78%), double coil; 5 cases (CR=80%), and reverse type, 3 cases (CR=100%). The cross bar shape, reverse and double coil type embolizations proved to be more effective in closing larger ductus (2.5±0.8 mm vs. 3.5±1.0 mm, p=0.0001). 4. There were two cases in which the coil had migrated into the distal pulmonary artery and one into the descending aorta on the day of coil embolization. Retrieval of the migrated coil and deployment of a second coil was successful in all 3 cases. In conclusion, transcatheter closure of the patent ductus arteriosus with Duct Occluder is an effective and safe therapeutic modality and we can extend its application to larger ductus with various modifications of its closing configuration.

P925

Wire-Stent to impede progressive dilation of the aorta: an experimental study.

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Patients with connective tissue disease (Marfan) are known to suffer from progressive dilation of predominantly the aorta, resulting in late aortic dissection, rupture or aortic valve dilation & regurgitation. This study aimed to establish the feasibility and safety of implanting specially performed wire-stents in the ascending and descending aorta in order to strengthen the vessel wall. Methods: Different wire-stents were developped and deployed in plastic life-size aortic arch models. The properties for succesfull deployment through a catheter (sheath) were determined. 5 normal young pigs (weight 25-30 kg) had different wire-stents placed throughout the arch. Four weeks after wire implantation (weight 45-60 kg), the pigs were examined by angiography and sacrified. The aorta was inspected for the degree of the wire-in growth, circulus formation, the influence on the aortic wall and vessels branching off the aorta. Findings: The wire stent in the ascending aorta was difficult to implant, and all went had migrated during the 4-week period. 1 wire-stent had migrated retrogradely through the aortic valve. In contrast, the wire-stents in the descending aorta were easily deployed, and retained their shape and position. There was no closing or obstruction to the flow, no obstruction at branching vessels, no evidence of distal thrombus embolisation. On histologic inspection the wire was well covered by intima. Conclusions: Because of significant pulsatile flow, a wire-stent in the ascending aorta was difficult to position, and migrated during follow-up. A more complex stent needs to be developped for the position. In contrast, the wire-stent in the descending aorta proved to be a low-cost, safe & effective way of strengthening the vessel wall.

P926

Minithoracotomy for Atrial Septal Defect closure, a cosmetic alternative to device closure for the developing world.

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From July 1994 to June 2000 61 children had surgical repair of isolated secundum atrial septal defects (ASDs) at our hospital through a right anterior mini-thoracotomy (4th to 5th interspace) without other incisions. The operation was cosmetic and rapid convalescence, and is presented as a cheaper alternative to transcatheter device closure of ASDs for developing countries. Repairs were all done on cardiopulmonary bypass at normothermia with a fibrillated heart. Average bypass time was 15 minutes (7 to 27 minutes). There was no operative or late mortality and no significant morbidity. Specifically, there were no phrenic nerve palsies, no wound infections, and no reoperations. Mean hospital stay was 5 days. Closing in this country indicates this is a

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

significantly cheaper option to transcatheter device closure. The anterior mini-thoracotomy is a safe and effective approach for repair of secundum ASDs and give a cosmetic result superior to more traditional approaches.

P927

Laser-Assisted Balloon Valvotomy for Pulmonary Atresia with Intact Ventricular Septum: Predictors of Initial Success and Biventricular Circulation

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Laser valvotomy with balloon valvoplasty may be effective in selected patients with pulmonary atresia and intact ventricular septum (PAIVS). We sought to determine factors that predict initial procedural success and achievement of biventricular circulation. Laser-assisted balloon valvotomy was attempted in 9 neonates and infants with PAIVS at a median age of 10 (range 3 to 270) days and weight of 3.4 (range 2.6 to 4.8) kg. Based on initial outcome, group I (n=5) comprised surviving patients with adequate forward pulmonary flow, while group II (n=4) comprised those with sub-optimum outcome including fatality, procedural failure or shunt insertion. The demographic, anatomic and hemodynamic variables were compared between the 2 groups. The atretic pulmonary valve was perforated and dilated in 7/9 babies. The procedure was abandoned in one and resulted in cardiac tamponade and death in another. Two patients required shunt insertion to improve systemic oxygenation. Group I patients had right ventricles that were larger (Lewis index: 12.4±1.5 vs 9.2±0.8, p=0.0016), of tripartite morphology (5 tripartite in group I vs 4 bipartite in group II, p=0.008), and not associated with ventricular coronary crossing (0 in group I vs 3 in group II, p=0.048). The initial outcome was not related to the age at intervention, hemodynamics, z-scores of tricuspid and pulmonary valve annulus or infundibular size. There was 1 septo-related in-hospital death. Of the 8 survivors, 5 (4 in group I, 1 in group II) achieved complete biventricular circulation. 3 after a second balloon pulmonary valvoplasty. The remaining survivor had a bipartite right ventricle that did not grow and was awaiting 1&1/2 ventricular repair. A tripartite right ventricle with absence of ventricle-coronary artery and a Lewis index of > 11 predict initial success and eventual achievement of biventricular circulation. Bipartite right ventricle may grow after the interventional procedure and be incorporated into the biventricular circulation.

P928

Closure of atrial septal defects with atrial septal aneurysm using the Amplatzer occluder – immediate and short-term outcome

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Background: Atrial septal aneurysms (ASA) occur in association with atrial septal defects (ASD) or patent foramen ovale (PFO), especially in patients presenting with stroke. Safety and efficacy of device closure in the presence of ASA has not been examined previously. Method: Echocardiograms of 89 consecutive patients who underwent device closure of ASD/PFO between February 1997 and February 2000 were studied for presence of ASA. Catheter procedure and outcome were examined. Results: 14 patients were identified to have ASA associated with ASD/PFO. Age range was between 1.4 to 66 yrs (median=31 yrs). Weight ranged from 8.7 to 113 kg (median=55 kg). 7 were male and 7 were female. 12 patients had ASD while 2 patients had PFO. Five patients had history of stroke. Multiple defects were present in 5 patients. Balloon sizing was performed by pull through technique. ASD size by TEE ranged from 0 – 21 mm (median=10mm). Balloon stretch diameter ranged from 4 – 28 mm (median=15 mm). Sixteen devices were placed in 14 patients. Size of Amplatzer device ranged from 7 – 26mm (median=14 mm). No complications were observed. Redundancy and excursion of atrial septum was eliminated in 8 patients and diminished in the rest. One patient had small residual defect immediately after closure (93% complete occlusion rate), which had resolved at 6 months follow up (100% complete occlusion rate at 6 months). No stroke was reported during follow up. Conclusion: ASA occurs with a significant frequency in patients referred for device closure of ASD/PFO. Successful closure of ASD/PFO with ASA is feasible using Amplatzer ASD occlusion device and is safe. Immediate and short-term results were similar to patients who did not have ASA associated with ASD/PFO.

P929

Follow-up After Transcatheter Device Closure of Ventricular Septal Defects in Infancy

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Follow-up After Transcatheter Device Closure of Ventricular Septal Defects in Infancy. Michael Rigby, Ramanjit Dhillon, Alan Magee. An increasing number of patients are undergoing transcatheter closure of ventricular septal defects (VSD). Since 1991, fifteen infants (mean 1-12 months) have undergone closure of a VSD using the 17mm Rashkind umbrella in twelve and the Amplatz ductal occluder in three. During this period, the procedure was unsuccessful in four because the defect was too large and two additional patients required surgery because of malpositioning of the device. When the remaining fifteen patients were assessed to determine the outcome of the procedure, four out of six with a perimembranous defect had partial or complete right bundle branch block. On Holter monitoring, all patients remain in sinus rhythm without atrial or ventricular premature beats. With transthoracic and transoesophageal echocardiography, a residual VSD was present in three, two of whom had a perimembranous defect. Mild tricuspid regurgitation was present in four. The Doppler derived right ventricular systolic pressure was 22–34 (mean 28) mmHg. No patient had left or right ventricular outflow tract obstruction. One patient has developed aortic incompetence and in three additional patients with a perimembranous defect, the left ventricular device is in apposition with the right coronary aortic leaflet. At cardiac catheterisation, small residual defects were confirmed in four patients (mean QP/QS 1.4:1, mean pulmonary arterial systolic pressure 25mmHg. Following transcatheter device closure of ventricular septal defects in infancy, all patients are symptom free. Right bundle branch block is present in nine patients, and four with a perimembranous defect have developed or have the capacity to develop aortic regurgitation or subaortic stenosis because of protrusion of the left ventricular device in the left ventricular outflow tract.

P930

Incidence of residual leakage after catheter intervention is higher in medium sized than in large persistent ductus arteriosus

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We reviewed the results of catheter closure of persistent ductus arteriosus (PDA) in 124 consecutive cases performed in our institution from 1995 to November 2000. 76 girls and 48 boys, median age 3 years (0.3–16.8 years) and median weight 15.1 kg (3.4–72 kg) were treated. Two cases had had earlier surgery for PDA and in two catheter closure had been performed using the Rashkind device. Rashkind devices were used in 7, Sideris in 2, Amplatz duct occluder in 18 patients and Cardioseal in one. 86 Cook coils, 15 PFM coils and 12 Gianturco coils were employed in the remaining 104 cases. On nine occasions coils embolized to the pulmonary circulation. All but one were retrieved. Reintervention was performed in five patients (4%). Surgical extraction of a 17mm Sideris device was needed due to malposition of the device before release. One cook coil impossible to unscrew from the delivery catheter dislodged to the left femoral artery and was surgically removed. The incidence of patients with complete closure of the PDA increased from 60% immediately after the intervention to 95% at follow up three years after treatment. A significantly higher proportion of residual leak was found three months or later after catheter intervention of middle sized PDAs (2.1–3 mm) compared to small (0.5–1.9 mm) or large PDAs (>3 mm) as measured by angiography (p=0.016). The higher rate of complete closure in large PDAs is probably due to the use of Amplatz devices in these cases in later years. We conclude that a high rate of PDA closure was achieved by catheter treatment (95%) at three years follow up. To further improve outcome a change of treatment strategy in patients with middle sized PDAs seems most important.

P931

Transcatheter Closure of Atrial Septal Defects (ASD) in Children Under Six Years of Age

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We report the single center experience with device occlusion of ASD using the Amplatzer Septal Occluder at preschool age, the typical age of surgical closure. Device occlusion was attempted in 41 patients with a median age of 4.2 years (1.4 – 5.9 years), median weight of 15.2 kg (8.7 – 33.5 kg) and a mean QP/QS of 1.9, and accomplished in 40. Seven patients had more than one defect. Defect size ranged from 2 to 16 mm by echo with the stretched diameter ranging from 5 to 24.5 mm. Seven additional interventions were

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

performed in 5 patients: coil occlusion of Blalock-Taussig shunts in 2, pulmonary artery stent in 2 and coil occlusion of patent ductus in 3. Two devices were implanted in one patient. Device size ranged from 4 to 26 mm (median 15 mm). Implantation from the right internal jugular vein was done in 2 patients. Procedure time was 40–190 min (median 110 min) with 11–42 min of fluoro time (mean 17 min). There were no device embolizations or other major complications. One patient developed transient AV block, which resolved at the end of the procedure and the marker band detached from the delivery sheath in one patient. Complete occlusion of the defects was achieved in 31/41 (77%) at 24 hrs, in 30/35 (86%) after 6 months and in 25/28 (89%) after one year. Multiple and larger defects were associated with residual shunts. There were no late complications. We conclude that elective transcatheter closure of ASD at preschool age using the Amplatzer Septal Occluder is safe and effective.

P932

The immediate and follow-up results of balloon angioplasty in 65 children with native coarctation

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The aim of this study is to review the results of our balloon angioplasty in 65 pediatric patients with native coarctation of the aorta. Our experience consists of 68 balloon dilation procedures in 65 pediatric patients (35 males, 30 females), 25 of them as isolated coarctation of aorta, others associated with various other cardiac anomalies. Their right arm systolic blood pressures were 141.1±26.2 mmHg (mean±SD). With balloon dilation, mean systolic gradient decreased from 54.3±18.9 mmHg to 17.1±12.3 mmHg; coarctation diameter increased from 3.1±1.4 mm to 7.2±2.1 mm after the procedure by angiography. Peak systolic gradient was measured successfully in 40 (62%) patients and 6 (9%) of these patients (10 day, 2.4,5,12 month, 6 year old) represented reoperation in the follow-up period between 3.5–6 months. Three of these 6 patients were redilated successfully by second balloon angioplasty. Procedure was partially successful in 22 (34.4%) patients. Balloon angioplasty and valvuloplasty were simultaneously performed in 7 patients and both procedures were successful in five of them. The mean follow-up duration was 23,2±5.8 months. The mean systolic gradient by echocardiography was 19.4±5.0 mmHg and mean systolic blood pressure was 113±15.5 mmHg in the last follow-up visit. Complication rates were remarkably low, with 1 early and 1 late aneurysm formation (3.1%) and transient pulse loss in 4 (6.1%) cases. One case died of rupture of a Berry aneurysm after a successful dilation. Due to reoccurrence of residual gradient or aneurysm formation 6 (9.2%) patients (associated with additional cardiac anomalies) required surgery. We conclude that balloon angioplasty is a safe and effective method of treatment in discrete native coarctation of aorta. Both balloon angioplasty and valvuloplasty procedures can be performed effectively to the patients with aortic coarctation and stenosis. There are controversies in the literature, however we believe that balloon angioplasty of discrete native CoA in newborn and infants under one year is effective and safe, and we recommending balloon angioplasty as the first choice as well as in older children.

P933

Distortion of magnetic resonance images by embolized coil

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Background. Although coil embolization for patent arterial duct(PDA) has become popular, it is not clear how far magnetic resonance images (MRI) after coil embolization is distorted around the embolized coil. The purpose of this study is to determine the spatial range of distorted MRI by various embolized coils in both horizontal and vertical direction. Methods. Using Gyroscan T5-NT(0.5T) (Philips Medical) and signa advantage(GE) (1.5T) with surface coil, MRI of phantom made of Acryl plate with a 3cm diameter lattice was investigated. The phantom was placed in 5mM Cupper Sulphate solution to visualize the lattice on MRI, in the center of which the following coil was attached : (1) Stainless steel coil, Gianturco(5mm) Jackson detachable coil (JDC) (5mm, 8mm) (2) platinum coil; fornade(5mm). (3) Inconel coil, Microye (5mm). MRI of T1 or T2 weighted spine echo parallel to the phantom plate with 10mm slice thickness were obtained. Results. Stainless steel coil distorted and lost the MRI images around the coil both in horizontal and vertical direction, proportional to the coil diameters and the number of coiling as well as to the strength of magnetic filed gradient, i.e. JDC(8mm) distorted 12cm wide in the horizontal plane and 14cm wide in the vertical direction(fig1). No distortion of MRI were found in platinum and Inconel coil. Conclusion. The commonly used stainless coil for PDA loses and distorts the MRI of organs located in axial and sagittal direction of the coil, while the Inconel coil shows no distortion and provide a chance for future diagnostic MRI examination of cervial and upper chest cavity.

P934

Percutaneous closure of Patent Ductus Arteriosus with Gianturco coils.

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We evaluated our immediate and short- term results of transcatheter coil closure (TCC) of Patent Ductus Arteriosus (PDA) using single or multiple Gianturco coils. From January 1996 to August 2000, TCC of PDA was attempted in 163 patients (114 female and 49 male), at median age of 6.2 years (6 months to 52 years). Coils were placed by the transarterial femoral route in all cases. The median PDA diameter was 2.5 mm (1 to 5 mm), 252 coils were employed varying from 1 to 4 coils per patient. Closure was successful in 156 patients, with immediate occlusion in 83 and after fifteen minutes in 73. We reviewed the echocardiographic follow up of 49 patients, with immediate success in 68%, after three months in 94% and 97% in 6 months. The most common complication was retrievable coil migration to pulmonary artery (21 patients). We conclude that coil closure of PDA is an effective, safe and low cost therapy.

P935

Coil occlusion of patent ductus (PDA) without detachment mechanism is safe and effective: experience of 200 consecutive cases.

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The single center experience of 200 consecutive transcatheter occlusion of PDA using long Gianturco coils without detachment mechanism is reported with regard to safety and efficacy. Transcatheter coil occlusion was attempted in 201 and accomplished in 200 patients with a median age of 2.8 years (1 month – 56 years) and a median weight of 13.2 kg (3.2 – 91.9 kg). The smallest ductus diameter ranged from 0.1 to 7.8 mm with a mean of 1.3 mm. Coils were selected and positioned to fit into the distal ampulla. Coils were delivered without snare or forceps technique. Aortography was performed 10 minutes after placement. color flow echocardiography was performed 2–12 hours and six months after placement. Coils with 4–6 loops were implanted transarterially in 90% (1.3 [mean 1.2) coils were implanted. Median fluoroscopy time was 12 min (1–37 min). Inadvertent embolization to the pulmonary artery occurred in 1%. Transient pulse loss was observed in 2%. There was no pulmonary artery or aortic arch stenosis. 75% of patients were discharged the same day. Complete closure was achieved in 82% after 10 minutes, in 93% after 2–12 hours, and in 98% after 6 months. There were no late complications. Conclusion: Transcatheter occlusion of the PDA using long Gianturco coils: safe and effective without distal lumen mechanism.

P936

Percutaneous balloon valvuloplasty in neonates and infants with pulmonary valve stenosis

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We report our experience with pulmonary valvuloplasty in children less than one year of age and review both immediate and mid-term follow up results. From May 1994 to May 2000, forty consecutive patients less than one year of age (25 female) with pulmonary valve stenosis (PVS) underwent catheter balloon valvuloplasty, at median age of 5 months (2 days to 11 months) and median weight of 6.2 kg (2.3 to 9.7 kg). The clinical presentation in the neonatal group (10 cases) was poor feeding (n=2), heart murmur (n=2), heart failure (n=1) and cyanosis requiring Prostaglandin E1 infusion in 5 cases. In the infant group (30 cases), 15 patients were asymptomatic, 9 presented fatigue or dyspnea and 6 had cyanosis. Percutaneous femoral vein access was used in all patients, dilation with single balloon in 29 cases, double balloon in 2 and progressive dilation in 9 cases. Immediate success (gradient less than 30 mmHg) was observed in 98% of patients. The complications were

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

infundibular spasm in 6 patients, arrhythmias in 2, low cardiac output in 2, vein thrombosis in one and death due to cardiac perforation in one. At the end of median follow up of 3 years (6 months to 6 years), 4 patients had recoarctation with reintervention in 3 and medical management in one patient with Ebsteins anomaly. Thirty one patients were symptoms free and five lost follow up. Our results suggest that catheter balloon valvuloplasty is an appropriate procedure for management of PVS in neonates an infants

P937

Role of intraoperative echocardiography in pediatric catheter intervention

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Background: Clinical advantages of interventional treatment for pediatric heart disease is less invasive comparing to cardiac surgery, however, recent reports advantages of cardiac interventional treatment could not reach 'no complication procedure'. We evaluated the clinical efficacy of intraoperative echocardiography in various settings of pediatric cardiac interventions. Since 1997 through 1999, 154 interventional catheterizations were performed, of which 28 procedures underwent intraoperative echocardiography in combination with fluoroscopy. These cohort were consisted of 17 cases with ASD closure, 10 cases with PDA closure, 2 cases with pulmonary atresia and intact ventricular septum, and 5 cases with stent implantation of pulmonary vein. Results: In cases with ASD, all intraoperative echocardiography underwent transesophageal and had advantages for the information of multiple septal defect, Eustachian valve and septal limbs. In patients having multiple defects, echocardiographic information was valuable for catheter approach through the largest defect and selection of device size. In 11 cases with PDA, intravascular ultrasound imaging from the descending aorta or main pulmonary artery revealed the precise location of coil into the vascular lumen. In 2 cases with neonatal PDA, transthoracic echocardiography guided the appropriate position of coil implantation and protrusion of coils. In cases with pulmonary atresia, guidewire puncture of atretic valve was performed under the guidance of transthoracic echocardiography. The optimal balloon catheter was monitored during the procedure to be fixed at the center of pulmonary valve. In cases with stent implantation, transthoracic echocardiography revealed the optimal site for stent implantation and the relationship to other pulmonary veins. Conclusion: Although our limited experiences were not reached any statistical significance for the support of efficacy of intraoperative echocardiography, this convenient technique would contribute safer procedure and reduction of avoidable complications related to cardiac catheter.

P938

Detachable coils (COOK) for easy closure of Fontan-fenestrations. Experience over 4 years.

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Closure of Fontan fenestrations using septal occluders (Amplatzer, Cardio Seal) has become an accepted method. We used detachable coils for closure with good results. 12 patients (age 5 – 17 years) with modified Fontan circulation and atrial fenestration (3 – 4 mm) were catheterized 50 months (range 23 – 207 months) after surgery. Closure of fenestration was thought to be necessary because of arterial desaturation due to increasing R-L shunt. After complete catheter study the fenestration was passed with a 4 F Berman balloon catheter and after inflation of the balloon the fenestration occluded. After 20 minutes closure under continuous measurement of oxygen saturation and venous pressure. If the pressure remained unchanged and the oxygen saturation increased, closure was thought to be possible. According to the measured size of the fenestration detachable coils (COOK) of adequate size were placed and the position controlled by angiography before detachement. Oxygen saturation increased from 88% (median) to 93 % (median). In two patients additional communications were detected after closure and were occluded in the same way. In two other patients the venous pressure did increase to 20 mmHg during balloon occlusion. Coil closure was therefore not performed. All patients stayed on Heparin for two days and on Aspirin for at least 6 months. All coils remained in position and showed no excessive thrombosis. In two patients a residual shunt was seen on echo for 5 months. Oxygen saturation remained stable and 5 – 10 % higher than before closure. In conclusion we think closure of fontan fenestration is an effective, easy to perform and cost saving way of dealing with this problem.

P939

The effect of transcatheter closure of atrial septal defects upon the cardiopulmonary response to exercise

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Background. Past studies have found that surgical repair of atrial septal defects (ASD) may impair the chronotropic response to exercise and usually produces little, if any, improvement in exercise capacity. Purpose: To examine the effect of transcatheter ASD closure upon exercise function. Methods: Seven prepubertal 7–48 (median 22) yrs performed peak exercise tests before and 1–7 (median 1.5) mos following ASD closure using the Amplatzer Septal Occluder. Results: Prior to ASD closure, the pts' exercise function was only mildly depressed. A single APC was observed in one pt. Following ASD closure, no change was detected in any of the exercise parameters studied. Three isolated APC's were observed in one pt and 2 isolated PVC's were observed in another. No more complex ectopy was detected. Conclusion: Transcatheter ASD closure does not promote exercise-induced rhythm disturbances and does not adversely affect the chronotropic response to exercise. However, the exercise function of children and young adults with ASD is usually well preserved and, in short term follow-up, is not enhanced by transcatheter ASD closure.

P940

Improved Patent Foramen Ovale Closure Results with a Modified Hybrid Sideris Device

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Aim: The aim of this study was to compare two different types of Sideris device for PFO closure in patients with cryptogenic stroke. Methods: From January 1995 to May 2000, 81 pts with history of paradoxical embolic stroke underwent PFO closure. They were divided into two groups (grp) according to the device used: Grp S (Standard Sideris device: occluder/connector occluder, n=34) and grp H (Hybrid Sideris device: occluder/inverted counter occluder, n=47). Results: There was no significant difference between the groups in balloon stretched diameter (12.4 vs 10.8mm). Pts in grp H were older (52 vs 43 yrs, p=0.01). Complications, reinterventions, occlusion rates and residual shunt were compared. There were no recurrent strokes. 4pts in grp S and none in grp H (11.8% vs 0%, p=0.02) underwent reintervention (3 surgical and 1 additional percutaneous device placement). Overall cumulative rate of success (effective occlusion, no reintervention) at 331+/-12 months follow up was 88% for grp S and 100% for grp H (p=0.02). Shunt was defined echocardiographically as either full occlusion (no shunt), effective occlusion (no or trace/small residual shunt: <4mm), or large residual shunt (color jet width >1mm). Effective occlusion was achieved immediately in 97% of grp S pts vs 100% for grp H (immediate full occlusion was 26% in grp S vs 14% in grp H). At 1 year follow up full occlusion was achieved in 46% of grp S vs 70 in grp H, effective occlusion was 91% in grp S vs 100% in grp H at 1 year. Large residual shunt occured only in grp S pts (3% immediately and 9% at one year). Conclusion: Transcatheter closure of PFO in pts with cryptogenic stroke is achieved with high success rate using the Sideris device. The use of Hybrid Sideris device is superior to the Standard, yielding higher full occlusion, effective occlusion and cumulative success rates.

P941

Stenotilation of middle aortic syndrome complicated by aortic rupture

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Middle aortic syndrome is an entity with multifactorial etiology. Recently, it is considered best to treat this disease with percutaneous angioplasty and stenting. We hereby report a case treated this way, complicated by aortic rupture. A 16 year old girl was referred for hypertension and a diagnosis of middle aortic syndrome from an MRI-scan. She was first catheterized in June 2000, receiving two Palmaz stents in tandem. They were dilated with an 8mm balloon, without complication. The patient came back for redilation in October 2000. We used a 12mm balloon with up to 8 Bar of pressure. That resulted in good expansion of the stented area, except for a short eng like stenosis. Here the diameter was unchanged, being 6,5 mm. The result being less than optimal, we decided to continue with a 12mm high-pressure

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

balloon. This resulted in a uniform diameter of 10–11 mm and with only 2 mm Hg in residual gradient. No vascular tears could be detected on angiography. Two hours after the procedure, the patient was complaining of chest pain. A CT-angiography revealed an aortic rupture at the site of the angular stenosis. There was a massive hemorrhage around the aorta, spreading down retroperitoneally and up to the subpleural space. In the Pediatric department we did not have a coverage wall stent, so the patient was transferred to the Radiology Department of the nearby adult hospital. From the other groin we placed a 32mm/58mm Hemoban wallstent. Due to the hemodynamic instability of the patient, the stent was placed without a preceding angiography. However, control after placement showed no residual leakage. Follow-up has showed a good result with normal blood pressure and good dimensions of the descending aorta.

P942

Percutaneous Implantable and Retrievable Pulmonary Arterial Band: Early animal experience

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Main objective. Feasibility study of percutaneous implantable pulmonary arterial band (PAB). Methods: 8 animal subjects (2 dogs/mean weight 6.9 kg, 6 lambs/mean weight 4.75 kg; 4 terminal, 4 subsequent). Percutaneous implantation of pulmonary arterial band" made up of a self-expanding nitinol wire mesh with a moderate luminal tunnel. Preliminary sizing was estimated from human measurements. Angiography and hemodynamics following PAB were documented acutely and after chronic implantation. Survivors were maintained with usual care and were studied 2 to 6 weeks after implantation at cath and at post mortem study. Results: Total of 11 successful implants, 2 at main pulmonary artery (PA), 9 in branch PA. Device size ranged from 7.5–14.5mm. PA size ranged from 5.5 to 11.5mm. 6 implanted devices resulted in an ideal PAB" while 5 were considered non-ideal. All ideal PAB show angiographic patency of the PAB (Figure 1) with pressure gradients from 21–35(mean 28.25)mmHg. All Non-ideal PAB show total or near total occlusion of vessel due to inability of the oversized device to assume appropriate configuration. Device/vessel ratio in ideal PAB" was 1.55 whilst in non-ideal PAB was 1.8. Angiography showed patency up to 6 weeks. None of the devices embolized. In fact, they were resistant to movement unless retrieved into the sheath. Post-mortem study showed a thin layer of fibrin on the device with a patent lumina. Histology did not show evidence of distal thrombi. Successful retrieval by direct vision using snares were possible in 4 devices up to 6 weeks after implantation. Histology revealed damaged intima but intact muscularis and adventitia. Conclusions: Percutaneous delivered PAB is feasible. With appropriate device to vessel size ratio. The device was retrievable at late follow-up leaving an intact vessel lumen.

P943

A new intracardiac ultrasound probe – Monitoring of transcatheter closure of septal defects: First experience in animal models

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Pre-interventional monitoring of transcatheter closure of atrial septal defects (ASD) utilizes fluoroscopy and transesophageal echocardiography (TEE). TEE might either be uncomfortable for the conscious patient in the cath lab or impossible in the animal lab evaluating new devices due to the physiological anatomy of the animals. Our aim was to examine the feasibility of monitoring catheter-based interventions with a new intracardiac ultra-sound (ICUS). A 10 French steerable ICUS catheter with longitudinal array (5–10MHz, AcuNav™ Acuson), capable of 2D imaging, PW-, CW-, and color Doppler was tested in a porcine and ovine model of ASD (n=4 each, weight: 20 – 60 kg, age: 2 to 8 month) where the foramen ovale was dilated by angioplasty leading to a ASD of 4 to 12 mm in diameter. The ICUS catheter was introduced via 11F sheath from the right jugular vein into the right atrium. Several interventions/procedures (puncture of the interatrial septum, angioplasty balloon ovale, placement and retrieval of prototype closure devices) were monitored. One single ICUS probe was used for all experiments which lasted between 1.5 and 6.5 hours. The image quality was equivalent to standard TEE, no mechanical interference was seen during any of the procedures performed. The ICUS probe allows from now on echocardiographic monitoring of the evaluation of interventional procedures in animal models at the level of conventional TEE. It may of advantage in clinical situations where a TEE approach is not well tolerated in conscious patients in the cath lab avoiding general anesthesia.

P944

Coil occlusion of systemic venous collaterals in hypoplastic left heart syndrome.

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Aim: After stage II reconstructive surgery for hypoplastic left heart syndrome (HLHS) there may be flow via systemic venous collaterals to the aorta, causing desaturation. Following one case, where an apparently insignificant collateral identified at catheterization before stage II required surgical ligation afterwards, we aimed to survey the frequency of such collaterals, and determine whether coil occlusion prevents the need for surgical ligation. Patients and Methods: Cardiac catheterisation was performed on 25 children with classical HLHS between July 1998 and November 2000: 10 before stage II at a mean(sd) age of 7.1(0.5) months and weight of 6.8(1.3)kg, and 15 before stage III at 35.5(1.2) months and 13.1(0.5)kg. Aortic oxygen saturations (SaAo) and pulmonary artery pressure (pPA) were recorded. Angiography was performed into the left internal jugular vein to look for venous collaterals. If present they were occluded with Cook MReye coils. Angiography was repeated to confirm occlusion, and SaAo and pPA remeasured. Results: No further collaterals were found before stage II, and 9/10 have undergone hemi-Fontan without complication. Collaterals were identified in 5/15 cases before stage III, and were successfully occluded with 1–3 coils without complication. Mean SaAo before occlusion was 81.2(2.2)%, in those with collaterals, compared with 86.9(1.1)% in those without (p=0.02), but there was no difference in pPA between the two groups. After coil occlusion mean SaAo rose to 84.6(1.9)% (p=0.03) and mean pPA rose from 13.8(1.7) to 15.4(2.0)mmHg (p<0.001). 10/15 have since undergone the Fontan procedure without complication. Conclusion: Angiography should be performed at catheterization before stage II and III surgery for HLHS, to exclude systemic venous collaterals if present, they may be safely and effectively occluded with coils to improve saturations and prevent the need for subsequent surgical ligation.

P945

Treatment of the aortic coarctation with the palmaz stents in children and adults, follow up for seven years with cwin helicoidal tomography

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The traditional treatment for the Aortic Coarctation has been surgical and later on with balloons. But now we know that, in several occasions, no matter the treatment, several patients would recoarcate, specially in the cases with some hypoplastic degree. In 1993, we started working in the first patient with the Takayasus Arteritis. Successful results were obtained with this new patient, who is still asymptomatic and gradient of zero. Along the last seven years, we have been working in 67 patients with aortic coarctation and hypoplastic descending aorta. The patients age ranged from 1.6 to 27 years (med 6.7 years). The diameter of the aortic coarctation was from 2.2 mm to 9.7 mm (med 5.7), and the gradients were from 30 mmHg to 70 mmHg (med. 50 mmHg). In 29 patients, predilatations had to be made with a balloon with a diameter of 5mm to get across with the device. In 64 patients only one stent was used for each one, and in three patients we used two stents (Palmaz 7/6). Results: Sixty five of all the patients were dilated or reopened successfully and the results are as follows. Of the 29 patients, two had been predilated with a balloon, and they presented the hemorragia of an aneurism, although just the dilatation with the balloon had been successful, which made us think in the implantation of the stents. In the other 38 patients one implantation was like a primary implant. In 65 of the patients, the dilatation was successful and the gradients were 0. In two of the patients, one is still with a gradient of 10 mmHg, and the other one with 25 mmHg and they are asymptomatic. Fifty seven patients have been followed up with cwin helicoidal tomography and we know the aortic arc looks normal, the stents on place, doesn't look stenosis, and the aneurisms have disappeared. Conclusions: The treatment of the aortic coarctation with the stents is the most impressive result, and the follow up should be with another kind of angiographic catheterization, like helicoidal tomography.

P946

Transcatheter PDA closure with four different devices (Gianturco,Gianturco–Grifka, Sideris: Regular device and the new patch). four years of experience in 137 patients.

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https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Hospital Central Militar Mexico (3F)Transcatheter PDA Closure for several years has been the most common way to correct these defects. we present four different systems to close pdaly the most common is the Gianturco Coil, and we used 96 coils in 93 patients, in there we used double coils. The diameter of the PDA is was 1.3 mm. to 5.5 mm. The most common type of PDA was type E. In 42 patients, we used 42 Gianturco Grifka Devices, and the diameters were as follows: one patient with 2.5 mm PDA diameter, and in 41 the diameter was from 4.2 mm. to 8.0 mm. In two patients we used two regular Sideris Devices and the diameter was, in the first patient, 14 mm., and in the second patient the diameter was 5 mm. Our last patient was a 43 years old female, with a pulmonary pressure equal to the systemic pressure The diameter of the ductus was 21–22 mm. and we used the new Sideris Device, whose name is the Patch RESULTS In the first 96 patients, from one day to four years of the closure, 100% remained succesfully occluded. In three patients, at six months of the closure, a new coil had to be used, and the occlusion was succesful. In two patients the coils were lost, which were unable to be recovered, the defect was finally occluded with bigger coils The patients are still asympetomatic, and the ductus occluded. The results with the Gianturco-Grifka were as follows: in 40 patients the P.D.A. was succesfully occluded, and some red residual leaks The most common PDA type was E. In one of our patients, the device have embolized five minutes after the implant, and was retrieved through the femoral artery, later the ductus was occluded with two coils, while in the second, when the device embolized is still unknown, with possibilities of happening in the fourth day after the implant, the device was also retrieved through the femoral artery (7 mm device), with another succesful double coil implantation. In the last three patients, two regular Sideris devices were used, the first a PDA type B with 14 mm., was succesful at the first try, and after one year, the ductus remained occluded; the second patient after three months, remained with a trivial leak. The last patient was occluded with the new Sideris device (Patch), and the ductus was succesfully occluded, after five months the patient was still asymptomatic. In every patient, the follow up has been through transthoracic echocardiography CONCLUSIONS The P.D.A. occlussion with several different devices demonstrate that all of them are effective and safe, with one device for every kind of defect. For example, the ductus type B can be occluded with the regular Sideris Device, and in some special cases (larger ductus) with the Patch, taking into account each technical problem for every device in the implant. Great care must be taken while choosing

P947

Complete occlusion of extravascular vascular anomalies using Amplatzer(r) occluders.

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Occlusion of unwanted vascular anomalies may be necessary to achieve and maintain circulatory stability Surgical ligation is usually successful, but transcatheter approach is less traumatic with a shorter hospital stay A variety of devices are available but not all suitable for large anomalies Amplatzer(r) Septal and PDA Occluders (ASO,APO) are now effective atrial septal defect (ASD) and patent ductus arteriosus (PDA) occluders under investigation. This study evaluate the efficacy of these devices to occluding large unwanted extracardiac vascular anomalies Between March 1998 and July 1999, seven patients age 0.4 – 16 y, mean 7.8 years had their anomalies occluded by catheter under general anesthesia The anomalies are: axillary artery fistula (n=2), aortopulmonary collateral (n=1), left cardinal vein (n=2) modified Blalock-Taussig shunt (n=1) and hemizygos vein (n=1) The occlusion in two patients (cardinal and hemizygos veins) was performed as an emergency after bidirectional cavo-pulmonary anastomoses Two devices were used in one of the patients with axillary artery fistula, and one device each in the rest (total 8, 2 ASOs and 6 APOs). All anomalies were occluded immediately except one (cardinal vein) which was totally blocked 2 months later There were no procedural complications or blood transfusion All but two patients (emergency procedures) were discharged 24 hours after the device placement. Beside being effecive ASD and PDA occluders, the Amplatzer(r) Occluders are excellent for transcatheter blockage of large unwanted extracardiac vascular anomalies

P948

A biopsytome with variable flexibility for transfemoral endomyocardial biopsy in infants and toddlers.

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Monitoring progress after heart transplantation by endomyocardial biopsy is generally an accepted approach. The biopsytome may be introduced via the femoral vein or the jugular vein. In the transfemoral approach, a long shaped sheath is usually used to guide the biopsytome into the right ventricle to biopsy the ventricular septum. The biopsytomes currently available are relatively stiff making it difficult at times to negotiate the bend in long sheath from the right atrium to the right ventricle, particularly in infants and toddlers. The stiff biopsytome may straighten the shaped sheath causing it to pop out of the right ventricle. This may be avoided by using a biopsytome that can made flexible while negotiating the right atrial – right ventricular(RA-RV) bend. The objective of this study is to evaluate a biopsytome of stainless steel construction with variable flexibility (Sparrow Hawk, A.I.C. Technologies Inc., Woburn, MA). The oval jaw head made from preoperatory hardened steel permitted sharper biopsytome in a floppy shaft without compromising axial force The jaw cup (3.1 by 3 by 3 mm) gave a very acceptable tissue volume. The biopsytome was introduced via a 6 or 7 Fr sheath negotiating the RA-RV bend without difficulty. All procedures were well tolerated without perforation or other myocardial injury This biopsytome of variable flexibility should be the device of choice for endomyocardial biopsy in infants and toddlers using the transfemoral approach

P949

Single Center Intermediate-Term Outcome of Transcatheter Secondum Atrial Septal Defect Closure Using the CardioSealy®" Septal Occlusion Device.

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Background. The CardioSealy®" septal occlusion device has been used in clinical trials since 7/96 for secondum atrial septal defect (ASD) closure. Methods. Cleveland Clinic data from patients enrolled in the prospective trial of the CardioSealy®" device were analysed for incidence of residual leak, the need for further intervention and major complications. Chest-X-ray, transthoracic-echocardiography (TTE) and electrocardiography were performed at 1, 6, 12 and 24 months post deployment. Results. Sixteen patients (14F/2M), median age 16 years (5–>66) with a Qp/Qs of 1.6 ± 0.5 and ASD size of 30 mm ± 2.5, had deployment of 22–>30mm CardioSealy®" devices. Fluoroscopic balloon stretched ASD diameter was 13.5mm ± 2.5 with device/balloon stretched diameter ratio of 2.1 ± 0.3. Immediate TEE demonstrated no leak in 1/16, trivial/small leak in 11/16 and moderate leak in 4/16 of patients. All patients have been clinically reviewed more than 12 months post procedure with a median follow-up of 2 years (0.99–2.95). TTE demonstrated a persisting leak more than trivial/small in 3/16 at 1 month, 1/15 at 6 months, 1/13 at 12 months and 1/11 at 24 months. The right ventricular end-diastolic dimension decreased by a median of 18.5% by 1 year. Device arm fractures were found in 3 patients by 6 months post implantation with 1 additional late fracture. There have been no clinical complications. Conclusions: The CardioSealy®" device can be implanted safely in hemodynamically significant ASDs with a subsequent reduction in the size of the right ventricle. The presence of arm fractures was not associated with functional failure of the device. There is a trend for residual leaks to decrease over the first 6 months and only 1 patient has a leak large enough to consider further intervention in the medium term.

P950

Balloon dilatation (BD) of Neonatal severe / critical pulmonary stenosis (PS): Single Center Experience.

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BD of PS differs in the neonate from the older child. We performed a retrospective review of our patients' (pts.) data, who underwent BD as neonates (< 30 days old) because of severe / critical PS, (defined as right ventricle pressure (RVP) > systemic or requiring prostaglandin (PGE1). Methods: Between July 1997 and November 2000, 36 pts. underwent BD. Seventeen pts. fit the inclusion criteria. Three pts. were on PGE1, 4 on ventilator, 1 on supplemental oxygen. Age range was 2 – 14 days (median 3.5 + 3); weight: 3.4 + .54 Kg (range 2.9 – 3.7). RVP decreased from 100 to 50 mm Hg, the ratio of RVP / systemic decreased from 1.47 to .57. There were no mortality or major complications. Two pts. required PGE1 for an additional 2–3 weeks. They were discharged with oxygen saturation (sats) in the low 80s (> 95% 3 weeks after discharge). One pt. with severe tetralogy of Fallot and kinked shunt underwent BD with increase in the sats from low 70s on oxygen to high 80s

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

on room air. Follow-up (F/U) period ranged from 8–24 months (median 10 ± 10). One pt who had BD of ductal/pulmonary atresia (PA) had residual severe PS with 90 mmHg gradient. She underwent a second BD with reduction of the gradient to 60 mmHg immediately post procedure and to 25 mm Hg on latest F/U. 2 pts with the most severe stenosis have > 3+ pulmonary insufficiency (PI). No pt has yet required surgery. Doppler derived gradient in all pts is < 25 mmHg. Conclusion: Neonatal BD offers a successful approach to the management of severe / critical PS. The residual gradient will likely regress or later BD can be performed. The PI seen after balloon is most likely related to the nature of the valve itself and is usually well tolerated.

P951

Antegrade valvuloplasty of critical stenoses in infants using a low profile high pressure balloon catheter

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In 1994, a high pressure (10 atmosphere), low profile (5.5 Fr shaft) was developed to deliver stents and dilate stenoses in infants. Over the past 6 years, the catheter (NuMed Inc., Hopkinton, N.Y.) was used in critical aortic stenosis (AS, n=10, mean age 47.7 days, range 1 hr – 82 days) and pulmonary stenosis (PS, n=15, mean age 18.4 days, range 1 – 89 days). Transvenous antegrade approach was used in all patients. Transcatheter guidewire (GW) approach was used in all PS. Umbilical arterial-venous GW loop was used in 2 AS. 0.018 inch GW was used all patients. Inflating the balloon (7–10 mm diameter) 2–4 times reduced the peak systolic gradient in AS for 57±7.4 to 13±2.5 mmHg, and in PS from 58.8±4.9 to 11.8±2 mmHg. Apart from brief hypotension and transient arrhythmia with the first inflation, the procedure was well tolerated without peripheral vascular injury, myocardial damage in most babies. One AS need reballooning and another surgical repair of the mitral valve damaged by the catheter procedure. Three PS with hypoplastic right ventricles needed aortopulmonary shunts. One of the remaining 12 PS underwent successful reballooning for recurrent stenosis. This general purpose high pressure balloon catheter permits effective and safe antegrade dilation of critical stenoses in neonates and infants.

P952

Variations in atrial septum and PFO morphology: Effects on transcatheter PFO closure techniques and outcome

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Purpose. To evaluate the effects of atrial septum and PFO morphology on transcatheter PFO closure techniques and outcome. Methods: Retrospective analysis of angiographic (angio) and transesophageal echo (TEE) data was performed on 17 pts (median age 42 yrs; range 24–66) who underwent PFO device closure from 4/00–11/00. Angio and/or TEE imaging was used on all pts to assess septal and PFO morphology. PFO diameters (non-stretched and stretched), device placement and residual shunt. Compliant angioplasty balloons were used to obtain stretched PFO diameter. CardioSEAL septal occluder devices (NMT Medical, Inc., Boston, MA) were implanted in all pts. Results: Septal morphology (SM) was flat in 7/17 pts and aneurysmal in 10/17. PFO morphology (PFOM) was simple (flap) in 5/17 pts and complex in 12/17. Complex PFOM included tunnel-shaped (8/12) and fenestrated (4/12) with fenestrated PFOs occurring only in pts with aneurysmal SM. Non-stretched PFO diameters were significantly smaller than stretched (mean/Std deviation: 4.5mm/1.1 vs 10.9mm/3.9, p<0.01). Overall device size:stretched diameter ratio (DEV SD) was 3.7:1 (range 2.2–9.1). DEV SD ratio was significantly larger in pts with aneurysmal SM and complex PFOM compared to pts with flat SM and simple PFOM (mean 5.9:1 vs 2.6:1, p<0.05). Device placement was successful in all pts and there were no complications. Effective closure (trivial or no residual shunt) at time of implant was achieved in 16/17 (94%). Conclusion. Variations in atrial septum and PFO morphology are frequently encountered and should be carefully assessed prior to transcatheter PFO closure. Modifications of current closure techniques, such as use of compliant angioplasty balloons to determine stretched diameters and use of larger devices for pts with aneurysmal SM or complex PFOM may lead to more accurate device placement, improved effective closure rates and fewer complications.

P953

Transcatheter closure of secundum atrial septal defects in pediatric patients with Amplatzer device.

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We report our clinical experience with the newly developed Amplatzer device in transcatheter closure of ninety-eight atrial septal defects (ASD). The mean age of the patients was 8.1+/-6.2 years (range, 2.5–33 years). They were selected according to the location and size of the defect by transesophageal echocardiography (TEE). All procedures were performed under general anesthesia with fluoroscopic and TEE guidance, following a routine hemodynamic evaluation in the catheter laboratory. The optimal device size was selected after the balloon sizing of the ASD's. One patient had pulmonary valve stenosis with a pressure gradient of 60 mmHg between right ventricle and pulmonary artery. A successful balloon dilation was performed to this patient during ASD closure. The patients received 24-hour heparin infusion and were discharged at 24 hours, after an evaluation with X-ray, ECG, and echocardiography. They were on 3–5 mg/kg/day aspirin and infective endocarditis prophylaxis for 6 months after the procedure. Reassessment was done at first month and every 6 months thereafter with echocardiography and Holter monitoring. Mean ASD size was 12.1+/-3.9 mm at TEE, and 18.3+/-4.1 mm at balloon sizing. The mean size of the device was 18.9+/-4.3 mm. The procedure time and the fluoroscopy time were 47.6+/-19.6 and 12.4+/-5.3 minutes respectively. Immediately after the procedure 3 patients (3.7%) had small (color Doppler reaching 1–2 mm beyond the disc of the device), and 15 patients (44.1%) had trivial shunts (TS < 1mm). TS remained in only 5 of them during discharge, and no shunt was observed at second evaluation. None of the patients had major complications. Junctional rhythm developed in a patient, and another patient had frequent supraventricular extrasystoles without symptoms. Amplatzer is an effective and safe device for transcatheter closure of ASD in pediatric patients, with especially very low rates of residual shunt and complications.

P954

Effective transcatheter perforation and valvuloplasty of pulmonary atretic valve using a coaxial radiofrequency catheter: a multi centre experience

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Pulmonary atresia with intact interventricular septum and adequate right ventricle may be successfully treated by transcatheter means. The imperforate pulmonary valve may be perforated using approaches varying from a simple stiff wire to an elaborate laser system. Puncture using radiofrequency (RF) energy is gaining popularity. We report the multi-centre experience of the clinical application of a 7 Fr coaxial RF catheter we developed and tested initially in an experimental lamb model. Under general anesthesia, six neonates weighing 2.0 – 3.6 (mean 3.2 ± 0.3) kg with pulmonary atresia and intact interventricular septum underwent anterograde transvenous perforation of the imperforate pulmonary valve using the coaxial RF catheter via a 5 Fr right Judkin coronary or cobra catheter placed just below the aortic valve as a guide catheter, the RF catheter was introduced and advanced to the valve. Using 2 – 10 watts RF energy the valve was punctured to allow immediate placement of a 0.014 coronary angioplasty guide wire. Replacing the guide and RF catheter with a balloon angioplasty catheter, valvuloplasty was completed in the newly perforated valve. Apart from transient arrhythmias and brief hypotension, all procedures were successful without myocardial perforation or peripheral vascular injury. Prostaglandin E1 infusion and/or mainpulmonary patency was maintained after the procedure. To date, no infants required a second procedure by surgery or catheter. In one patient, oxygen saturation improved from 68% after the procedure to over 90% in the subsequent months without further intervention. We conclude that RF perforation to facilitate valvuloplasty of the imperforate valve in pulmonary atresia using the 7 Fr coaxial RF catheter is not only successful and effective, but safer by permitting balloon dilation of the pulmonary valve immediately after puncture.

P955

Ventricular septal defect closure using the gianturco-grifka vascular occlusion device

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https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

The Gianturco-Grifka Vascular Occlusion Device ("Grifka bag") has been used for occlusion of abnormal vessel supply in arteriovenous malformations and in patent ductus arteriosus too large for standard coil embolization. The device consists of a nylon sack within which filler wire coils are placed. The bag-like configuration allows for placement of the device into pouch-like structures, such as those seen in some ventricular septal defects. We report the use of this device on six patients with paramembranous ventricular septal defect with tricuspid pouch ("aneurysm of membranous septum"). One of six had a prior VSD patch repair with residual defect at the paramembranous area. There were 4 males and 2 females; median age = 5 yrs, and median weight = 20 kg. All had restrictive, hemodynamically small ventricular septal defects not contiguous with the aortic valve. Placement of the Grifka bag was successful in 5 of 6 patients. Complete immediate closure of shunt was obtained in 3 of 5 patients. Two of 5 had residual tiny ventricular septal defect at the rim of the bag. These two patients underwent supplemental placement of Gianturco coils to occlude the shunt, with partial success. There were no untoward complications. All patients were discharged one day after the procedure. Echocardiographic follow-up after 6 - 423 days (median = 172 days) demonstrated findings similar to the immediate post procedure echocardiogram with no migration of the device. Summary: The Gianturco-Grifka Vascular Occlusion Device is an alternative and safe method for closure of selected paramembranous ventricular septal defects.

P956

Percutaneous closure of atrial septal defect with the Amplatzer Septal Occluder in adults.

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This study reviews our experience with transcatheter closure of atrial septal defect (ASD) in adults using the Amplatz Septal Occluder. From June 1998 to October 2000, 53 patients (34 female, 19 male; mean age 44.2 years (19–71)) had closure of a ASD. Procedure was done under general anesthesia with fluoroscopic and transesophageal echocardiographic (TEE) guidance. Patients received antibiotics and heparin during the procedure followed by aspirin and endocarditis prophylaxis for 6 months. Clinical evaluation and transthoracic echocardiogram were done at 24 hours, 1, 3 and 12 months. ASD sizes on TEE ranged from 8–26mm (mean=17.3), stretched diameter was 12–36mm (mean=23,9) and size of the shunt was 1,2 5.1 (mean=1,9 1). Devices sizes ranged from 12–36mm (mean=23.7). The device was successfully deployed in 46/53 patients. The fluoroscopy time was 7–38min (mean=12,8) and procedure time 12–135min (mean=30.6). We failed in 7 cases (defect too large in 5, technically impossible in 2). There was no mortality but 2 patients had major complications: air embolism in a coronary artery and tamponnade 24 hours later. In another patient, thrombus was detected on the device but disappeared at 48 hours and the patient was put on coumadin. Intracardiac shunt was detectable in all patients immediately after the procedure. Two patients had residual shunt of more than 2mm at 24 hours that persisted at 1 month and 1 had small shunt at 24h with complete disappearance at 1 month. 22 patients have completed the 12 month follow-up and are clinically asymptomatic. Conclusion: Amplatz Septal Occluder can be easily and safely deployed in adults with excellent short and intermediate term results.

P957

Cardiac troponin t in detection of myocardial injury during routine cardiac catheterization procedures in pediatric patients

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This study aims to investigate whether intracardiac catheterization produces myocardial damage on pediatric heart. Five blood samples were collected (basal, immediate post procedure, at 4, 12 and 24 hours after the procedure) for troponin T and creatine kinase MB (CKMB) from 48 consecutive patients (age: 5.34+/-6.03 years). The patients are studied in groups of congenital heart defect (group A, with pulmonary hypertension, group B, cyanotic patients, group C, others), age, duration of procedure, medication taken (patients treated for congestive heart failure versus patients without this treatment), and number of contrast injection runs. The peak troponin levels (PTL) in groups A, B, and C were 0.32, 0.05, 0.02 ng/ml respectively ($p<0.06$). The clinically significant increase of troponin T (from 0.01 to 0.32 ng/ml) was only observed in group A. The mean peak CKMB level was 16.3 7.3, and 5.1 ng/ml in groups A, B, and C respectively with clinically significant increase of CKMB in all the groups ($p<0.05$). Patients with procedure time >30 minutes, and 4.1 year old patients had higher PTL than the ones with shorter procedure time, and older patients ($p=0.013$ and $p<0.001$ respectively). Mean PTL of patients receiving treatment for congestive heart failure was 40.3+/-40.39 ng/ml) higher than of patients without treatment (3.03+/-0.01 ng/ml; $p<0.001$). The number of injection sites was not correlated with PTL. Younger patients with more complex cardiac pathology, pulmonary hypertension, and especially compensated cardiac failure are under risk of myocardial damage during cardiac catheterization.

P958

Perforation of the pulmonary valve by radiofrequency followed by pulmonary balloon valvuloplasty in the treatment of patients with pulmonary atresia with intact ventricular septum

Silva, CMC; Abojamra, PA; Gomes, L.F.G., Mattos, R.B, Lima, V.C, Paula, A.A., Carvalho, A.C.C
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Perforation of the pulmonary valve by radiofrequency followed by pulmonary balloon valvuloplasty in the treatment of patients with pulmonary atresia with intact ventricular septum. Silva, CMC; Abojamra, PA; Gomes, L.F.G., Mattos, R.B, Lima, V.C, Paula, A.A., Carvalho, A.C.C. Universidade federal de São Paulo – EPM – São Paulo- Brazil. The treatment of patients (pts.) with pulmonary atresia (PA) and intact septum remains with surgical results not as good as for others types of congenital heart disease. Recently pulmonary valve (PV) perforation with radiofrequency followed by pulmonary balloon valvuloplasty (PBV) have been an alternative for these cases. The aim of this study was to analyze the outcome of our initial experience with this technique. Between May 1996 to July 1998, 7 patients (pts) with disease underwent pulmonary perforation with radiofrequency followed by PBV in our center. All cases were initially select by echocardiography, had a congenital RV parent infundibulum and absent RV to aorta connections with dependent coronary circulation. These echo findings were confirmed by RV angiography before procedure. All pts but one were female. Their age ranged from 3 days to 4 years, with a mean of 1yr1mo. Five pts previously underwent surgery (Blalock Taussig Shunt). Their weight ranged from 2.5 to 15 Kg, mean of 6.6kg. RV pressure was suprasystemic in all. The circular annules ranged from 9.8 to 19, mean of 13.4 mm, Z value ranged from -0.4 to +3.11. PV perforation was possible in all pts, but one (case6), although PBV was possible only in 5 (4.38%). The energy used ranged from 10 to 50watts. The procedural time ranged from 2hs40min to 6hs40min. As complication 1pt. had persistent bleeding at the site of puncture that led to death 36hs later, a pericardial effusion (1pt), resolved spontaneously (1pt) and a venous thrombosis (1pt). In conclusion the perforation of the PV by radiofrequency followed by PBV is a promising alternative in the surgical treatment in the most favorable cases of PA with intact ventricular septum.

P959

Intraoperative stenting of pulmonary artery stenosis

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Background: Transcatheter placement of balloon-expandable stents has proven useful in the treatment of pulmonary artery (PA) stenosis in children. We report our initial experience with intraoperative PA stent placement as an adjunct to surgical repair. Methods: Patients who underwent combined surgical reconstruction of the right ventricular outflow tract and intraoperative stenting of the pulmonary artery were identified via retrospective chart review. Intraoperatively, Palmaz stents (Cordis Corporation) were mounted on ZMED (B. Braun Medical, Inc.) or PowerFlex (Cordis Corporation) balloons and delivered under direct visualization, without use of fluoroscopy or guide wires. Results: Nine stents were implanted in six patients (median age 8.5 years [range 5.5- 13], weight 31.2 kg [16.7–67.2]). One patient required bilateral stents and two required multiple stents in the same vessel. Three patients had aortic compression of the PA not amenable to surgical repair and three had long-segment ostial obstruction or vessel kinking. All stents were prepared and deployed in less than 15 minutes. Using maximal balloon inflation size as the final diameter, the mean PA size increased from 8 mm to 14 mm ($p<0.001$). Two patients had the proximal stents trimmed after expansion. Patients with aortic compression of the PA had successful relief of the compression. Minor complications included three balloon ruptures (without sequelae), and one stent migration during inflation necessitating a second stent to treat residual stenosis. Conclusions: In patients who require both RV outflow reconstruction and relief of PA obstruction, a combined approach using intraoperative stenting is feasible, safe and effective. Benefits include decreased radiation exposure and shortened length of hospital stay when compared with standard transcatheter

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

techniques. When compared with surgical angioplasty, intraoperative stenting appears to offer the potential to reduce cardiopulmonary bypass time, although this requires substantiation.

P960

Aortic angioplasty for native aortic coarctation in adolescents and adults: an experience from a brazilian institution

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There is little data on the outcomes of balloon dilation for native CoA (NCoA) in adolescents and adults. In this study, we report our experience with this procedure. Between 01/86 and 08/2000, 22 patients underwent balloon angioplasty for discrete NCoA (20.2 (9.0 years, 17 Males). Seventy four percent had hypertension (BP (140/98 mmHg), 72% were symptomatic (NYHA I = II) and 31% were on medication. Success was defined as an improvement of at least 20% of the smaller diameter of the CoA area along with a reduction in the peak gradient (PG) < 20 mmHg across the obstruction. The balloon diameter was chosen according to the isthmus size (ratio of 0.92 (0.1). The PG decreased from 55.4 (17.8 mmHg to 6.4 (8.2 ($p < 0.01$) immediately after. One patient kept a PG > 20 mmHg. Although the mean diameter of the CoA site increased from 7.8 (3.4 to 14.5 (3.2 mm ($p < 0.01$) (mean percentage change of 110.0 (89.9%). 5/22 (23%) did not reach the 20% increase (in all the PG became < 20 mmHg). Nine percent (2 cases) developed small aneurysms which did not increase in size over time. There were no major complications. At a mean time of 3.7 (3.8 years, subsequent catheterization in seven patients showed maintenance of a reduced PG (11.7 (10.2 mmHg; p = NS compared to immediately after). Most of patients (59%) were not hypertensive (p=NS), 33% were symptomatic and none were on medication. Balloon dilation of discrete, symmetrical NCoA in adolescents and adults is a safe and efficacious procedure to reduce the PG across the obstruction. Although mid-term outcomes are good, residual hypertension is common.

P961

Stent implantation for native coarctation of aorta (NCoA). Initial experience at a brazilian institution

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Stent implantation for native CoA has been employed in adolescents and adults. In this study we report our initial experience with this procedure. From 03/98 to 07/00, 8 patients underwent stent implantation for discrete NCoA (26.7 (6.4 years, 5 female). All had hypertension (BP (140/90 mmHg) and were on medication, and 3 were symptomatic (NYHA I). One patient had undergone a PDA surgical closure. Nine stents (Palmaz-Schatz or CP-Numed) were used. The balloon diameter was chosen according to the isthmus size (ratio of 1.1 (0.1). In one patient (previous PDA surgery), the stent was not fully expanded due to a tight suture line (iatrogenic CoA) and was referred to surgery. One patient had 2 stents implanted due to initial stent slippage. The peak systolic gradient decreased from 46.6 (16.2 to 5.4 (5.6 mmHg ($p < 0.001$) immediately after. The mean diameter of the CoA site increased from 5.7 (1.3 to 17.1 (3.7 mm ($p < 0.001$) (mean percentage change of 213 (93%). There were no major complications or aneurysm formation. At a mean time of 13.4 (7.1 months, subsequent catheterization in 4 patients and spiral CT in 1 showed maintenance of the initial results with no recoarctation or aneurysm formation. Six patients were not hypertensive (p= 0.001), 1.3 were asymptomatic (p=0.07) and 4 were on no medication (p= 0.04). This initial experience reflects our initial learning curve with some technical problems. The procedure is safe and efficacious with good short-term outcomes. Immediate diameter gain at the CoA site seems to be greater and residual hypertension less frequent when compared to balloon dilation alone in this population. Larger numbers of patients and longer follow-up are required.

P962

Bio-degradation of coil material and recanalization of Major Aorto Pulmonary Collaterals after transcatheter embolization

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Aim of this retrospective investigation was to follow up children after coil embolization of aorto pulmonary collaterals with tungsten coils. Method and Materials: In 49 children (aged 3 weeks to 9 years) with congenital heart disease transcatheter coil embolization was performed in 99 aorto-pulmonary collaterals using 152 tungsten coils. Chest X-rays were obtained on day one, 3, 6, 12 months and then yearly after intervention. Re-catheterization was performed prior to planned staged correction surgery or in suspition of newer collateral development. Results: all MAPCAs were successfully closed. after 6 to 12 months, chest X-rays revealed a decrease of radiopacity and reduction of coil width in 39% of all collaterals. After a mean follow up of 25 months (17 to 51 months) there was a complete loss of visibility in 67.5% and a decrease of radiopacity in the remaining 32.5% of coils. Re-catheterization was performed in 41 collaterals. After a mean interval between implantation and re-catheterization of 28,4 months, recanalization occurred in 58.3% of all collaterals, independently of loss of visibility or decrease of radiopacity. Conclusion: Tungsten is bio-degradable. Best fitting exponential function predicts a complete loss of radiopacity of 99.5% of implanted tungsten coils within 10 years. In more than 50% of MAPCAs recanalization occurs after a mean of 28.4 months independently of the degree of metallographic visibility.

P963

Mitral septal annulus motion is reduced by percutaneous atrial septal defect closure with an Amplatzer(r)ty(tm) device

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The effect of atrial septal defect (ASD) closure by an Amplatzer(r)ty(tm) device on LV function was studied by conventional and Doppler tissue echocardiography. Thirty-eight patients (age 22.1 (20 years) were studied pre- and post-ASD closure (device size 23.5 = 6.3 mm). Results (Table): Although LV size and fractional shortening increased post-closure with a change in diastolic mitral inflow indices observed only in late diastole, the mitral septal annulus motion decreased in systolic, early and late diastole. Positive correlation was found between the percentage of the decrease in both systolic (r = 0.32, p < 0.05) and early diastole (r = 0.31, p < 0.05) mitral septal annulus motion and the ratio of the device size to BSA. Conclusion: The abnormal reduction in mitral septal annulus motion following ASD closure suggests that the Amplatzer(tm) device may have a negative effect on longitudinal LV systolic and diastolic function.

P964

Assessment of myocardial injury after pediatric cardiac catheterisation by troponin assays and other markers

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To assess for potential subclinical myocardial damage, all patients undergoing cardiac catheterisation at our institution since 2/2000 were prospectively examined regarding clinical, electrocardiographic (ECG), and biochemical markers before and after the investigation. 45 of 193 patients (23%) turned cardiac troponin T (cTnT) positive, 83% of them right after completion of the procedure. The risk decreased with age and increased with duration of the investigation. For neonates, it was 103 fold the risk for teens. It doubled every 30min. The highest cTnT values were measured after great arteries valve manipulation. Yet, like clinical course, ECG changes and other biochemical markers, the type of procedure had only minor discriminatory significance. In conclusion, young age and duration of the procedure appear to be the two most powerful predictors for frequent minor myocardial lesions following cardiac catheterization as detected by a cTnT rise. Follow-up studies are needed to assess immediate and prognostic relevance.

P965

Initial use of the intratherapeutics, Inc. IntraStent™ Double Strut™ biliary endoprosthesis in the treatment of congenital heart disease

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Stent design and structural limitations are obstacles for the pediatric interventional. The IntraStent™ (IS) Double Strut™ (DS) design makes it preferable to the Palmaz stent for use in children and adults with CHD. The IS-DS is composed of laser cut stainless steel, with a unique coil geometry designed for flexibility, radial strength, and avoiding stent shortening. The maximum expanded diameter is 10 mm, with the Large Diameter (LD) modification

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

expansion to 18 mm. Multiple stent lengths are available. From June-Nov 2000, we implanted 11 BTI-DS LD stents in 6 infants & children (11 mo-16 yr, 6.5–47.5 kg) with pulmonary artery, RV-PA homograft, or pulmonary venous obstruction. Expanded stent diameters were 4 mm to 18 mm introduced through 7–11 Fr sheaths. RESULTS: Pre/post _ mmHG LPA RPA RV-PA PV 21/2 17/3 25/7 18/3. Conclusions: 1) The BTI-DS & LD stents are effective in CHD. 2) The flexible design, minimal stent shortening and small introducer sheaths make this stent promising in infants and children.

P966

Availability & versatility of Amplatzer(r) occluder devices in treating children & adults with common & unusual CHD: importance in establishing a new interventional program.

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The Amplatzer Occluder Devices (AOD): ASO for ASDs, PFO for PFOs, and MVSDO for VSDs & other conditions are limited to clinical test sites in the US. After the 1st 7 mos, 110 cases were performed (65% interventional). Since AOD availability, 17/64 pts cathed (26.6%) were referred for AOD from 3 states & 2 countries. ASD closure (14 pts, age 2–62 yrs, wt 11–77 kg) using ASOs from 6 to 32 mm. 5 required 2 ASOs, 1 required transhepatic delivery, and 'custom' 36 mm device was used to close a 'fenestrated' ASD. Success rate at 24 hr was 100% in all 14 pts. Two pts had PFO closure after a neurologic event; 1 required 2 devices to close multiple fenestrated PFOs. Contrast echo was + with Valsalva in 9. Last, a 5mm MVSDO was used to close severe prosthetic paravalvular leak: BP 110/40, 77yr/y, to 110/62, 88 with mild AI. Conclusions: 1) AOD are versatile & allow interventionalists to treat common & unusual CHD that otherwise would have required surgery. 2) availability of AODs encourage referrals from a large geographic area, and 3) over 25% of all cases involve AODs.

P967

Coil embolization of aortopulmonary collateral vessels and surgical shunts in congenital heart disease

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To evaluate the result of nine children undergone transcatheter occlusion of aortopulmonary vessels or shunts, a retrospective review of clinical manifestation, radiologic findings, and cardiac catheterization data of nine consecutive pediatric patients were performed. Gianturco coil via transarterial placement were used to embolized all these cases. Eight of nine were successful embolization required in total occlusion immediately in six cases, only residual shunt in two. There is no coil dislocation or requirement for surgery immediately after the procedure in all cases. There is no intermediate or late complication. The results demonstrate that coil embolization is an effective and safe procedure for aortopulmonary collateral vessels and surgical shunts in children with complex congenital heart.

P968

Long term results of balloon pulmonary valvoplasty in patients less than 6 months of age.

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To evaluate acute, medium and long (up to 15 years) term results of balloon pulmonary valvotomy (BPV) performed in the neonatal and early infancy period. Patient records, catheterization data, cineangiograms and echocardiograms of 20 consecutive neonates undergoing attempted balloon dilatation were reviewed. Dilatation was accomplished in 18 of 20 attempted. Mean peak systolic gradients from right ventricle to pulmonary artery were as follows: before BPV, 62 mmHg (SD 23), immediately after, 11mmHg (SD 10)($p < 0.0001$), at one year follow up 27.4 mmHg ± 25 b($p<0.0001$) (n=15), at 5 years, 28 mmHg ± 16mmHg (n = 17), at 10 years, 16.8mmHg ± 7.8 mmHg (n=16), and at 15 years follow up, 15.5mmHg ± 5.5mmHg (n=12). We conclude that balloon pulmonary valvoplasty provides long term relief in infants and neonates with severe pulmonary stenosis and supports the maturation of the right ventricle.

P969

Closure of patent ductus arteriosus (pda) with the 'duct-occlud'-system: a single center experience

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Interventional closure of PDA has become accepted as the primary line of treatment. Several devices are currently used depending on PDA size and shape, but also on operator preference. We report our experience with the 'Duct-Occlud' device (PFM, Koeln, Germany) that we used during the last 5 years. During this period we attempted to close a small to medium sized PDA in 88 patients (pts), aged 0.4–40 (median 3.2) years and weighing 4.6–78.3 (median 14.0) kg. Mean Qp/Qs ratio (±SD) was 1.4±0.5 (range 1.0–4.2), and systolic pulmonary artery pressure was 22±6 mm Hg (range 14–55). PDA-diameter was 4.7±2.1 mm at the aortic and 1.4±0.8 mm at the pulmonary end. After assessment of hemodynamics and ductal size we inserted one (n=68) or more 'Duct-Occlud'-coils, another procedure was necessary in 20 pts. Procedure duration was 137±46 min. X-ray fluoroscopy time 21.4±14.2 min. The PDA was closed with 'Duct-Occlud'-coils in 69 pts (86%). 4 pts had secondary surgical PDA-closure, and 7 pts with residual shunts are waiting for late spontaneous closure or another procedure. Coil embolization occurred in 4 pts; coils could be retrieved in all pts and did not require further measures. Comparison of our results during early and late periods of experience with the device showed significant improvement with success and complication rates in the late period (table, *p<0.05). We conclude that the small to medium sized PDA can be closed effectively and safely with the 'Duct-Occlud' system. Operator experience with the device leads to higher success and lower complication rates.

P970

Buttoned device modifications: influence on feasibility, safety and effectiveness

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Since the initial human use of the buttoned device in 1989 to transcatheter occlude ostium secundum atrial septal defects (ASD), the device has undergone a number of modifications to improve its performance. These cohorts of buttoned device closures were reviewed to test the hypothesis that these modifications are useful in increasing effectiveness and reducing complications. Results of single button (1st, 2nd, and 3rd generation, 1988–1993, N=185, cohort-1), double-button (4th generation, 1993–1997, N=423, cohort-2) and centering-on-demand (Rotoglide 4th generation with centering mechanism, January 1998-July 2000, N=65, cohort-3) buttoned devices from the international and US/FDA trials were examined and the data are shown in Tables I and II. Whereas the cohort-3 (COD device) was similar to the other two cohorts in terms of ASD size, the implantation feasibility and effective occlusion rates improved and troubleshooting problem abolished. Although the re-intervention rates are low, the follow-up duration is too short to accurately evaluate this issue. The data presented confirm our hypothesis that device modifications improve device performance. Experience in a larger number of patients and evaluation of longer-term follow-up results are necessary to confirm the safety and efficacy observed in the initial small sized cohorts.)

P971

Multiple coil occlusion for pda using 0.052 inch gianturco coils

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Purpose: To evaluate the efficacy of multiple coil occlusion of PDA using 0.052 inch Gianturco coils (.052 coil, Cook). Subjects: Eight PDA patients whose age, body weight, minimum diameter, and Qp/Qs ranged from 6 to 212 (median 113) months, 5.9 to 67.5 (32.0) kg, 2.3 to 5.6 (4.0) mm, and 0.7 to 1.8 (1.5), respectively. The youngest patient had a PDA with a minimum diameter of 2.3 mm and a Qp/Qs of 0.7, complicated by a small ASD and systemic PH. Angiographically all ductuses were conical in shape with a good aortic ampulla. Methods: In cases 1–3, we usually deployed a .052 coil from the aortic side. Subsequently, we added a detachable PDA coil (Cook) with a loop diameter of 5mm (MWCE-3-PDA5 or MWCE-5-PDA5). In cases 4–8, two .052 coils were deployed simultaneously from both the aortic and pulmonary sides. The loop diameter/the length of coils was 6/8, 8/8, 8/10, or 10/15. To deploy the .052 coils, we used a JF bustome (Cook) to create a detachable mechanism as Gritka et al reported. Results: Coils were successfully deployed in every case without complications. Complete occlusion was achieved immediately or within a few days after deployment. Fluoroscopy time including diagnostic catheterization ranged from 21 to 45 (32) minutes. Conclusions: Multiple coil

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

occlusion using a 052 coil is a safe, effective, and economical method of closing moderate to large sized PDA with good aortic ampulla

P972
Recanalization of pda – case report.
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PDA is sensitive to prostaglandin, blood pressure, oxygen saturation and other factors only in first days of life. These factors stimulate closure of PDA. After complete closure PDA is unobstructive and recanalization is impossible. We present case of 7 years boys with Non-Hodkin lymphoma. During intensive chemotherapy, hemorrhage to respiratory tract and respiratory failure appears. He required machine ventilation during 9 days period. He, as all patients during chemotherapy was in program of monitoring of cardiotoxicity. Echocardiography was performed a few time before this incident by 4 independent physician and recorded on VHS. There were no changes in echo examinations. The typical signs of PDA were observed in control echocardiography performed after respiratorotherapy. We performed the repeat analysis of previous echo examination – there was not a sign of PDA. Cardiac catheterization confirmed diagnosis and coil closure of PDA was successfully performed. This is the first case of recanalization of naturally closed PDA. We can directly explain the mechanism of this recanalization.

P973
Amplatzer Duct Occluder vs. Gianturco Coil for Transcatheter Closure of the Patent Ductus Arteriosus
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The Amplatzer Duct Occluder (ADO) is designed for transcatheter closure of a broad range of patent ductus arteriosus (PDA) sizes, especially larger channels amenable to Gianturco coil closure. The indications for use of the ADO vs. coils are unclear. From 4/1/00–11/20/00, all pts undergoing PDA transcatheter closure have been considered for ADO implantation. ADO was used for all PDA>4 mm diameter; in PDA< 4 mm, device choice was operator dependent. Procedure time and outcome were compared between pts who underwent ADO vs. coil closure. Catheterization for PDA closure was performed in 42 pts, ADO was used in 28 pts, Gianturco coils in 14 pts and 2 pts were not closure candidates (both with severe pulmonary hypertension). In ADO pts, narrowest PDA diameter ranged from 1.7–10 mm (median 3.1). Smallest ADO diameter was a median of 2.6 mm > PDA diameter (range 0.4–4 mm). Complete closure was seen at 24 hrs by echo in 25/28 pts (96%), 1 pt had a trivial residual leak. There were no instances of pulmonary stenosis or coarctation. Six-month follow-up is available in 6 pts, none have a residual shunt. In coil closure pts, narrowest PDA diameter was a median of 1.5 mm (range 1.2–3.7). 1 coil was used in 6 pts, 2 coils in 5 pts, and 3 coils in 3 pts. Complete closure was seen in 13/14 pts (93%) at 24-hour follow-up. Procedure time was 50% shorter in pts who underwent ADO vs. coil closure p<0.01. In conclusion,

Coronary Artery Disease/Kawasaki Disease

P974
A national survey of pediatric cardiologists regarding clinical management of kawasaki disease in the US
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To compare the current practice of US Pediatric Cardiologists with the 1994 guidelines of the American Heart Association (94-AHA) for the management of Kawasaki disease, a multiple-choice survey was sent to three practicing in US fellowship programs. Opinions of 97/350 (28%) physicians practicing in 29/40 (73%) programs are summarized. Years of practice in pediatric cardiology (average ± SD) was 13.2 ± 10 for respondents vs. 13.6 ± 10.5 for non-respondents (p=0.69). In contrast to 94-AHA guidelines, 10% of respondents do not use high-dose ASA in the acute phase of KD and another 12% are aware of colleagues who do the same. Another 50% recommend clinical trials to evaluate ASA dose. Harada's criteria (validated in Japan) for patient selection to administer IVIG are followed by 3%; another 18% feel that similar criteria need to be established. In contrast to 94-AHA guidelines, 70% advise follow-up for risk-level I patients. For risk-level II patients, only 20% follow the 94-AHA no-follow up option. For risk-level IV patients, 90% prefer periodic stress-echo or stress-perfusion scan vs 20% who prefer periodic rest echo or stress ECG (94-AHA does not provide clear recommendation). For asymptomatic patients without coronary involvement and who do not have risk factors for coronary artery disease, 24% do not consent patients for healthy lifestyle habits. For persistent coronary aneurysms, 36% perform cardiac catheterization periodically and 61% do so only if coronary symptoms, stress echo, or stress myocardial perfusion abnormalities are present. Conclusion: The responding opinion was in concert with data from the most recent literature. The current trend in Kawasaki disease management in US teaching institutions suggests a need for an update in the 94-AHA guidelines.

P975
Brain natriuretic peptide – a useful biochemical marker of myocarditis in patients with kawasaki disease.
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Myocarditis of Kawasaki disease (KD) can be diagnosed by positive cardiac uptake of gallium 67. However, sufficiently specific and sensitive biochemical markers of it have not been reported. To determine whether brain natriuretic peptide (BNP) can be useful, we investigated BNP concentration, ECG and 2DE findings. Sixty-nine cases of KD (aged 2 months to 8 years) were studied. The blood samples, ECG and 2DE records were obtained before the treatments were started (on the 4.0 th day of the disease, mean) and in the convalescent phase (12.6th day). The plasma BNP concentration was measured by immunoradiometric assay, and 73.2 (range 0.6 to 641) pg/ml in the acute phase. We checked for the ECGs showing decreased QRS complex voltage, ST segment elevation or depression, abnormal Q wave, and T wave flattening or inversion, which helped to represent the myocarditis of KD. The group whose BNP was over 50 pg/ml in the acute phase showed abnormal ECGs more frequently than the group BNP less than 50 (21/29 vs 5/40, P<0.0001, odds ratio 22.1). T wave amplitudes in standard limb leads were measured in both phases, and the differences (convalescent – acute) were calculated. We regarded the sum total of these differences as the quantity of flattening T wave", then we examined the correlation between the BNP level and the "total supposed T wave voltage". The correlation was significant (r=0.960, P<0.0001, n=69). There was no correlation between BNP and LVEF. WE conclude that plasma BNP is a useful biochemical marker of myocarditis of KD. When the titer is over 50 pg/mL, the patient probably have abnormal ECGs and most likely to have myocarditis."

P976
Value of qualitative determination of cardiac troponin t in differential diagnosis of acute chest pain in emergency room
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Objective: To probe a quick, sensitive and specific method to differentiate the high/low risk of patient with acute chest pain(CP) in emergency room (ER). Methods: Seventy patients with acute CP were included. History and electrocardiograms were obtained, and CK was determined quantitatively. The qualitative determination of cardiac troponin T (TnT) was done, and repeated during the follow-up. The end-point of follow-up was acute myocardial infarction (AMI), cardiac sudden death, or other diseases definitely diagnosed. Results: (1) TnT was positive in 34 patients, the positive rate was 48.6% (2) In patients with AMI or unstable angina pectoris (UAP), the positive rate of TnT was higher than that of CK, at the first examination. (3) In UAP patients with positive TnT, the quantity of nitroglycerin infused intravenously per minute higher, the time of being observed and followed up in ER longer, and the rate of cardiac events higher than the UAP patients with negative TnT results. (4) In patients with non-coronary artery disease, TnT was positive in 2 patients, and they all had other evidence of myocardial damage. Conclusion: The qualitative determination of TnT could be used as a quick and effective method to differentiate the high/low risk if patients with CP in ER.

P977
Doppler characteristics of transmitral diastolic flow in kawasaki disease – a predictor of coronary artery involvement

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

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Kawasaki disease is a febrile illness notable for the development of coronary vasculitis. Identification of the patient at high risk for coronary artery aneurysm has been assumed to be important. To predict coronary artery involvement, we examined diastolic cardiac performance in 51 patients that may reveal the evidence of myocardial injury. All patients were less than 2 years old and had no evidence of any other cardiovascular disease. Echocardiographic examinations were performed in acute (within 10 days) and subacute stage (beyond 21 days of illness). From the Doppler tracings of transmitral flow, peak velocities during early rapid ventricular filling (peak E) and atrial contraction (peak A) were measured. Each area under the E wave (E area) and A wave (A area) were measured as velocity time integral. The ratio of the peak E to peak A (peak E/A ratio) and the ratio of E area to A area (E/A area ratio) were calculated. Acceleration and deceleration time of the E wave were measured respectively. From the simultaneous tracings of LV outflow, isovolumic relaxation time was also measured. The patients were divided into two groups with coronary artery involvement (n=22) and without coronary artery involvement (n=29) based on 2-D echocardiographic findings. Doppler characteristics of acute stage in each group were analyzed by multiple logistic regression test. Analysis determined peak E/A ratio as the most significant predictor of coronary artery involvement (p < 0.05). The risk of coronary artery involvement increases as the peak E/A ratio decreases (odds ratio 0.971; 95% confidence interval 0.943 - 0.997). In conclusion, Doppler pattern of transmitral diastolic flow in acute stage could be one of substitute means to predict coronary artery involvement in Kawasaki disease.

P978

Myocardial ischemia following surgical repair of anomalous left coronary artery from pulmonary artery (ALCAPA)

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Excellent long-term outcome has been reported for children who are subjected to surgical repair of ALCAPA. Late postoperative myocardial ischemia has been scarcely reported. The situation described hereby is rare, only one detected in a large series of 83 patients treated in our institution. A ten year old girl was admitted for investigation of a presyncopal episode. Surgical repair of ALCAPA had been performed by the age of 10 months. At that time, she had signs of low cardiac failure, LV shortening fraction of 20% and LV ejection fraction at 39%. Four years later, she had a complete recovery of cardiac function, with LV shortening fraction of 32% and ejection fraction of 62% in the absence of any perfusion defects on thallium-201 myocardial scintigraphy. Current data corresponding to 10 years of follow-up include alterations of repolarization on ECG, important heart rate-dependent ST segment depression on Holter analysis and a persistent perfusion defect at the anterior and lateral LV walls. Cardiac catheterization showed patent coronary arteries but diffusely hypokinetic LV, suggesting inappropriate development of the coronary microcirculation. The patient is now under medical treatment with beta-blockers, diltiazem and nitrates besides being included in a supervised cardiac rehabilitation program. On the basis of these findings we would like to suggest that these patients should be carefully monitored over time for early detection of unsuspected myocardial ischemia.

P979

Dipyridamole stress ultrasonic myocardial tissue characterization in patients with

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The purpose of this study was to assess the feasibility of using dipyridamole stress integrated backscatter for evaluation of myocardial ischemia or damage in patients with Kawasaki disease. Dipyridamole stress integrated backscatter was used in 31 patients with coronary artery lesions due to Kawasaki disease, in comparison to Tl-201 myocardial imaging. All patients underwent echocardiography at rest and after dipyridamole stress at three left ventricular wall segments in the short axis view, anterior interventricular septum (AS), posterior wall (PW), and inferior wall (INF). At rest, there was no significant difference of integrated backscatter in the regions with normal or abnormal distribution on Tl- 201 imaging. After dipyridamole stress, in contrast, the cyclic variation of integrated backscatter in the regions with abnormal distribution became significantly smaller than that in the regions with normal distribution in each segment: 3.6 ± 1.2 vs 6.2 ± 1.7 dB in AS, 3.3 ± 1.1 vs 8.0 ± 1.9 dB in PW, and 4.0 ± 1.4 vs 7.3 ± 1.6 dB in INF (p< 0.001). One hour after stress the cyclic variation recovered to the level at rest in all patients. When values below 5.0dB during stress were defined as abnormal, the sensitivity of abnormal cyclic variation in integrated backscatter was 75 % in PW, 91 % in INF. Specificity was 91 % in PW, and 90 % in INF, in comparison to Tl-201 imaging. Dipyridamole stress integrated backscatter can provide sensitive discrimination of regions of myocardial ischemia or damage in patients late after onset.

P980

Two unusual cases of coronary artery aneurysms not due to typical typical Kawasaki disease

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We report 2 cases of giant coronary artery aneurysms originating from unusual etiologies. Case 1 was a 2.5-year-old boy who suffered a myocardial infarction with right and left coronary artery aneurysms noticed after he was brought to the hospital unconscious. This patient has never been diagnosed as having Kawasaki disease (KD). However, at 1.5 years of age, he had a fever for 3 days accompanied by exanthema and hyperemia of the bulbar conjunctiva. The patient's coronary lesions were similar to those of Kawasaki disease in morphology and location. Case 2 was an 11-year-old girl who had giant coronary artery aneurysm with multiple systemic artery aneurysms. She had an enlargement in the temporal region and was diagnosed as having multiple cerebral aneurysms seen by angiograms at 4 year of age. The aneurysm of her superficial temporal artery was removed at that time. At 9 year of age, she underwent echocardiography because of chest oppression and was diagnosed as having a giant right coronary aneurysm. Later coronary angiography revealed that the circumflex stenosis appeared to be obstructed by a thrombus. The pathologist found congenital defect of media hemangiopoietiferative organization in the aneurysmal vessel wall. The first case is considered to suffer from atypical KD diagnosed from only 2 major symptoms of KD. The second case is the first case diagnosed as having a congenital coronary artery aneurysm from the pathological examination.

P981

Unusual congenital coronary anomalies. Diagnostic and therapeutic implications

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We present our experience with 6 cases of unusual coronary anomalies and their implication in the medical and clinical management. This is a retrospective study based upon a six-year experience. There were 3 unusually high originating ALCAPAs, 1 ALCAPA, a functionally from LV originating left coronary artery and an inter-aortico-pulmonally running right coronary artery originating from the undersurface of the aortic arch. The study supports the importance of the careful echocardiographic assessment of the coronary artery origins in suspected congenital heart disease, and the need for further diagnostic studies in case of doubtful echocardiographic findings.

P982

Treatment of kawasaki disease with moderate dose (1 gm/kg) of intravenous immunoglobulin

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To determine initial treatment failures and incidence of coronary aneurysm in Kawasaki disease (KD) treated with moderate dose (1 gm/kg) of intravenous immunoglobulin (IVIG). Retrospective review of all patients with a diagnosis of KD who had initial treatment with 1 gm/kg of IVIG at a tertiary care hospital (1994-1998). Thirty-one (76%) of 41 patients completely responded to a single treatment with moderate dose (1gm/kg) of IVIG (group A). Retreatment with a second dose of 1 gm/kg of IVIG was required in 7 patients (17%) who had persistent fever more than 48 hours after the initial treatment (group B). Three patients (7%) required 3 doses of 1 gm/kg of IVIG due to persistent fever after the second dose (group C). Aneurysms of coronary artery were detected in the subacute phase 19%, 29% and 100% in group

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

A, group B, and group C respectively. After 1-year follow-up, the incidence of coronary aneurysm had been reduced to 3%, 0% and 57% in respective groups. Only 1 patient in group C developed a giant aneurysm of the right coronary artery. We conclude that the long term beneficial effect of the moderate dose (1gm/kg) is comparable to the high dose (2gm/kg) regimen of IVIG in most of the patients. This moderate dose regimen may be more practical in the countries where the cost of IVIG might be greatly concerned. The limitation of this regimen is the high incidence of coronary aneurysms in few patients in the group who need more than 2 doses of IVIG. Further study is though needed to identify factors at the time of initial treatment which could predict the patient in this high risk group.

P983

Plasma adrenomedullin levels in kawasaki disease .

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Adrenomedullin (AM) is a potent vasodilating and natriuretic peptide originally isolated from human pheochromocytoma. Kawasaki disease (KD) is an acute febrile illness in young children, characterized by systemic vasculitis preferentially affecting coronary arteries. We hypothesized that plasma AM levels are increased reflecting coronary artery vasculitis in KD. To elucidate this hypothesis, we measured plasma AM levels by radioimmunoassay in six patients with Kawasaki disease (5 male, 1 female, 0.4–2.6 years, 1.2+-0.8 years) at before and 3 days after high dose intravenous immune globulin therapy and at recovery phase (2 weeks later). In all patients, white blood cell count (WBC) and serum C-reactive protein (CRP) levels increased before treatment (WBC 16500+-4509/ ul, CRP 11.1+-4.1). Compared with normal subjects (5.5+-0.5 fmol/ml), plasma AM levels were markedly elevated before treatment. Highest levels of each patients were ranged 58.2 to 141.9 fmol/ml (50.5+-35.4 fmol/ml). Specifically, plasma AM levels were remarkably higher in 2 patients who had been detected the coronary artery dilatation by echocardiography (125.6 and 141.9 fmol/ml, each). We believe that the rise in plasma AM in KD is due to the cytokine induced increase of AM expression in vasculature. Marked elevation of plasma AM at acute phase of KD may help to diagnose the coronary artery inva

P984

Neutrophils and mononuclear cells express vascular endothelial growth factor in acute kawasaki disease: its possible role in progression of coronary artery lesion

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Kawasaki disease (KD) is a syndrome of systemic vasculitis of an unknown etiology that is complicated by coronary artery lesions (CAL), leading occasionally to cardiac ischemic sequelae. To examine whether vascular endothelial growth factor (VEGF) is responsible for CAL in KD, we determined serum VEGF levels by ELISA and peripheral blood mononuclear cell (PBMC) and neutrophil VEGF expression by immunoblot analysis. Significantly increased levels of VEGF were demonstrated in acute KD, as well as in other vasculitis syndromes ($p < 0.0001$). In the 10 KD patients with CAL, serum VEGF levels were maximal approximately 2 weeks post onset, when CAL generally develops, and were significantly higher than in 20 patients without CAL (mean 474 and 241 pg/ml, respectively, $p = 0.0015$). During the same period, immunoblot analysis revealed maximal VEGF expression in PBMC, corresponding to serum VEGF levels in most patients and being particularly marked in patients with CAL ($p < 0.01$). Neutrophils expressed VEGF only in the early stage of acute KD, and declined rapidly in the majority of KD patients regardless of the presence of CAL, showing a strikingly different expression pattern than that for PBMC. Predominant VEGF expression by PBMC was also demonstrated in patients with other vasculitis syndromes, and only faintly in normal controls. The results suggest that VEGF is generated dynamically in KD, presumably reflecting its disease activity. Neutrophil-derived VEGF may play a role in regulating early vascular responses, whereas PBMC-derived VEGF may contribute to later vascular injury.

P985

Coronary artery fistula detecting after corrective surgery for ventricular septal defect and pulmonary stenosis

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A five-year old patient with a coronary artery fistula between the left anterior descending coronary artery (LAD) and the right ventricular (RV) cavity is presented. He underwent primary repair for ventricular septal defect (VSD) and pulmonary stenosis (PS). Postoperative routine color and continuous wave Doppler echocardiography showed a sistolodiastolic flow signal indicating drainage into the right ventricle from the left anterior descending coronary artery. There was no residual ventricular septal defect and pulmonary stenosis. Postoperative cardiac catheterization showed step-up of oxygen saturation at RV resulting pulmonary to systemic flow ratio was 1.8:1. Selective coronary arteriography revealed a fistula between the left anterior descending coronary artery and the right ventricular outflow tract. In our considerations the coronary artery fistula is not an acquired lesion as a result of interventional operation. The fistula was congenital and it was appeared after decompression of right ventricular pressure by operation. Discovery of coronary artery fistula in our patient showed that the importance of postoperative routine detailed echocardiography.

P986

Kawasaki disease with coronary artery aneurysms 5 years experience

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Between Nov 1995 and Nov 2000 KD was diagnosed in 15 pts (age 3mo-8y, boys-11, girls-4). In 12 pts CAA were seen in 2-small, in 4-medium, in 5 giant with thrombi in 3 of them. In 12 pts IVGG infusion was started 10 days after onset and in 3 before with. IVGG was given again in dose 100mg/kg/day in 5pts and 50mg/kg/day in 10. IVGG infusion was repeated in 6 -total dose of IVGG 2.5 - 5 g/kg. In 3 pts with giant CAA thrombi were found in aneurysms. In all of them tissue-type plasminogen activator (t-PA) was used with good result. One pt with giant CAA without thrombi died during acute stage. During the follow up period CAA regressed in 3 with small, in 2 with medium. Became smaller in 2 with giant and in 1 with medium. Conclusions: 1. The thrombolytic therapy with t-PA in pts with thrombi in CAA is efficient and safe 2. Echo successfully demonstrate thrombi and their disappearance during thrombolytic therapy. 3. The small and medium CAA showed a tendency to regressed to normal diameter and giant became smaller.

P987

Percutaneous transluminal coronary angioplasty for early developed coronary arterial stenosis due to kawasaki disease in infant.

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It has been reported that percutaneous transluminal coronary angioplasty (PTCA) had no indication for localized stenosis(LS) after many years from the onset of Kawasaki disease (KD). However, LS due to KD may develop within 1 or 2 years after the onset. We report the efficacy of PTCA for early developed LS in 2 infants with ischemic sign. One patient was 2 year 2 month-old boy, weighed 11.5 kg. Selective coronary angiogram (CAG) also 1 year and 10 months revealed 89% LS at the left anterior descending branch (LAD). Under general anesthesia, he underwent PTCA through 5F guiding catheter. The balloon size was 2.0 mm. LS improved from 89% to 0%. The other patient was 2 year 7 month-old girl, weighed 12.4 kg. CAG after 2 year and 3 months revealed 80% LS at LAD. Under general anesthesia, she underwent PTCA through 6F guiding catheter. The balloon size was 2.5 mm. LS improved from 80% to 21%. Intravascular ultrasound before and after PTCA was performed. Pre and post total cross sectional arterial area excluding vessel wall were 14.8 mm2 and 34.5 mm2, respectively. Lumen area was increased from 1.06 to 2.26 mm2. Intima-media thickness was decreased 1.88 to 1.09 mm. Compression on vascular wall by the balloon brought to enlargement of the lumen area. It is suspected that tissue of intimal thickening is soft within 2 years. In 2 patients, no complication was occurred and ischemic sign was disappeared. Re-stenosis was not detected in the follow-up CAG after 6 months. PTCA for early developed LS is effective, although the indication is limited.

P988

What is the feasibility of imaging coronary arteries during routine echocardiograms in children?

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Background: In the last 2 years we have examined 7 patients in whom echocardiogram identified life-threatening abnormalities of the coronary arteries (CA). No reference exist regarding the subset of children in whom CA can be imaged. The purpose of this study was to prospectively evaluate our ability to image origins and branches of CA. Methods: 100 children who did not have significant heart disease were studied. Mean age was 3.8 +/- 6.4 years. Examinations were performed by registered pediatric cardiac sonographers utilizing size-appropriate transducers on HP 5500 systems. CA origins were called with clock-face reference in standard views. Results: Right CA was imaged in 96%, with origin most commonly seen at 10-30 o'clock (range 9-1 o'clock). Color flow was demonstrated in 48% of R.C.A. Left CA was imaged in 98%, with origin most commonly seen at 1-30 o'clock (range 2:30-5:00). Bifurcation was seen in 78%. Color flow was imaged in 50% of L.C.A. Four abnormal CA were identified (Right CA from left coronary sinus n=2, circumflex from right CA n=1, small left CA to pulmonary artery fistula n=1). Five additional normal variants were identified (Ramus intermedius n=3, posterior descending from ramus n=1, sinus node branch from left CA n=1). Conclusions: Detailed CA anatomy can be assessed by echo in most children. Incorporation of CA imaging into routine echo exams is feasible. Since abnormal CA have been shown to be associated with sudden death, routine screening appears feasible with potentially life saving results.

P989

Myocardial tomoscintigraphy after arterial switch operation: comparison with coronary artery angiography

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Coronary artery obstruction is a major complication of arterial switch operations. Such lesions are detected by selective coronary artery angiography. Decision making in asymptomatic patients with coronary artery obstruction remains difficult. Our aim was to assess myocardial perfusion after arterial switch operation with myocardial tomoscintigraphy. Sixty-one Tl-201 myocardial tomoscintigraphy were performed in 45 pts after arterial switch operation (6±3 yrs). Selective coronary artery angiography performed in all pts was considered abnormal in case of >50% stenosis. Myocardial tomoscintigraphy was abnormal if there were one or more perfusion defects on stress images. Specificity and positive predictive value of myocardial tomoscintigraphy to predict coronary artery lesions were 79 and 74% whereas sensibility and negative predictive value were 69 and 73%. Nine pts with coronary artery lesions had normal myocardial tomoscintigraphy at the first evaluation in 4 of them repeated tomoscintigraphy showed perfusion defects. Twelve pts with coronary artery lesions and perfusion defects underwent surgical revascularization (6 angioplasties and 6 bypass grafts), myocardial tomoscintigraphy performed 6 months after surgery was normal in all of them. Sequential myocardial tomoscintigraphy are warranted in patients with coronary artery lesions. Prognosis of pts with perfusion defect and normal coronary artery angiography needs to be defined.

P990

Long-term outcome of the giant aneurysms in Kawasaki disease: comparison among the therapeutic regimens

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To assess the prophylactic effect of the therapeutic regimens to occlusion of the giant aneurysm, 102 ones in 69 patients with antiplatelet drugs (aspirin and dipiridamole, [a]) and anticoagulant (warfarin, [w]) through the three eras mainly using steroids (S), aspirin (A) and gamma-globulin (G) respectively, were evaluated by the serial CAGs (1.57, 3.49, 5.82 and 6.52 years as average after the illness for the first to the fourth CAGs, re- spectively). In results, [aw] as the prophylaxis did not any occlusion, regardless of the therapy in the acute stage. Regimen (S+[wa]) significantly inhibited the occlusion in comparison with Regimens (S+[a]) and S without prophylaxis) in log-rank test ($p<0.05$ and $p<0.01$, respectively). Regimen (S+[a]) significantly showed higher incidence of the occlusion than Regimens (G+[wa] and G+[a]) ($p<0.001$ and $p<0.025$, respectively). Regimen (no special therapy except for antibiotics administration +[wa]) significantly inhibited the occlusion in comparison with Regimen (S without prophylaxis) ($p<0.05$). Among the same therapy in the acute stage, [aw] as the prophylaxis had a tendency to inhibit the occlusion in comparison with [a], except for aspirin therapy in the acute stage in which [a] had almost same effect as [wa]. In conclusions, steroids as the therapy in the acute stage showed higher incidence for the occlusion of giant aneurysm. Combination of antiplatelet drugs and anticoagulant was the most effective prophylactic therapy to occlusion.

P991

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P992

Efficacy of end-diastolic images using quantitative gated single-photon emission computed tomography (QGS) in detecting areas of myocardial ischemia in children with Kawasaki disease

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The present study compares the clinical efficacy of end-diastolic images using either technetium 99m-sestamibi or tetrofosmin quantitative gated single-photon emission computed tomography (QGS) in detecting areas of myocardial ischemia in children with Kawasaki disease. QGS images were obtained for 9 patients with Kawasaki disease (4 males and 5 females, mean age and range: 16 years and 14 (9-18 years) with patients engaged in ergometer exercise and at rest. Left ventriculography and selective coronary angiography were also performed for all 9 patients. The two different QGS images were expressed as polar maps (Bull's eye maps), 1) gated end-diastolic images 2) non gated stress images. Defect areas were defined as a percent uptake (uptake ratio of myocardium uptake) of less than 70%. Of all 9 patients, 7 segments showed coronary stenosis and 21 segments showed a coronary aneurysm on selective coronary angiography. In the gated end-diastolic images, the values for sensitivity and specificity of detecting myocardial areas with coronary stenosis were 85.7% (6/7) and 28.6% (4/14), respectively. On the other hand, in the non-gated stress images, these values were 57.1% (4/7) and 57.1% (4/7), respectively. Thus, the sensitivity of detecting myocardial areas with coronary artery stenosis was greater for the gated end-diastolic images than the non-gated stress images in our group of children with Kawasaki disease. In conclusion, gated end-diastolic QGS images closely reflect myocardial blood perfusion in the diastolic phase during cardiac performance. Such images are useful for detecting areas of myocardial ischemia with coronary artery stenosis in children with Kawasaki disease.

P993

Elevated BNP level in patients in acute phase of atypical Kawasaki disease.

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Background: Intra-venous injection of gamma globulin remarkably reduces coronary artery lesions (CAL) of Kawasaki disease (KD). In atypical KD,

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

however, there are still difficulties in preventing CAL because of the delay of the diagnosis and the consequent delay of the therapy. There has been reported that atrial natriuretic peptide (ANP) and brain natriuretic peptide (BNP) elevated in the acute phase of KD.We examined the usefulness of ANP and BNP to diagnose the atypical KD.Patients and methods.We examined ANP and BNP in 10 atypical KD patients, and 30 patients with acute febrile diseases other than KD as a control group. Patients with less than 4 of 6 major criteria, or less than 3 major criteria with CAL were diagnosed as atypical KD. Differences in the data between the two groups were analyzed using unpaired Student's t test. Results. BNP was higher in the atypical KD group than those in the control group (38.2 +- 28 pg/dl (mean +- SD)vs(p<0.001)) vs 8.8 +- 17.3), while there was no difference in ANP between the atypical KD group and the control group (47.44+-22.3 vs 34.5 +- 28.8 (p=0.21)) ANP elevated more than 40 pg/dl in 6 of 10 patients with atypical KD and 10 of 30 patients in the control group. Meanwhile, BNP elevated to more than 20 pg/dl in 6 of 10 patients with atypical KD and in 3 of 30 patients in the control group.The sensitivity and specificity of ANP for the diagnosis of the atypical KD were 60.0% and 67.7%, respectively. Those of BNP were 60.0% and 90.0%, respectively. Conclusions. BNP is useful to diagnose an atypical KD patient in acute phase.

P994

Prediction score for the coronary artery lesions in the patient with Kawasaki disease at 3, 4 days after the first intravenous administration of gamma-globulin

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Background: High-dose intravenous gamma-globulin (IVGG) therapy is effective in preventing coronary artery lesions (CAL) in Kawasaki disease (KD). However, in spite of IVGG therapy, we often experience the patients whose coronary arteries develop into coronary artery aneurysms. We previously designed a prediction score that was composed of the following factors to detect a high-risk group for CAL at 3, 4 days after the first administration of 400mg/kg/day IVGG therapy. These factors are (1) body temperature more than 37.5 degrees, (2) neutrophils more than 7,000, (3) CRP more than 30% of the previous highest values and more than 3.0mg/dL, and during the course of illness, (4) the minimum albumin less than 3.0g/dL, (5) the maximum GPT more than 40IU/dL. The patients with more than 3 positive factors were defined as a high-risk group for CAL. Methods. (1) We examined retrospectively the validity of this score in 79 KD patients with CAL and 55 patients without CAL. (2) We conducted a prospective trial as follows. We started 5 days administration of IVGG in 32 patients with acute KD, and at 3, 4 days after the first administration of IVGG, we determined the high-risk patients using this score. For the high-risk patients, we treated with additional stronger therapies. We examined prevalence of the patients with CAL in this protocol. Results: (1) In the retrospective study, the sensitivity and specificity of this score at 3days after the first administration of IVGG were 91.7% and 70.9%, respectively. Those at 4 days after the administration were 92.9% and 80.0%, respectively. (2) In the prospective trial, we have no patients with CAL. Conclusion: This prediction score is useful to predict CAL after IVGG therapy.

P995

Systemic heterogeneity of endothelial function after Kawasaki disease

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Coronary arteritis associated with Kawasaki disease (KD) raises concern about the premature development of arteriosclerosis. Accordingly, we investigated endothelial function in the epicardial, resistance CAs, and femoral arteries (FAs) after KD during long-term observation. We assessed the responses of left epicardial and resistance CAs to serial intracoronary infusions of acetylcholine (final concentrations, 0.1 and 1 microml/L) and nitroglycerin in subjects by using quantitative angiography and a Doppler flow wire system. Three age-matched groups were evaluated: 8 control subjects (group 1), 10 KD patients with normal left CA from the onset (group 2), and 9 KD patients with a persistent or regressed aneurysm in the left anterior descending CA (LAD) (group 3). Acetylcholine (1 microml/L) changed the LAD area to 114.0+/-2.6%, 72.7+/-5.9% (P< 05 versus group 1), and 88.9+/-4.3% (P< 05 versus groups 1 and 2) of baseline in groups 1, 2, and 3, respectively, with a similar degree of increased coronary blood flow in each group. Nitroglycerin increased the LAD area to 143.5+/-7.7%, 132.3+/-3.9%, and 120.8+/-5.6% (P<.05 versus group 1), respectively. Newly, we evaluated the reactive hyperemia- or sublingual nitroglycerin-induced FA dilatation by high-resolution ultrasound in two age-matched groups: 13 controls, and 15 KD patients with persistent or regressed aneurysms in 4 and normal CAs in 9 patients. There were no differences in the FA responses to reactive hyperemia or nitroglycerin between the two groups. Results demonstrate a persistent endothelial dysfunction in the 'uninvolved' epicardial but neither in resistance CAs nor in FAs after KD, suggesting systemic heterogeneity of endothelial function in this disorder.

P996

Congenital coronary artery anomalies in children

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The aim of the study was to analyse symptoms, diagnosis and treatment of the coronary artery anomalies in children. The origin and coronary artery pattern was estimated in all standard echocardiografic examinations. Congenital coronary artery anomalies were found in 27 children. In 18 isolated coronary anomalies were recognized: anomalous origin of the left coronary artery from the pulmonary trunk (ALCA) in 15, anomalous origin of the right pulmonary artery from the pulmonary trunk in 1, coronary artery fistula in 4. In 9 children coronary abnormalities coexisted with congenital heart diseases: in 4 children with Tetralogy of Fallot, 3 neonates with transposition of the great arteries, 1 with truncus arteriosus and 1 in pulmonary atresia with intact ventricular septum. 11 infants with ALCA demonstrated angina spells, signs of myocardial ischemia or infarction in ecg. Features of congestive heart failure were observed in all infants with ALCA and 1 newborn with coronary artery fistula. In 23 children the diagnosis was established due to echocardiography and confirmed by angiocardiography in 16. In 4 patients the diagnosis based on angiography. 14 children underwent surgical correction. Conclusions: 1. The spectrum of clinical presentation in children with coronary artery anomalies vary from asymptomatic to life threatening. 2. The anomalies of coronary arteries can be defined by echocardiography. 3. Urgent surgical treatment is indicated in infants with ALCA and hemodynamically significant coronary artery fistula.

P997

Influence of Epstein-Barr virus infection on outcome of coronary artery lesions in patients of Kawasaki Disease

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Purpose. Kawasaki disease(KD) may be linked to primary infection by certain common viruses. KD concurrent with Epstein-Barr virus(EBV) suggests the possibility of an etiologic agent related to the KD rather than to the EBV infection itself, but unclear to influence on coronary complications. To establish whether infection with EBV contributed to the outcome of coronary artery lesions in patients with KD. Methods. Retrospective studies were performed on 96 cases(age 2.46±1.94 years) of KD evaluated with serologic studies of EBV(EBV EA IgM, EBV EA IgG, EBNA IgG) at admission. We evaluated the clinical features and coronary outcome between control group with KD and EBV associated group with KD. Results. On serologic studies of EBV, 62 patients (64.6%) with one more than positive results were EBV associated group with KD, in 30 patients with recent EBV infection and in 32 patients with previous EBV infection. There were no significant differences between control group and EBV associated group in age, sex, and other clinical findings. Eighteen patients (18.7%) had abnormal baseline echocardiograms, 13 patients(21.0%) of them were in EBV associated group and 5 patients(14.7%) in control group, and there was no significant difference between two groups. Significantly 11 of 13 patients(84.6%) with coronary lesion had positive result of EBNA IgG suggesting previous infection. There was no significant difference in retreatment and recurrence incidence. After retreatment, in 5 of 6 cases with EBV associated group and in no case with control group had cardiac complication. Conclusion. Previous or chronic EBV infection may influence on the occurrence of coronary lesion in patients with KD, even though patients with cardiac complication improved later.

P998

Interaction between human HGF and MMP-9 in Kawasaki disease

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https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Kawasaki disease (KD) is characterized by systemic arteritis, which often causes coronary involvement. MMP-9 is an enzyme that contributes to extracellular remodeling in several disease states including KD, and is regulated by various cytokines. Human HGF is one of the strongest angiogenesis factors produced by various cells including fibroblasts. To clarify the mechanisms of vascular remodeling in KD, it is important to know the interaction of MMP-9 and hHGF. We, therefore, investigated the plasma levels of MMP-9 and hHGF, and the regulatory mechanism of them in KD. (Subjects) 30 KD patients (group KD: M 19, F 11; 3m–5y), 10 healthy controls (11 M 5, F 5, 5m–3y) and 10 febrile controls (F: M5, F5; 10m–4y). (Methods) Plasma MMP-9 and hHGF were measured by ELISA. After treatment with plasma samples or cytokines including hHGF, the levels of mRNA for MMP-9, hHGF and cMET in HUVEC and fibroblasts were detected by RT-PCR (quantitative or not). (Results) Plasma MMP-9 and hHGF levels markedly increased during all phases in KD (MMP-9: 1130.8 ± 12.1 ng/ml, F 100.9 ± 87.1, KD pre-IVIG 304.3 ± 269.8, post-IVIG 330.5 ± 116.5, 1 m 77.7 ± 73.4 and hHGF, 0.09 ± 0.05 ng/ml, 0.32 ± 0.16, 0.92 ± 0.49, 0.37 ± 0.19 and 0.45 ± 0.69, respectively). There was a significant positive correlation between MMP-9 and hHGF. The assessed levels of mRNA for MMP-9 in HUVEC were significantly higher in KD pre-IVIG phase and stimulated by hHGF in a dose dependent manner. IL-6 enhanced hHGF expression in fibroblasts. Messenger RNA for cMET was constitutively expressed in HUVEC. (Conclusion) It was suggested that hHGF produced by fibroblasts stimulated by other cytokines, could regulate the synthesis of MMP-9 by endothelial cells in KD.

P999

Serum levels of procalcitonin in patients with Kawasaki disease

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Procalcitonin (PCT) is a new parameter of inflammation, the clinical usefulness of which is currently being evaluated. The present study measured the serum levels of PCT in patients with Kawasaki disease (KD) (n=25), and compared them with those in patients with systemic autoimmune disease (n=10), bacterial infection (n=12), or viral infection (n=17), as well as in healthy control children (n=18). Serum procalcitonin levels in patients with KD (2.1±0 ng/mL) were similar to those in patients with bacterial infection (2.2Å±2.9 ng/mL) but were significantly higher than in those with systemic autoimmune disease (0.4Å±0.4 ng/mL) or viral infection (0.4Å±0.3 ng/mL) and healthy controls (0.2Å±0.1 ng/mL) (p=0.001, 0.016 and <0.0001, respectively). Serum procalcitonin levels in KD were greater during the acute phase than the subacute phase of KD and the convalescent phase of KD (p=0.0001, <0.0001). Serum procalcitonin levels were significantly greater in KD patients who developed coronary aneurysm (7.2Å±3.8 ng/mL) than in those who did not (1.4Å±1.7 ng/mL) (p<0.005). Procalcitonin level is increased in acute KD. Procalcitonin may be a useful clinical predictor of the severity of KD, and may also be useful for differentiating KD from systemic autoimmune disease.

P1000

The management of the pregnancy and delivery in patients with coronary arterial lesion due to kawasaki disease >4 cases report

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The grown-up patients with coronary arterial lesion due to Kawasaki disease have been increasing, and some of the female patients have reached reproductive age. There are two important problems. When we manage these patients. One is the way of the delivery, and the other is the anticoagulant therapy during their pregnancy and delivery. We have managed four patients with coronary arterial lesion after Kawasaki disease. Two of the four patients had undergone coronary artery bypass grafting because of stenotic lesions. One patient had stenotic lesions of coronary arteries and other had dilated lesions. Selective coronary angiography by upper extremities' approach was performed for planning of delivery in two patients after 20 weeks. Three patients had the assisted vaginal delivery with extradural anesthesia. Only one patient who had stenotic lesions underwent caesarean delivery because of her progressive symptoms at 33 weeks. All of them had no major complication during delivery, and no ischemic changes on electrocardiograms. All neonates were healthy. One had been taking isosorbide dinitrate, and one had been taking low-dose aspirin(5 mg/day). These drugs were not affecting maternal and neonate during pregnancy. Pregnancy and delivery are possible in the patients with stenotic lesions, if they have no ischemic sign. Caesarean section should be considerable, if the patient is symptomatic. Extradural anesthesia reduce pain during delivery, and assisted vaginal delivery shortens the second period of delivery. Recently it has been reported that the use of low-dose aspirin is effective for habitual abortion and sterility. Low-dose aspirin may be recommended as anticoagulant therapy to suppress any thrombo-embolic tendency. Furthermore, it is impossible to suspect acute myocardial infarction. The emergency support system must be prepared for the patients who may have acute.

P1001

Evaluation of the efficacy of early treatment of Kawasaki disease before day 5 of illness

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Evaluation of the efficacy of early treatment of Kawasaki disease before day 5 of illness. Fong N.C., Hui K.W., Li C.K., Chiu M.C. Hong Kong SAR, China. To evaluate the efficacy of treating Kawasaki disease earlier than day 5 of illness with the standard dose of immunoglobulin and aspirin. This was a case control study of patients with Kawasaki disease admitted to Princess Margaret Hospital, Hong Kong during the period from 1994 to 1999. The Case group was consisted of 15 patients that received treatment earlier than day 5 of illness while the Control group was of 66 patients who were treated on or after day 5 of the disease. The 2 groups were matched by age and sex. All patients received aspirin 80–100mg/kg/day until fever subsided and immunoglobulin 2g/kg. Treatment efficacy was assessed by the duration of post-treatment fever and the incidence of coronary artery aneurysms. Proportion of febrile patients 48 hours after the first dose of immunoglobulin in the Case and Control group were 33% and 7% respectively(P=0.0032). The fever and other physical signs subsided with additional cases of immunoglobulin. The incidence of coronary artery aneurysms in Case and Control group were 13% and 9%(P=0.1). Treating Kawasaki disease before day 5 of illness may require a second dose of immunoglobulin although the incidence of coronary artery aneurysms was not increased. Additional doses of immunoglobulin may increase the chance of transfusion acquired infections. Further studies should be done to clarify this observation.

P1002

Myocardial Revascularization in Children with Kawasaki Disease. Diagnostic and Surgical Aspects in 2 cases

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The myocardial revascularization (MR) in children with coronary artery disease was initiated in our institution on March 95 and performed in 6 children; 2 of them had diagnosis of Kawasaki disease. The first case was a 20 months old boy, with Kawasaki disease symptoms; Echo showed good left ventricle function and a giant aneurysm and thrombus in right coronary artery (RCA). Angiographic study confirmed the giant aneurysm in RCA and showed also a small one in left anterior descending (LAD) coronary artery. He was submitted to a right internal mammary artery (RIMA) to RCA anastomosis and resection of giant aneurysm. With 45 months of asymptomatic evolution, the child was submitted to a routine angiographic study that showed RIMA to RCA with no obstruction and LAD occlusion. He was submitted to LIMA to LAD anastomosis, with no cardiopulmonary bypass, with good recovery. The child is well and asymptomatic 5 years after the first operation. The second case was a 10 years old boy, with Kawasaki disease symptoms and acute inferior myocardial infarction 30 days prior to the operation. Angiographic study showed aneurysms in RCA and LCA, with inferior wall dysfunction. He was submitted to a RIMA to RCA and LIMA to LAD anastomosis, with good evolution. With 15 months of follow up, after inespecific symptoms, he was submitted to an angiographic study that showed RIMA to RCA with no obstruction and left coronary artery and LIMA to LAD anastomosis occluded. He was submitted to a new operation, using the LIMA to directly connect the LAD to the main LCA. The child is well and asymptomatic 6 months after the second operation, with negative radioisotopic myocardial perfusion studies. In conclusion, MR in children with Kawasaki disease and coronary aneurysm seems to be a safe and effective method, despite the need of redo in both cases. Longer follow up and angiographic controls required to better evaluation.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P1003

Gamma globulin re-treatment in Kawasaki disease

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Gamma globulin (GG) re-treatment is still controversial. The objectives of this study were to evaluate the therapy protocol of GG re-treatment. Patients: From Jan. 1994 to Apr. 2000, 185 consecutive typical patients (pts) were admitted to this hospital without coronary artery dilatations at admission. One hundred forty-two pts (77%) of them were treated with GG. One hundred eighteen of them were selected as high-risk pts by Iwata's risk score system and 108 pts were treated with single 2g/kg of GG within 9 days of illness. Seventy-three pts showed defervescence (<37.5 degrees centigrade) within 3 days after initial GG. Of the 108 pts, 19 pts had one time re-treatments, 8 pts had two times and 3 pts had three times or four times re-treatments without steroids. GG were used sulfonated GG (Sulf-GG) or PEG treated GG (PEG-GG) at random. Doses of GG re-treatment were determined according to the d-value (blood culture and d-C-reactive protein values (d-value) were pre values – post values after GG therapy). GG re-treated were repeated till their fever under 37.5 degrees centigrade. Results: Thirteen pts (male/female = 12/1) of 185 had coronary artery lesions (CALs), which were detected at 10.6+-3.4 days of illness. These CALs were transient dilatations and not aneurysms. High-risk male pts used Sulf-GG had more re-treatments and CALs significantly higher than PEG-GG. Conclusion: The first detected days of CALs were generally 10 days of illness, and re-treatments should accomplish in the end of 10 days of illness. Repetition of full re-treatment of PEG-GG can reduce coronary aneurysms.

P1004

Multidetector-row Computed Tomography for detection of coronary artery lesions in children with Kawasaki Disease

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Multidetector-row Computed Tomography (MD-CT) is a new helical CT which can visualize each coronary artery by reconstruction. One study with MD-CT requires only a few minutes. MD-CT was performed in 20 patients (16 patients with coronary sequelae documented by previous angiography, 3 with transient change on acute-phase echocardiography and 2 with normal coronary artery in acute-phase echo). In 2 youngest patients we could not reconstruct a total coronary image because they could not withhold respiration as directed. Remaining 16 patients could be assessed successfully in terms of 1) coronary diameter, 2) coronary wall thickness, 3) coronary artery calcification, and 4) patency of coronary artery. Coronary flow direction could not be judged in the so-called re-canalized lesions. Ten patients also underwent coronary angiography around the time of MD-CT. Of 8 patients who showed coronary calcification on MD-CT, 2 could not be confirmed by fluoroscopy and/or coronary angiography. Two patients exhibited progressive coronary stenosis on MD-CT over previous angiographies. Subsequent angiography confirmed progression of stenosis and/or calcification, and a bypass surgery was performed. In 13 patients, MD-CT was assessed without concomitant angiography. MD-CT suggested coronary calcification in 1 patient who had no coronary change during acute-phase echo. Another patient exhibited progressive coronary stenosis on MD-CT over previous angiography. Due to the patient's reluctance, angiographic confirmation is not obtained. There was no complication with MD-CT. We conclude that MD-CT is safe and sensitive method for detection of coronary artery stenosis and calcification after Kawasaki Disease.

P1005

Role of MPO-ANCA in Candida albicans extract induced vasculitis as a model of Kawasaki disease

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Kawasaki disease (KD) is the most frequently acquired systemic vasculitis in childhood. Coronary arteritis is life threatening complication of KD. Recently, some patients with KD show high titer of MPO-ANCA, which is a kind of autoantibody to myeloperoxidase in cytoplasm of neutrophil. This autoantibody is known to be related some kind of vasculitis syndrome, such as microscopic polyangiitis and Churge-Strauss syndrome. We have established experimental model of systemic vasculitis in mice induced by an intraperitoneal injection of Candida albicans extract which produced from the yeast isolated from the feces of a patient with KD. In this model coronary arteries are more frequently involved and histological feature of vasculitis was expressed as proliferative and/or granulomatous inflammation. For close resemblance of both distribution and histological feature of vasculitis, this model has been accepted as the animal model of KD. In our model, the average level of MPO-ANCA in coronary arteritis positive mice was much higher than those in negative mice. These results indicate that MPO-ANCA may be closely related to development and/or extension of coronary arteritis in this model. Next, we applied the experimental system to MPO deficient mice in order to analyze the roles of MPO and MPO-ANCA in this model. We had expected that vasculitis had not developed and titer of MPO-ANCA in sera had not increased in MPO deficient mice. However, coronary arteritis developed even in MPO-deficient mice. There was no histological difference between MPO-deficient mice and Wild mice. In addition, MPO-ANCA titer in sera tended to be higher in vasculitis positive mice than in vasculitis negative mice. These results suggested that some antigens other than MPO involved in MPO-ANCA production induced by C. albicans extract treatment.

P1006

Biochemical markers of myocardial injury in acute phase of Kawasaki Disease

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Background: In acute phase of Kawasaki Disease (KD), subclinical myocarditis is observed. In the recent studies, the heart- type fatty acid-binding protein (H-FABP) can be used as an early indicator of myocardial injury. Objective: To investigate the degree of myocardial injury in patients with acute phase of KD using non-invasive serological tests, we studied the serum levels of several biochemical markers for myocardial injuries. Methods: we sequentially measured the serum concentrations of H-FABP, myoglobin (MYO), cardiac troponin-T (cTnT), cardiac troponin-I (cTnI), and myosin light chain-I (MLC-I), before and after treatment with intravenous gammaglobulins (IVGG) in 20 acute KD patients (mean age, 2.6 +-1.4 years, M/F=14/6). Results: The mean levels of H-FABP were 4.3 +-1.9 ng/ml before IVGG and decreased to 3.7 +-1.5 ng/ml (p<0.05) after 1 month. The H-FABP significantly elevated in 10.4%(4/38) before IVGG treatment. Despite clinical symptoms and signs improved after IVGG, the serum levels of H-FABP elevated in 15.4%(4/26). The mean levels of MYO were 27.8 +-9.5 ng/ml before IVGG and decreased to 19.4 +-7.2 ng/ml (p<0.05) after 1 month. However, the levels of MYO elevated in only 3.8% (1/26) that normal range before IVGG. The cTnT, cTnI, MLC-I were in normal range. Conclusion: Subclinical myocarditis can be detected by serum H-FABP and MYO. H-FABP seems to be a more reliable biochemical marker for the early detection of myocardial injury in acute phase of KD. IVGG may exert an anti-cellular-necrosis effect on acute myocarditis in KD.

P1007

Cardiovascular insufficiencies during acute stage of kawasaki disease

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This retrospective study is to evaluate the patients of Kawasaki disease (KD) complicated with severe cardiovascular insufficiencies, which include CHF, shock, systemic arterial hypotension (SAH), peripheral gangrene (PG) and multiorgan failure (MOF). The correlation with the IVIG therapy or coronary arterial aneurysm was also evaluated. During the period of Aug 1, 1991 to July 31, 2000, 405 hospitalized patients were diagnosed as KD in this hospital during the acute stages. All were treated with IVIG 2gm/kg single dose, and aspirin 100mg/kg/day. Vital signs were monitored every 30 mins to 1 hr throughout the IVIG therapy. All were followed up with echocardiograms for at least 3 months. Seven patients were found to be suffering from severe cardiovascular insufficiencies (1.7%), including 4 of CHF, 4 of SAH, 2 of shock, 2 of MOF and 1 of PG. Six were complicated with CAA and 4 had CAA bigger than 7.0 mm. Particularly, 3 of the 4 SAH incidences were found to be temporally correlated with the IVIG therapy. Shock or MOF occurred in 3 of these 4 SAH patients. Although the IVIG therapy can reduce the complication of CAA, some severe CVI still exist, especially when SAH presents.

P1008

Comparison of dipyridamole stress tc-99m-sestamibi SPECT and coronary angiography in patients with Kawasaki disease

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https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

To compare dipyridamole stress Tc 99m-sestamibi single-photo emission computed tomography (SPECT) with coronary angiography in patients with Kawasaki disease. Forty-one consecutive patients (29 boys, 12 girls) were divided into 3 groups according to coronary angiography. Group A consisted of 2 patients (1 boy and 1 girl, aged 10.3 and 1.9 years respectively) with coronary stenoses. Group B consisted of 30 patients (18 boys and 2 girls, aged 0.7–15.3 years, mean of 3.8 years) with coronary aneurysms. Group C comprised of 29 patients (25 boys and 9 girls, aged 1.3–13.8 years, mean of 7 years) with normal coronary angiograms. There was no significant correlation between Tc-99m-sestabimi SPECT and coronary angiography to detect coronary stenosis ($p=0.5$) or aneurysm ($p=0.18$). In conclusion, there is significant discordance between coronary angiography and Tc 99m sestabimi SPECT.

MAY 30 Time: 14:00–15:30

Session 6
Cardiomyopathies/Myocarditis/Heart Failure

P1009

Left ventricular restrictive dysfunction in children with dilated cardiomyopathy and its clinical implications

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To evaluate left ventricular (LV) diastolic function in children with idiopathic dilated cardiomyopathy (DCM). LV diastolic function was assessed in 48 children with DCM (26 male, 22 female, aged 3m–14yrs, 6.1±4.4 yrs) and 48 age and gender matched normal controls by using Doppler echocardiography. Mitral flow and pulmonary vein flow (PV) were recorded and measured at the initial enrollment. All patients were followed serially. The results showed that deceleration time of mitral E wave (DT) in patients was shorter than controls (92±27ms vs 128±40ms, $P<0.01$). Mitral E/A ratio increased and the ratio of PV peak systolic velocity to peak diastolic velocity (S/D ratio) decreased in patient group (both $P<0.05$). According to our own criteria, 20 (42%) patients showed LV restrictive diastolic dysfunction (RDD), including 16 (33%) with a shortened DT, 14 (29%) with an increased E/A ratio and 14 (29%) with a decreased S/D ratio. By multinominal logistic regression, RDD related to calculus of left atrium and LV elevation of pulmonary systolic pressure, and did not related to LV ejection fraction. But patients with RDD had higher NY cardiac function scores ($P<0.01$). All patients were followed for 6–57 months (mean 24 m). Nine (45%), 2(10%), 9(45%) and 0 of 20 patients with RDD died, deteriorated, unchanged and improved respectively while 1 (4%), 4(14%), 8(28%) and 15 (54%) died, deteriorated, unchanged and improved, respectively, in 28 patients without RDD ($P=0.05$). We concluded that 42% of children with DCM had RDD, which was related to the NY cardiac function scores and clinical outcome.

P1010

Left partial ventriculectomy in a ten year year old child with end stage dilatated cardiomyopathy

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A ten year old girl with dilatated myocardiopathy was referred for severe heart failure, complicated with acute pulmonar oedema, left atrium thrombosis and vascular cerebral embolism. Parenteral treatment and assisted ventilation were necessary. The child improved, had total neurological recovery but kept on NYHA class III. Hemodynamic explorations showed major dilatation of the left ventricle (VG ed 67mm) and catheterization confirmed low cardiac index (2,300/min/m2), moderate mitral valve regurgitation but normal pulmonary pression. Heart transplantation was indicated but not accepted because of the geographic conditions that did not allowed immunosupressive therapy and follow up. Palliative surgery was then discussed and left partial ventriculectomy was performed following the BATISTA procedure. After surgery a short inotropic support was necessary and the child was extubated on the third day. The left ventricle remained dilatated (VG ed 58mm) and hypokinetic but a catheterism showed improvement of cardiac index (3,350/min/m2). Two months later the child was well equilibrated with oral treatment. An exercise ECG was performed, showing submaximal performances. Seven months after surgery she had a new decompensation that could be related to medication interruption. Parenteral inotropic support and vasodilatators were administrated. The situation improved and the child could be stabilized in NYHA II with oral therapy. Two years after surgery she remains well equilibrated. The conclusions in this case were that left ventricular remodeling was possible in children with end stage heart failure without major perioperative complication. The benefits were significative in the early time after surgery with improvement in our functional class. The evolution did not avoid relapse of a new decompensations few months later although it was still accessible for medical treatment. This procedure may be indicated in children where heart transplantation is not possible.

P1011

Cardiac involvement in patients with metabolic disorders (md) Echocardiographic follow-up study.

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The aim of this study was to analyse retrospectively the cardiac involvement in patients (pts) with MD treated in our Centre and evolution at follow-up. Material and methods: 2M pts, 11M, 12F, median age at diagnosis 2 yrs (0.1–35) were studied between 1980 and 2000 by echocardiography at diagnosis and at follow-up (for 7 pts, median 6yrs). 3d pts had organic acidemia group (gr.1), 4M- ammonia ideopathies (gr.2), 3d- urea cycle defects (gr. 3), 3M- d. of metabolism of carbohydrates (gr.4), 6-d. of beta-oxidation (gr.5), 12-mitochondrial d. (gr.6), 5F-lysosomial storage d. (gr.7), 1F-microlithiasis d (gr.8). Results: 25 cases (9.7%) 11M, 12F, had cardiac impairment: 21 – hypertrophic cardiomyopathy (HCMP), obstructive in 2, 5-dilated CMP (DCMP); 4- H-DCMP, 2 with chronic pericarditis; 3- mitral and/or valvar anomalies. Cardiac involvement was more frequent in gr 5(66.7%) and 6 and 8 (13–15.7%). Evolution at follow-up: 6 children (26%) died at 1m–4 yrs, 4 with DCMP and 2 with HCMP, one due to metabolic decompensation. 5 pts with HCMP improved after a specific metabolic treatment, one on a combined metabolic and cardiac therapy. One child worsened and the remaining pts are stable. Conclusions: Cardiac involvement of variable severity is noticed in 10% of our pts, regressive in some cases after specific treatment of the basal disorder.

P1012

Myocarditis in children – clinical course and prognosis.

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The aim of study was to identify prognostic features of children with biopsy-proven myocarditis (myo). The clinical profile of 55 pts with myo aged 2 mo - 17,5 yrs (x=10+/- 5,1 yrs) was reviewed to detect any factors that might be predictive for their outcome. In all pts were done: echocardiographic evaluation of LV diameter and function, chest X-ray, exercise and 24-h Holter ECG, endomyocardial biopsy. Follow-up ranged from 6 mo to 5 yrs (x=2.6+/- 1.2 yrs). There were 49 (89%) survivors and 6 (11%) nonsurvivors. The 1-year mortality was 6%, 7% and the 2-year 8%, 2%. All deaths were within first 3 years. Survivors and nonsurvivors were compared with regard to the age at presentation, mean time between clinical onset and diagnosis, functional class (NYHA), LV diameter and shortening fraction, cardiomegaly (CTR), ST-T changes and ventricular arrhythmias. Conclusions: 1. Clinical course of myocarditis in children is variable and influences the prognosis. 2. The risk factors for death included: the mean time between clinical onset and diagnosis, NYHA class, LV diameter and function, cardiomegaly. 3. The coexistence of acute myocarditis and DCM or ARVD predicts poor outcome.

P1013

Beta-receptor blocker therapy on children with heart failure: clinical experiences with carvedilol

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Pediatric patients with congestive heart failure, receiving standard anticongestive therapy without any improvement are considered for heart transplantation. A new therapeutic option for children could be the additional treatment with b-receptor blockers e.g. Carvedilol (C), which have been shown to reduce morbidity and mortality in adult patients. Patients and Methods: Fourteen patients (2 month up to 18 years) with congestive heart failure due to dilated cardiomyopathy (n=10) and congenital heart disease

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

(n=4) were treated with slowly increasing doses of oral C (initially, 0.09 up to 0.70 mg/kg/day). They have been treated with digoxin, angiotensin-converting enzyme inhibitors and diuretics. Ejection fraction (echocardiography), Brain Natriuretic Peptide (NT-proBNP), clinical symptoms (modified Ross Score) and electrocardiography (heart rate, QT duration) were determined before carvedilol therapy and then monthly up to six month.Results: Ejection fraction increased from 34 (7–48) to 54 (10–75, p<0.05) % during treatment with carvedilol. NT-proBNP decreased from 663 (221–2008) to 461 (211–3058) fmol/ml. Clinical symptoms (Ross Score) decreased from 5,4 (3–10) to 2.6 (0–6) points (p<0.05). Mean heart rate decreased by 13% (0 to 23%; p<0.05). Mean QT duration with heart rate correction calculated by Bazett (QTB) and Fridericia formula (QTF) decreased from 420 ms (372–507 ms, QTB) to 385 ms (323–440 ms, QTB; p<0,05) and from 372 ms (315–466 ms, QTF) to 353 ms (318–435 ms, QTF, p<0,05).We observed side effects on 5 of 14 patients, however there were no side effects which would have led to a modification of the therapy. Conclusion: Heart function, clinical symptoms, humoral and electrocardiographic parameters improved under therapy with carvedilol. Our first clinical data indicate that oral carvedilol in addition to standard therapy, constitutes an effective treatment in pediatric patients with congestive heart failure.

P1014

Involvement of the heart in newborns of diabetic mothers.

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Background: A hypertrophic cardiomyopathy with congestive heart failure has been described in detail more recently in newborns of diabetic mothers. For this cardiomyopathy is typical asymetrical septal hypertrophy, dynamic subaortic stenosis, and myofibrillar disarray. It generally regresses over 6 or 12 months after birth. It has been estimated that 30 % of newborn of diabetic mothers have cardiomegaly, that 5 to 10 % have congestive heart failure. Diabetic hypertrophic cardiomyopathy does seem to be more commonly associated with poor maternal glucose regulation. Aim of study: Our purpose was to determine whether left ventricle mass (LVM) of newborns of diabetic mothers differ from LVM of healthy newborns. Methods: Echocardiographic evaluations were performed after birth, in 1th and 6th month of life in 30 healthy newborns (group 1) and in 20 newborns of diabetic mothers (group 2). The following variables were measured: left ventricle mass (LVM), left ventricle mass index (LVMI), left ventricle end-diastolic volume (LVEDV), mass volume index (LVMVI) and relative wall thickness (RWT). Results: A significant higher LVM and LVMI was found after birth in newborns of diabetic mothers (group 2) in comparing with healthy newborn (group 1). These abnormalities are more evident in newborns of mothers with poorer glycaemic control during pregnancy. The parameters characterizing the left ventricle increases significantly in 1th month and 6th month in group 1 but not in group 2.Conclusion: The results are useful in interpretation of echocardiographic examination of left ventricle in newborn and infants of diabetic mothers.

P1015

A cohort study of dilated cardiomyopathy in childhood: predictors of outcome

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Objective: To study the correlation of clinical features and echocardiographic and electrocardiographic measures with outcome in paediatric patients with dilated cardiomyopathy in order to define early predictors of outcome. Methods: Retrospective study of the total cohort of 31 paediatric patients with dilated cardiomyopathy referred to a regional centre of cardiology between 1977 to 2000. Original ECG and M mode echocardiographic traces were remeasured for 8 electrocardiographic and 30 echocardiographic measures at presentation, 4 months follow-up and latest visit. Multivariate correlation analysis was used to correlate with death or heart transplantation. Results: 16 patients with dilated cardiomyopathy died or required heart transplant (median time 0.96 years). Out of the total cohort of 31 1-year survival was 78% and 5-year survival 55%. Patients who were younger than 2 years at the time of presentation had a better survival (p=0.02). There were no ECG features that correlated with non-survival. The echocardiographic measures showing early (4 months follow-up) correlation with outcome were: left ventricular ejection fraction (p=0.009), systolic left ventricular posterior wall-to-cavity ratio (p=0.048), systolic and diastolic volume of the left ventricle, corrected by body surface area (p=0.013 and p=0.028 respectively), and left atrium to aorta ratio (p=0.021). Failure to improve beyond cut-off values of ejection fraction <0.26, systolic left ventricular wall-to-cavity ratio <0.20, end-diastolic volume >190 ml/m2 BSA or end-systolic volume >150 ml/m2 BSA and left atrium-to-aorta ratio >1.8 all predicted non-survivors early with high specificity and sensitivity. Conclusion: Early identification of those patients whose best chance of survival is heart transplantation is possible using relatively simple echocardiographic measures.

P1016

Effects of carvedilol on left ventricular function, mass, and scintigraphic findings in isolated left ventricular noncompaction: a case study.

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Despite an increasing awareness and interest on isolated left ventricular noncompaction, there is still no information regarding the effects of medical treatment. A 4-month old infant with congestive heart failure due to isolated left ventricular noncompaction underwent carvedilol, non selective beta- blocker, treatment. Hemodynamic analysis and imaging with several modalities, including echocardiography, magnetic resonance imaging (MRI), and single photon emission computed tomography with Tl-201, I-123-beta- methyliodophenyl pentadecanoic acid (BMIPP), and I-123 -metaiodobenzylguanidine (MIBG), were performed before and 14 months after treatment, before and after carvedilol. left ventricular ejection fraction increased from 30 to 57%. Remarkable reductions were observed after treatment in left ventricular end-diastolic volume (from 47 [147% normal] to 28 ml, [130% normal]), end-systolic volume (from 23 to 12 ml), mass (from 39 to 23 g), and end-diastolic pressure (from 13 to 8 mmHg), respectively. Present wall thickening determined with cine MRI increased after carvedilol in the segments corresponding with noncompacted myocardium. Nuclear studies demonstrated that, before treatment, there was a mismatch between Tl and BMIPP uptake, indicating preserved myocardial perfusion but compromised oxidative fatty acid metabolism at the area of noncompaction. Before carvedilol, sympathetic nerve dysfunction at noncompacted areas was demonstrated with MIBG, a radiolabeled norepinephrine analogue taken up by the sympathetic neuronal terminals. Nearly normal uptake 15 minutes after injection of MIBG and obviously decreased uptake 4 hours later indicated preserved cardiac innervation but impaired neuronal function storing MIBG in presynaptic vesicles of noncompacted myocardium. After carvedilol therapy, there was no more mismatch between Tl and BMIPP, and the neuronal function was improved demonstrated on MIBG delayed images. In conclusion, carvedilol demonstrated marked favorable effects on global and regional left ventricular function, hypertrophy, and both metabolic and adrenergic abnormalities in isolated left ventricular noncompaction.

P1017

Left ventricular non-compaction in children – does the isolated form differ from that associated with cardiac malformations?

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Left ventricular non-compaction (LVNC) is a poorly described cardiomyopathy with uncertain outcomes. This study compares the presentation and outcome of children with isolated LVNC (ILVNC) with those who have associated cardiac malformations (LVNC+CHD). We reviewed all children with LVNC seen in our hospital between 1990 and 2000. LVNC was diagnosed when prominent trabeculations and deep recesses of the left ventricular myocardium were seen on ventricular angiography. Non parametric statistical methods were used, with continuous data expressed as [median (25th – 75th centile)]. Of 39 children identified, 25 (12 male) had ILVNC and 14 (9 male) had LVNC+CHD. The malformations included atrial and ventricular septal defects, aortic stenosis, aortic coarctation, hypoplastic right heart syndromes, double inlet LV and double outlet RV with atrioventricular septal defect. Children with ILVNC presented later [155 (60 – 780) days versus 2 (1 – 60) days, p<0.01] and the majority (80%) presented with congestive cardiac failure (CCF). 14% of the LVNC+CHD group presented with CCF, but most presented with symptoms of their malformation. The five year survival from birth for LVNC and LVNC+CHD was 58% and 86% respectively (p=0.37). A larger proportion of the ILVNC group have died or undergone cardiac transplantation (52% versus 14%, p<0.05). Among surviving patients cardiac projections have cardiac dysfunction (fractional shortening <20% or restrictive physiology) at last follow-up (75% for ILVNC versus 33% for

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

[VNC+CHD, p=0.67] We conclude that LVNC can be associated with a variety of cardiac malformations. Children with LVNC commonly present with CCF at an older age, and are more likely to die or require transplantation. The presence of LVNC with other malformations does not appear to dominate outcome and should not preclude aggressive surgical management.

P1018

Long term follow-up of myocardial changes following Kawasaki disease (KD) with coronary aneurysm using repeated endomyocardial biopsy

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Purpose: To investigate the long-standing myocardial abnormalities and the relationship between coronary artery lesions (CAL) and myocardial damage, serial coronary arteriography and EMB were performed in the patients with KD, followed up more than ten years after onset. Patients and method: They included 12 patients with giant coronary aneurysm (G-AN), the male to female ratio was 7: 5 and 16 patients with CAL ; the male to female ratio was 9:7. All patients were followed up more than ten years after onset. Episodic coronary arterial lesions were analyzed semiquantatively with MIPRON and histopathology with histomorphometric method to calculate the percent area of myocytes, fibrous tissue and fatty tissue and small vessel changes. Results: In the long term follow-up EMB: fibrosis, degeneration , disarray and inflammatory cell infiltration were noticed in 67 %, 54 %, 33 % and 25 %, respectively in the patients with G-AN. One of the cases with KD who had CAL ,revealed massive inflammatory cell infiltration and myocytolysis in the subsequent study, which suggested chronic myocarditis. Conclusion: Myocardial changes in the patients with G-AN were relatively mild on light microscope but still remained and ultrastructural changes as macrocytopathy in late stage. Some cases of KD may develop chronic myocarditis leading to a cardiomyopathy-like state. Further approaches should be mandatory to clarify the significance of the myocardial sequelae of KD with close attention not only to CAL but also to the myocardial changes in long standing cases of the disease.

P1019

Prospective longitudinal assessment of late anthracycline cardiotoxicity in Wilms' tumour(WT) survivors - can diastolic data predict the outcome?

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Anthracyclines (Anth) are highly effective in the treatment of childhood malignancies, but are cardiotoxic. It has been suggested that alterations in diastolic function may permit manifest systolic abnormalities. Most previous reports have been cross sectional studies of heterogeneous diagnostic groups. We performed a longitudinal, prospectively study of 37 well-characterized WT survivors (age at treatment: 4.1 ± 2.1 yrs, Anth dose: 303 ± 78 mg/m2, range 60–468). Echocardiograms were performed on 7.1 ± 3.7 and 11.1 ± 4.7 yrs after the last anthracycline treatment and compared to values obtained in 100 healthy controls. All values are expressed as SD units (Z-scores) derived from the control population, and adjusted for body surface area. At the second assessment, both early (E) and atrial (A) peak velocities were low (Z scores: -0.65 and -0.38, respectively), but E/A ratio was normal. The average isovolumetric relaxation time (IVRT) and deceleration of early filling (Edec) were both increased (Z-scores: 0.57 and 0.50, respectively). Paired serial data revealed a significant increase in E velocity (from -0.99 to -0.68, P=0.004). A velocity was unchanged, and E/A ratio increased from -0.36 to 0.03 (P=0.004). Furthermore, a prolongation of IVRT (from 0.18 to 0.56, P<0.001) and Edec (from -4.2 to 0.6, P<0.001) was seen at the reassessment. Anthracycline dose did not correlate with neither of the diastolic parameters. E velocity at first evaluation correlated with fractional shortening at the second evaluation (r=0.32, P=0.002), otherwise diastolic parameters failed to predict systolic dysfunction at second assessment. Anthracycline treatment is associated with significant changes in LV relaxation, but long-term follow-up does not show severe or restrictive diastolic dysfunction.

P1020

Mode and age at death in children with hypertrophic cardiomyopathy (HCM).

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We studied the incidence, mode, and age at death in a group of infants and children with HCM during a period of 25 years ending December 1995. We included all patients with primary HCM and those secondary to systemic diseases not likely to shorten the life span. Children with associated heart defects producing more than mild hemodynamic derangement were excluded. There were 43 patients with a mean age of 5.3 + 5.8 years (range 1 month – 22 years), 20 were male. The follow-up period was 6.6 + 5.6 years (range 1 month - 20.6 years). Twelve had secondary forms; 6 had Noonan's syndrome; 3 LEOPARD syndrome, and one patient each with Friedreich ataxia, Williams syndrome, and Noonan' disease. Total mortality was 21 percent (9 patients), with an annual rate of 3.7 percent. Death was sudden in 5 and caused by congestive heart failure in 4. Patients dying suddenly were older (13.9 + 7.4) than those with congestive heart failure (3.2 + 2.9); (p<: 0.05). Two of the sudden deaths occurred in patients with Wolff-Parkinson-White syndrome. One of them had had radiofrequency ablation of the anomalous pathway. Another sudden death, happened in a child with obstructive HCM who had a permanent DDD pacemaker implanted because of persistent symptoms despite pharmacologic treatment. Sudden death occurred while asleep in 2 and during ordinary activities in the rest of the patients. We conclude that the annual rate of death in young patients with HCM does not differ much from that quoted for adults in referral centers. Infants and young children usually die in congestive heart failure, while older children tend to die suddenly. However, in this series, none died during strenuous activities.

P1021

Non-compaction of ventricular myocardium: report of seven cases

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Non-compaction (NC) of ventricular myocardium is characterized by increased trabeculation and intertrabecular recesses. We report seven cases of NC diagnosed by echocardiography in the last five years at our institution. Age ranged from 2mo -32 years. Associated heart diseases (corrected transposition of great vessels, patent ductus arteriosus, congenital mitral regurgitation) were probably responsible for NC in 3 cases, and it was primary in the other 4. Five patients presented in congestive failure, one had growth retardation while NC was incidentally detected. Dysmorphic impurities and strokes were associated with primary NC in one case each. Ventricular dysfunction was present in all. Our patients died of congestive heart failure. No arrhythmias or embolism were noticed in these 7 patients. In conclusion, NC of ventricular myocardium is a rare heterogeneous cardiomyopathy, its occurrence in disorders of bone synthesis is intriguing.

P1022

Acute myocarditis with dilated cardiomyopathy caused by parvovirus B19

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Human parvovirus B 19 can cause a broad spectrum of clinical manifestations including erythema infectiosum (or fifth disease), intrauterine fetal death, aplastic crisis and anaemia in immunincompetent patients. Myocarditis caused by parvovirus B 19 was described in an infant and has been found in the hearts of infected fetuses dying of hydrops fetalis. We observed acute onset of myocarditis with dilated cardiomyopathy after parvovirus B 19 infection in a 5 year-old boy. Diagnostics included routine cardiac investigations and heart catheterization with biopsy of the endomyocardium. The diagnosis was made by DNA-hybridisation and polymerase chain reaction investigations of serum and myocardial biopsy. This case report suggests that parvovirus B 19 infection can cause life-threatening myocarditis in childhood and that diagnostic assays like polymerase chain reaction should include parvovirus B 19. To our best knowledge there are only very few cases described with myocarditis associated with parvovirus B 19 infection.

P1023

ECMO and transcatheter left heart decompression in an infant with acute severe left heart failure

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We describe a unique technique for support of the heart during an episode of acute, severe heart failure. A 14 month-old infant presented with a 1-week history of poor appetite, low-grade fevers, vomiting and progressive respiratory

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

distress. Significant cardiomegaly with severe left ventricular (LV) dilatation and dysfunction was noted. Endotracheal intubation and mechanical ventilation were required during transport from an outlying hospital. Inotropic support with epinephrine, dopamine, and milrinone was initiated. Despite this, she developed progressive low cardiac output with episodes of ventricular tachycardia, eventually suffering ventricular fibrillation that did not respond to electrical defibrillation. During CPR she was placed on ECMO via cannulation from the right neck and immediately taken to the catheterization lab. With transesophageal echocardiographic guidance a transseptal puncture was done and progressive static balloon dilatation of the atrial septum was performed. The initial LA pressure of 35 mm Hg was reduced to 9 mm Hg. Pulmonary edema fluid cleared almost immediately with instantaneous improvement in lung compliance. Endomyocardial biopsy of the right ventricle confirmed acute myocarditis. The patient subsequently required 5 days of ECMO support. At the time of decannulation, her LV function was markedly improved. She was discharged after three weeks, with full neurologic recovery and normalization of left heart function, on only a small dose of diuretic and was taken off of the transplant list two weeks later. We conclude that aggressive therapy is warranted in infants who present with acute severe heart failure. ECMO cannulation from the neck with transatrial decompression of the left heart is an effective way of mechanically supporting the failing heart of a child without median sternotomy. The capacity for complete resolution of severe of myocarditis and acute heart failure is again demonstrated.

P1024

A case of restrictive cardiomyopathy with ventricular tachycardia treated by amiodarone and pacing

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Background: Amiodarone(AMD) is used as the antiarrhythmic agent in patients with life threatening recurrent ventricular arrhythmias with hemodynamic instability. We report a restrictive cardiomyopathy (RCM) patient with VT who has been treated by AMD and DDD pacing. Case report: A 28-year-old RCM patient admitted due to persistent palpitation and bradycardia of three hours' duration. A chest X-ray film revealed no cardiomegaly. An electrocardiogram showed a sinus rhythm, at a rate of 75, with prominent P waves and prolonged PR. Echocardiography visualized enlarged LA and near-normal thickness of the IVS and LVPW. Systolic function was normal reflected by ejection fraction. Cardiac catheterization showed elevated pulmonary artery wedge pressure and LV diastolic pressure. Both right and left ventricular volumes were small. His bundle electrogram disclosed prolonged HV time. Sinus node function was normal from the results of SNRT, CSNRT and SACT. HV block was revealed by the high frequent atrial stimulation at a rate over 120. The oscillasional electrogram revealed the normal duration of LAS and the normal voltage of RMS although filtered QRS was prolonged. Mitral inflow and LV outflow were recorded by echocardiography after DDD pacing was implanted. He was paced with AV synchronously at a fixed AV delay of 175msec. The Holter monitoring performed because he felt palpitation after he was paced, revealed VT at a maximum rate of 180 with consecutive 19 ventricular complex. Not only palpitation has been diminished also no VT has been seen on Holter monitoring since disopiramide was changed to AMD. He showed great capability of ergometer exercise with five minutes endurance before he discharged. Conclusion: It is controversial whether AMD is used in patients who have serious heart failure with non-sustained VT after pacing.

P1025

Prevention of recurrences idiopathic pericarditis by Colchicin in two children.

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A two children with severe form of idiopathic recurrent pericarditis are reported. On several occasions during a period of 2 years in first cases and 5 months in second case they were shown to be dependent on corticosteroid therapy and became cushingoid. After corticosteroids were substituted with colchicin, no further relapses occured during a period of 12 months in first case and 9 months in the second case. In accordance with the last published pediatric results, colchicine represents an effective and well-tolerated alternative therapy for recurrent idiopathic pericarditis and might replace prolonged administration of corticosteroids.

P1026

Real-time power spectral analysis of heart rate variability in Duchenne-type progressive muscular dystrophy

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Aim: To assess heart rate variability – HRV- in pediatric subjects affected by DMD, by means of real time power spectral analysis and to compare it to a healthy control group. Methods: We have conducted a cohort study comparing a group of 20 DMD subjects with normal cardiac ultrasound and no evidence of acrhythmia with a control group. An ECG monitoring (HP 78354 C) has been performed for 10 minutes with the subject at rest, supine, 1 hour after breakfast, between 08.00 and 10.00 a.m. Data from the monitor have been analyzed on real time using the software previously described (Comput Methods Programs Biomed, 1998). Two different frequency bands have been considered for spectral analysis: low frequency band – LF – between 0.02 and 0.15 Hz and high frequency band – HF between 0.15 and 0.5 Hz. The power spectral density (PSD) in the two bands and the LF/HF ratio, as indicator of sympathovagal balance, have been calculated. Results: HR was significantly higher in the DMD group (mean 95±16 vs 68 ± 10(SD) p<.05). LF did not vary significantly. HF was lower, but not significantly, in the DMD subjects compared to the healthy ones (0.064, 0.04 (SD) ABU vs. 0.10 ± 0.02 (SD) ABU; p= NS). LF/HF ratio was significantly higher in the DMD (5.27 ± 2.36 vs 0.65 ± 0.39, p<.05). Conclusions: The increased HR in the DMD subjects is a result in line with previous findings. The markedly increased LF/HF ratio in the DMD group indicates a disturbance in autonomic balance characterized by either an increased sympathetic activity or, more likely, by a decreased parasympathetic modulation. These preliminary results support the hypothesis of cardiac impairment risk in DMD.

P1027

Pericardial effusion associated with hypothyroidism in children

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Hypothyroidism develops in case of inadequate hormone production by the thyroid gland or in case of an inadequate response of thyroid gland to thyrotropin. In thyroid hormone deficiency sometimes a disorder of the cardiovascular system may dominate. The authors described in two patients the clinical picture of two girls with severe hypothyroidism where cardiovascular symptoms predominated in the clinical picture. They paid special attention to the diagnosis and causal treatment of the disease. In the conclusion the authors discuss the causes of development of hypothyroidism in childhood and draw attention to the importance of collaboration of the pediatric endocrinologist and cardiologist in the diagnosis and treatment of disease.

P1028

Late anthracycline cardiotoxicity after childhood cancer. Prospective longitudinal assessment

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More young adults are survivors of childhood cancer than of any congenital heart lesion. Most survivors have received anthracyclines (A) and reports suggest that more than half have late cardiac abnormalities. The literature on cardiotoxicity relates to cross sectional studies of heterogeneous diagnostic groups. We assessed 120 ALL survivors (age at treatment 4.7±2.8 yrs; A dose 90 to 270 mg/m2; follow up 6.2±2.0 yrs) and 97 Wilms tumour (WT) survivors (age at treatment 4.1±2.5 yrs; A dose 60 to 468 mg/m2; follow up 7.2±3.9 yrs) and 100 normal controls. Compared to normals, both groups had reduced fractional shortening (FS). (ALL 32.2±4.4, WT 30.8±4.9, normal 35.9±4.2%, p<0.001), accounted for by increased LV end systolic stress (LVESS) (ALL 49.4±13.5, WT 51.9±13.1, normal 42.2±9.1 g/cm2, p<0.001), whereas contractility assessed independently of loading conditions was normal. Total dose and dose intensity were risk factors for increased LVESS in WT but not in ALL. At reassessment 4.6 years later, sudden death or heart transplant had occurred in two patients. No other patient had clinical heart failure. Paired serial data revealed increasing LVESS, decreasing LV wall thickness and decreasing FS correlated with higher A dose, but not with cancer diagnosis, follow up duration, gender or growth. In the observation interval, cardiac indices seemed to improve for patients who had received lower doses and progress for those who had received higher doses with a cut-off cumulative dose of > 250 mg/m2, so that there was no deterioration in LVESS or LV contractility overall. Though a few subjects with low dose had impaired cardiac performance, the rarity of deterioration in cardiac function beyond 10 yrs follow-up after < 250 mg/m2 is reassuring. Late surveillance might focus particularly on patients who have received > 250 mg/m2.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P1029
Prediction of the risk for sudden death in young patients with hypertrophic cardiomyopathy by ambulatory blood pressure monitoring and heart rate variability.
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Young patients with hypertrophic cardiomyopathy (HCM) are at high risk of sudden death. Objective of the study is to predict the risk for sudden death of patients with HCM. We examined ambulatory blood pressure (BP) and heart rate variability in 6 patients (age 12–18y, 3 males and 3 females) and 56 healthy age-matched controls. Among the HCM patients, sudden death during follow-up, near death with ventricular tachycardia, and non-specific discomfort sensation were found each, while the rest three complained no symptom. In both cases sudden death or near death occurred in the morning. An ambulatory BP was measured for 24 hours at intervals of 15 minutes during daytime (from 6:00 to 23:00) and of one hour during nighttime. Time-domain and frequency-domain indices of heart rate variability (HRV) every one hour were determined from 24-hour Holter recordings. Ambulatory BP monitoring revealed that 3 out of the 6 patients showed a drop of systolic and diastolic pressures for 30 to 60 minutes in the morning (3 of 3) and in the late afternoon (1 of 3). All these patients were symptomatic with either sudden death, syncope, or other manifestations. Incidence of subjects with a pressure drop was significantly higher in patients than those in controls (4 of 52, p=0.0132). Analysis of HRV revealed that high-frequency power corrected by mean RR interval and low frequency power were significantly lower in patients during pressure drop periods than those of the normal controls. These data suggest that patients with HCM showed abnormal autonomic function at some times a day which overlapped with the timing of symptoms and that analysis of ambulatory blood monitoring and HRV may predict the risk of future occurrence of syncope with time of error in patients with HCM.

P1030
Echocardiographic assessment of left ventricular function in the rabbit.
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Kaplanova J., Babu A., Cech V., Kvasnicka J. Echocardiographic assessment of left ventricular function in the rabbit. Cardiac doxorubicin toxicity is a dreaded complication of cancer treatment but also a useful experimental model of cardiomyopathy. In small animals with high heart rates, the use of ultrasound examination is limited by the need of the high sampling frequency (and cost) of the equipment (UE). In this pilot study we tried to find out whether an inexpensive UE could be used in doxorubicin studies in the rabbit. WINCMED CSM 800 with the sampling rate of 47 frames/s (fps) was used. The rabbits were sedated by ketamine and examined using the 5 MHz probe. The data from the UE were captured by an ATI All wonder" video capture card enabling a capture frame rate of 30 fps. These videos were then saved onto Compact Disc-Recordable (CD-R) media, reviewed frame by frame with the help of Video Snap" software package (shareware license) and evaluated by help of the Osiris" software (freeware license). In order to overcome a possible error caused by the frame rate discrepancy between the UE fps and fps of ATI capture card, three diastolic and corresponding systolic images were randomly extracted from various cycles. Ejection fraction (EF) was calculated from manually indicated areas. In 9 animals of the mean weight of 3.21±0.17 kg and heart rate of 230±11/s the left ventricular (LV) end-diastolic volume was 2.34±0.47 ml, end-systolic volume 0.65±0.21 and EF 70.3±6.1. In the first animal treated by 8 doses of doxorubicin 3mg/kg/week the respective values of the LV parameters were 2.5, 1.5 and 45. The obtained data suggest that measurement of the LV function in the rabbit by inexpensive UE and widely available software could provide data, comparable with those, obtained by highly sophisticated and more appropriate techniques.

P1031
Cardiac Troponin T: It's role in the diagnosis of clinically suspected myocarditis and chronic dilated cardiomyopathy.
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Objectives: The study objectives were: 1) to compare the cardiac troponin T (cTnT) in the clinically suspected or biopsy- proved acute myocarditis and chronic dilated cardiomyopathy as pediatrics and 2) to find out whether cTnT could replace endomyocardial biopsy. Background: Myocarditis and dilated cardiomyopathy are clinically difficult to differentiate. Endomyocardial biopsy is seemed to be useful. However, its inconvenient, some risk especially in pediatric patients, limitation in sensitivity and time consuming make the endomyocardial biopsy less valuable. Noninvasive, prompt, sensitive and inexpensive means to diagnose acute myocarditis are interesting. Methods: Every cases with clinically suspected myocarditis, dilated cardiomyopathy, and 15 cases with moderate left to right shunt and CHF (group 3) who were presented at Dept of Pediatrics, Siriraj Hospital, Mahidol University, Bangkok, Thailand during July 1999 – June 2000 were included. History, physical examination, ECG, CXR, echocardiogram, cTnT, CK-MB mass and/or endomyocardial biopsy were studied. Gold standard to diagnose myocarditis are endomyocardial biopsy (Dallas criteria) and/or recovery from cardiovascular problems within 6 months of follow up period. Results: Nine patients were diagnosed myocarditis (group1) and 10 were dilated cardiomyopathy (group 2). Mean serum cTnT were 0.49 +/- 1.09, 0.13+/- 0.31, and 0.01+/- 0.01 ng/ml in group 1, 2 and 3 respectively. The mean CK-MB mass level for group 1, 2 and 3 were 24.64+/- 20.61, 14.64+/- 15.06 and 2.88+/- 1.69 ng/ml. Both cTnT and CK-MB mass levels were statistically higher in group 1 than 2, group 1 than 3 but insignificantly higher in group 2 than 3. Four cases in group 1 had endomyocardial biopsy and 50% found evidence of myocarditis. Conclusion: Measurement of cTnT and CK-MB can probably differentiate myocarditis from dilated cardiomyopathy. Histology might not be necessary for the diagnosis of myocarditis in pediatric patients.

P1032
The role of Fas/FasL and apoptosis in the development of virus myocarditis in mice
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Methods: twenty Balb / c mice/group inoculated with Coxsackie virus B3 and five mice/group injected with saline were sacrificed at 7, 10, 14, 21 and 28 days post-inoculation (p.i.). Terminal transferase-mediated dUTP-fluorescein nick end-labeling (TUNEL) assays were used to detect apoptosis in myocardium. The expression of Fas and FasL protein and mRNA in myocardium were determined by immunohistochemistry, reverse-transcription polymerase chain reaction and in situ hybridization, respectively. Results: The percent of apoptosis myocytes increased significantly after the infection 7 to 14 days than 21 to 28 p.i. (P<0.05). Fas mRNA and protein expressed mainly in myocytes and FasL mRNA and protein expressed mainly on infiltrating lymphocytes increased remarkably from 7 to 14 days compared with control group (P<0.01). Conclusion: cytotoxic T lymphocytes mediated apoptosis in myocardium through Fas / FasL pathway might play an important role in the development of VM.

P1033
The normal value of Doppler tissue image (DTI) and the usage in dilated cardiomyopathy in children
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The cardiac muscle movement velocity of systolic early diastolic and late diastolic stage of mitral valve, left ventricular posterial wall (LVPW), cardiac apex (CA) and ventricular septum were measured of 203 normal children from newborn to 14 years of age by using DTI. The five rules were found: (1) The velocity increased with age. (2) The velocity was not influenced by heart rate. (3) The velocity of endocardiac muscle was faster than that of systolic stage. (4) The velocity of diastolic stage was faster than that of CA. (5) The velocity of LVPW was faster than that of ventricular septum. The DTI of 40 cases with dilated cardiomyopathy (DCM) were measured. Early diastolic velocity of mitral valve ring (MVRDmV) was decreased in 100%. The other cardiac functions such as ejection fraction shortening fraction, cardiac index, peak velocity of pulmonary artery and aorta were also measured and compared with the change of DTI. The DTI change was more sensitive and reliable than other cardiac function test.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P1034

Inherited metabolic diseases and heart disorders in children

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Metabolic diseases are a number of metabolic derangements resulting from inherited disorders. Although systemic, cardiac involvement is poorly described. We describe heart disease in patients with lysosomal storage disease (LSD), mitochondrial disease (MD), and peroxisomal disease (PD) diagnosed according phenotypic, biochemical and enzimatic studies. 132 patients with LSD, 3 with MD and 2 with PD were investigated and 25/132, 3/3 and 1/2 respectively had clinical evidence of heart disease corroborated by color Doppler echocardiographic. LSD were classified in group 1, mucopolysaccharidosis (MPS): MPS II, 5/11, MPS III, 4/8, MPS IV, 3/6. Group 2, salidosis (S): SII, 1/1. Group 3, gangliosidosis (GM): GM I, 1/3; GM II (Sandhoff disease), 3/92. Group 4, mucolipidosis (ML): ML II, 1/2, ML III, 2/2. Group 5, sphingolipidosis Niemann-Pick type A, 1/1. Group 6, glucogenosis (G): G II (Pompe's disease) 2/3. In LSD 19/25 patients had myocardiopathy (MCP) hypertrophic 17, dilated 2; valvulopathy (V) isolated 4/25 and associated with MCP 10/25; mitral insufficiency was present 2.3 times more often than aortic insufficiency. Group 1 (MPS), group 4 (ML) and group 6 (G) cardiac compromise was more common than the others groups. Sandhoff disease, prevalent in a region of Cordoba, 2 patients had congenital heart disease (CHD) (ventricular septal defect and coarctation of the aorta) and 1 patient had dilated MCP. MD were three different disorders, 1 patient with mitochondrial disease associated with MELAS mutation and a Barth syndrome-like illness, had dilated MCP and pulmonary hypertension. 1 patient had COX deficiency with hypertrophic MCP and 1 patient with MERRF-MELAS overlap with mitral insufficiency. PD were 2 patients with punctate condrodysplasia, 1 of them had CHD (major aorto pulmonary collateral). Metabolic heart disease was present in 29/137 (21%) patients and 21/30 (70%) had MCP, 15/30 (50%) had V and 3/30 (10%) had CHD. These observations sugest that metabolic studies should be performed in all children with cardiomyopathy as the prevalence of metabolic disorders is high in this population. This may help to define therapeutic strategy and to improve genetic counseling.

P1035

Endomyocardial fibrosis in children

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Ten children with endomyocardial fibrosis(emf) submitted to surgical treatment between 1978 and 1997 are described. Seven were male and 3 female ranging in age from 4 to 15 years(mean 11). All were in the final stage of heart failure. Three had biventricular disease, 6 had involvement of the right ventricle alone and 1 had emf confined to the left ventricle. There were 2 deaths (30%) in the postoperative period due to low cardiac output. The 7 survivors were followed up for a period ranging from 12 to 166 months (mean 72 months). Two late deaths have occurred resulting from heart failure and infectious endocarditis. Five (50%) children were alive. Two required 3 reoperations for valve prosthesis dysfunction. One patient is in functional class IV and 4 in class II to III despite intensive medical treatment. It is concluded that surgery for emf is an essencially palliative procedure and, especially in children, results of surgical treatment are far less satisfactory than desired.

P1036

Plasma brain natriuretic peptide(BNP) level in patients after Fontan type operation

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Plasma brain natriuretic peptide (BNP) level in patients after Fontan type operation. Background. BNP is secreted from the ventricle and is elevated in variety of cardiovascular pathophysiological states. A study showed that plasma BNP level was positively correlated with atrial pressure in adult patients with heart failure, suggesting that BNP is secreted from the atrium. Purpose: Since right atrial pressure is elevated and its volume is increased in patients after Fontan type operation, we hypothesized that plasma BNP level may be high which is brought about from its secretion from the right atrium. Thus, we studied to confirm our hypothesis. Methods and results: Subjects were 24 Fontan patients, including 21 patients after direct atrial-pulmonary anastomosis. On postoperative cardiac catheterization, we sampled blood from SVC, IVC, PA, PV and Ao and measured BNP levels in each sample. We studied the relationship between hemodynamic data and BNP levels. BNP levels in Ao ranged from 8.4 to 569 with a mean of 106 pg/ml. Atrial and ventricular filling pressure averaged 12 mmHg and 7 mmHg, respectively. End-diastolic volume and ejection fraction of the main ventricle averaged 98% normal and 0.58, respectively. BNP levels increased at PA above the mixed vein, and at Ao from PV. BNP level at Ao was positively correlated with atrial pressure. In 3 patients with TCPC, BNP levels did not increase between mixed vein and PA. Conclusion: 1) Plasma BNP level in patients with Fontan operation is high, and it has no relation with ventricular function. 2) BNP is secreted not only from the ventricle but also from the atria, and plasma BNP level increases with high atrial pressure.

P1037

The epidemiology of childhood cardiomyopathy

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Background: Basic information about the epidemiology of childhood cardiomyopathy (CM) would assist in understanding possible aetiologies and planning of medical services for this group of patients. Methods: The National Australian Childhood Cardiomyopathy Study is a population-based study which includes all children in Australia with primary CM who presented at 0–10 years of age between the years 1987–1997. Cases were collected from all pediatric cardiologists and pediatric cardiac centers as well as from adult cardiologists, regional paediatricians, cardiac transplant centres and coronial records. Study performa were completed by the same 3 investigators who undertook a series of site visits to each centre and viewed all available medical records and cardiac imaging. Cases were classified according to WHO guidelines. The mean annual incidence for each CM type was obtained by dividing the mean number of newly diagnosed cases each year during the study period by the mean at-risk population during this time based on data obtained from the Australian Bureau of Statistics. Results: A underlying infective, genetic, syndromal or metabolic explanation was available for up to 60% of dilated CM cases who had early cardiac histology available. 54% of hypertrophic CM cases, 22% of restrictive CM cases and 54% of unclassified CM. Conclusion: The peak incidence for all childhood CM except restrictive CM is during the first year of life. The early onset of these conditions suggests a genetic, rather than acquired aetiology, in the great majority of cases.

P1038

Carvedilol for myocardial failure in pediatric patients: Do infants and young children behave like little adults?

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Due to the maturation of the myocardium and the systems involved in drug metabolism and elimination, young children with organ failure may show a different response to the β-receptor blocker carvedilol (C) compared to adolescent patients. We performed a prospective, open study using C in addition to digoxin, ACE-inhibitors and diuretics in pediatric/adolescent patients of different age with heart failure and investigated clinical, pharmacogenetic and pharmacokinetic parameters. Six infants/children (6 weeks to 4 y) and 6 adolescents (12 to 19 y) received increasing doses of oral C (initially 0.09 followed by 0.18, 0.35 and 0.70 mg/kg/d/day). Ejection fraction (EF) was determined before and monthly up to 6 months. Pharmacokinetic parameters and heart rate were measured at 15 predefined time points and pharmacogenetic testing was performed for CYP2D6 before C therapy. In adolescents, EF over 6 months was initially depressed, dropped below baseline values after 1 month, improved with a delay and increased over baseline after 4 months. In contrast, in infants/children EF increased immediately without initial depression from baseline 1 month after onset of C. Pharmacogenetics revealed no differences between both groups but pharmacokinetic/pharmacodynamic modeling using heart rate showed that infants/children had an increased sensitivity (EC50: 4.1 ± 2.02 versus 10.9 ± 1.97 ng/ml; C: $p<0.05$) and a comparable efficacy (Emax: 23.5 ± 7.81 versus 21.1 ± 10.0 l/min/m²) towards C heart rate reduction. Therefore, as maximal heart rate reduction occurs at lower C concentrations in infants/children, an earlier protection from excessive adrenergic stimulation may lead to an earlier improvement of ventricular function than in adolescents. We conclude that infants/children with myocardial failure benefit from β-receptor blocking therapy mainly because they experienced early ventricular improvement without initial myocardial depression.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P1039

Influence of exercise on QT dispersion in children previously treated with anthracycline

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We sought to clarify the serial changes in QT dispersion (QTD) during exercise in children previously treated with anthracycline. According to the progress in the treatment for malignant lymphoma or acute leukemia, evaluation of the risk factors from cardiovascular viewpoint is inevitable. Seven patients aged from 6.9 to 17.8 years (median, 9.6 years) were compared with 12 controls aged from 6.6 to 15.3 years (11.0 years). All of the patients treated only by chemotherapy including anthracycline ranged 165 to 450 mg/m2 in dose-calculation. The Sycaptom-limited cycle ergometer exercise testing was performed with ramp incremental protocol. QTD was assessed with a simultaneously recorded 12 leads electrocardiogram at four different points: rest, during exercise (at the maximum heart rate), 2 minutes after exercise, and 5 minutes after exercise. QTD in control group significantly decreased during exercise compared with the value at rest, then returned to the initial value by 2 minutes after exercise (40.0±8.9 at rest, 29.3±5.2 ($p < 0.0001$) during exercise, 39.0±5.7 at 2 minutes after exercise, 36.7±5.9 at 5 minutes after exercise). QTD in patient group, however, showed no change during exercise but increased at 2 minutes after exercise and returned to the initial value by 5 minutes after exercise (44.0±4.0, 45.1±8.3, 61.1±12.2, 44.0±17.4). As the results, the values during exercise and at 2 minutes after exercise were significantly higher in the patient group, although there was no difference on the initial values between the groups. Our results demonstrated that the patients treated with anthracycline probably remain some degree of heterogeneity in the myocardium regardless of their symptomatology. Therefore, periodic cardiovascular evaluation should be considered for all such patients.

Cardiac Imaging: CT, PET, MRI-MRA

P1040

Systemic-to-pulmonary collateral blood flow in congenital heart disease with decreased pulmonary blood flow: usefulness of CT evaluation

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Purpose: To evaluate the relationship between decrease in pulmonary blood flow(PBF) and increase in systemic-to-pulmonary collateral blood flow in congenital heart disease(CHD) with decreased PBF using computed tomography(CT). Subject and Methods: 11 patients(7 TOF, 2 pulmonary atresia and 1 interruption of right pulmonary artery) and 22 age-matched healthy persons were included. The cross-sectional area(CSA) ratio of pulmonary artery(PA) to pulmonary vein(PV) at each lung, defined as PA/PV ratio, was calculated from the diameters measured on CT. The ratio below [mean PA/PV ratio-2SD] of controls was considered as criteria of decreased PBF and increased pulmonary venous flow through systemic-to-pulmonary collaterals in patient group. The collaterals were classified as bronchial artery(BA), intercostal artery(IA) and internal mammary artery(IMA). The CSA of collaterals were recorded also. Difference in terms of PA/PV ratio between patients with collaterals and without those was assessed. Relationship between the PA/PV ratio and CSA of collaterals at each lung was evaluated. Results: Of 22 lungs of patient group, 15 lungs were met with the criteria defined above. Of these, the BA at 14 lungs(73%, 3.9 mm2 to 38.9 mm2), the IA at 9 lungs(60%, 0.8 mm2 to 63.6 mm2) and the IMA at 7 lungs(47%, 7.1 mm2 to 28.3 mm2) were found. The PA/PV ratio between patients with collaterals and without those was different for all 3 types of collaterals($p < 0.05$, t). Reciprocal relationship between PA/PV ratio and CSA of collaterals was revealed as: BA($r = -0.642$), IA($r = -0.638$) and IMA($r = -0.475$). Excellent reciprocal relationship between the PA/PV ratio and sum of CSA of collaterals at each lung was demonstrated also($r = -0.864$). Conclusion: CT may be useful to serve the semiquantitative information for systemic-to-pulmonary collateral blood flow in CHD with decreased PBF.

P1041

Spiral ct staging of pulmonary arteries in patients with complex congenital heart defects.

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Purpose: Evaluation of the usefulness of spiral CT in diagnostics of anomalies of pulmonary artery branches in patients with complex heart defects. Methods: 7 patients, aged 4–45 years, mean 19,4 years, with complex cyanotic heart defect associated by anomalies of pulmonary arteries and without ASD were examined. Besides routine non-invasive investigations, heart catheterization was performed. For difficulties with performance or interpretation of pulmonary angiography, spiral CT was done. Results: In 3 patients with complex cyanotic heart defect CT excluded agenesis of LPA or RPA and performance of Blalock-Taussig shunt was possible. For unfavorable spiral CT diagnosis in 4 cases (3 patients – hypoplasia of pulmonary branches, 1 patient with pulmonary atresia III type, VSD, hypoplasia of LPA and RPA as well as MAPCAS with HP) palliative surgical treatment wasn't undertaken. Conclusion: Spiral CT imaging is valuable in the diagnostics of pathology of pulmonary arteries in patients with complex heart defects.

P1042

Progression of myocardial ischemia and fibrosis in the pediatric patients with hypertrophic cardiomyopathy. Serial evaluation using electron-beam ct

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To study the development of myocardial damage in young patients with hypertrophic cardiomyopathy (HCM), serial electron beam CT (EBCT) findings were retrospectively analyzed. Nine consecutive patients (six males and three females) with HCM were serially examined with EBCT. Mean age and the follow-up period (±SD) were 14.5±3.8 and 4.3±2.2 years. The scanner used is IMATRON C-100 or C-150 (Imatron CA). The scan was performed at the pre-contrast phase and in early and late phases after an intravenous injection of contrast medium. Incremental CT numbers (increase of CT number from pre-contrast level) were measured at the lumen and the myocardium by setting a region of interest. The early and late M/L (mean myocardial CT numbers in the left ventricular myocardium and lumen) were analyzed as a parameter for quantitative assessment of myocardial enhancement. The area of early defect (ED: early M/L<0.25) and late enhancement (LE: late M/L>0.80) were assessed as the findings of the myocardial ischemia and fibrosis. In the initial EBCT study, ED were detected in all patients. ED were typically seen in subendomyocardial area. LE were seen in 4 patients (44%). LE were usually recognized as focal patchy stained lesions in the myocardium. In the follow-up study, all patients have ED and 8 patients (88%) have LE. Three patients progressed to dilated-phase HCM during the follow-up period. They all had marked LE in the initial study. Subendomyocardial ischemia was detected in most patients. The presence of LE by EBCT study, which was done in the early stage of disease, may be a poor prognostic factor. EBCT is an effective method to evaluate the myocardial damage in the

P1043

The use of thin-section computed tomography in the evaluation of pulmonary hemodynamics in patients with congenital heart disease

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Objective: Thin-section CT was carried out to predict the pulmonary hemodynamics and reversibility of pulmonary vascular obstruction (PVO) in 36 patients with congenital heart disease (CHD) with left-to-right. Materials and methods: According to the pattern of background lung density, the percentages of increased attenuation area (lobular and nonlobular (conformed or not with shape of lung lobule) low attenuation areas (LAA) were measured. The number of tortuous collateral-like vessels were count. The hemodynamic data obtained by cardiac catheterization were compared with the CT data. Results: The mean pulmonary arterial pressure and Rp/Rs ratio were correlated with the nonlobular LAA ($r = 0.53, 0.61$) and the number of abnormal vessels ($r = 0.38, 0.44$). The Qp/Qs ratios were significantly correlated with the increased attenuation area and lobular LAA ($r = 0.35, 0.38$). Pathologic correlation in a patient received lung transplantation revealed nonlobular LAA being intimal thickening, collateral-like vessels being dilatation lesions and lobular LAA being bronchial constriction. Conclusion: With thin-section CT, it is possible to noninvasively evaluate pulmonary hemodynamic changes in CHD patients with shunt. Non-lobular LAA and collateral-like vessels in CT were presumed to be the portion of irreversible PVO. CT is helpful for discriminating inoperable cases due to PVO and for selecting the surgery site in controversial cases with left-to-right shunt disease.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P1044

Three-dimensional CT angiography of aortic arch anomalies in neonates and infants with congenital heart disease.

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Objective: The purpose of our study was to investigate the usefulness of three-dimensional (3D) helical CT in the evaluation of aortic arch anomalies in neonates and infants with congenital heart disease. Materials and Methods: Forty two patients were examined with helical CT angiography. They ranged in age from 4 days to 18 months (median age, 2.4 months) and weight from 2.4 to 8.2 kg (median weight, 4.1 kg). Image acquisition was performed after sedation with oral chloral hydrate. 3D images were rendered using the multiplanar reformations, maximum intensity projection, and shaded-surface display. All images were reviewed for various types of aortic arch anomalies, the shape and spatial relation of the aortic arch, central pulmonary arteries, and the parent duct or arteriosus. Surgical confirmation was available in all patients. Results: In all 42 patients 3D CT angiography showed congenital abnormalities of the aortic arch such as the following categories: coarctation of aorta in 25 patients, right aortic arch with aberrant left subclavian artery in 6 patients, left aortic with aberrant right subclavian artery in 3 patients, interrupted aortic arch in 3 patients, double aortic arches in 2 patients, right aortic arch with left descending aorta in 1 patient, persistent fifth aortic arch in 1 patient, and aortic atresia related with hypoplastic left heart syndrome in 1 patient. Three-dimensional rendition demonstrated the abnormal aortic arch and spatial relation of the great arteries. Conclusion: 3D CT angiography is a relatively noninvasive, readily available imaging technique and can be a primary diagnostic modality for the evaluation of aortic arch anomalies in neonates and infants with congenital heart disease.

P1045

Evaluation of stenotic lesions in patients with congenital heart disease before balloon angioplasty using enhanced CT imaging

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Objective: The purpose of this study is to clarify the usefulness of enhanced CT in the evaluation of stenotic lesions in patients with congenital heart disease (CHD). Patients: Thirteen patients with CHD (TGA 3, TOF 3, CoA complex 2, DORV 2, SRV 2, HLHS 1) were enrolled in this study (mean age was 4.7 years old, range: 0.5 to 12.5). They all needed balloon angioplasty for stenotic vessel lesions (BT shunt stenosis 2, PA branch stenosis 8, Re-stenosis of CoA 2, PV stenosis after Fontan procedure 1) after the surgical interventions. Methods: The CT equipment was Toshiba X-viger TSX-012A for dynamical CT scanning. Total dose of 2.0ml/kg Iopamidol was injected in 30 seconds. Scanning was started 5 or 10 seconds before the first point of injection and obtained at 2mm slice thickness. All patients underwent balloon angioplasty. The morphology and diameters of stenotic lesions were assessed by angiography and compared with CT imaging. The images of post interventions were also obtained in 7 patients. Results: All 13 stenotic lesions were clearly visualized on horizontal view. There was a good correlation between the diameters obtained by CT and by angiography ($r=0.74$, $p<0.01$). Post-interventional images obtained by CT in 7 patients also showed good correlation with those by angiography ($r=0.84$, $p<0.05$). Conclusions: Enhanced CT imaging was an useful modality to assess vessel anatomy before balloon angioplasty of patients with CHD. It was also suitable for repeated follow-up studies because it is noninvasive.

P1046

Evaluation of the pulmonary venous connection with the atrium in the visceral heterotaxy by the helical ct

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Background: In the Heterotaxy heart, some patients after Fontan type operation get pulmonary venous obstruction (PVO) due to the morphological characteristics of those heart. We think that PVs connection with the atrium and the angle of PV with the atrium will cause PVO after Fontan operation, and the stenosis of PVs will occurs at the two parts, the orifice of PVs in the atrium and PVs at the back of the atrium. The helical scanning computed tomography (HCT) was analyzed about those points. Methods: enhanced HCT was performed in 32 Heterotaxy patients (asplenia 22 and polysplenia 10, aged: 0 day to 9 years). The path of PVs, the drainage patterns of the PVs connected with the heart via a single orifice in the posterior wall of the atrium, and the parallel pattern of PVs with the posterior wall of the atrium near the inferior vena cava were evaluated. Results: In 28 patients PVs from both lung couldn't behind the atrium at drained into the atrium, which are longer than normal PVs because of a long way round. In 8 patients the PVs connected with the atrial part of the heart via a single orifice in the posterior wall of the atrium is seen. The parallel pattern of PVs with posterior wall was seen in 20 patients. conclusion: It is likely for heterotaxy PVs to get obstructed mophologically if there is no problem at remaining lateral connections IVC to SVC, extracardiac conduit has to be chosen as the procedure.

Cardiac Imaging: Echo-2-D, 3-D, TEE

P1047

Evaluation of cardiac beta-adrenergic receptor responsiveness in children by DES

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Objective: To evaluate cardiac beta-adrenergic receptor(beta-AR) function and responsiveness in children. Methods: Left ventricular ejection fraction(EF),fractional shortening(FS), end systolic volume index(ESVI), ratio of systolic blood pressure and ESVI(SP/ESVI) and change of left ventricular posterior wall thickness (ΔPWT%) etc. in 30 children with beta-AR hypersensitivity, 15 children with dilated cardiomyopathy and 30 normal children were measured by dobutamine stress echocardiography (DSE). Results: Before the pharmacological stress, the values of SP/ESVI and ΔPWT% were higher in beta-AR hypersensitivity group($P<0.05$), and EF,FS,SP/ESVI and PWT% were lower and ESVI was higher in dilated cardiomyopathy group ($P<0.05$) compared with those in controls. After the pharmacological stress, EF, FS,SP/ESVI, ΔPWT% increased significantly in beta-AR hypersensitivity group, EF, FS,SP/ESVI and ΔPWT% also increased in normal group ($P<0.05$), while there values in dilated cardiomyopathy group changed little($P>0.05$). Conclusion: DSE might be used to evaluate cardiac beta-AR function and responsiveness in children.

P1048

The value of nitroglycerin echocardiography

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Objective: To study the technique of nitroglycerin echocardiography(NE) and color kinesis(CK) to assess viable myocardium. Method: 30 patients with stable coronary artery disease (CHD) were studied using NE and CK.Nitroglycerine was infused from 0.4 μg/kg.min to 2μg/kg.min consequently. Left ventricular wall motion was analyzed by dividing the ventricular wall into 16segments. And a wall motion score index (WMSI) was used. The detected hibernating myocardium segments by NE was compared with the actual improvement after coronary revascularization. Results: The results showed that after NE, the WMSI decreased from 1.57±0.27 to 1.35±0.20, ($P<0.001$).The WMSI decreased from 1.62±0.36 to 1.37±0.19,($p<0.001$) .The sensitivity, specificity, and accuracy of NE+CK in identifying hibernating myocardium were 80.95%,79.71%,and 80.53% respectively. The only hemodynamic response was a drop in systolic blood pressure(SBP) and no much change in diastolic blood pressure, heart rate(HR),and HR×BP . no patient was forced to stop the test by side effects. Conclusion: NE+CK has considerable value in detecting hibernating myocardium. NTG is useful for its accuracy and reliable, especially for its anti-ischemic effect.

P1049

Doppler flow velocity measurements to assess changes in inotropy and afterload: a study in healthy dogs

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To noninvasively assess changes in inotropy and afterload, we evaluated changes in aortic blood velocity waveform. Ascending aortic blood flow was measured by continuous wave Doppler echocardiography before and after the administration of an inotrope and a vasodilator in eight healthy dogs. Data were collected in the baseline, at three different doses of epinephrine (0.1, 0.5

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

and 1 ug/kg/min) and nitroprusside (0.4 and 8 ug/kg/min) administration, and after a simultaneous infusion of both drugs in various combinations. Epinephrine infusion caused increases in peak velocity (PV), mean acceleration (MA), velocity time integral (VTI) and minute distance without a significant change in afterload. Acceleration time (AT) and ejection time showed a slight tendency to decrease with an increase in inotropy, but with no significance. Nitroprusside infusion produced dose-dependent decreases in blood pressure and index of systemic vascular resistance (ISVR), which was associated with increases in PV, MA and minute distance, and with a decrease in AT. The combined infusion of nitroprusside and epinephrine, unless ISVR was elevated, produced synergistic effects on PV, MA, VTI and minute distance. However these Doppler parameters tended to diminish with an elevation in afterload. ISVR obtained during nitroprusside infusion had a better correlation with both PV and MA than with VTI or the Doppler time intervals. Our study suggests that Doppler measurement of aortic blood flow velocity and acceleration may be used for the noninvasive assessment of changes in inotropy and afterload.

P1050

Auto-estimation of the propagation velocity of left ventricular diastolic flow : feasibility study

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The propagation velocity (Vp) of left ventricular (LV) diastolic flow derived from color M-mode doppler echocardiography is expected to be a useful index of LV diastolic function. Current methods to subjectively determine the slope of the maximal velocity affect the accuracy of comparison. This study sought to evaluate the feasibility of auto estimation of Vp of LV diastolic flow. Software was designed to recognize different color and bright of pixel. Color doppler first aliasing technique was adopted in the study. After locating ROI at mitral valve orifice to near LV apex, the slope of the peak velocity of early diastolic flow could be obtained with custom-made software. A color M-mode doppler images of LV diastolic flow and its Vp were obtained from apical two and four chamber views in 25 patients with congenital heart disease, with mean age of 5 years old. Within 24 hours of the doppler examinations, cardiac catheterization was performed. -dp/dt and tau of LV were calculated from pressure tracings. There was no significant difference between Vp(56.25±11.5 cm/s) obtained from apical four chamber view image and Vp (57.5±10.98 cm/s) obtained from apical five chamber view image. -dp/dt of LV.

P1051

Noninvasive assessment of left anterior descending coronary artery flow reserve in normal children during supine bicycle doppler echocardiography

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Coronary flow velocity reserve (CFVR) measurement have provided useful clinical and physiologic information. CFVR is usually assessed by pharmacological interventions. However, pharmacological interventions provide only limited information on CFVR under physiological conditions. To assess CFVR during exercise, transthoracic Doppler echocardiography was performed at rest and during supine bicycle exercise. Study subjects consisted of 18 normal children (13–17 years). Echocardiographic studies (Aloka SSD Pro5mind SSH) were performed at rest and during a submaximal exercise on a supine bicycle ergometer. The electrocardiogram, heart rate, and systolic and diastolic blood pressures were monitored throughout the exercise test. Peak diastolic velocity in left anterior descending coronary artery (CFV) was recorded by pulsed Doppler under the guidance of color Doppler flow mapping. CFVR was calculated as the ratio of maximal CFV during exercise to basal CFV. Heart rate and systolic and diastolic blood pressures increased during exercise (77 ± 6 vs 137 ± 18 beats/min, 122 ± 8 vs 173 ± 26 mmHg, and 67 ± 9 vs 86 ± 9 mmHg, respectively, $p < 0.001$). CFV was recorded satisfactorily in 16/18 (89%) at rest and in 14/18 (78%) during exercise. The maximal CFV increased significantly from rest to peak exercise (27 ± 5 vs 52 ± 8 cm/sec, $p < 0.001$). Mean CFVR was calculated as 1.91 ± 0.38. In the present study using high frequency transthoracic echocardiography, we demonstrated the changes in coronary flow velocity during exercise. Success rate in the measurement of CFVR was high enough for the clinical applications. Therefore, transthoracic Doppler echocardiography has a possibility of assessing CFVR during exercise stress.

P1052

Do all patients of tetralogy of fallot (tof) need angiography before surgery?

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This study was undertaken to assess the adequacy of 2-D echocardiography in patients with TOF before total correction so as to avoid angiographic studies. Between the years 1997 -2000, 80 consecutive patients (age 8m-7y) of TOF, in whom complete anatomy could be defined on 2-D echocardiography & color Doppler studies were operated on the basis of echocardiography alone. In the initial 15 cases the findings were further confirmed on perioperative transesophageal echocardiography (TEE). Any discrepancies between the echocardiographic and surgical findings were noted. Alterations in surgical procedure and the outcome were analysed. Additional muscular VSD's were detected on intra-operative TEE in two patients, one needed surgical closure. Major abnormalities were missed on echo in 3 cases, these were single LCA and LAD from RCA in 2 and 1 case respectively. Minor abnormalities missed in 18 cases were LSVC, PDA, aberrant LAD, DPA stenosis and sternal regurgitation in MPA in 8, 5, 2, 2 and 1 cases respectively. In two cases the VSD on echo was diagnosed as perimembranous but at surgery it was doubly committed. LPA stenosis was over diagnosed in one case. In none of these cases the surgical plan or the postoperative course was altered due to the discrepancies. In our experience major abnormalities were missed in 3.5% cases only and did not result in alteration of the surgical outcome. Most of the patients of TOF in infancy and pediatric age group can be safely operated on echocardiographic studies. However if the imaging is not adequate, additional VSD, pulmonary artery anomaly or aortopulmonary collaterals are suspected angiographic studies are mandatory.

P1053

Model-based echocardiography powered by analogical cellular neural network computer (cnn-um) technology.

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Introduction: Congenital heart diseases represent complex 3D structures, therefore the reconstruction using the classical 2D or 3D methods is often difficult. The consequences for the treatment are ominous. In contrast with the conventional voxel-based 3D echocardiography, this method performs an appropriate 3D object reconstruction by storing all of the important geometric and anatomical data of the heart. The new method is presented. Method: The pixel-level information (derived from position sensed or controlled 2D image slices) is processed by the unit, based on cellular neural network (CNN) technology. CNN-UM analogical computer makes a correct object contour tracking and content-context based real time recognition from the received images. Getting data from these images, the CNN-UM develops a moving polygon based 3D model. Visualization takes place using a 3D-accelerated layout (OpenGL) of a 3D graphics workstation. Beyond the delineation, the system builds up a geometric database, which contains anatomical and geometrical information (sizes, volumes, areas without manual measurement). Using virtual reality 3D view, the specialist can view these complex structures from different views and angles, therapeutic procedures can be simulated beforehand, using even 3D models of interventional devices. Accurate quantitative measurements for the proper indication can also be achieved. Conclusions: 1.Conceptionally different from the conventional 3D imaging techniques. 2.Visualization of cardiac structures beyond the present limitations. 3.Treatment simulation before any real procedure. 4.Automatic determination of geometrical values. 5.Improvements of safety and efficacy.

P1054

Neonatal congenital heart disease: clinical and color flow mapping studies on 668 patients.

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The study was to evaluate neonatal congenital heart disease (CHD) by clinical analysis and color flow mapping (CFM), in order to investigate its distribution and to explore the relationship between it and clinical features. 668 cases of neonatal CHD detected by CFM in recent 13 years were analyzed. Among them, 512 noncyanotic CHD and 156 cyanotic CHD were found.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Incidence of cardiac CHD in both cases group were shown at below in the table. The clinical data showed that common symptoms in the noncyanotic group included heart murmur (169), tachypnea (156), cardiomegaly (148), and hypoxia (98), while in the cyanotic group were cyanosis (66), heart failure (20), tachypnea (12) and heart murmur (12), etc.. Conclusion: CFM is the most useful tool to arrive at a specific diagnosis of CHD in the neonates.

P1055

The use of exercise echo-doppler to unmask svc obstruction in post-operative congenital heart disease

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Background: The symptoms associated with SVC obstruction are often non-specific and the obstruction is difficult to diagnose because non-arterial flow and venous collaterals may keep resting pressures low. In this study we report the use of exercise (Ex) echo-Doppler to unmask significant SVC stenosis. Patients: Five patients (Age: 14–26 yrs) were studied. Four had atrial repair of TGA (3 Mustard, 1 Senning). Two of the post-op TGA patients had angioplasty and one had previous surgery for SVC stenosis. The 5th patient had repair of TAPVR to SVC/RA and subsequent surgery and angioplasty for SVC stenosis. The patients' symptoms included chest pain (2), dyspnea and wheezing (2), facial edema (2), headaches (2) syncope (1), hot upper body (1), occasional cyanosis and acrocyanosis (1). One patient had a pacemaker implanted because of his symptoms; another was seen by an ENT specialist for wheezing. Methods: Subjects were exercised submaximally (40–120 watts) on a semi-recumbent bicycle ergometer and echo-Doppler was performed. Using both suprasternal and apical views, the SVC and SVC/atrial junction velocities were obtained by pulsed Doppler at rest and peak exercise. Results: Mean SVC velocity was 1.47 m/sec (0.8–1.7) at rest and 2.19 m/sec (1.6–2.75) at peak exercise. This translates into a pressure gradient of 12 mmHg across the stenotic area. In normal subjects tested in our laboratory the peak exercise echo-Doppler SVC velocity is less than 1 m/sec. All patients had further angioplasty and/or stent placement and are clinically better. One patient had a repeat exercise study following intervention and has shown remarkable improvement with resting SVC velocities falling from 1.7 to 0.8 m/sec and peak exercise velocities falling from 2.75 to 1.4 m/sec. Conclusion: This study reports the clinical utility of submaximal exercise and echo-Doppler to unmask significant SVC obstruction.

P1056

Anomalous origin of the pulmonary artery from the aorta: echocardiographic diagnosis

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Purpose of the presentation: draw attention to the misleading clinical and echocardiographic features of anomalous origin of a pulmonary artery from the aorta. Anomalous origin of a pulmonary artery from the aorta is a rare congenital anomaly often presenting as an isolated lesion. Two-dimension (2 D) echocardiographic features have been described but misdiagnoses are not unusual. We describe 3 cases presenting in the neonatal period. Only one had a correct diagnosis made on arrival by 2 D echocardiography. In the other 2, a diagnosis of persistent fetal circulation and of pulmonary atresia, VSD and arborization anomalies of the pulmonary arteries were made. Surgical correction was undertaken in all 3 persons after the diagnosis with no early nor late mortality. All 3 pts are progressing normally at a follow-up of 1 month and 8 yrs respectively. Conclusions: anomalous origin of a pulmonary artery from the aorta is a rare condition which require very careful echo studies. Surgical correction is feasible and needs to be performed in the first few weeks of life in order to prevent irreversible pulmonary vascular disease.

P1057

The effect of transannular patch on right ventricular function after repair of tetralogy of fallot. Assessment by acoustic quantification echocardiograph

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The current trend in the surgical repair of tetralogy of Fallot (TOF) in infancy is to use a transannular patch for reconstruction of the right ventricular (RV) outflow tract. However, the influence of the transannular patch on long term survival rate is controversial. Its effect on RV function is also unclear. The purpose of the present study was to determine whether the later RV function is related to the type of RV outflow tract repair. In most previous studies on RV function, the RV was regarded as a single chamber. In this study, we evaluate the RV function by Acoustic Quantification Echocardiography dividing it into two components: the RV inlet and the infundibulum. The following parameters were obtained for each component. (1) maximal area index (Amax/m=2) (2) minimal area index (Amin/m=2) (3) fractional area change (Amax–Amin/Amax) (4) peak filling rate (dA/dtmax) (5) peak ejection rate (–dA/dtmax). The study group consisted of 24 patients, 2 to 5 years (mean 4.0) after total surgical repair of TOF and 7 normal children. Outflow tract repair was carried out in two different ways: (1) muscular resection and pulmonary valvotomy without transannular patch (n=5) (NTAP), (2) transannular patch with a homograft monocusp (n=19) (TAP). Amax/mm2 of RV infundibulum in patients with both NTAP and TAP was significantly greater compared to control subjects. Amax/mm2 of RV inlet in patients was significantly greater with TAP but not with NTAP compared with control subjects. There were no significant differences between patients and control subjects in the parameters except for Amax/mm2 of the both components. In patients, there were no significant differences in each parameter for both components between NTAP and TAP. The function of the two components of RV in patients after surgical repair of TOF is not significantly impaired or is not dependent on the type of o

P1058

Cor triatriatum in children – diagnostically difficult congenital heart defect.

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Cor triatriatum (CT) is a rare and diagnostically difficult congenital heart defect. We present 15 patients with CT aged from 0.1 to 12 (mean 3.4) years. They presented hypotrophy, recurrent respiratory infection, heart failure and presystolic murmur. Heart catheterization was performed in 10 pts, but the final diagnosis was established by echocardiography. In two pts, CT was accompanied by other cardiac malformations: VSD in one and PS in the second pt. All pts were operated on. In one additional rare anomalous pulmonary venous connection to RA was suspected but not confirmed during surgery. In another 3 pts left superior vena cava draining to the LA was found during surgery. In postoperative period died two pts – one because of coexisting malformation of central nervous system. Conclusions: Cor triatriatum is severe congenital malformation which require early surgical treatment. Echocardiography with Colour Doppler is the best method of its diagnosis. Special attention of presence of venous connections to the atria must be done.

P1059

Myocardial velocity gradient of interventricular septum and posterior wall in patients with surgically repaired tetralogy of Fallot.

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Myocardial velocity gradient (MVG) is a new index that can assess myocardial wall thickening and thinning motion independently with whole cardiac motion. Purpose: The purpose of this study is to evaluate the MVG values from patients with surgically repaired tetralogy of Fallot (TOF) comparing the results with those from normal subjects and asses the relation with the results obtained from cardiac catheterization. Methods: Study subjects consisted of 29 patients with surgically repaired TOF (aged 5 months to 15 years) and age matched 30 controls. Color coded M-mode tissue Doppler imaging were recorded on all subjects in the short axis view, and MVG in interventricular septum (IVS) and left ventricular posterior wall (PW) was calculated using a novel software (Hitachi version 1.4.7, Tokushima). Systolic thickening motion (Svg), early diastolic thinning motion (Evg), late diastolic thinning motion (Avg) were recognized on both walls, and peak values of each motion were analyzed. Results: Svg on both walls and Evg in posterior wall were lower in TOF than those in control group ($p<0.01$). On the other hand, Evg in IVS was higher in TOF than that in control group ($p<0.05$). Although left ventricular shortening fraction showed no significant relation with left ventricular ejection fraction (LVEF), Svg in PW showed positive relation with LVEF ($r=0.44$, $p<0.05$). Evg in IVS showed positive relation with right ventricular end-diastolic volume ($r=0.65$, $p<0.001$). Conclusion: These data

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

indicate that thickening wall motion in left ventricular wall is weak in TOF patients, and systolic velocity gradient in posterior wall can estimate LVEF even in the patients with right ventricular volume overload (RVVO). Besides, relaxation of IVS is affected by RVVO and the degree of RVVO can be estimated by early diastolic velocity gradient in IVS.

P1060

Transthoracic 3d-echocardiographic assessment of lvot anatomy in patients with transposition of the great arteries.

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Background: In patients with transposition of the great arteries (TGA), an associated left ventricular outflow tract (LVOT) obstruction may contraindicate the arterial switch operation (ASO). Pre-operative 2D-echocardiographic imaging does not always ensure an accurate anatomy definition of the LVOT. In addition, the Doppler-derived LVOT pressure gradient may overestimate the severity of the obstruction. 3D-echocardiographic imaging is now regarded as one of the most promising diagnostic tool to improve the anatomic definition of congenital heart malformations. Purpose: Aim of our study was to assess the feasibility and utility of transthoracic 3D-echocardiography (3D-TTE) in evaluating the LVOT anatomy and planimetric area of the pulmonary valve (PV) in patients with TGA and LVOT obstruction. Methods: In this study 6 pts (1M, 5F, age 3 days – 4 months) underwent 3D-TTE examination. Accuracy of the 3D-echo diagnosis was assessed by cardiac catheterization (5 pts) or surgery (1 pt). The suspected LVOT obstruction was related to PV anomaly (2 pts), subpulmonary stenosis from fibrous tissue tag (1 pt), mitral valve abnormality (1 pt) and stenosis at subvalvar and valvar level (1 pt). Results: 3D-TTE reconstruction was adequate in 5/6 pts. The suspected LVOT obstruction was confirmed in 5/6 pts (83%). One pt showed bicuspid PV with normal PV area. 3D-echo anatomic reconstruction accurately correlated with hemodynamic and intraoperative findings. In the 2 pts with mixed LVOT obstruction, 3D-echo accurately defined the obstructive contribution of surgically remediable lesion as well as its dynamic and fixed components. Surgical 3D-echo-guided options were adequate in all pts (4 ASO and 2 Rastelli operations). Conclusions: 3D echocardiography seems to be a very useful tool to evaluate the anatomy of the LVOT, thus improving the surgical management of patients with TGA and suspected LVOT obstruction.

P1061

Growth-related normal values of the movement of the atrioventricular ring: insight into physiologic changes of ventricular long-axis function

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Objective: To investigate the growth-related changes in atrioventricular displacement and its relation to parameters of ventricular function and geometry, we prospectively studied 195 healthy children, aged 3 months to 18 years, with two-dimensionally guided M-mode echocardiography. Methods: We measured left ventricular dimensions and shortening fraction according to the standard approach. In addition, in an apical four-chamber view, tracings of the mitral annulus motion at the left lateral (MAL) septal (MAS) and posterior (MAP) positions, and tricuspid annular motion at the right lateral (TAL) position were then obtained with simultaneous ECG and phonocardiography recordings. Results: Height correlated well with long axis dimensions both in systole and diastole (MAL r=0.9 resp. r=0.87, MAS r=0.9 resp. r=0.9, MAP r= 0.92 resp. r=0.93, TAL r=0.92 resp. r=0.93). Early diastolic amplitudes correlated linearly with height at all positions (r=0.74–0.77). Amplitudes during serial comparison showed a marked decrease during the first two years of age for all mitral positions and a further decrease for the TAL. Conventional shortening fraction of the short axis shows a low correlation with the longitudinal shortening fraction at all but the left lateral position (MAL 0.08, MAS 0.21*, MAP 0.30*, TAL 0.27*, *p<0.05). A weak correlation was found between age and longitudinal shortening fraction in the following positions: MAL -0.31*, MAS -0.35*, TAL -0.51*. *p<0.05. Sphericity index shows a parabolic correlation with height, reaching its nadir for children with a height of 120 cm. Conclusion: Long axis parameters can be measured reliably unveiling maturational changes in ventricular function and geometry. Our data suggest that this process is not complete until late childhood, when adult geometry develops. Long axis function plays an essential role during ventricular adaptation to changing load conditions.

P1062

Three-dimensional Color Doppler Echocardiography for assessment of aortic stenosis: an 6Dia vitro study

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Background: Accurate quantitative evaluation of aortic stenosis (AS) remains a challenge in the pediatric population due to the many limitations of current invasive and noninvasive methods. Three-dimensional color Doppler echocardiography (3DCDE) has the potential to measure cross-sectional effective flow area (EFA) distal to the stenotic valve and thus provide a simple and direct assessment of the severity of AS. Methods: An AS model was created by using 3 stenotic porcine bioprostheses, resembling tri-commissural, bi-commissural and uni-commissural stenoses. The prostheses were mounted in a flow phantom driven by a pulsatile flow pump. Twenty four flow states (cardiac output: 1.2 to 7.2 l/min; peak velocity: 1.44 to 6.03 m/sec) were studied. The reference effective flow area (EFA) was determined: EFA = peak flow rate by ultrasound flowmeter/peak CW Doppler velocity. The 3-D color Doppler data set was acquired using a GE SyvernV ultrasound unit interfaced with a TomTec 3-D system. The EFA was obtained by measuring the cross sectional color Doppler area of the vena contracta, the narrowest flow area distal to the stenotic valve, from the 3-D color Doppler data set. Results: the EFA by 3DCDE was observed to overestimate reference EFA (mean difference = 0.33 ± 0.28 cm2, P = 0.048) mainly due to the dependence of 3DCDE on Doppler color gain and power. Using minimal color gain and power and high flow velocity filter (18.9 cm/sec), EFA by 3DCDE correlated (r = 0.95, P < 0.05) and agreed (mean difference = – 0.02 ± 0.16 cm2, P > 0.05) well with reference EFA although underestimation was seen for small EFA and overestimation for larger EFA (Figure). Conclusion: With proper adjustment of the instrument setting, 3DCDE provides good estimates of EFA for quantitative assessment of pediatric AS.

P1063

Normal values for left anterior descending coronary artery flow velocity assessed by transthoracic Doppler echocardiography in healthy children

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Recent advances in Doppler and color echocardiographic techniques enable us to estimate coronary flow dynamics even in children. To assess left anterior descending coronary artery (LAD) peak flow velocity and to determine its relations to age and heart rate, a large number of healthy children were studied using high frequency transthoracic Doppler echocardiography. The study group consisted of 264 healthy children (1 month to 21 years old). Subjects were arbitrarily divided into 4 age group: 1 month to < 1 year (n = 52), 1 to < 4 years (n = 57), 4 to < 7 years (n = 56), 7 to < 21 years old (n = 99). LAD peak flow velocities were measured by Doppler echocardiography (Aloka SSD ProSound SSD0). LAD peak flow velocities were calculated considering the angle between the Doppler beam and the coronary flow direction. LAD peak flow velocity significantly decreased against the age (r = -0.64, p < 0.001) and increased relating to heart rate (r = 0.63, p < 0.001). Multiple linear regression analysis showed that LAD peak flow velocity was associated with age and heart rate (LAD peak flow velocity = 21 – 0.36 (age) + 0.15 (heart rate), r2 = 0.402, p < 0.001). The upper limit of normal values (+2SD) for LAD flow velocity in each age group were determined as 55 cm/sec in < 1 year, 48 cm/sec in 1 to 4 years, 42 cm/sec in 4 to 7 years, and 40 cm/sec in 7 to 21 years. As this study revealed, whenever functional evaluations of coronary lesions are conducted in children, it is important and necessary to consider the age-related coronary flow dynamics.

P1064

Myocardial performance index combining systolic and diastolic myocardial performance in doxorubicin treated patients, and its correlation to conventional echo/doppler indices

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This study was designed to evaluate the utility of myocardial performance index (MPI) in anthracycline cardiotoxicity. The MPI measures the ratio of total time spent in isovolumic activity (isovolumeric contraction time and

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

(isovolumetric relaxation time) to the ejection time, thus giving a global index combining systolic and diastolic myocardial performance. In this study, MPI was measured in 38 doxorubicin treated children (aged 108.5±55.51 months, 23 male, 12 female) in sinus rhythm, and 32 age matched controls, and was compared with conventional Doppler echocardiographic parameters. The isovolumetric contraction time was prolonged (38,37±24,45 vs 26,37±15,53, p< 0,02) and ejection time was shortened (231,91±28,87 vs 256,21±19,55 p<0,001) in doxorubicin treated patients compared with that in normal children. The isovolumetric relaxation time did not show significant difference between patients and control group(60,11±10,92 vs 61,06±12,12 p>0,05). Myocardial performance index was significantly increased in doxorubicin treated patients compared with that in control groups (0,42±0,07 vs 0,34±0,06 p<0,001), and significant correlation was observed between MPI and fractional shortening, ejection fraction, and left ventricular end diastolic and end systolic diameters(respectively, r= − 0,508 p<0,002, r= − 0,532 p<0,001, r=0,467 p<0,005, r=0,606 p<0,001). Also a weak correlation was found between MPI and duration of the disease and patient age(r = 0,393 p< 0,02, r = 0,379 p<0,02). However, there was no correlation between MPI and cumulative doxorubicin dose (r= 0,311 p>0,05) and diastolic Doppler parameters in doxorubicin treated patients. These data suggest that MPI may be a useful parameters in monitoring left ventricular dysfunction in anthracycline treated patients.

P1065

Assessment of usefulness 3D and 2D ECHO measurements in determination of ASD II dimension before interventional closure with the Amplatzer device

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The aim of the study was a verification of usefulness of 3D ECHO transthoracic estimation of ASD II dimension comparing to 2D TTE and TEE techniques, for sizing of Amplatzer device. Out of 14 children, (3 to 17 years) treated with Amplatzer device, examined using 2D TTE and TEE in 8 cases the results of 3D ECHO (TomTec Echoscan) were estimated. Results of measurements of maximal diameter in 2D were compared to calculated diameter obtained from circumference measurement in 3D and balloon stretched diameter before closure. Results: diameters were in 2D TTE 7–11 mm, in 2D TEE 9–16 mm, in 3D 10–24 mm and stretch 8–34 mm. In all but 2 cases results of 3D were in good correlation to which diameter. Underestimation in these two cases resulted from poor quality of 3D reconstructed picture. Conclusion: Calculated diameter of ASD II using ECHO 3D is useful for the assessment of the predicted waist size of the Amplatzer device.

P1066

Dobutamine stress echocardiography in children at risk for coronary ostia after surgical intervention.

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The purpose of this study was a preoperative evaluation of the left ventricular function with dobutamine stress echocardiography (DSE) in children after operations with reimplantation coronary arteries. The study group included children after arterial switch operation (21pts), Ross operation (1pts) and left coronary sy replacement (5pts). All patients underwent DSE according to the standard protocol. Dobutamine was infused in 3-min stages with doses of 5 to 40ug/kg/min and atropine 0.02mg/kg,when needed. Echocardiographic images were obtained in 4 views using 16-segment model. A positive test response was defined as a new or worsened wall motion abnormalities. RESULTS:All studies were performed without major complications.3 pts complained of palpitations, 2 pts headache,4 pts had arrhythmia.31 of 38 studies were normal, 1 were non diagnostic. In 4 pts test was positive (2 pts after Ross oper.,2 pts after ALCA) with electrocardiographic abnormalities as ST-T depression. CONCLUSIONS: 1 Dobutamine stress echocardiography is feasible,safe and well accepted technique in children. 2 This method can be used in routine follow-up in children after surgical intervention with reimplantation coronary artery.

P1067

Echocardiographic findings of the anomalous origin of the left circumflex artery from the right pulmonary artery – a case report

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The purpose of this paper is to report echocardiographic findings of a case with anomalous origin of the left circumflex artery form the right pulmonary artery. The usefulness of the echocardiography in the diagnosis of the disease is also discussed. The patient was 23 years old male. He had a history of end-to-end anastomosis of the coarctation of the aorta at 62 days of age. There had been no particular problem in the post-operative course. The treadmill test showed abnormal ST-T changes in leads II, III, aVF, V5 and V6. Thallium-201 myocardial scintigraphy at baseline and after exercise showed mild re-distribution at apex and anterior wall of the left ventricle. By the coronary angiography the anomalous origin of the left circumflex artery from the right pulmonary artery was diagnosed. By two-dimensional and color Doppler echocardiography, the proximal portion of the left circumflex artery running from the right pulmonary artery were well visualized. The right coronary artery was dilated, but the left main trunk and left anterior descending artery were normal. Myocardial contrast echocardiography using SH/TA508 showed low perfusion area at the apex and lateral wall of the left ventricle after adenosine triphosphate injection. Re-implantation of the left circumflex artery to the posterior aspect of the ascending aorta was performed. Post-operatively, the orifice of the translocated coronary artery was visualized by echocardiography. Myocardial contrast echocardiography showed no perfusion defect. These results suggest that echocardiography, including myocardial contrast echocardiography, is useful in the anatomical and functional diagnosis of the anomalous origin of the left circumflex coronary artery from the right pulmonary artery. When coronary artery lesion is suspected, it is important to evaluate the coronary arteries carefully.

P1068

Detection of intracardial stenosis necessitating revision after repairs, using intraoperative TEE in pediatric patients with complex cardiac defects

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In several reports, intraoperative TEE could detect residual abnormalities necessitating revision in about 7% of the cases that underwent intracardiac repairs. Most of them were VSD leakages and residual stenoses in LVOT or RVOT. Although intracardial stenosis is rare, we could successfully repair the stenosis based on TEE findings in 2 cases of complex cardiac defects. The purpose of the study is to demonstrate how useful and reliable intraoperative TEE can be in detecting residual abnormalities, especially intracardial stenosis. We reviewed anesthetic, surgical records and TEE findings recorded on videotapes, of 56 intracardiac repairs that managed various types of defects from October 1998 to October 2000. The studied patients ranged from 3.1 to 79.6 kg in weight and 16 days to 17 years in age. A biplane Toshiba pediatric TEE probe with shaft diameter 7.8 mm was used for most small patients less than 15 kg, and a multiplane Toshiba adult-size probe for larger patients. Revisions were required in 6 cases. In 2 cases, reasons for revisions were intracardial stenosis, and in remaining 4 cases VSD leakage or residual RVOT obstruction. All TEE diagnoses were confirmed during revisions. Only TEE could detect residual abnormalities and especially intracardial stenosis during Senning operation and TCPC. Moreover, it was remarkably effective in providing precise anatomical and flow evaluations. All patients requiring revisions showed good outcomes. No complications related to probe manipulation were seen in this study. In conclusion, intraoperative TEE is useful to identify mechanism, severity and precise locations of residual abnormalities, especially intracardial stenosis, helping surgeons to complete revisions briefly, and to confirm effectiveness of revisions.

P1069

Quantification of left atrial volume using three dimensional echocardiographic reconstruction

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Purpose: To assess the dynamic changes in left atrial (LA) volume estimated by three dimensional (3D) echocardiographic reconstruction and compare the results with those calculated by conventional M-mode echocardiography and those obtained by angiocardiography. Methods: Thirteen subjects with congenital heart disease (aged 6 months to 30 years) underwent examination by echocardiography and cardiac catheterization. 3-D reconstruction of LA cavity surfaces was performed using a novel software (Echo-pac, GE Vingmed) based on sequential image data of one cardiac cycle obtained from three apical imaging planes: four chamber, two chamber and long axis plane.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

M-mode recordings of LA dimensions were done at the level of the aortic valve and the LA volume were estimated. Results. Reconstruction of LA cavity was produced well in all subjects and continuous LA volume changes were well evaluated. Although there was a fair correlation between LA volume measured with M-mode and angiocardiography (r=0.89, p<0.0001), there was a strong correlation between LA volume measured with 3-D reconstruction and angiocardiogram (r=0.99, p<0.0001). Conclusion: LA volume estimates based on 3D reconstruction using this algorithm have less bias than those of conventional M-mode imaging. The present algorithm for 3D reconstruction facilitates a feasible and repeatable fast assessment of LA volume in the clinical setting.

P1070

Comparison of ventricular volume determination by 3D-echo, MRI, and angiography in excised porcine hearts

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Ventricular volume determination may be important for the management of patients with congenital heart disease. Three-dimensional echo (3D-echo), magnetic resonance imaging (MRI) and angiography can be used for quantifying ventricular volumes. The aim of this study was to compare the accuracy of these 3 methods in an animal model. The AV-valves of 8 excised porcine hearts were sewn up. The coronary arteries were perfused with Kaxedin fixative for elastic conservation. Ventricles were filled with different volumes (saline/contrast medium) and their real volumes were compared with the 3D-echo, MRI and angiographic measurements. For each imaging technique the volumes were calculated using a multiple slice method after manual tracing of cavity borders. 3D echo volumes correlated very well with real volumes (LV: y=1.3+0.9x, r2=0.96; RV: y=0.4+0.95x, r2=0.94) and underestimated true volumes mildly (LV: -4.8±5%; RV: -9.6±6%). These differences were independent of the transducer position. MRI was superior to 3D-echo in assessing true ventricular volumes (LV: y= 1.5+1.1x, r2=0.98; RV: y=0.1+0.9x, r2=0.96). MRI volumes were almost alike true volumes (LV: -2.9±5%; RV: 1.9±3.3%). Angiography showed considerable overestimation of true volumes and a high variability (LV: 14.4±9.3%; RV: 57.4±40%) and the correlation was less well compared to 3D-echo and MRI (LV: y=2.1+1.2x, r2=0.96; RV: y=33.6+0.7x, r2=0.62). Conclusion: RV and LV volumes can be determined with a high degree of accuracy by 3D-echo and MRI. Volume determinations by angiography were less accurate and showed the greatest variability and systematic difference from real volumes.

P1071

Quantification of pulsatile flow through major vessels using digital 3D echocardiographic video study

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Accurate non-invasive determination of regional blood flow is an important goal. We evaluated a 3D digital color Doppler technique to accurately quantify pulsatile laminar flow in an in-vitro model. We developed a model in which forward and reverse pulsatile flows of identical magnitude oscillate in a closed-circuit 20mm tube designed to mimic great vessel flow. Forward flow in one limb is reverse flow in the other. Flow was calibrated with an ultrasonic flow meter over 9 stroke volumes (15 – 55 ml/beat) at 60 beat/min. Gated 3D transcostal color Doppler imaging was performed with a 3–4 MHz multiplane TEE probe connected to an ATL HDI5000 ultrasound system. Raw Doppler scanline data were transferred to a workstation where stroke volume was calculated on a Gaussian surface perpendicular to the flow direction to compute flow forward or reverse, or both in the same image. There was good correlation between the reference data and 3D computed stroke volumes, R = 0.99 and P = 0.0001 for both forward and reverse flows. There was also excellent correlation between the forward and reverse flow matched against each other for each cardiac cycle.

P1072

Level of inhaled anesthesia affects LV function and mass measurements in an echocardiographic measured murine model

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Levels of inhaled anesthetic used during echos of transgenic murine models for cardiomyopathy may affect heart function, and thus echo measurements. Thirteen wild-type mice [C57BL/6J], ages 12–41 weeks, weighing 24.7–35.5g, were imaged by transthoracic echo with a 15MHz linear array transducer under steady state 1.5% or 2% Isoflurane sedation. LV mass was corrected for cardiac muscle density (1.055) and normalized by Meeh's formula: BSA(cm2) = 12.3 x (weight in grams)E2/3. Heart rates were slightly lower at 1.5% vs 2% Isoflurane, and LV diastolic dimension was larger (4.45 ± 0.37mm [SD] vs 4.0 ± 0.37 mm, p<0.05). Fractional shortening (37.8% ± 5.38% and 39.8% ± 5.38%, p<0.001) was significantly depressed. BSA corrected LV mass ranged from 0.67–1.17mg/cm2 and 0.89–0.79mg/cm2 (p<0.05, two-tailed t-test, paired means), an apparent increase in mass at 1.5% Isoflurane. Isoflurane's cardio-depressant effects may increase LV diameter in diastole and with extrapolation of wall thickness not detectably thinned (because of resolution problems) can yield a (most likely) artifactual change in cardiac mass.

P1073

Transesophageal echocardiography and mitral valve repair in children

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Transesophageal echocardiography (TEE) is essential for decision making and peri-operative monitoring of patients undergoing valvular heart surgery. There are very few reports on its use in children. To evaluate the diagnostic accuracy of TEE and its value on surgical decision and peri-operative management of mitral valve (MV) repair, we reviewed 47 examinations performed in 28 children (CH). Median age of the CH was 11 years (range: 3.5 y – 18 y). In all TEE, 5 chamber, 4-chamber, 2-chamber anterior, 2-chamber posterior and short-axis views were recorded. Pre-operative and post-operative TEE records were compared with trans-thoracic echo (TTE), surgical morphology and follow-up data. TEE showed rheumatic MV disease in 22 CH. 19 had mitral regurgitation (MR), 2 had mitral stenosis (MS) and 6 had mixed MV disease. Three CH had congenital MV disease and 2 had MV disease due to endocarditis. TTE diagnosis was modified by TEE in 4 CH. In all CH, TEE achieved better anatomical definition. TEE confirmed good surgical result in 22 CH, mild to moderate residual lesions in 5 and significant residual MS detected during MV repair in revision of the surgery and a prosthetic MV implantation. During follow-up by TTE, intra-operative TEE diagnosis of mild MR in 1 was changed to trivial MR in 1 CH and a more significant residual lesion was detected in 2 CH. Specificity of 95.2% and sensitivity of 83.3% was achieved by TEE. We conclude that MV repair is a good therapeutic solution in CH and TEE is very useful to guide surgical management.

P1074

Comparison of 3D versus 2D digital color Doppler methods for assessing flow volumes through the main pulmonary artery and its branches: Studies in a physiologic in vitro pontine pulmonary artery model

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We compared biplane 2D and 3D digital color Doppler (DCD) methods for calculating pulmonary artery (PA) and PA branch flow. Main PAs and their branches surgically excised from 3 pigs (48–58kg) were connected to a pulsatile pump with a reference ultrasonic flow meter. 8 flow volumes (20–55 ml/beat) were generated. An ATL HDI 5000 system with a multiplane transesophageal transducer (MTEE) was used to acquire 3D datasets on 180° rotations encompassing the main PA and branches. Images were analyzed perpendicular to the direction of flow by a Gaussian Theorem method using an SGI workstation. Orthogonal biplane 2D DCD data was obtained and analyzed on a Toshiba PowerVision with an on board ACM calculation for flow volumes. Main PA flow volumes by both 3D and biplane 2D methods correlated well with reference data (3D: r = 0.98; 2D: r = 0.92). Combined flow volumes in the right and left PA branches also agreed well with reference (3D: r = 0.97, 2D: r = 0.94). Results by the biplane 2D ACM method showed wider variability (2D: mean difference = 1.1 ± 6 ml/beat) than 3D (mean difference = 0.2 ± 2 ml/beat).

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P1075

Intraoperative transesophageal echocardiography for congenital heart surgery

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Purpose: Intraoperative Transesophageal Echocardiography (ITEE) can assess cardiac function and integrity of repair for patients with congenital heart disease (CHD). Those in favor argue that immediate corrective re-interventions can avoid the expense and hardship of a subsequent repair operation. Those against, argue that the increased manpower resources are not cost effective. We review our experience with ITEE as a perioperative diagnostic tool in patients with CHD. Methods: From 1991 to 2000, 914 patients underwent ITEE as a diagnostic adjunct to intracardiac repair. Patients were grouped by diagnosis and procedure. Patients requiring re-intervention (n=85) based on ITEE findings were analyzed. Results: Based on the findings and with the intention to improve the results, we re-intervened at the time testing in 85 cases (9%). Ten patients were returned to the operating room to repair problems, which were missed or underestimated at the initial operation. One patient suffered an esophageal perforation, and 7 had premature pacing removal due to ventricular or hemodynamic problems. Conclusion: ITEE is an effective perioperative diagnostic tool for patients with CHD. Liberal use can: (1) delineate the pre-operative diagnosis, (2) assist the surgeon during crucial parts of the operation including immediate re-intervention, and (3) assure the integrity of the repair.

P1076

Echocardiographic evaluation of bilateral patent ductus arteriosus: 15 years experience.

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Bilateral patent ductus arteriosus (bPDA) is an uncommon form of pulmonary to systemic blood supply associated with complex congenital heart disease. We retrospectively reviewed all patients with diagnosis of bPDA in our institution from January 1985 to November 2000, to describe echocardiographic findings, associated cardiac pathology, and outcome of affected patients. Eleven newborns were encountered as having bPDA (4 females, 7 males). 2D and color Doppler echocardiography identified accurately bPDA in all patients, permitting differentiation from aorto-pulmonary collaterals. Confirmation of bPDA by autopsy, angiography, MRI, or surgical inspection was available for all. Heterotaxia syndrome with complex intracardiac pathology was present in 7, pulmonary atresia with VSD in 1, and complex univentricular hearts in 3. Pulmonary atresia and non-confluent pulmonary arteries (PA) was present in 9. Of the remaining two, one had interrupted aortic arch type B and the other aortic atresia with double aortic arch (DAA). The aortic arch was left sided in 8 patients, right sided in 2, and DAA in 1. Three neonates with heterotaxia syndrome expired (complex cardiac). Surgical palliation was performed in the remaining 8. Six of the 8 died despite surgical intervention (aorto-post PA re-anastomosis and Blalock-Taussig shunt (1), Norwood operation (2) bidirectional Glenn (2), and heart transplant (1). Only 2 are currently alive, both after Fontan operation. At follow-up, 6 infants with surgical anastomosis of discontinuous PA's developed significant branch PA stenosis at the site of ductal insertion. Conclusions: Echocardiography permits accurate assessment of bPDA. bPDA is primarily identified in complex lesions associated with pulmonary atresia, non-confluent PAs, and heterotaxy. bPDA is associated with a high incidence of branch PA stenosis at follow up and a poor clinical outcome.

P1077

Diagnostic value of contrast echocardiography in the examination of congenital or acquired heart disease

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Echocardiographic contrast agent SHU 454 provides micro bubbles in a solution of galactose and SHU 508A is specially manufactured galactose microparticles and palmitic acid. Gas bubbles are known to have a limited stability in fluids. SHU 454 is absorbed in the capillaries of the lung after intravenous injection and does not reach to the left side of the heart; in contrast SHU 508A are not absorbed in the capillaries of the lung and does reach the left heart. In this study we aimed to demonstrate the role of peripheral venous injection of SHU 454 and SHU 508A in the diagnosis of heart disease. Material And Methods: Four hundred thirty patients (age range 1 month- 17 years) were involved in the study (January 1994- April 2000). The subjects had right heart lesions (270 patients; ASD,VSD, pulmonary arterio- venous fistula, tetralogy of Fallot) and left heart lesions (60 patients; aortic and mitral valve insufficiency, coronary arteria-venous fistula, coronary artery ectasia). The echocardiographic examination was performed to evaluate apical four chambers, parasternal short and long axis views. The dosages of SHU 454 were 0.5ml/kg/injection (max 10ml/ inj) and SHU 508A was 0.5-2 ml/ injection five times. Result: Each patient received single injection. SHU 454 and SHU 508A gave comprehensive information about outlet of right ventricle, pulmonary vascular structure and anatomy in tetralogy of Fallot, about surgical indication ASD,VSD, silent PDA and valve morphology in other anomalies. In conclusion: SHU 454 and SHU 508A provide better cardiopulmonary evaluation compared to conventional methods and stand as candidate to replace conventional agents, possibly to further create new diagnostic capabilities.

P1078

Longitudinal echocardiographic assessment in preterm newborns from birth till term.

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To evaluate the echocardiographic evolution parameters of preterm newborns (PTNB), adequate and small for gestational age (AGA and SGA), from birth till term, 61 PTNB, divided in Group1- 29AGA (GA:29.9+/-1.9w and BW:1296+/-223g), and Group2- 32SGA (GA:30.1+/-2.2w and BW:1047+/-214g) were prospectively evaluated through serial echodoppler-cardiographic examinations (weekly), since the 3rd day of life until term (39th week of corrected GA), and 30 term NB-AGA (control group), with GA=39.1+/-0.9w and BW=3290+/-294g, were evaluated only in the 2nd day life. Measurements were obtained, per unit of weight: aorta (AO), left atrium (LA), left ventricular dimension at end diastole and systole (LVDD, LVSD), thickness of interventricular septum and posterior wall at end diastole (IVS, LVW), left ventricle mass and volume (MASS LVV), right ventricular dimension at end diastole (RVDD) and left cardiac output (CO). Growth curves of each parameter were built based on longitudinal data analysis. The Growth curves of groups 1 and 2 were compared to each other, and at term with the control group. Statistical analysis were based on longitudinal data analysis and generalized least square technique. The significance was set at $p<0.05$. PTNB groups 1 and 2 showed a reduction of the values of AO, LA, LVDD, LVSD and RVDD, while MASS, LVV and CO increased along the postnatal period. The growth curves of groups 1 and 2 were similar except the AO measurement, which was significantly bigger in the group 2 At term, groups 1 and 2 showed significantly bigger measurements of LA ($p<0.0001$), LVDD ($p<0.0001$), LVSD ($p<0.0001$), LVV ($p<0.0001$) and CO ($p<0.0001$) than the control group. The PTNB, AGA and SGA, showed similar echocardiographic measurements along the postnatal period. However, at term they had a left heart volumetric overload and left cardiac output bigger than to an NB at birth. This suggests the presence of an hyperdynamic state in these PTNB.

P1079

Estimation of pulmonary artery pressure by contrast-enhanced Doppler signals and comparison with catheter measured pressures.

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Determination of pulmonary artery systolic pressure (PASP) is essential for the diagnosis, the time and the type of management of patients with congenital heart disease (CHD). Usually cardiac catheterization that is expensive and invasive. Determination of pulmonary artery systolic pressure (PASP) is essential for the diagnosis, the time and the type of management of patients with congenital heart disease (CHD). Usually cardiac catheterization that is expensive and invasive technique, is required for accurate measurement. A number of non-invasive methods for the assessment the PASP have been developed., one of them is the estimation of PASP using by contrast enhanced tricuspid regurgitation Doppler signals (TRDS). In the study; right ventricular systolic pressure (RVSP) and PASP of 36 patients (19 girls, 17 boys, aged 5 months to 15 years) with CHD, were estimated by TRDS before and after galactose solution (GS) and compared with catheterization measurements. Significant TRDS (+ Images) were obtained in 9 of 36 patients before GS (25 %), and 23 of 36 patients (64 %) after GS. TRDS were increased significantly by contrast agent. Estimated RVSP and PASP were significantly different from

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

the measured pressures before and after GS. There were significant correlations between the estimated RVSP and PASP and measured RVSP after GS. Estimated pressures were underestimated. In conclusion, it will be better to use the estimated PASP on the patients with significant TRJDS for the classification of PASP.

P1080

Effects of persistent ductus arteriosus over echocardiographic parameters in preterm newborns : a longitudinal study.

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PDA occurs often in PTNB and can cause serious complications along its postnatal evolution. The evaluation of some echocardiographic parameters can indicate the magnitude of the hemodynamic repercussion of PDA, being important to detect which of them are important and the possible variations in the presence of PDA. To evaluate the PDA repercussion over preterm newborns(PTNB) echocardiographic measurements along the postnatal maturation process. 61 PTNB, with mean gestational age(GA) 30 +/- 2w and mean birthweight(BW) 1,2 +/- 0,2Kg, were included in this prospective longitudinal study being accomplished serial echocardiographics examinations (weekly) since the 3rd postnatal day until term (39th week corrected GA). Measurements, per one of weight, were obtained from aorta (AO), left atrium (LA), LV end systolic and end diastolic diameter (LVSD,LVDD), interventricular septum and posterior wall thickness (SS, PW), LV mass and volume (MASS, LVV), left cardiac output (CO) and LA/AO ratio. Growth curves of these parameters were built through technique of longitudinal data analysis. In the presence of PDA the echocardiographic parameters variation were compared to the growth curve lines obtained through generalized least squares techniques, and the significance was set at p<0.05. In the presence of PDA there was a medium increase of 0,61 +/- 0,27 mm/Kg of the LA (p=0.0271), 0,88 +/- 0,26 mm/Kg of the LVDD (p=0.0154), 0,56 +/- 0,25 mm/Kg of the LVSD (p=0.0271); 0,20 +/- 0,07 mm/Kg of the SS (p=0.0088), 0,20 +/- mm/Kg of the PW (p=0.0072), and 20,3 +/- 9,0 ml/min/Kg of the CO (p=0.0701). The other analyzed parameters didn't change even the LA/AO ratio. PDA in PTNB causes a significant increase of the left cardiac dimensions and the left CO, being the early detection of these changes an indicator of treatment need, before the occurrence of severe hemodynamic repercussions.

P1081

IVUS findings of pulmonary artery and aorta in Williams syndrome

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Purpose: The vascular luminal morphology of Williams syndrome has not been well known. We studied intravascular ultrasound (IVUS) in 5 patients with Williams syndrome and 1 with isolated peripheral pulmonary artery stenosis to clarify the luminal morphology of these patients. Methods: Instruments used for this study was 0.014"200A with 3.5Fr 30MHz ultrasonic catheter(Boston Scientific Co.). Patients' ages ranged from 11months to 2years old. The IVUS findings were compared with the maximum pressure gradient of PA and Ao. Results: The maximum pressure gradient at pulmonary artery(PA) ranged 3 to 60mmHg. The maximum pressure gradient at the aorta(Ao) ranged from 0 to 20mmHg. IVUS images were obtained in 4 patients from the PA and 5 from Ao. Results: The thickness of the intima-media complex(IMC) of the PA and the ratio of IMC/PA diameter measured by IVUS ranged from 0.6 to 2.4mm and 0.07 to 0.27, respectively. The thickness of the IMC of the Ao and the ratio of IMC/Ao-diameter ranged from 0.6 to 1.2mm and 0.08 to 0.28, respectively. In PA, patients with thicker IMC have greater pressure gradients. However, in Ao, the thickness of the IMC did not seem to be related with the pressure gradient. In conclusion, thickened IMC of PA and Ao may be a primary etiology of making the stenosis rather than the secondary change from the aforehead.

P1082

Truncus arteriosus with intact ventricular septum diagnosed by echocardiography

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Truncus arteriosus with intact infundibular septum is extremely rare. We presents a unique case diagnosed by two-dimensional and color Doppler echocardiography at age of six years. The echocardiographic examination revealed situs solitus, D-ventricular loop, a single great artery arising from both ventricles with a single semilunar valve. The bicuspid truncal valve was incompetent. The truncal valve was embraced by the anterior and posterior limbs of the septal band. No ventricular septal defect was present. Two-dimensional echocardiography and color flow Doppler imaging has greatly increased the accuracy of evaluation of these cases. Now a definitive diagnosis of truncus arteriosus is feasible non-invasively.

P1083

Utility of continuous transesophageal echocardiographic monitoring in catheter interventions for congenital heart disease

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The increased utilization of therapeutic catheter interventions necessitates the need for imaging modalities that can facilitate safety and effectiveness of these procedures. Transesophageal echocardiography (TEE) has become a standard tool for intraoperative assessment of congenital heart disease. We report our experience of continuous TEE during complex catheter interventions in a series of patients with congenital heart disease. Thirty-three continuous TEE were performed on 30 patients (ages 1 day – 76 yrs, median age – 13; weight 3.2 kg – 91 kg, median weight – 42 kg.). Diagnoses consisted of secundum ASD (14 pts), VSD (6 pts), S/P Fontan operation (3 pts), PDA (2 pts), and 1 pt each with Shone's syndrome with mitral stenosis, DORV with hypoplastic LV, pulmonary atresia with intact ventricular septum, subaortic stenosis post Ratchli for corrected transposition, tricuspid stenosis and SVC syndrome. Percutaneous intervention procedures performed were ASD closure (using Amplatzer, Cardioseal, and Sideflex devices), VSD closure, Gianturco coil placement, balloon dilations (tricuspid and mitral stenosis), vein occlusion of Fontan fenestration, Cardioseal closure of Fontan fenestration, perforation and dilation of atretic pulmonary valve, stent placements in stenotic Fontan pulmonary circuit, subaortic stenosis and SVC syndrome, test occlusion of potential ASD-dependent lesions (ASD with pulmonary hypertension and DORV with hypoplastic LV). There patients had thrombus observed at the distal intracardiac end of the introducer sheath that were successfully aspirated without evident complication. Continuous TEE is a valuable tool for accurate diagnosis of size, severity, number, location of defects, and for determining presence of associated lesions. It facilitates proper catheter and device placement, providing views unobtainable from biplane fluoroscopy. Radiation time and cumulative dose of contrast media is minimized. Furthermore, potential complications are identified and prompt intervention undertaken.

P1084

Is LV dysfunction and/or hypertrophy completely reversible following adequate early repair of isolated coarctation of the aorta?

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Purpose: To investigate whether LV dysfunction and/or hypertrophy in infants with isolated coarctation of the aorta (CoA) is completely reversible following adequate repair of the coarctation during the first year of life. Methods: Conventional 2D, Doppler and M-mode echocardiography was performed on 16 patients (mean age 11.6 (SD 3.0) yrs, wt 39.1 (SD 12.0) kg, 12M) who underwent repair of CoA at a mean age of 87 days (range 0–357) and had no clinical or echocardiographic evidence of residual or recurrent coarctation. The mean follow-up period was 11.6 (SD 3.0) yrs. The results were compared with those of 20 normal controls (mean age 12.9 (SD 3.8) yrs, wt 48.1 (SD 15.9) kg, 11M). Results: All results are expressed as mean±SEM. There was an increase in LV shortening fraction (37.2±1.3 vs 32.0±0.9%, p=0.002), mitral/mitral relaxation time (58.1±1.7 vs 48.9±1.5ms, p=0.0003), peak E velocity (1.10±0.04 vs 0.89±0.05m/s, p=0.002) and peak A velocity (0.69±0.04 vs 0.54±0.03m/s, p=0.003) and a decrease in meridional end-systolic wall stress (ESWS, 60.5±2.8 vs 51.8±2.9g/cm2, p=0.010) and LV radius:thickness ratio (2.58±0.10 vs 3.18±0.11) in the patients compared to controls. There was no significant difference in the heart rate-corrected mean velocity of circumferential fibre shortening/ESWS relationship between the 2 groups. Conclusion: Whilst the patients had normal LV contractility, their increased systolic performance, reduced ESWS, mildly impaired LV filling and increased LV wall thickness suggest that LV dysfunction and hypertrophy may not be completely reversible despite adequate repair of isolated CoA during the first year of life.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P1085

Tissue Doppler echocardiography provides new insights into LV longitudinal function following early repair of isolated coarctation of the aorta.

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Purpose: To investigate whether pulsed tissue Doppler echocardiography (TDE) provides new information about LV longitudinal function following adequate early repair of isolated coarctation of the aorta (CoA). Methods: Pulsed TDE of the medial (MMA) and lateral (LMA) mitral annulus in the apical 4-chamber view and conventional echocardiography were performed on 16 patients [mean age 11.5 (SD 3.0) yrs, wt 39.1 (SD 12.6) kg, 12M] who had undergone coarctation repair. The mean age at operation was 87 days (range 8–357) and mean follow-up period 11.6 (SD 3.0) yrs. No patient had clinical or echocardiographic evidence of residual or recurrent coarctation. Comparison was made with a control group of 20 normal children [mean age 12.9 (SD 3.8) yrs, wt 48.1 (SD 15.9) kg, 11M]. Results: All results are expressed as mean±SEM. There was a decrease in the MMA peak systolic velocity (8.9±0.2 vs 7.9±0.2cm/s, p=0.006) and peak early diastolic velocity (Ea, 14.3±0.6 vs 10.4±0.7cm/s, p=0.027) and a borderline decrease in the LMA peak systolic velocity (8.9±0.5 vs 10.1±0.4cm/s, p=0.057) in the patients compared to controls. The LV shortening fraction (37.2±1.3 vs 32.0±0.9%, p=0.002) and mitral inflow peak E (110±4 vs 99±5cm/s, p=0.002) and peak A (49±4 vs 24±3cm/s, p=0.005) velocities were increased in the patient group but the E/A ratio was not significantly different between the 2 groups. The mitral inflow/annular peak early diastolic velocity ratio (E/Ea) was increased for the medial (10.1±0.8 vs 11.4±0.7cm/s, p=0.0001) and lateral (6.2±0.4 vs 8.6±0.2cm/s, p=0.0008) annulus in the patient group. Conclusion: Pulsed TDE revealed reduced peak systolic and peak early diastolic velocities of the MMA suggesting that longitudinal function of the LV may be abnormal on medium-term follow-up of children with isolated CoA repaired during the first year of life.

P1086

Dobutamine stress echocardiography in mild to moderate pulmonary stenosis

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Mild to moderate pulmonary valve stenosis (PS) is considered a benign disease with excellent prognosis. However the response of such pts to strenuous exercise, is not clearly defined. Objectives: The purpose of this study was to investigate the pathophysiologic responses of pts with valvar PS during dobutamine stress echocardiography (DSE) and possibly assess eligibility for competitive sports. Methods: We examined 20 pts, 12 male and 8 female, median age 10 years (range 4–25). Of these 15 pts had native PS, and 5 had a previous valvuloplasty procedure. Echocardiographic measurements of the peak instantaneous (PIG) mean (MG) Doppler gradient across the pulmonary valve (PV), and cardiac index (CI) were performed at baseline and after intravenous infusion of 5, 10, 20, 30, 40, and 50 mcg/kg/min of dobutamine, with heart rate (HR) and blood pressure (BP) monitoring. Data were analyzed using ANOVA for repeated measures and expressed as mean ± SD. Results: PIG and MG across the PV at least doubled during DSE. Two pts with baseline PIG of 47 and 46 mmHg, and PIG during DSE of 124 and 105 mmHg, respectively, underwent successful balloon valvuloplasty with significant decrease of both PIG and MG. Conclusions: DSE in pts with mild to moderate PS reveals significant changes in the PIG and MG. The increase in both PIG and MG measurements aid in the management of individuals with borderline indications for valvuloplasty. DSE may be useful in determining eligibility for participation in competitive sports.

P1087

Echocardiographic estimation of pulmonary arterial resistance indexes from the jet volumes of ventricular septal defect

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By color Doppler echocardiograms from the parasternal short-axial view at the aortic valve level, the shape of means from a jet of ventricular septal defect (VSD) looked as a right circular cone. The volume of the cone (V) is equal to 1/3 pi (h2-h1)(r22+r2-r1+r22), where h2-h1 is the distance between the center of VSD and the center of cone's base, while r1 and r2 are diameters of the base of cone and VSD. From nineteen patients of VSD, aged from three months to twelve years (4.61±2.41), their pulmonary arterial resistance indexes (PVRI), calculated from their catheterization hemodynamics, were 2.73±1.33 Wood·m²/m2. During the same study time, their volumes of cone from the VSD jet, were 1.75±2.06 cc. Linear regression study revealed an inversely relation as the equation PVRI=2.61−0.16*V expressed. We concluded that if the axis of cone, the real size of VSD, and temporal accordance could accurately identified, PVRI would be calculable using this inverse relation.

Arrhythmias, Electrophysiology, Sudden Cardiac Death

P1088

Acute arrhythmogenicity of chemotherapeutic agents in children

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Chemotherapeutic agents have been reported to cause severe arrhythmias and sudden death in the first 24 hours after administration. In this prospective study we determined the magnitude of acute arrhythmogenicity of these agents in children. Thirty patients with diverse malignancies [leukaemia n=15, Wilms tumour n=3, brain tumour n=3, lymphoma n=2, others n=7] were studied with Holter monitor 24 hours before, during and in the first 24 hours following the first dose therapy. Two patients experienced conduction disturbances (phases of second-degree sinoatrial and atrioventricular block) during a 4-hour period corresponding to a 30mg/m2 daunorubicin infusion. Eight patients experienced ventricular ectopy (>10 beats/hour) (VE), short salvos (4–10 beats) of ventricular tachycardia (VT) and/or short salvos (4–7 beats) of supraventricular tachycardia (SVT). Six had leukemia (therapy: daunorubicin + vincristine), one had a lymphoma (therapy: vincristine + cyclophosphamide) and the last one a brain tumor (therapy: carboplatine + procarbazine). Two patients with leukemia had posttreatment arrhythmias (1 VT and 1 SVT). The 6 other patients had posttreatment arrhythmias (1 with VT, 1 with VT, 1 with SVT and 3 with SVT and VE). No patient had life-threatening arrhythmias. Predictive factors and prognostic value of these disturbances could not be demonstrated. Conduction disturbances (2%) and arrhythmias (20%) are common respectively during and after infusion of chemotherapeutic agents in cancer children. There are no acute or long-term adverse consequences related to their appearance.

P1089

Investigation on the mapping method and the underlying mechanism of idiopathic left ventricular tachycardia in children.

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To determine the mapping method of ILVT on the curative efficacy in pediatric patients, by which to investigate the underlying mechanism. A total of 16 consecutive pediatric patients aged 2 to 17 years (mean 7.6±4.5) with idiopathic left ventricular tachycardia(LVT). 9 female and 7 male were included in this study, the tachycardia of one of which could not be induced by programmed stimulation , so the ablation was given up. The remaining 15 underwent radiofrequency ablation with radiofrequency energy in 7 of them (Group A) sites for radiofrequency energy delivery were selected on the basis of pace mapping. In the other 8 (Group B), the radiofrequency current was delivered according to the endocardial activation mapping. In all the 15 patients the tachycardia could be induced and terminated by programmed stimulation repetitively, whereas the tachycardia appeared and stopped suddenly. In 2, entrainment could be demonstrated by pacing the right ventricular apex. The ventricular tachycardia was successfully ablated in 14 of the 15 patients during the initial session. The total successful rate was 93.3%. VT recurred in 5 children, in 2 of them, VT was successfully ablated during the second session. The ablation site in the 14 children with successful ablation was located at the septum in 13 patients, and at the posterior lateral free wall in 1 patient. In 6 of the 7 children in Group A, the tachycardia was successfully ablated, the successful rate was 85.7%. In 5 of them, VT recurred, the recurrence rate was 83.3%. In all the 8 children, a very similar or identical pacing map to that of the tachycardia could be obtained in the current delivery site. The tachycardia was successfully ablated in all of the 8 children in Group B, the successful rate was 100%, none of them recurred. In this group, the successful ablation sites were characterized by a pre-existing of the P potential that preceded the local ventricular electrogram and occurred 20 to 30 milliseconds before the onset

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

of QRS during tachycardia in 4 patients. In 3 of them, current was delivered at the sites recording the P potential , in 2 ,the ablation was successful, in one, it was not. In the patient with unsuccessful ablation guided by P potential and the remained 4, it was successful when current was delivered at the site of earliest endocardial activation maximum the P potential preceding it. Pacing at the successful ablation sites during sinus rhythm produced a similar, but not identical QRS configuration to that of the tachycardia. Conclusions ? ILVT can be induced and terminated by programmed stimulation . ? Endocardial activation mapping can increase ablation success rate of ILVT effectively and prevent recurrence.? The success of the ablation a obtained at the site earliest endocardial activation preceding the QRS. Pacing at the successful ablation sites by endocardial activation during sinus rhythm produced a similar, but not identical QRS configuration to that of the tachycardia; ? The mechanism of the ILVT is assumed to be reentry involving the Purkinje fibers

P1090

Arrhythmias and sinus node dysfunction after intra-atrial lateral tunnel versus extracardiac conduit fontan procedures

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Background: Atrial arrhythmias occur frequently after the Fontan procedure. Although it has been hypothesized that the extracardiac conduit (ECC) Fontan may lead to fewer arrhythmias than the intra-atrial lateral tunnel (ILT), systematic comparisons are not available. Methods: Abnormalities from ECG and 24 hour Holters were compared between 19 ILT and 19 ECC patients. Mean age at surgery was similar 34±20 months (ILT) vs 40±19 months (ECC), however, mean follow-up was longer for ECC patients: 34±17 months vs 18±18 months (ILT). All patients had undergone an intermediate surgical stage with either a bidirectional Glenn shunt (2 in the ILT and 12 in the ECC group), or a hemi-Fontan procedure (17 ILT and 7 ECC). Results: 4/19 ILT patients (21%) and 10/19 ECC patients (53%) had sinus node dysfunction (SND) on ECG or Holter (p<0.05). Of those, no ILT, but 4 ECC patients required pacemaker placement for symptomatic bradycardia (p<0.05). All 4 of the ILT patients with SND had undergone hemi-Fontan, while of the 10 ECC patients with SND, 4 had undergone hemi-Fontan and 6 had a bidirectional Glenn. No patient in either group had documented or suspected tachyarrhythmias. Conclusions: Despite the theoretical advantages of ECC in preventing arrhythmias, in this small paired group both asymptomatic and symptomatic SND were significantly more frequent after ECC compared to ILT. Further studies with equivalent follow-up are needed to compare these 2 surgical approaches before one can be considered superior in the roles for arrhythmia prevention.

P1091

The electrocardiographic t wave as a marker of repolarization dispersion in premature infants receiving an ikr channel blocker

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Background: we set out to evaluate the T wave (T peak to T end)as a marker for myocardial repolarization dispersion in premature infants receiving cisapride, an IKr channel blocker. Methods. Sixteen non-ventilated premature infants mean gestational age 33.6 weeks (range 30.9–36) and mean post-natal age 21.7 days(range 6–51) were enrolled. A digital 12 lead electrocardiogram (Marquette 8 Cellige) was obtained prior to and 2 days after administering cisapride 0.8 mg/kg/day. Serum electrolytes were measured concurrently with the ECG recordings. The following ECG parameters were measured before and while on cisapride: QT, QTc Bazett, QT dispersion, R-R interval, Tpeak-Tend, Tpeak- Tend /Q to Tpeak, T wave area, T wave maximum voltage in Limb leads and QRS-T angle. A paired t test and analysis of variance was used to compare the variables before and during treatment. Results: expressed as before vs during therapy, mean (standard deviation): QTc 429(65)ms vs 454(29) ms p<0.001; Tp-Te/QT 0.22 0.06) vs 0.35(0.16) p<0.001. QRS-T angle: 40.8°(22) vs 43°(30); ns; T voltage: 0.24(0.08)mV vs 0.23(0.07)mV. QT dispersion: 48.9(16)ms vs 42.5(19)ms. No significant difference in T wave voltage, angle or QRS-T angle was detected with use of the IKr blocker. However, the QTc was significantly prolonged and the Tp-Te/Q to Tp ratio significantly increased. Serum electrolytes were normal in all. Conclusion: The interval delineated by the peak and end of the T wave and expressed as the ratio T peak - Tend/Q to T peak may represent regional dispersion of repolarization across the ventricular wall and may be a potentially useful clinical marker in the assessment of arrhythmic risk.

P1092

Congenital long qt syndrome with prenatal onset of ventricular tachycardia and av block

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Background. Congenital long QT syndrome with prenatal onset carries a poor prognosis. The therapy should be tailored to the defects of specific ion channels. Methods. In utero diagnosis of long QT syndrome was based on the intermittent AV block and tachycardia with AV dissociation, and positive family history. Postnatal confirmation was made in all patients. A guarded therapeutic approach was made in the last 2 patients. Results. Four patients were identified. The first two patients (case 1&2) were noted to have ventricular tachycardia and pseudo-AV block at the mid-trimester. Long QT syndrome was diagnosed soon after birth. Both received chronic propranolol (2 -3 mg/Kg/day) and pacemaker implantation, but both died during infancy. The case 3 was referred due to prenatal bradycardia and congenital ventricular tachycardia. EKG showed long QT interval and pseudo- 2:1 AV conduction. Lidocaine shortened the QT interval and resumed 1:1 AV conduction. Isoproterenol shortened the QT interval further. Therefore, the baby received high-dose mexitil only. He experienced rarely functional AV block without any ventricular tachycardia (follow up 7 months). Case 4 was referred at the 24th gestation week due to intermittent AV block and was the sibling of case1. The mother received the protocol guarded by fetal echocardiography. The fetus developed ventricular tachycardia after lidocaine and the tachycardia converted after discontinuing adenosine. At the 28th gestation week, the fetus developed ventricular tachycardia that was controlled by maternal administration of propranolol. The ratio of the duration of ventricular tachycardia to that of sinus rhythm was maintained at 10 to 15%. The baby was delivered at full term and the EKG showed corrected QT interval 0.60 sec and intermittent pseudo- 2:1 to 3:2 AV block. He subsequently received oral propranolol and experienced no ventricular tachycardia. But, the degree of pseudo-AV block aggravated at the age of 2 weeks. Nocerandil decreased the QT interval and again improved the conduction. Conclusions. Before the establishment of molecular diagnosis, selective Na channel block or blocker therapy as adjunctive K channel opener can be tailored by guarded therapeutic trial and may improve the outcome of long QT syndrome with prenatal onset.

P1093

Is there still a place for surgery in the treatment of supraventricular arrhythmias?

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Radiofrequency catheter ablation for the treatment of supraventricular tachyarrhythmias has revolutionized the management of arrhythmias with fine results. Although when the supraventricular tachyarrhythmias occur in association with congenital heart defect that need surgical correction, the treatment of arrhythmia can be considered to be done at the same procedure. We report the simultaneous surgical treatment of 2 children with atrial flutter and ASD. Two children where admitted with a diagnosis of atrial flutter and ASD. The first one was a 2 years-old boy and the second one a 6 months-old girl. Both had surgical repair which consisted of: Transatrial incision beginning at the inferior edge of the ASD to the tricuspid valve ring, transmural incision from the medial-superior edge of the ASD to the tricuspid valve ring and closure of the ASD with a patch of bovine pericardium. The children are in sinus rhythm with no recurrence of atrial flutter. The follow-up now is 10 and 4 months. We concluded that children with atrial flutter and ASD can have simultaneous surgical treatment with total cure.

P1094

Risk factors for late arrhythmias after fontan operation

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Objective. Arrhythmias (Arr) are frequent late complications after Fontan operation (FO) leading to a serious deterioration of patients' condition. However, their causes remain disputable. The aim of this work was to reveal the risk factors for late Arr. Methods. Since 1990, 128 patients were examined (range 0,5 to 22 years (mean 4.8±3.4 years) after FO. The examination

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

included repeated ECG, 24-hours Holter ECG monitoring, and cardiac catheterization. Clinical and hemodynamic parameters before and after surgery in patients with/without AR were compared. Risk factors were revealed by means of multifactor correlative analysis. Results: Different AR took place in 31 (24%) patients. In 3 (10%) of them AR continued since the time of operation, in 5 (20%) happened during the first year afterwards, in 8 (26%) in 1–5 years, and in 12 (38%) more than 5 years after surgery. AR included different ectopic rhythms (35.5%), paroxysmal supraventricular or ventricular tachycardia (16%), atrial flutter (13%), sinus or nodal bradycardia (13%), complete atrioventricular block (6.5%), frequent supraventricular or ventricular extrasystoles, as an independent rhythm disorder (16%). The following risk factors for the AR were revealed: 1. Several shunts before FO ($r=0.72$, $p<0.05$); 2. Elevated mean pulmonary arterial pressure ($r=0.71$, $p<0.05$); 3. Initial anomaly or insufficiency of atrioventricular valves ($r=0.73$, $p<0.02$); 4. Total number of risk factors for FO ($r=0.80$, $p<0.01$); 5. Aorto-pulmonary anastomosis, as a surgical method ($r=0.74$, $p<0.05$); 6. Early post-operative AR ($r=0.90$, $p<0.02$); 7. Time passed after the operation ($r=0.80$, $p<0.02$). Conclusions: Late AR after FO operation could be dependent on initial clinical and hemodynamic parameters, method of surgery, as well as the time passed after the procedure.

P1095

Rhythm disturbances after conservative surgery of ebstein's anomaly

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Background. The arrhythmias remain an unsolved problem in Ebstein anomaly. The aim of the study is to investigate the evolution of arrhythmias after surgical repair. Methods. Among 162 operated patients with Ebstein's anomaly, 85 pts presenting pre operative arrhythmias were studied. Mean age was 32±15 years. 24 (53%) had paroxysmal supraventricular tachycardia, 12 (27%) atrial fibrillation or flutter, 8 (18%) ventricular preexcitation (Wolff Parkinson-White syndrome), 1 a non sustained ventricular tachycardia. Surgical technique included detachment of the tricuspid anterior leaflet with division of the muscular bands between the leaflet and the ventricular wall, mobilisation and suture of the leaflet on the atrio-ventricular annulus, associated with longitudinal right ventricular plicature. Results: Three were late hospital deaths (3%). A pace maker was implanted in 6 patients (7%). During a mean follow-up of 57±50 months (4 to 226), there were six additional deaths, three of these were sudden. Among the surviving patients 8 (12%) continued to have symptomatic arrhythmias and 14 (21%) had a permanent sinus rhythm. Of the 24 patients with preoperative paroxysmal supraventricular tachycardia and of the 12 with atrial fibrillation or flutter preoperatively, 9 and 2 of the surviving have had no further episodes of arrhythmia. The incidence of arrhythmia with or without symptoms was reduced to 27% of the surviving patients. Conclusion: Arrhythmia is not totally abolished after surgery. Conservative surgery with anterior leaflet detachment is efficient on the accessory pathways but not on atrial fibrillation. Patients with Ebstein's anomaly and arrhythmia are clinically improved significantly after surgery.

P1096

Neonatal adenosine and verapamil sensitive ventricular tachycardia

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Purpose: Two neonates with adenosine and verapamil sensitive VT demonstrate a benign prognosis with spontaneous resolution. Case 1 presented at two days with spontaneous sustained VT. The VT did not respond to propranolol, but was terminated with adenosine. Procainamide caused transient suppression. Transesophageal electrophysiologic study (TE-EPS) could not induce the tachycardia in the baseline state or during an isoproterenol infusion. The tachycardia then remitted spontaneously and TE-EPS at that time demonstrated a sustained monomorphic (LBBB) VT. The tachycardia could be terminated with burst atrial pacing. It could not be simulated with programmed extra stimulation. The child was treated with intravenous and then oral verapamil. At one year of age, the tachycardia had not returned, the verapamil was discontinued and a repeat TE-EPS demonstrated no inducible arrhythmia. She remains arrhythmia free at 3 years. Case 2 is a premature baby boy with sustained VT which responded to adenosine. At twenty-two days of life, TE-EPS demonstrated a reproducibly inducible sustained monomorphic ventricular tachycardia of undetermined morphology. The tachycardia could be terminated with burst atrial pacing and with adenosine. The tachycardia continued paroxysmal for the next week occasionally requiring doses of adenosine for termination. The VT was then successfully suppressed with oral verapamil. At one year of age, the verapamil was discontinued; follow-up TE-EPS was normal with no inducible arrhythmia. The child has had no recurrences at 4 years. Conclusion: These two neonates with adenosine and verapamil sensitive VT demonstrate a benign clinical course with spontaneous resolution. In case 1 the LBBB morphology suggests an origin from the RV, while in case 2, the undetermined morphology does not suggest a specific origin. The VT's response to both adenosine and verapamil suggests cAMP-mediated triggered activity as the underlying mechanism.

P1097

Eag and holter monitoring in isolated congenital complete atrio-ventricular block.

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Background: Low heart rate is a frequently used indication for the need of pacemaker intervention in patients with isolated congenital complete atrioventricular block (CCAVB). The objective of this study is to compare heart rates before pacemaker implantation, between paced (PM) and non paced (NPM) isolated CCAVB patients. Methods: Retrospective evaluation of Z-scores, adjusted for age and use of atrial and ventricular (ECG), and minimal and maximal (Holter) heart rates in 149 CCAVB patients prior to PM implantation (n=113). Results: The average Z-score for the atrial rate was +0.51 (n=50) in PM and +0.60 (n=22) in NPM group (no significant difference), the average Z-score for the ventricular (average) rate was -0.91 (n=83) in PM and -0.93 (n=30) in NPM group (no significant difference). Minimal heart rate was -1.94 (n=61) in PM and -0.86 (n=25) in NPM group (not significant). Maximal heart rate was -0.51 (n=62) in PM and -0.02 (n=26) in NPM group, which differs significantly (p=0.05). Conclusion: The maximal heart rate seems to be the only reliable predictor for the need of future pacemaker intervention in the isolated CCAVB patient and should therefore be considered an indication for pacemaker implantation when significantly lowered.

P1098

Pacemaker therapy in isolated congenital complete atrio–ventricular block.

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Objective: Evaluation of the effect of pacemaker (PM) therapy on heart size and function in patients with isolated congenital complete atrio-ventricular block (CCAVB). Background: Patients with CCAVB eventually qualify for pacemaker(PM) implantation, however timing of PM implantation remains controversial. Methods: Retrospective evaluation of left ventricular end-diastolic diameter (LVEDD), shortening fraction (SF) and cardiothoracic ratio (CTR) in 149 CCAVB patients, prior to and after PM implantation. Results: LVEDD shown an average increase of 0.48 %ile/month in non PM patients (NPM), and an average decrease of 0.88 %ile/month in PM patients (PM). SF shows an average increase of 0.10 %/month in NPM, and an average decrease of 0.32 %/month in PM. CTR shows an average increase of 0.02 %/month in NPM, and an average decrease of 0.19 %/month in PM. The difference between NPM and PM is significant ($p=0.05$) for all variables. Symptomatic patients show no significant change in LVEDD after PM (from 66.5 %ile before to 68.5 %ile after PM). Asymptomatic patients do show a significant ($p<0.001$) decrease in LVEDD after PM (from 78.4 %ile before to 70.1 %ile after PM). CTR does not differ significantly between symptomatic and asymptomatic patients before PM (58 % and 57 % respectively). CTR does differ significantly ($p<0.001$) between symptomatic and asymptomatic patients after PM (52 % and 46 % respectively). Conclusions: Heart size and SF are increased in most patients with untreated CCAVB. PM implantation is associated with decrease in heart size and normalization of SF in most patients. Indications for PM therapy in children may require re-evaluation in asymptomatic patients with increased cardiac size and decreased cardiac function. While pacing may improve cardiac function, cardiomyopathy might not be prevented in a subset of patients.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P1099
Post-operative junctional ectopic tachycardia
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Introduction Junctional Ectopic Tachycardia (JET) is a transient complication occurring in the post-operative period after open-heart surgery. JET is a tachycardia whereby the QRS frequency is faster than the p wave frequency except when there is a retrograde P wave on the ECG. The frequency is higher than the maximum of the normal frequency for the age of the patient. JET is described as a life threatening complication with a high mortality rate, even up to 50%. However, there are inconsistencies in the literature concerning the outcome and mortality rate of JET. Therefore we set up a retrospective study to learn what the incidence and outcome was of JET in our centre. Setup: To determine the incidence and mortality rate of post-operative JET we examined retrospectively the post-operative records of 246 open-heart operations for congenital heart defects in patients under 18 years of age in 1998 and 1999. Results. From the 246 patients operated in this period only 15 developed a JET (6.5%). Five patients (2.2%) had a hemodynamic significant JET and needed treatment. They were treated with cordarone or propafenon. Ten patients (67%) were not treated. JET developed on the same day of surgery or the day after. The duration of a JET ranged from several hours to 5 days. No patients suffering JET, treated or not, died. Post-operative JET was strongly associated with very young age. There was no significant relation between the occurrence of post-operative JET and prolonged the bypass machine during surgery. Tetralogy of Fallot or Double Outlet RightVentricle had a high incidence of post-operative JET compared to other malformations. Conclusion: Post operative JET is not by definition a life-threatening complication.

P1100
Sinus node reentrant tachycardia in a newborn
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Comparison of troponin-T release in infants with cyanotic and acyanotic heart disease following cardiopulmonary bypass surgery. Does cyanosis cause more myocardial injury? Uzun, O., Barth, J., Parsons, J.M., Dickinson, D.F., Gibbs, J.L., Wilkinson, J.L. Leeds, United Kingdom: We aimed to define preoperative, postbypass values of troponin T in children with cyanotic and acyanotic heart disease, and its correlation with operative, postoperative recovery variables and outcome. 74 children aged 1day to 15 years undergoing cardiopulmonary bypass were prospectively studied. Blood samples were taken after anaesthetic induction, 4 hours post-bypass, then at regular intervals for a further 120 hours. Preoperative, peak and final levels of troponin T were compared between cyanotic and acyanotic patients. Troponin T showed higher values preoperatively in cyanotic and sick infants. It peaked at 4 hours, declined gradually over 48 hours, but remained detectable at 120 hours even in patients with uncomplicated recovery. Younger age, cyanosis, and decreased urine output were all correlated with higher postoperative values. Of the two infants who died, one showed highest preoperative value 0.98mcg/L, and a peak value of 14.98mcg/L, and in the other levels continued to rise beyond 48 hours. Elevated levels of Troponin T above 5mcg/L after 24 hours was associated with a longer and complicated postoperative recovery. There was no significant difference in preoperative recovery, duration of ventilation, and hospital stay between cyanotic and acyanotic patients. Preoperative detection of elevated serum troponin T levels may allow to identify high risk infants. Serum troponin T levels greater than 5 mcg/L at 24 hours postoperatively may indicate complicated recovery.

P1101
Significance of ventricular late potentials in children with mitral valve prolapse
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The purpose of this study was to determine the incidence and significance of late ventricular potentials (LP) in children with mitral valve prolapse (MVP). Methods. 151 consecutive children with MVP (12.2±3.1 years) and 164 healthy subject (12.3±3.7 years) were examined. All patients (pts) underwent 24 h ambulatory ECG monitoring and echocardiography. The analysis of time domain signal-averaged ECG (SAECG) was performed at filter settings of 25–250 and 40–250 Hz. Children with MVP were followed prospectively for a mean of 64 months. Results: Pts with MVP had a significantly higher prevalence of ventricular arrhythmias (VA) than in controls (42% vs. 13%, p<0,0001). Three (2%) pts with MVP had runs of ventricular tachycardia (VT) during 24-h ECG monitoring compared with one (0.6%) from control group (NS). Pts with MVP and VA were significantly older compared with those without VA (13.1±2.4 vs. 11.5±3.4 years, p<0.002). LP were more frequently observed in MVP group than in healthy children (17% vs. 3%, p<0,0001). Abnormal SAECG results were more common in pts with MVP and VA (27%) compared with those without VA (10%, p<0.02). There was no significant correlation between an abnormal SAECG and the presence of VT and age of pts with MVP. During follow-up, VT occurred in 24 children with MVP (3.3/100 subject-years). Fourteen of these pts had LP at SAECG. The sensitivity of LP for the identification of children with MVP who developed VT was low (54%) although specificity was high (91%). Conclusions. MVP is associated with increased occurrence of VA and LP in children. Abnormal SAECG is specific but not sensitive marker for development nsVT in children with MVP.

P1102
Qt dispersion and ventricular arrhythmias after repair of tetralogy of Fallot
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Ventricular arrhythmias (VA) are a significant cause of late mortality and morbidity following repair of tetralogy of Fallot. Abnormalities in ventricular repolarization have been studied in patients who are susceptible to severe VA. To determine whether any association exists between QT dispersion (QTd) and the presence of VA following repair of tetralogy of Fallot, we have studied a group of 74 patients (mean age = 11.3±3.9 years). Post repair follow-up time ranged from 1.7 to 16.6 years (mean = 8.1±2.9 years). VA were identified by 24h Holter monitoring and considered significant when equal or superior to Lown modified grade 2. QT dispersion was manually performed by 2 blinded observers on 12 leads ECGs and defined as the difference between the maximal and the minimal QT intervals occurring in any of the 12 leads and classified as abnormal when longer than 40 ms. All patients were in sinus rhythm. Significant VA were detected in 6 (8.1%). One patient had syncope, but no documented VA. QTd was found abnormal in 5 of these 6 patients, as well as in another 25 who did not have documented VA. Test sensitivity and specificity were respectively 83 and 63%, predictive negative value of QTd was 98%. We found that in patients operated for tetralogy of Fallot, QTd might be a valuable non-invasive marker for identifying low risk for VA.

P1103
Atrial tachycardia from enhanced automaticity in children: results of initial medical management
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Ten patients (age 0-9 years) with the diagnosis of automatic atrial tachycardia (AAT) during August 1997-August 2000 were reviewed. Three patients had paroxysmal (repetitive) AAT and the tachycardia was incessant in six (defined as presence of AAT for more than 90% of the time). The type of AAT in one patient was unknown. Four patients presented with congestive heart failure (CHF), one with pre-syncope, one with palpitation, and four were asymptomatic. Six patients (60%) had depressed left ventricular ejection fraction. All patients with CHF had incessant AAT with atrial rate > 220/min and ventricular rate > 200/min at admission. After treatment with antiarrhythmic medications, all patients had adequate control of the AAT (9 had complete elimination of AAT and 1 partial control). Amiodarone (alone, or in combination with digoxin) was effective in 5 of 6 cases (83%), although complete elimination of the AAT was usually delayed (median = 5 days, range 30 minutes to 17 days). Other effective medications were digoxin, digoxin + propranolol and steroid (all in patients who did not have CHF on presentation). At the time of this report, 3 patients had no AAT off antiarrhythmic medication, 5 patients still receive treatment (with good control) and 2 patients died from sepsis during the same admission even though AAT was controlled. All surviving patients had normal ventricular ejection fraction on follow-up. AAT in children is rare, but when it occurs in persistent form at fast rate, it is usually associated with CHF and is difficult to treat. Amiodarone (+/- digoxin) effectively controls the arrhythmia in majority of cases.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

although full effect may take several days. With successful treatment, most patients do well and some can be taken off the medication(s) without recurrence of the arrhythmia.

P1104

Sustained t-wave alternans after repair of tetralogy of Fallot.

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Objective–To determine T-wave alternans (TWA) prevalence and characteristics late after transatrial-transpulmonary repair of tetralogy of Fallot. Design–Prospective cross-sectional study. Patients– Forty-nine subjects who had consecutively undergone transatrial-transpulmonary repair of tetralogy of Fallot. Median age was 14.9 yrs(11.5–20.9). Median age at repair 1.6 years (0.2–4.9). Median follow-up post-repair was 11.6 yrs (9.4–17.2). All patients were NYHA class I. None had symptomatic arrhythmias. Methods- TWA was evaluated during bicycle exercise. Patients also had a standard ECG, chest X-ray and 24-hour Holter recording. Results. Median QRS duration was 120ms(80–150). Sustained TWA was detected in 7/31(23%) of those with adequate tests. In these 7, median onset HR was 120(98–155). Median HR threshold as a percentage of predicted maximum HR (220-age) was 58%(48–77). Sustained TWA prevalence was not significantly different compared to normals (7/31 vs 9/85,p=0.3). Onset HR in the TOF group was significantly lower: mean(SD):122(20) vs 159(12);p<0.05). In the TOF group with sustained TWA, 4/7 occurred at <60% predicted maximum HR vs 1/9 normals (p<0.05) and 5/7 had an onset HR <120 vs 0/9 normals (p<0.03). There was no significant difference in age, gender, transannular patch use, restrictive RV physiology, QRS duration, QTc, QT-QRS dispersion nor non-sustained VT detected on Holter in those with and without sustained TWA. Conclusion– Sustained TWA prevalence late after repair of TOF using minimal ventriculotomy is not significantly higher than normal. However the onset HR is significantly lower. Further study is required to determine whether or not sustained TWA at a lower onset HR indicates malignant arrhythmia risk.

P1105

Implantable loop recorders document cardiac rhythm for pediatric patients with syncope but failures can occur

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Syncope (S) of unknown cause is an infrequent but troubling problem in children. The new implantable loop (IL) recorder allows ECG documentation during symptoms. We report 7 pediatric patients, 2–16 yr, 6/7 normal structural hearts, who underwent IL procedures for one or more indications subsequent S (n=5), too young to activate standard external loop recorder (n=1), no warning before S (n=4). Standard manufacturer recommended techniques were employed to ensure optimum ECG sensor before subcutaneous implantation. During 5–180 d(mean 86) follow-up after IL 5 of 7 pts had recurrent S and in 3 of 5 a diagnosis was made [ventricular tachycardia, 1, sinus rhythm 2] by IL. In 3 pts, the automatic recording mechanism failed. The VT was recorded by pt activated mode but the auto mode failed. T wave oversensing caused inappropriate triggering of the auto recording mechanism and filling of the memory in 3 pts. Of the 2 pts in whom inadvertent pt activated events occurred, 1 had full memory which prevented a pt activated recording during S. Monitoring continues in 4 pts (1.5–6 mo after implant). Conclusions. IL are valuable for diagnosis of unexplained S in pediatric pts but the automatic activation feature appears to have limited utility. Problems of T wave oversensing and inadvertent pt activation suggests that frequent interrogation and clearing of memory is needed.

P1106

Natural history of asymptomatic Wolff-Parkinson-White syndrome in children

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Background: Although most asymptomatic patients with the Wolff-Parkinson-White (WPW) electrocardiographic pattern have a good prognosis, some die suddenly. Recently, catheter ablation therapy has been performed for asymptomatic WPW syndrome in children. Purpose: The purpose of this study is to investigate the prognosis of children with asymptomatic WPW syndrome in cardiac mass screening. Objects and Methods: All students (elementary school students 21,420, junior high school students 25,504) were performed 2 times a group of cardiac examination at 6 and 12 years old, from 1987 to 1998 in Osaka city, Japan. All students were interviewed and had physical examination and 12 lead ECG. Fifty-three patients had been diagnosed WPW syndrome at cardiac mass screening. Twenty-five patients were diagnosed at 6 years old and 28 patients were diagnosed at 12 years old. Children had been followed up from 1 to 12 years. They had been performed physical examination and 12 lead ECG every year. Results. Type A were 12, type B were 40 and type C was one patient. Results of electrocardiograph was that mean of heart rate was 74 per minute, mean PQ interval was 0.1 seconds and mean QRS interval was 0.11 seconds. Three patients (5.7%) had tachycardia events for follow-up period. However, 24-hours recordings and exercise electrocardiography examination did not demonstrated supraventricular tachycardia in two patients. One patient (1.8%) lost capacity for preexcitation and anterograde conduction over the accessory pathway, which produces the WPW pattern. Sudden death rate is none. Conclusions: These results indicated that the rates of sudden death and supraventricular tachycardia attack are low in elementary and junior high school students with asymptomatic WPW syndrome. In most patients with asymptomatic WPW syndrome in children, they may not need to have catheter ablation therapy.

P1107

Current management of supraventricular tachycardia in infants.

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The main aim of this investigation was to study effective antiarrhythmic drug therapy in neonates. The effectiveness and safety of digoxin, atenolol and sotalex, given in severe recurrences, were compared in 18 infants < 1 year (7 (38%) < 1 month) with resistant and ectopic supraventricular tachycardia between 1995 and 2000. Thirteen patients had permanent tachycardia (72%) including 8(44%) patients with atrioventricular reentrant tachycardia in WPW. Two patients were after surgical correction of congenital heart disease (arterial switch operation and total correction of anomalous pulmonary venous connection). Digoxin was the drug of first choice in 18 patients at a dose of 11–14 mg/kg/d. It was effective in 6 (44%). Sotalex was used as first line therapy after the failure of digoxin and atenolol, at a maintenance dose of 1.2–9.5 mg/kg/d and was effective in 6 (33%). Atenolol was used in 6 patients a dose of 1–3 mg/kg/d. It was effective in 3 (16%). In one patient with ectopic supraventricular tachycardia a combination of digoxin (12 mg/kg/d) and atenolol (2 mg/kg/d) was effective. Prophylactic therapy was maintained for 6 to 12 months and only 3 patients had to increase after withdrawal, one of them having a drug resistant permanent junctional reciprocating tachycardia. Patients had no ventricular fibrillation, significant sinus bradycardia or heart failure. Conclusion. sotalex was found to be safer and more effective in patients with digoxin resistant supraventricular tachycardia. Dose of antiarrhythmic drugs in the neonate was very individual. Sotalex may be proposed as first line therapy for prophylaxis of recurrent SVT combined with other premature atrial and ventricular contractions in infancy.

P1108

The diagnosis and therapy of rhythm disturbances in paediatric populations

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The aim of this study was to evaluate the frequency of SVPT diagnostic values of Holter, as well as the application of Adenosine. During 1998–2000 at Paediatric Clinic in Sarayköy 1303 pts were hospitalised. The retrospective evaluation was done on 17 (1.3%) pts with registered rhythm disturbances. 8 boys, age 1–15 years. The I Group included 8 pts with SVPT, II Group 9 pts with other rhythm disorders. The rhythm disturbances at admission have been diagnosed on ECG in 12/17 (70.5%) pts and by Holter in 5/17 (29.5%). ECG has diagnosed SVPT in 8/17 (47%) pts, 3/8 (37.5) pts at admission, and in 5/8 (62.5%) by Holter. The mean heart rate during the SVPT was 251/min (range 226–281/min). Conversion of SVPT in SR successfully has been performed in 3/8 pts by IV application of Adenosine dose 0.25 mg/kg and in 5/8 with antiarrhythmic drugs of II and IV generation. Conclusion. the dg of SVPT is possible not only by ECG but with Holter as well. The application of adenosine with the aim to convert SVPT in to SR is possible, achievable, safe and powerful.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P1109

Increased temporal qt interval variability in the patients with post-operative tetralogy of Fallot

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Recently, it has been known that increased temporal QT interval variability reflects ventricular repolarization inhomogeneity and may be a predictor of ventricular arrhythmias and sudden cardiac death. We aimed to determined whether the temporal QT interval variability changes in the patients with postoperative Tetralogy of Fallot (TOF) with or without ventricular arrhythmias. Study population included thirty one patients with postoperative TOF with no ventricular arrhythmia (arrhythmia negative group), eighteen patients with ventricular arrhythmia (arrhythmia positive group), and thirty one healthy children (control group). The 24-hour ambulatory electrocardiography were digitized by high speed analog-to-digital converter (DWT 3001, USA). We extracted arrhythmia-free electrocardiograph of ten minute duration recorded during resting midnight to 6a.m. We measured beat-by-beat R-R intervals and QT intervals using the technique of template matching strategy. The QT variability index (QTVI) was calculated using the formula, $QTVI = \log(Var(QT/Mean(QT)^2)/(Var(RR/Mean(RR)^2))$. The QTVIs of the arrhythmia positive (-0.479 ± 0.284, p<0.0001) and negative group (-0.661 ±0.267, p<0.00011) were significantly higher than that of the control group (-1.200 ± 0.373). Although the mean QTVI value of the arrhythmia positive group was higher than that of the arrhythmia negative group, it did not reach statistical significance. Conclusively, the patients with postoperative TOF, whether or not they have ventricular arrhythmias, have increased temporal QT variability and thus may have ventricular repolarization inhomogeneity.

P1110

Cardiac rhythm in right atrial isomerism

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We sought to establish in a large cohort of children with right atrial isomerism the nature of atrial rhythm and prevalence of arrhythmia. Standard baseline electrocardiograms performed in 110 patients with right atrial isomerism performed at a median age of 1 day (range 1 day to 5.7 years) were reviewed. The type, timing and precipitating factors of arrhythmia that occurred over a total follow-up duration of 2.8 (range 0.01 to 25) years were noted. All except 2 patients had a sinus rhythm. Of these, 88% (95/108) had single while 12% (13/108) had multiple P-wave morphology. For those with a single P-wave morphology, the frontal P-wave axis was 0° to 90° in 62% (59/95), >90° to 180° in 23% (22/95), and superior in 15% (14/95). There was no relation between cardiac anatomy and P-wave axis or morphology. The other 2 patients had concordantly complete heart block and junctional rhythm. Cardiac arrhythmia was documented in 16 (14.5%) patients, 14 of which had supraventricular tachycardia, and 1 each of atrial flutter and transient complete heart block. Precipitating factors included open-heart surgery (n=4) and cardiac catheterization (n=2), both were absent in 10 patients. Management included adenosine (n=8), vagal manoeuvres (n=2), digoxin (n=2), and one each of DC cardioversion, amiodarone and sotalol. Freedom from arrhythmia was 90% at one, 85% at five and 78% at ten years. Anomalous pulmonary venous connection was the only risk factor (p=0.04) for arrhythmia in patients offered surgery (n=73). Neither arrhythmia nor abnormal P wave morphology or axis is a risk factor for the 5% overall mortality. Variations in P wave morphology and axes are common in patients with right atrial isomerism. While cardiac arrhythmia is not uncommon, it does not appear to influence mortality.

P1111

Clinical efficacy of treadmill exercise test for patients with wpw syndrome

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Background: Although it is rare, WPW syndrome can be a cause of sudden death as a result of pseudo-VT caused by conduction of atrial flutter to LV through an accessory pathway. There is no reliable method by which to predict the risk of WPW syndrome. Thus, a noninvasive method that could identify an accessory pathway having a short refractory period, which allows orthodromic conduction at the time of the short R-R interval, would be useful for cases of WPW. Purpose & Method: To assess the refractory time of orthodromic conduction through an accessory pathway during strenuous exercise among children with WPW syndrome, we performed a Double Master EKG, a treadmill exercise test, and 24-hour Holter recording. Twenty-four children with WPW Syndrome, including 14 boys and 10 girls whose ages ranged from 3 to 17 years, were studied. Results: The delta wave disappeared in 19 out of 24 cases tested by the treadmill exercise test, in 4 out of 24 cases tested by 24-hour Holter recordings, and in none out of 24 cases tested by Double Master EKG. In 2 of the 4 cases of Holter monitoring in which the delta wave disappeared, the delta wave disappeared without tachycardia. Conclusion: The disappearance of the delta wave in these tests indicates that the effective refractory period in orthodromic conduction through the accessory pathway becomes longer than the R-R interval of the sinus rate during exercise. It was clear that the treadmill test was superior to the Double Masters test for assessing the effective refractory period. Twenty-four hour EKG is good for assessing the inducible refractory period of the accessory pathway, and could be useful to decrease the risk of sudden death in children with WPW syndrome. The double Master EKG does not appear useful for this purpose.

P1112

An infant case with a malignant form of brugada syndrome

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A 6 month-old Japanese infant (male), one of a pair of dizygotic twins was admitted to our hospital, because of recurrent episodes of cyanosis after crying. The other twin died unexpectedly during sleep at the age of 4 months. The 12-lead ECG at admission showed coved-type ST segment elevation in leads V1 and V2R and normal corrected QT intervals. Non-sustained polymorphic VT were recorded 734 times a day by Holter monitoring. Recurrent VF, family history of sudden death, ST segment elevation in leads V1 and V3 and absence of organic heart disease established a diagnosis of Brugada syndrome. To our best knowledge, this infant is the youngest patient reported as having Brugada syndrome. Intravenous administration of propranolol increased the amplitude of ST segment elevation in leads V1 and V2 as well as the incidence of polymorphic VT. Immediately after continuous infusion of MgSO4, polymorphic VT developed with syncope. Intravenous injection of a Class IB antiarrhythmic drug (mexiletine) did not change the amplitude of ST segment elevation or J wave, resulting in only a slight decrease in the incidence of polymorphic VT and VF. In contrast, continuous infusion of a b-adrenergic agonist (isoproterenol), as well as intravenous injection of a parasympathetic antagonist (atropine), dramatically decreased the amplitude of ST segment elevation and J wave, and totally suppressed polymorphic VT and VF. The prophylactic implantation of an implantable cardioverter defibrillator (ICD) was performed in this patient. Combined oral administration of a b1 adrenergic agonist, a parasympathetic antagonist and quinidine has successfully suppressed recurrences of VT or VF for 1 years, which may have the potential to decrease the incidence of VT or VF as an adjunctive therapy under a prophylactic implant.

P1113

Time domain heart rate variability in healthy children.

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The aim of the study was evaluated time domain heart rate variability normal values in healthy children. 364 children aged from 4 to 18 years (mean 11 +/- 4) 189 boys, 175 girls were divided in 10 groups from 4 to 12 years every two years girls and boys together and from 12 to 18 years every two years either boy separable for gender minimum 27 children in each group. The analysis was performed from 24- hours Holter recording. Following parameters were calculated (ms): mean R-R, SDNN, SDANN, SDANNI, rMSSD, (%) pNN50. Statistical analysis was performed using ANOVA procedure confirmed by LSD test. The minimal level of significance accepted was 0.05. Results: mean R-R, SDANNI increased significantly with age. SDNN, SDNNI were significantly different between the youngest and the oldest groups. There were no statistical differences in rMSSD, pNN50 between groups. Conclusions: 1. mR-R, SDANNI, SDNN, SDNNI increased with age 2. In our study rMSSD, pNN50 were independent parameters.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P1114

Spectral domain heart rate variability analysis in healthy children

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There were 369 pts aged 4– 18 yrs (x = 11±4) 189 boys, 175 girls divided in 10 groups, from 4 to 12 yrs every 2 yrs girls and boys together and from 12 to 18 yrs every two yrs girls and boys separable, min. 27 pts in each group. Spectral analysis was performed using algorythm FFT from short – term (5 minutes) Holter recording during day and night. The following parameters were calculated: ULF, VLF, LF, HF, TP, ratio LF/HF in following units: ms2, logarithm natural (ln) and normalized units LF nu, HF nu. Results. VLF (ms2), ratio LF/HF were significantly higher in older then in younger groups during day as well as at night. LF values (ms2, nu) during day and LF nu at night were statistically higher in oldest than in youngest pts. There were no statistical differences between groups in relation to TP ms2. HF in during the day, HF nu and LF nu at night, HF nu during day were significantly lower in older than in younger groups. Conclusions. 1. LF, VLF spectral power values increased with age. 2. HF spectral power values during childhood decreased after 14 years of age.

P1115

Arrest is relatively rare in infancy. Is there an anaemic explanation?

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We studied the morphology of the atrioventricular(AV)node in 40 normally structured hearts obtained at autopsy from patients without a history of cardiac arrhythmia in the following age ranges: <1 year(n=19), 1–12years(n=11), and 12–20 years(n=10). The AV septo-junctional area was removed en bloc and serially sectioned at 10–micron thickness at right angles of the AV annulus. The length of the compact node and rightward and inferior extensions were calculated. Computer-assisted three-dimensional reconstructions were made of 6 hearts. The ratio of the right extension to the compact AV node showed a statistically significant increase with age. In addition, with increasing age the geometry of the AV node changed from a half-oval to a spindle shape, concomitant with development of a distinct so-called muscular AV septum. Furthermore, the left- and right inferior extensions spread wider apart, the septate area containing connective cells enlarged and showed an increase in fibrofatty tissue. It is concluded that these age-related changes may have clinical relevance. The increase in length of the inferior extensions may set the scene for AVN reentry and could explain why this condition is more frequent in young adults than in infancy.

P1116

Postoperative nonsustained ventricular tachycardia predicts poor outcome after cardiothoracic surgery in infants

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Bedside parameters may predict risk of a poor outcome after cardiothoracic surgery. These may include physiologic, hemodynamic, and laboratory findings until acute cardiac failure, poor cardiac function, or organ system deterioration. We sought to determine if tachyarrhythmias in the first 72 hours after operation were associated with a poor outcome in infants (< 12 months). Methods: A nested case-cohort analysis of 319 infants from 543 consecutively monitored patients who underwent cardiothoracic surgery was performed. Each patient was prospectively monitored in the first 72 hours after surgery for selected tachyarrhythmias, namely nonsustained and sustained supraventricular (SVT) and ventricular tachycardia (VT), and junctional ectopic tachycardia (JET). Known predictors of poor outcome were also reviewed. Poor outcome was defined as an intensive care unit death and/or the use of a mechanical assist device. Results: Of the 319 patients, 75 (23.5%) had at least one tachyarrhythmia, nonsustained VT was most commonly (11.0%), followed by nonsustained SVT (7.5%), JET (5.6%), and sustained VT (3.1%) and SVT (2.8%). A poor outcome occurred in 34 (7.5%) of the infants. By univariate analysis of arrhythmias, only nonsustained VT was associated with a poor outcome (relative risk 3.3, p=0.001), the positive predictive value was 20% while the negative predictive value was 94%. Multivariate modeling revealed younger age (p=0.02), total bypass time > 75 minutes (odds ratio 3.1, p=0.05), and nonsustained VT (odds ratio 3.5, p=0.02) to be significant risk factors for a poor outcome. Conclusion: Nonsustained ventricular tachycardia in the first three postoperative days is an independent predictor of outcome after cardiothoracic surgery in infants.

P1117

Permanent pacemaker therapy in children with corrected transposition of great arteries

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The aim of study was analysis the problems with permanent pacing in the cTGA patients. Material: between 1980–2000 in 13 children (10 boys and 3 girls) with cTGA pacemaker was implanted. The age for initial implantation ranged from 2 weeks to 13 yr (mean 5.8 yr.). Results: 5 pts had isolated cTGA, 8 pts had other associated cardiac anomalies like VSD, ASD, PS, AI, dextrocardia, dextroversion. Complete a-v block was found in 11 pts (4-congenital, 4-post surgery, 3– acquired); second degree Mobitz type a-v block was found in 2 pts, in both post surgery. The age for initial implantation ranged from 2 weeks to 13 yr. Two children died because of heart failure. For the first implantation epicardial system was used in 12 children: VVI-2, VVIR-7, DDD-3. The endocardial VVI system was used in 1 child. During the reoperation endocardial approach was failure to attempt in 2 pts due to unreliably high threshold and problems with lead stability. Five children had more than 1 lead placed. Five leads had malfunction due to high threshold (all were epicardial), 1 due to lead fracture and 1 – isolation break. In 1 child endocardial DDD system was implanted successfully, but after 12 hr ventricular screw-in lead displaced. An average lead life was 5.6 yr. Reimplantation for pacemaker depletion was necessary in 4 pts with average life 8.6 yr. Conclusion: Endocardial lead implantation in cTGA patients is difficult but possible. High threshold is a main reason for reoperation.

P1118

Permanent cardiac pacing in children: 20 years of experience

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The aim of study was to present our experience with permanent pacing in children. Material: between 1980–1999 in 247 children pacemakers were implanted. The indications for therapy were as follows: a-v block II/III° – 139 pts, sinus node dysfunction – 46 pts, long QT syndrome – 11 pts, vasovagal syncope – 2 pts. Results: the follow up period ranged from 1 day to 16 years (mean 5.8 yr). For first implantation epicardial system was used in 198 pts: VVI- 68 pts, VVIR – 52 pts, DDD – 8 pts. Eighteen children died, 17 with heart failure, 1 with myocardial cytopathy. Thirty seven children have still first implanted pacemaker. In 63 pts reoperation was necessary: pacemaker depletion – 44 pts, lead failure – 15 pts, high threshold – 6 pts, pacemaker failure 2 pts, infection – 3 pts, pacemaker dislocation – 1 pts. In 23 of them epicardial system was changed for endocardial: VVI for VVIR 10 pts, VVI for DDD 6 pts. The endocardial system was used in 139 pts for first implantation: VVI - 36 pts, VVIR - 44 pts, DDD 51 pts, AAI – 8 pts. Seven children died, 4 with associated cardiac defect, 3 with LQTS. First implanted pacemaker still works in 95 children. In 31 pts reoperation was necessary: pacemaker depletion – 7 pts, lead failure 5 pts, pacemaker failure 5 pts, infection 2 pts, lead dislodgement 8 pts, accidentally left ventricular lead puncture – 3 pts, right ventricular perforation by the lead 1 child. In 8 children VVI mode was changed to VVIR, in 1 to DDD. Conclusions: permanent pacing in children is much more troubleshooting than in adults. Endocardial system worked longer than epicardial. The quality of leads and pacemakers as well as doctors experience determined system proper function.

P1119

Permanent cardiac pacing in children with long qt syndrome

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Children with LQTS are at high risk for sudden cardiac death. The therapy of choice are still beta-blockers. Permanent cardiac pacing is another treatment option that has been reported. The purpose of the study was to review our experience with permanent pacing in LQTS children. Pacemakers were implanted in 24 children with corrected QT (cQT) > 0.44 s to 0.74 s (mean 0.55 s). In 5 of them complete a-v block was diagnosed. In 5 pts. family history

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

was positive. Before treatment 14 of 18 study children experienced either cardiac arrest (n=8) or syncope (n=11), in 11 pts polymorphic ventricular tachycardia was documented. Age for pacemaker implantation ranged from 30 days to 16 yr 4 mo (mean 5 yr 2 mo). After the pacemaker implantation all pts were on beta-blockers. Maximal pacing rate was 70 to 180 /min. We have used VVIR mode in 7 pts, DDD in 6 and AAI in 5. Follow up period ranged from 2 mo to 12 yr 8 mo (mean 5 yr 2 mo). In children with DDD and AAI cQT interval shortened to mean 0.49 s (from 0.44 to 0.56). 11 of 18 pts are asymptomatic on pacing and high doses of beta blockers but in 2 pts complex ventricular ectopy persists. One girl had recurrent syncopes, changing of pacing rate to 100 /min and increasing dose of beta blockers was effective in this case. 5 children died suddenly in spite of proper pacing and pharmacotherapy, 2 of them during physical effort, 1 at rest. One boy died when beta blockers were stopped. We lost from follow up 1 boy, another 1 died in pyrotechnic accident. Pacemaker therapy in children with LQTS prevents bradycardia, makes possible beta blockers treatment and may shorten QT interval. Permanent cardiac pacing with beta blockers therapy is effective in the most of high risk children with LQTS but does not provide complete protection.

P1120

Permanent cardiac pacing in children with sinus node dysfunction

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Among 200 pts. who needed permanent cardiac pacing between 1980–2000 in 36 cases (18%) the indication for pacemaker implantation (PI) was SND. The diagnose of SND was established by routine and Holter ECG. In 16 (4.2%) children SND was diagnosed before (3pts) or after (12pts) surgical repair (TOF– 4 pts, ASD II– 3 pts) or palliation (TGA-Senning – 2 pts, complex defects-Fontan – 3 pts) of CHD. Normal heart had 17 pts (47%), myocarditis –4pts (11%). Age during first PI ranged 10 mth–16yrs. The type of implanting pacing system dependent on the type of SND, normal a-v conduction, presence and type of CHD, age and body size. One of 36 pts with SND the indication for PI was symptomatic sinus bradycardia in 25 pts (69%) and bradycardia – tarmycardia syndrom (BTS) in 11 pts (31%). BTS (SB such AT and/or VT and/or SVT) had 5pts with idiopathic SND, 2 pts with post-operative SND and 1 with myocarditis. During 4 mth–8 yrs follow up in 32 pts (89%) the first implanted pacing systems still works correctly. In 4 pts (=4 reoperation were performed because of battery depletion, exit block or a-v conduction disturbances. At the end of follow up period pacing systems worked in mode VVI or VVIR in 21 pts (59%), DDD or DDDR in 16 (44%) and AAI in 1, with endocardial leads in 25 children (69%) and epicardial in 11 pts (31%). Conclusions: In our experience 18% of pediatric pacemaker recipients were children with SND. CHD pts after surgical procedures involving the atrium are at the higher risk for TBS and may require PI prior to administration of AA drugs. The decision for choosing a permanent pacing system in children with SND must be individualized.

P1121

Ecg abnormalities in children with marfan syndrome: is qt dispersion a useful measurement?

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Background: Marfan syndrome (MFS) is a dominantly inherited connective tissue disorder caused by an abnormality of fibrillin. Commonest cardiac abnormality in children is aortic root dilatation, which can lead to sudden death. Cardiac arrhythmias are thought to be an important cause of morbidity but the incidence in childhood is unknown. QT dispersion (QTD) has been proposed as a simple non-invasive measurement of dispersion of repolarization available from the 12-lead surface electrocardiogram (ECG). Little is known about QT dispersion in children. AIM: The aims of this study are 1. to assess the prevalence of ECG abnormalities in children with Marfans syndrome 2. to assess the usefulness QT dispersion in predicting arrhythmia. Method: Data were obtained retrospectively from the Paediatric MFS surveillance project in Wales (1993 –2000) and Bristol Cardiac database (1996–2000). 12 lead ECG was analysed for average RR, QT, QTc intervals and QT dispersion and reported on the ECG recorded after starting beta-blocker therapy. Symptoms and the results of Holter monitoring were noted. Results: 73 children were identified (age: 14 days to 19 years) of which 55 children had 12 lead and/or ambulatory ECGs, 17 patients had 24hour ECGs and 8 children reported symptoms. 80%(35/44) had ECG abnormalities of which 17% (6/35) had evidence of dysrhythmias. 41% (7/17) had abnormal Holter monitoring and 14%(8/44) reported symptoms. QTc intervals were within the normal range. 51%(24/44) of patients with Marfan syndrome had prolonged QTD. There was no significant difference between QTD pre and post beta-blockade. Conclusion: ECG abnormalities are a common finding in children with Marfan syndrome. Our study suggests that QT dispersion is prolonged in these children. This may be secondary to abnormal conducting tissue in the presence of abnormal fibrillin.

P1122

Intraoperative ablation for unresponsive supraventricular tachycardia during surgery for congenital heart disease.

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Supraventricular tachycardia is one of the major late complications of surgical correction of congenital heart disease, especially when atrial enlargement occurs. We have evaluated the results of intraoperative ablation in a group of 18 pts with congenital heart disease and chronic supraventricular tachycardia (mostly intraatrial reentry) unresponsive to conventional therapy. All the procedures have been consecutively performed between september 1999 and october 2000. In 14 pts ablation has been performed during redo operation (Fontan conversion to total cavo-pulmonary connection in 13pts) and in the remaining pts during elective surgical correction of their congenital heart disease. The mean age at operation was of 25 yrs (2–51yrs). In 10 pts was performed cryoablation and in 8 a radiofrequency ablation. The 18 pts had ablation of all possible critical sites (mean 4 linear lesions) in right atrium, and a generous atrial reduction. Six pts had implant of an atrial wire. Two patients died during the first postoperative month. All the procedures were acutely effective. During mean follow-up of 9 mths we have observed a relapse of atrial tachycardia in 2 pts both responsive to medical treatment. In conclusion intraoperative ablation is a promising treatment of unresponsive atrial tachycardia in those pts whom required a concomitant surgical procedure for their congenital heart disease.

P1123

Long Term Benefits Of Active Fixation, Steroid Eluting Ventricular Leads In Children

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Pacing therapy is usually a lifetime therapeutic decision and low threshold characteristics and mechanical stability are important features of an ideal pacemaker, especially for pediatric patients. We report our experience and long-term results with a steroid eluting active fixation ventricular lead in pediatric patients. Active fixation steroid eluting leads were implanted in 48 patients, among which 24 were Telectronics Accufix II DEC model 033–212 ventricular leads. Eighteen of them (11 male/ 7 female; 10.6+/-4 years), who were followed for a mean period of 4.3+/-0.8 years were included in the study. Pacemaker mode was DDDR in three patients, and VVIR in the remaining 15 patients. Mean threshold value was 0.5 V at implant, which increased to 0.7 V in the first month. It remained stable (0.62–0.67) until 2.5 years, after when a slight rise in the mean threshold value was observed, however the differences were statistically insignificant (p>0.05). Pacing lead impedances did not differ significantly throughout the study either (p>0.05). Pulse generator replacement was done due to end of life in 5 patients at 4 years or later from implantation. The leads were kept in place in all the patients. In the remaining 6 patients who were followed for at least 4 years the mean cell impedance was 1790+/-560 ohms, and the mean magnet rate was 86+/- 3 ppm as their last visit. The mean longevity of the generator was determined as 30.5 months. Steroid eluting active fixation ventricular leads have low chronic stimulation threshold values, allowing lower outputs, thus saving generator energy and increasing the longevity of the pacing system. These features have definite advantages in pacing therapy of pediatric patients.

P1124

Sustained Ventricular Tachycardia in Children – Multicenter study in Japan –

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The purpose of this study is to know the best treatment of sustained ventricular tachycardia (SVT) in children. Sixty three cases (37 males, 26 females) of SVT, mean age: 9.6+/-4.2 years (m+/-SD), 10 of them had organic heart

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

disease were included in this study. The mean heart rate of SVT is 184+/-37 bpm, left bundle branch block morphology in 32, right bundle branch block in 18, polymorphic in 12, unknown in 1. The first documentation of SVT was heart wrenching in 27, syncope 13, palpitation 8, heart monitor 4, chest pain 3, nausea 2 and others in 6. SVT was induced by exercise in 39 of 56 (70%), programmed stimulation 9 of 23 (39%), isoproterenol infusion 5 of 9 (56%). Late potential was recorded inside of 28 cases by SAECG. Catheter ablation (CA) was selected in 30, conventional pharmacological therapy (P) in 39 and 14 received no therapy (N). Heart rate of SVT was significantly higher in CA (192+/-49) than N (130+/-21) (p<0.0001), and P (170+/-30) than N (p<0.0003). During follow up of N, 12 of 14 remained asymptomatic, 1 disappeared SVT and 1 decreased SVT rate. In 39 P, SVT was suppressed by P in 14, disappeared in 7, sudden death in 2, 3 had no symptom and the other lost in follow up. In 30 CA, abolished SVT in 8 and decreased SVT rate in 2. All of sudden death cases had syncope and followed up by P. We conclude that first choice of therapy for SVT in children is CA, because of high success rate. The indication of CA from this study is SVT rate over 170 bpm, patient had syncope, difficult to control by medication and of single SVT focus.

P1125

Single pass lead VDD pacing in children: effect of postures and physical activities on stability of P wave sensing

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Introduction: We designed this study to evaluate the P wave sensing stability in relation to postures and physical activities in children with VDD pacing system. Methods: The surface ECG and telemetered atrial electrograms were simultaneously recorded in 10 children (mean age 10.1 years) with a single lead VDD pacing system during different postures and activities: supine prone, right lateral, left lateral, sitting, squatting, deep inspiration and expiration, coughing, arm swinging, walking on treadmill at 1, 2, 3, 3.5mph, down stairs, upstairs, cycling at 25 and 50 rpm. The amplitude of the atrial electrograms was measured manually (Resu 6). The mean P wave amplitudes of the group at supine position was 1.68+/-0.52mV and was no significantly different (P= NS) when comparing to different postures and activities. However variation in the P wave amplitudes occurred considerably within individuals. From all the patients the lowest P wave, highest P wave and lowest-highest P wave difference were 0.64+/-0.39mV, 1.79+/-0.5) mV and 1.15+/-0.43 mV respectively. There was no particular posture or activity identified with the lowest P wave amplitude in the group. However in more than half of the patients the prone position, walking at 3.5 mph downstairs and cycling at 50 rpm showed decrease in P wave amplitudes. Despite the changes in P waves no atrial undersensing occurred in all patients throughout the test. Conclusions : The P wave amplitudes vary considerably within an individual in relation to different postures and physical activities. P wave sensing test during various physical maneuvers may be needed to ensure reliable VDD pacing in children.

P1126

Ventricular tachycardia late after repair of congenital cyanotic heart disease.

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In the long term postoperative stage of patients with congenital cyanotic heart disease, ventricular tachycardia (VT) is a serious complication and is associated with an increased risk of sudden death. We performed an electrophysiologic study (EPS) and applied catheter ablation and/or cryoablation to VT in 7 patients who received corrective surgery for tetralogy of Fallot (4) or double outlet right ventricle (3). They were underwent radical surgery at an age of 3 to 17 years with a mean of 7.6 years. VT occured at an age of 12 to 36 years and an average of 13.6 years had elapsed after corrective surgery. Programmed electrical stimulation induced VT in all the 7 patients, sustained VT in 6 and nonsustained VT in 1. Earliest activation sites of VT were identified at the site of myotomy in the right ventricular outflow tract and at the margin of the interventricular septal defect. Six patients had multiple VT foci. Five patients underwent catheter ablation (CA) for VTs, but CA was unsuccessful or only temporarily effective. Three patients, one of them following the failure of CA, underwent cryosurgical ablation for VTs. In 2 patients, arrhythmogenic foci were identified, and VTs were ablated by cryoablation. In another patient, no VT focus was identified by EPS, and cryoablation failed to ablate VT. VT originated after corrective surgery for congenital cyanotic heart disease, located at the right ventricular myotomy scar, and surgical cryoablation will be certainly effective for VT rather than CA if VT foci is precisely determined by EPS.

P1127

Epicardial pacing in infants and small children below twelve years of age.

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Background: Traditionally permanent endocardial (EP) pacing is preferred over epicardial pacing, however it may be difficult in infants and small children due to small subclavian vein (SV) size and thin subcutaneous tissue. We present our experience of EP in 68 children below 12 years of age, performed between 1991 – 2002. Methods: Mean age at the time of pacemaker implantation was 6.5±3.9 years (range 9 months – 12 years), 27 cases were below 5 years. Indications for pacing included congenital complete heart block in 18 and post operative complete heart block in 50 patients. Follow up period ranged from 1 to 84 months (mean=26.3± 24.6 months). Results: The SV was accessed percutaneously in 65 (Left SV in 43). Cephalic vein was used in 2 cases and internal jugular vein in 1. All patients were given ventricular pacing (8 rate-responsive). Mean acute threshold was 0.65 ± 0.22msec. The pulse generator was implanted subcutaneously in older children (n=30) and subpectorally in smaller children (n=38). Complications occurred in 18 patients. Three patients had generator extrusion, (the generator was placed subcutaneously in these), two patients had pacemaker infection, all 5 patients were managed with debridement and skin flap. Early capture failure occurred in 4 cases, due to cuff block in 2 and lead displacement in 2. Three patients had capture failure after 1 year due to increase threshold and were managed by increasing the pulse width and amplitude. One patient had lead fracture. In 3 patients pacing lead stretched over a period of time due to growth of children, in 2 lead repositioning could be done successfully. Elective generator change was done in 5 patients, one of these was given DDD pacemaker. Three patients had sudden cardiac death within 1 year of pacemaker implantation. Conclusion: Transvenous EP is feasible and safe in small children with the currently available pacing systems. Subpectoral insertion of the pulse generator should be preferred.

P1128

Notched T waves on holter recordings enhance detection of patients with LQT2 (HERG)mutations

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Background: The 2 genes, KCNQ1 (LQT1) and HERG (LQT2), encoding cardiac potassium channels are the most common cause of the dominant long-QT syndrome (LQTS). Besides QT interval prolongation, notched T waves have been proposed as a phenotypic marker of LQT5 patients. Methods: The T wave morphology of carriers of mutations in KCNQ1 (n=155) or HERG (n=57) and of 100 control subjects (C) was analyzed from Holter-ECG recordings. Averaged T wave templates were obtained at different cycle lengths, and potential notched T waves were classified as grade 1 (G1) in case of a bulge at or below the horizontal whatever the amplitude, and as grade 2 (G2) in case of a protuberance above the horizontal. The highest grade obtained from a template defined the notch category of the subject. Results: T wave morphology was normal in the majority of LQT1 and C subjects compared with LQT2 (92%, 96% and 19% respectively, p<0.001). G1 notches were relatively more frequent in LQT2 (28% vs 8% [LQT1] and 4% [C], p<0.01) and G2 notches were seen exclusively in LQT2 (6.5%). Predictors for G2 were young age, missense mutations, core domain mutations in HERG. Conclusion: This study provides novel evidence that Holter recording analysis is superior to the 12-lead ECG in detecting G1 and G2 T wave notches. These repolarization abnormalities are more indicative of LQT2 vs. LQT1, with G2 notches being more specific and often reflecting HERG core domain missense mutations.

P1129

Familial polymorphic ventricular tachycardia unlinked to chromosome 1q42

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Background: Polymorphic ventricular tachycardia (PVT) is characterized by exercise/adrenaline-induced ventricular arrhythmias and is associated with sudden

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

death in patients. Familial forms with an autosomic dominant pattern of inheritance have been reported. So far, this cardiac disease has been linked to 1q42 in 3 families. Methods: The phenotype and genotype of 26 members of a family with PVT were ascertained. Subjects were considered as affected in the absence of structural heart disease, in case of exercise stress test: induced ventricular bigeminy or polymorphic ventricular tachycardia with or without syncope or in case of an adrenergically-triggered sudden death. Linkage analysis was performed using D1S179, D1S235, D1S2680, D1S2670 and D1S304 markers at 1q42. The 4 main long QT syndrome genes KCNQ1, HERG, SCN5A and KCNE1 were screened for mutations by PCR-SSCP. Results: Twenty-six family members were collected. Ten subjects were considered as clinically affected including a 26-year old male and a 28 year-old female who died suddenly during an effort. The diagnosis of a 9 year old boy was undetermined due to the presence of isolated extrasystoles at stress test without bigeminy. The genotype could not be ascertained in some cases because their young age. Among the affected subjects aged from 14 to 54 years (5/8 males), only four experienced stress-induced syncope. No abnormal conformation underpinning such the disease were found in KCNQ1, HERG, SCN5A or KCNE1 by PCR-SSCP analysis. In addition, the affected patients did not share any common allele for all the markers studied at 1q42 locus. Conclusion: Thus familial form of exercise-induced PVT does not map to chromosome 1q42. This finding supports the hypothesis that at least one other gene is responsible for this disease.

P1130

Postoperative junctional ectopic tachycardia

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Junctional ectopic tachycardia (JET) can be a life-threatening arrhythmia in children after open heart surgery. Various risk factors such as metabolic disturbances, use of inotropics and AV-nodal injury have been reported as causative factors and different treatment strategies have been proposed. In a retrospective study we analyzed the data of 39 postoperative JET-patients from different centers (1993–2000). Median age at operation was 3 months (range 4d–11months). 36/39 pts underwent closure of VSD/AVSD as part of the operation and 3/39 pts had transient AV-block prior to JET. At onset of JET, electrolytes (Mg, Ca, K, Na) were with in normal limits in 36/39 pts. Mean onset of JET was 4 hr (range 0–72 hrs), upper heart rate 220/min (170–260) and median duration of JET was 37 hrs (12–108). Treatment consisted of amiodarone +/– hypothermia or digoxin in 23 pts, hypothermia +/– digoxin in 9 and digoxin only in 5. Atrial pacing as adjuvant therapy was used in 10/39 pts. In 3 pts JET was not treated. Four patients (2 VSD, 1 Tetralogy, 1 AVSD) died during the episod of JET. Conclusion: postoperative JET is associated with transient AV-block and surgery near the AV-node, metabolic disturbances appear unimportant. Combined therapies are often necessary to control JET.

P1131

Atrial flutter and atrial fibrillation after surgery for congenital heart disease

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Objectives. To review the clinical characteristics and the management of atrial flutter and/or atrial fibrillation (AF/Afl) after surgery for congenital heart disease (CHD). Methods: Since 1978, thirty-two patients have had episodes of AF/Afl after surgery for CHD at our institute. They included 6 patients after Fontan operation (atriopulmonary connection, APC) (group A), 15 with (group B) and 11 without (group C) atrial volume and/or pressure overload at the onset of AF/Afl. Results: Age at the onset of AF/Afl and its follow-up period was 13.0±7.1 and 6.5±5.1 years, respectively. The interval from the definitive operation was 11.2±13.3 years. In group A, cardioversion (CV) was successful in 5/6 (83%). These patients could not be controlled with antiarrhythmic agents (AA). Conversion from APC to total cavo pulmonary connection (TCPC) with Maze or cryoablation was effective in 2/3 (66%). Four patients (66%) were alive and 2 (33%) died suddenly. In group B, CV was successful in 9/11 (81%). AA were effective in 7/10 (70%). Surgical relief of overload of the atrium was effective in 4/5 (80%). Thirteen patients (73%) were alive, 2 (13%) died due to heart failure. Five patients (33%) had no recurrence for more than one year. In group C, CV was successful in 9/10 (90%). AA were effective in 3/11 (27%). Catheter ablation was performed in 1 but failed. Six patients (54%) were alive, 2 (18%) died suddenly. Three (27%) had no recurrence for more than one year. Conclusions: Post-surgical AF/Afl in CHD patients is difficult to control with AA. Conversion to TCPC and relief of atrial overload in groups A and B can be achieved with low morbidity and mortality and is effective treatment of intractable arrhythmia in some.

P1132

Results of treatment radiofrequency transcatheter ablation in patients with permanent junctional reciprocating tachycardia

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Objectives. Permanent junctional reciprocating tachycardia (PJRT) occurs primarily in young patients and causes nearly incessant tachycardia that is frequently refractory to pharmacological treatment. Radiofrequency transcatheter ablation (RFA) appears promising as safe and effective therapy in children. Methods: Between May 1998 and September 2000, thirty-seven patients with PJRT underwent RFA. The age of the patients ranged from 4 to 60 years (mean age 22.1 +/- 11.5 years), including 16 children less than 16 years of age. The length of treatment of tachycardia was from 6 months to 36 years (mean age 10.2 +/- 8.1 year). Results. The site of the earliest retrograde atrial was right posteroseptal in 26 patients (70%), left posterior in 4 (11%), right midseptal in 3 (8%), right posterolateral in 2 (5%), and multiple (right posteroseptal and right posterolateral (n=1) and left lateral, left posterolateral and right posteroseptal (n=1) in 2 (5%). Thirty-six accessory pathways (PA) were successfully ablated with a mean of 4 +/- 3 (mean, 3). RFA of a mean duration of 58 +/- 12 s. Only one patient with right posteroseptal AP could not be ablated. After a mean follow-up of 19 +/- 14 months (mean, 12, range 2 to 50 months) 34 patients remain asymptomatic. There were recurrences in two patients after the initial successful ablations (during the first month), and both were ablated in a second ablation procedure. All patients with depressed left ventricular function showed a marked improvement after successful ablation. Conclusions: Our study supports the concept that RFA is a safe and useful treatment for patients with PJRT. Radiofrequency current should be the treatment of choice in these patients. AP with decremental conduction properties is localized in the posteroseptal zone. Cessation of the arrhythmia after successful ablation results in recovery of left ventricular dysfunction.

P1133

Ten year follow up of steroid-eluting epicardial leads versus non-steroid epicardial leads in pediatric patients

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Introduction: Transvenous leads have been used in children who require pacemaker therapy due to elevated thresholds with non-steroid (NSE) epicardial (EPI) leads. However, concern for vascular and valvular integrity with lifelong pacing favors an initial EPI approach. Although steroid-eluting (SE) leads demonstrate low pacing thresholds in the short term, performance with chronic use is unknown. We compared 10 year performance of SE and NSE EPI leads in a growing pediatric population with and without CHD. Methods: From 1990–1999, 37 patients (pts) (age: newborn-18 yrs, median 3 yrs) received 54 Medtronic (MDT) SE lead models 10295A, 10295B and 4965. 31 ventricular (VEN), 20 atrial (ATR). Of these pts, 31 had CHD, 6 no CHD. Another 24 pts (ages 1–28 yrs, median 8 yrs) received 32 NSE leads/T models 6917, 5069, 5071, CPI 4320. 25 VEN, 7 ATR. Of these pts, 20 had CHD, 4 no CHD. Pacing thresholds (THR), impedance (IMP) and performance were surveyed at implant and over a 10 year (median 5 yr) follow-up. Results: IMP were comparable for all leads. Fracture or dislodgements occurred in 2 SE (4%) and 4 NSE (16%) leads. Based on chronic pacing THR, mean energy requirements with SE (2 +/- 2.7 uJ) were significantly lower than NSE (7.1 +/- 6.2 uJ) leads throughout the study ($p<.05$). At 2.5 v output, the final mean pulse width THR for SE leads was not significantly different from initial implant values for either ATR (.09 vs .07 ms) or VEN (.1 vs .1 ms) sites, or among patients with and without CHD. Conclusion: SE reduce battery drain and show stable, very low THR over time. SE EPI leads can be safely used in any age growing child with or without CHD.

P1134

Coincidence of long QT-Syndrome and epilepsy

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https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Long QT-syndrome (LQTS) is a rare cardiac arrhythmia and it may lead to the misdiagnosis of epilepsy. Early diagnosis is important because of different prognostic and therapeutic consequences. The coincidence of LQTS and epilepsy has been described only in very few cases. We report of two 12- and 14- year old brothers with LQTS. The way to the correct diagnosis was tortuous in both patients. The elder brother had had EEG pattern characteristic of epilepsy for several years before LQTS was diagnosed, and EEG signs were progressive after diagnosis and treatment of both diseases had been established. He showed exercise-induced syncope first and the diagnosis of Rolandic-epilepsy was made because of typical unilateral epileptiform discharges in EEG. Anticonvulsive drugs proved ineffective. After further syncopes the detection of ventricular extrasystoles on exercise in ECG lead to β-blocker therapy without diagnosing QT-syndrome at that time. Therapy diminished the frequency of syncopes. Three years after his first symptoms his younger brother had ventricular fibrillations and was successfully defibrillated and resuscitated. Now the elder brother was cardiologically reassessed and in both brothers the diagnosis of LQTS established. Despite β-blocker therapy four months later the elder brother had a severe syncope and needed ICU support over 4 days. Both brothers received automatic cardioverter-defibrillation and have been free of symptoms for 3 months. On EEG, the elder brother has continued to show typical epilepsy pattern now bilaterally. Nonetheless he was put on anticonvulsive drugs.

P1135

Clinical Evolution of the Wolff-Parkinson-White Syndrome in Children

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The Wolff-Parkinson White syndrome of congenital in origine and his importance is in posibility of complication-reciprocating tachycardia and atrial fibrillation, which take happend in childhood. The purpose of this investigation is trying to make decision for best treatment in children with Wolff-Parkinson-White syndrome and recurrent tachycardia. We evaluated 26 patients (pts) below the age of 17 (average value 5.8+-5.6 years) M:F=19:17, who followed up during 1-14 years (average value 3.6+-2.7 years). All of them have had detail physical examination, EKG, Roentgenographic picture and Echocardiography. At first 24 hour Holter EKG monitoring and two electrophysiologic examinations. From 26 children 7 pts (27%) have had congenital heart disease: mainly as cardiomyopathy 2(5%), Ebstein anomaly (EA), EA and ASD, VSD, MVP and mild a pulmonary stenosis one of each (3%). Other 19 pts (80%) was without structural heart disease. During long term period of follow up only 4 (15%) children had or had episode of tachycardia. Only one attack of tachycardia had 15 (38%) and 2 or more attacks had 19 (64%) pts. In group A (0-5 years) without recurrent tachycardia were 9 and with recurrent tachycardia were 8 (NS). In group B (6-17 years) there were not of tachycardia recurrence in 4, but were present in 11 pts (p<0,05). In both groups medical treatment were similar: in group A the first choice of treatment were Dilacor and after that Verapamil and Propranolol. In group B the most frequently were given Verapamil, than Propranolol and Amiodarone. The last one was the best in tachycardia control in children with Wolff-Parkinson-White syndrome. In conclusion, children with the first presentation of reciprocating tahycardia and White-Parkinson-White syndrome, are in great risk for recurrence of tachycardia, despite medicamental treatment. Sinradio-frequency ablation have important role in the therapeutic aproach, specially in children over 5 years of age.

P1136

Ventricular preexcitation and sudden death in children and young adults

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Prevalence and clinicopathologic features of ventricular preexcitation (VPE) were investigated in a series of 273 sudden death (SD) cases in the young (<35 yrs). Site of accessory pathways was predicted by 12-lead ECG. Right and left AV rings together with interatrial and AV septal junction were studied by serial sections. Twenty (7.6%), all male, mean age 24 yrs, had VPE of whom in terms of Wolff Parkinson White (WPW) syndrome and 2 in terms of Lown Ganong Levine (LGL) syndrome. SD occurred at rest in all but 3 and 6 had previous symptoms. In the 2 LGL pts pathologic substrates consisted of AV node hypoplasia and right sided atrio-Hisian tract respectively. In the 18 WPW pts, 10 total accessory pathways consisting of ordinary myocardium were found (7 left lateral, 2 right posterolateral, and 1 septal). These pathways were close to the endocardium (mean distance 750 micron) and very thin (mean thickness 310 micron). Moreover, 4 pts (50%) showed an isolated acute atrial myocarditis which was polymorphous in 3 and lymphocytic in 3. In conclusion, VPE accounted for 3.6% of SD in young people and was not preceded by warning symptoms in 40%. A left sided accessory pathway was the most frequent substrate, and its subendocardial location supports the feasibility of catheter ablation. Isolated atrial myocarditis may act as a trigger of paroxysmal atrial fibrillation that leads to SD.

P1137

Epinephrine provocation and the congenital long QT syndrome: a novel clinical test

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Background: Approximately 30% of individuals with genotyped long QT syndrome (LQTS) fail to manifest significant QTc prolongation (QTc > 460 ms). Since sympathetic stimulation is a common arrhythmogenic trigger in LQTS, epinephrine provocation may aid in the identification of at-risk individuals. Methods: 29 patients (pts) with congenital LQTS (age 12 - 45 years, mean 25 years, 18 females) from 16 different kindreds and 22 age- and gender-matched control subjects were studied at baseline and during gradually increasing rate of epinephrine infusion (0.05, 0.1, 0.2, and 0.3 mcg/kg/min). 12-lead ECG was monitored continuously and QT, QTc, and heart rate were measured. Genotype was established by PCR amplification and DNA sequencing of the three most common LQTS genes: KVLQT1 (LQT1), HERG (LQT2), and SCN5A (LQT3). Results: The baseline QTc was greater in LQTS (502 ms) than in controls (439 ms, p < 0.001). However, 7/16 LQT1 subjects had a non-diagnostic QTc (< 460 ms) while 11/22 controls had QTc > 440 ms. During epinephrine infusion, every LQT1 subject displayed paradoxical prolongation of the uncorrected QT interval whereas controls, LQT2, and LQT3 pts tended to shorten their QT intervals (p < 0.001). The maximum delta-QT (epinephrine QT minus baseline QT) was +3 ms (controls), +93 (LQT1), -9 (HERG), and -47 (LQT3). Epinephrine-triggered T wave alternans and/or non-sustained ventricular tachycardia occurred in 4/29 LQTS pts but 0/22 controls. Conclusion: Epinephrine-induced paradoxical prolongation of the uncorrected QT interval appears to be pathognomonic for LQT1. Epinephrine challenge distinguishes concealed LQT1 subjects manifesting an equivocal QTc at rest from normals. Thus, epinephrine provocation may improve the diagnostic accuracy of the LQTS clinical evaluation and strategically direct molecular genetic testing.

P1138

Paroxysmal complete atrioventricular block with consecutive syncope during head-up tilt testing.

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A 17 year-old adolescent was referred for cardiac investigation because of one previous syncope. Physical examination, ECG, chest X-ray and echocardiography showed no abnormalities. Twelve-lead ECG, treadmill exercise testing and 24-hour Holter monitoring revealed right bundle branch block and paroxysmal, high-degree AV block. During head-up tilt testing the patient suffered from similar syncope with clonic convulsions. That was due to a paroxysmal complete AV block, which is a very rare manifestation of neurocardiogenic syncope. A dual chamber (DDD) pacemaker was implanted and the patient has been asymptomatic thereafter.

P1139

Benign cardiac arrhythmias in healthy newborn infants.

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Background: Benign disturbances in neonatal cardiac rhythm are relatively common. The aim of this study is to confirm incidence and types of cardiac arrhythmias in healthy, full-term neonates. Methods: Twenty five healthy, full-term newborn infants (15 male, 10 female) 1 day old, were examined by 24-hour ECG ambulatory monitoring (Holter). Results: Average minimum heart rate was 84.7 ± 12.4 beats/ min (range=55-115, 95% confidence interval (CI)= 79.2-90.3). Average maximum heart rate was 187.5 ± 19.4 beats/ min (range= 156-212, 95% CI= 178.8-196.1). Based on this data, transient sinus bradycardia noted in 20 (40%) neonates and sinus tachycardia in 4 (16%)

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

neonates. Premature atrial contractions (PACs) observed in 22 (88%) neonates with average level of 9.5 ± 18.2/ hour and unusual dispersion (range = 0.5–80.2 median= 3.5 95% CI= 1.4–17.6). In most cases PACs were isolated, uniform, infrequent, observed at resting state. Frequent PACs (maximum PAC/ hour 360) was noted in 5 (20%) newborn. Blocked PACs (n = 2), abnormally conducted PACs (n = 1), PACs out of resting state (n = 1), atrial bigeminy (n = 1) and presence of PACs and premature ventricular contraction (n = 1) were uncommon. Conclusion: Transient sinus bradycardia due to exaggerated vagal tone is common in the asymptomatic newborn infants without underlying disease. PACs is the most common arrhythmias diagnosed in this neonatal population, therefore it can be considered normal finding, demanding no treatment.

P1140

Effects of Atrial Flutter vs Sinus Rhythm on Exercise Tolerance In Grown-Up Congenital Heart (GUCH) Patients

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Background: Atrial flutter (AFL) is common in GUCH patients and leads to deterioration of patients' effort tolerance and Functional Ability Indices. Methods: Exercise tests using modified Bruce protocol were performed in 20 consecutive GUCH patients who presented with symptomatic atrial flutter (AFL) during and again 24–48 hours after DC conversion to sinus rhythm (SR). Diagnoses were one ventricle 9 (5 with Fontan type surgery), transposition of great arteries 4 (3 had Mustard and 1 arterial switch), hypertrophic cardiomyopathy (HOCM) 3, closed atrial septal defect 2 and other lesions 2. Age at study was 21–62 years, 11 female. The first AFL attack was in 4 and recurrent in 16 patients. Results: TABLE. During AFL, the 5 patients with Fontan dropped SBP by 16±5.9 mmHg lower than the other 15 patients, P=0.001, but the mean exercise duration was not different. Exercise test was limited by near-syncope in 4/5 Fontan and 1 arterial switch patients. Heart rate was over 200 bpm within 0.5 to 12 minutes in 4 patients who were not taking antiarrhythmic medicine. With sinus rhythm exercise was terminated mainly because of fatigue or breathlessness. Conclusion: Atrial flutter causes dramatic reduction in exercise tolerance in GUCH patients and results in syncope and hypotension. Marked improvement in effort tolerance occurred after restoring sinus rhythm. Thus, it is important to return and maintain sinus rhythm in GUCH patients.

P1141

Electrocardiographic parameters predicting arrhythmic risk after surgical correction of tetralogy of Fallot.

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Background. After surgical correction of Tetralogy of Fallot, depolarization and repolarization abnormalities of ventricular myocardium are related to an increased risk of life-threatening ventricular arrhythmias. Aim of the study: Identification of the ECG-derived parameters related to an increased arrhythmic risk. Methods: 88 patients (45 males, 43 females) operated for Tetralogy of Fallot (ToF) and followed up at our institution were evaluated. ECG parameters were calculated using computerized off-line analysis. Patients were divided into two groups according to the presence (group A, 16 patients) or the absence (group B, 72 patients) of ventricular tachyarrhythmia, both non-sustained or sustained, during clinical follow-up or 24-hour ECG monitoring. p<0.05 was considered significant. msec represents milliseconds ; ns represents results not found to be significant. Conclusion: In patients who have undergone surgical correction of ToF, QRS dispersion, mean and max QT duration and max JT duration were shown to be significantly associated with an increased risk of ventricular tachycardia. QT and QTc dispersion were not shown to be associated with an increased risk of arrhythmic events.

P1142

Predictors of Refractory Tachycardia in Infants with SVT

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Supraventricular tachycardia (SVT) is the most common chronic arrhythmia of infancy. Prophylactic medical therapy is complicated by lack of predictors of SVT recurrence. We retrospectively reviewed 42 infants with SVT <1 yr of age presenting between 1/1/95 and 31/12/99. SVT was defined using accepted criteria for accessory pathway or AV node reentry. Patients with structural heart abnormalities other than patency of the foramen ovale or ductus arteriosus were excluded. Variables assessed as potential predictors of SVT recurrence are listed below. Outcomes were defined as freedom from SVT recurrence (Simple, n=22) on no prophylactic medication (n=5) or on the first medication prescribed (digoxin, n=23; sotalol, n=10; propranolol, n=5; procainamide n=3), or refractory SVT episodes needing further intervention (Complex, n=19). Significant differences were observed between the groups (Simple vs Complex, mean+/-SEM) with respect to Age at presentation (50.4+/-33.2 vs 10.2+/-2.5 days, P= 0.006), initial echocardiographic left ventricular ejection fraction (0.55 (7/18 vs 7/16 patients, P=0.044), median R-P interval in SVT (101 vs 125 ms, P<0.001); and median SVT R-P/cycle length ratio (0.455 vs 0.543, P<0.001). The groups did not differ significantly with respect to fetal SVT (1/23 vs 5/19 patients), ECG ventricular preexcitation in sinus rhythm (5/23 vs 7/19 patients), median SVT cycle length (220 ms, both groups), sinus rhythm cycle length immediately after first SVT conversion (424+/-52 vs 404+/-52 ms), digoxin as initial prophylactic therapy (10/22 vs 13/18 patients), hospital stay (6.2+/-1.2 vs 14.1+/-4.7 days), or follow-up duration (593+/-92 vs 838+/-113 days). We conclude that recurrent SVT in infancy is associated with younger age and/or ventricular dysfunction at presentation, and with slower ventriculoatrial conduction in SVT as indicated by longer R-P intervals on ECG.

P1143

Follow-up study of 88 pediatric patients with ventricular preexcitation: Clinical and electrophysiological implications of age at presentation

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Patients: 88 pediatric patients with mean age at diagnosis of 5.3 months, male/female ratio 1.62 with ventricular preexcitation at baseline ECG, were followed-up for 7.2±5.8 years using clinical examination and ECG monitoring. No patient was lost at follow-up. Results: 25 of 88 patients (27%) had an associated congenital heart disease. 5 patients(22%) had mitral valve prolapse and 4 patients (16%) had Ebstein anomaly. Anomalous pathway location according to reported criteria was as follows: 56.8% left ventricular free wall (LVW), 23.6% right ventricular free wall (RVW), 2v66 posteroseptal (PS) and 9.1% anteroseptal (AS). Fifty-five patients (62.5%) became symptomatic experiencing atrio-ventricular re-entry tachycardia (AVRT) at a mean age of 5.0.6 months. Thirty-one (56%) of these symptomatic patients developed AVRT during the first year of life. 83% of these patients experiencing ARVT in the first year of life were symptom-free at right years follow-up. In contrast only 39% of patients who developed ARVT beyond one year of age were symptom-free at 8 years follow up. Age at diagnosis in symptomatic patients according to anomalous pathway localization was: 6.4 months for LVW, 33.2 months for PS, 62.5 months for RVW and 85.1 months for AS. No patients experienced cardiac arrest or aborted sudden death. Discussion: Our population resembles that in the literature data, in regard to associated anomalies, pathway distribution, age at symptoms onset, pattern of early onset and late withdrawal of AVRT. Nonetheless our symptomatic patients showed a wide variability of age at diagnosis, ranging from 8.4 months for LVW to 85.1 months for AS pathways. We believe that variability could be ascribed, to the electrophysiological behaviour of both the anomalous pathway and the ventricular myocardium. AVRT, like atrio-atrial tachycardia, needs a critical length of the pathway/arrhythmia wave front velocity ratio to be maintained. This ratio seems to favour an earlier onset of symptoms for left-sided pathways. Factors implicated in the production of this difference in clinical behaviour include ventricular remodelling, supported by afterbirth left ventricle development and hypertrophy, and right ventricle myocardial regression, due to enhanced apoptosis, with fibrosis and conduction slowing of right-sided pathways.

P1144

Unconventional approach to cardiac pacing: transatrial lead placement in a patient with Fontan circulation and complex arrhythmias (Case Report)

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Pacemaker (PM) treatment of complete AV-block (AVB) in patients without regular venous drainage to the heart is usually accomplished by epicardial lead placement. Complicating atrial arrhythmias such as atrial flutter (AF) may induce electrode and PM dysfunction due to side effects of the medical and

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

electrical therapy. Then alternative therapeutic approaches are necessary to ensure lead- and PM performance. Casereport: A 12 year old boy with DORV, I. TGA and pulmonary stenosis is PM-dependent since his sixth year of life because of postoperative AVB. After completion of the Fontan circulation recurrent atrial flutter lead to altered physical performance. Side effects of medical and electrical treatment as well as epicardial rear ripple formation lead to severe threshold rise of the ventricular lead, which resulted in exitblock related syncope and fast battery depletion although high capacity PMs and new epicardial leads were implanted. Placing new electrodes transvenously via the interatrial Gorex-septum by means of transseptal puncture failed. Therefore 2 bipolar transvenous active fixation leads were implanted per media sternotomy into the right atrium and the single ventricle. Since the intraoperative open heart cardioversion AF is controlled medically and by the PM including programmed stimulation (follow-up 11 months). Conclusion: Operative transatrial placement of pacing leads can be an unusual, but effective approach to cardiac pacing in patients without typical venous access after surgery for congenital heart disease and that be an alternative to epicardial placing.

P1145

Incessant supraventricular tachycardia due to Wolff-Parkinson-White syndrome in an infant.

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Supraventricular tachycardia (SVT) associated with an accessory connection is the most common tachyarrhythmia in children. The majority of infants who experience SVT as newborns will stop having episodes of tachycardia by the end of the first year. Evidence of Wolff Parkinson White (WPW) syndrome on ECG is the only clinical parameter that predicts the risk of recurrence of SVT later in child hood. In this study we report an infant with incessant tachycardia due to WPW syndrome. A 2-month-old boy was referred to our unit for incessant tachycardia diagnosed at 20 day of life. Electrocardiogram demonstrated supraventricular tachycardia with a rate around 200 bpm. Echocardiogram revealed LV dysfunction without any congenital heart disease. Medical management with multiple antiarrhythmic drugs including digoxin, procainamide, sotalol, propafenone and IV amiodarone remained uncontrolled. The electrical cardioversion attempted for three times was also not successful. Therefore radio frequency ablation was decided after three weeks of amiodarone treatment. A left anterolateral pathway was eliminated successfully after a total of three applications. No recurrence of tachycardia was noted at the end of a 8-month follow up period and the LV function returne to normal. The infant is currently on no medication and is in excellent health. To our knowledge, WPW syndrome presented with incessant tachycardia in infancy has not been described previously.

P1146

Antibradycardia pacing in patients with congenital heart disease: Experience with automatic threshold determination and output regulation (Autocapture)

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The efficacy and safety of Pacesetter's Autocapture-algorithm (AC) with beat-to-beat capture confirmation, automatic pacing threshold determination and output adjustment was confirmed for transvenous leads in adults without congenital heart disease, but there is little experience in patients (pts) with congenital heart disease (CHD). Method: Our institution surveys 20 pts. (m=6, f=14) who received a transvenous (group A: n=15, m/f=5/10) or epicardial (group B: n=5, m/f=1/4) single (n=5) or dual chamber (n=15) AC-pacemaker (PM). A safe AC-function is ensured when a sufficient evoked response (ER) and a little lead polarisation (LP) are present. Therefor these parameters were reevaluated from 109 follow-up (F/U) PM-interrogations. The results are summarized (median±SE[range]). Group A: n=90 interrogations, age at PM implantation: 22.7±20.2[0.66–67.1]years, F/U: 34.0±22.5 [0–714]days, ER: 13.9±6.1[1.6–29.3]mV, LP: 1.57±0.92[0.39–4.96]mV. Group B: n=19 interrogations, age at PM implantation: 6.5±32.0 [0.1–66.6] years, F/U: 53.0±126.6[3–419]days, ER: 4.64±2.23[0.47–7.74]mV, LP: 1.0±1.02[0.86–4.93]mV. In all transvenous systems AC functioned correctly (in 1 pt AC was activated late after 616 days). Only 1 epicardial system preserved AC-function for 53 days after implantation. Conclusion: These data suggest, that the application of the AC-Algorithm is safe even in pts with CHD when transvenous leads are used. If appropriate AC-function is possible with epicardial leads needs individual verification.

P1147

Pacemaker treatment of recurrent atrial flutter in a patient with complex congenital heart disease: initial experience with the Medtronic AT500 (Case Report)

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The Medtronic AT500 is a new active implantable medical device designed to prevent, detect and treat atrial tachyarrhythmias (AT) including atrial flutter (AFl). Its application in patients (Pt) with operated congenital heart disease and recurrent AFl is limited. Casereport: A now 28 year old man with d-transposition of the great arteries and ventricular septal defect (VSD) suffered from recurrent AFl since his 14th year of life. A Senning procedure and VSD closure had been performed with 8 years, but significant pulmonary vascular obstruction disease remained causing mild cyanosis via a small baffle leakage. A transvenous dual chamber pacemaker (PM) was implanted at the age of 23 years because of sick sinus syndrome, but recurrent AFl persisted and lead to paralyses cerebral embolism. Medical treatment and several DC shocks did not control the rhythmic problem. On the basis of intention to treat at the end of life of the implanted PM a Medtronic AT500 and a new atrial lead were implanted. Within the early postoperative period (4 weeks) the new device detected 3 periods of AFl, the last sustained and was terminated with burst stimulation during follow up examination. Therapeutic automatic treatment algorithms were initiated. During the following 3 weeks another 3 periods of AFl were detected correctly but automatic termination failed. Mild ventricular far-field sensing was present but did not disturb AFl detection. Conclusion: This initial experience with the Medtronic AT500 in a pt with complex congenital heart disease and recurrent AFl shows that reliable tachycardia detection is possible. Manual termination of AFl was achieved with the device, yet refinement of the automatic treatment algorithms is still ongoing.

P1148

Atrial fibrillation in Japanese children

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Background: Pediatric atrial fibrillation (Af) is rare. Objective & Method: To disclose the characteristics of the pediatric atrial fibrillation in Japan by questionnaire to the regional center hospitals. Results: The number of patients with Af under 20 years old is 36 since 1978 (congenital heart disease (CHD) 20, idiopathic 13, cardiomyopathy 2 and other 1). The preoperative patients with CHD include the atrial septal defect 2, tricuspid atresia (TA) 2, and postoperative cases do status of post Fontan procedure 7, tetralogy of Fallot (TOF) 2, ventricular septal defect 2, atrial septal defect 1, post-Senning's procedure 1, tricuspid regurgitation 1, Ebstein's 2, endocardial cushion defect 1 and others 2. Idiopathic cases include 11 male and 2 females. The average age of the onset in idiopathic cases is 13.9 (9–17.9) years old. One of them is a familial case who combined Alport syndrome. Treatments against the atrial fibrillation was digoxin (45% effective), cateopyramide (26%), beta-blocker (36%), cibenzoline (50%), mexiletine (0%), verapamil (0%), flecainide (0%) and current defibrillator (30%). The rate of resolution of Af after surgical repair (atrioseptal enlargement 2, mitral valve replacement 2, Maze 1, Damus 1, conversion to total cavopulmonary connection) was 75%. Anticoagulant therapy was given to 80% of the cases accompanied with the underlying disease, and to 69% of the idiopathic cases. Cerebral embolization was occurred in the only postoperative patients with TOF and with pacemaker implantation who did not take anticoagulant therapy. Conclusion: Idiopathic Af is dominant in male. The idiopathic Af is good in prognosis, but that of patients with underlying disease is not curative.

P1149

Cardiorespiratory endurance in children with complete atrioventricular block (CAVB)

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Children with CAVB fail to increase their ventricular heart rate during exercise. Inadequate rate response may be compensated by increased stroke volume or a larger peripheral oxygen extraction. We investigated the cardiorespiratory endurance under maximal exercise in 19 children (8 female, 10 male) with CAVB, aged 5.-17 years (median 9). CAVB was congenital in 9, acquired in 5 and of uncertain cause in 4 children. Systolic

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

ventricular function was normal in all, and 10 patients had a permanent pacemaker (PM). Exercise testing was performed on a treadmill using a modified Bruce protocol. We assessed the heart rate, total endurance time, maximum oxygen uptake (VO2max) and the ventilatory anaerobic threshold (AT). We compared the results with an age matched control group (N=42). During exercise the heart rate increased by 69 ± 46% in the patient group and by 85 ± 20% in the control group. Exercise time was significantly reduced in the CAVB-group (17.44 min vs 20.14 min ; p= 0.001); VO2max was lower (33.3 ml/kg/min vs. 42.2 ml/kg/min, p< 0.001), and AT was reduced in CAVB-children (18.8 ml/kg/min vs. 24.7 ml/kg/min, p= 0,001). CAVB patients with PM did not perform significantly better than those without PM. Cardiorespiratory endurance performance is significantly reduced in children with CAVB, no matter they are paced or not. The impaired maximum oxygen uptake and aerobic capacity of CAVB- patients suggests, that their inadequate rate response is not compensated by larger stroke volumes or peripheral oxygen extraction.

P1150

Radiofrequency catheter ablation of the left side accessory atrioventricular pathways by retrograde approach in children

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21 children with left side accessory atrioventricular pathways (AAP) aged from 7 to 15 years underwent radiofrequency (RF) ablation procedures by retrograde approach. Ablation catheter electrode was inserted through right femoral artery, aorta, left ventricle to atrioventricular ring. 12 of them had AAP with preexcitation and 13 – concealed AAP. All procedures were done under general intravenous anesthesia with Propofol. 21 patient (87,5%) was successfully treated by single RF ablation procedure using retrograde approach. In two patients transseptal approach was used during the next procedure. In one patient transseptal approach was successful, in one – failed and this patient was treated by closed heart cryoablation during thoracotomy. In one unsuccessful case (girl, 13 years old) procedure was interrupted due to spasm of the right femoral artery with temporary ischemia of the foot. CONCLUSION. Retrograde approach is useful and safe for left side AAP ablation in children from age 7 years and over.

P1151

Idiopatic ventricular tachycardia. follow-up of a pediatric population

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The occurrence of ventricular tachycardia (VT) in patients without associated heart defect is rare. Prognosis is usually good, even if there are no long-term studies published. We describe 12 pediatric patients with VT, in which we excluded structural anomalies of the heart. Mean age was 136±105 months (range 1–288); mean age at diagnosis was 63±47 months (range 1 day –132 months). Mean follow-up has been 71±60 months (1–156 months). According to QRS morphology during VT, patients were divided into two groups: 6 pts with left bundle block (LBB) morphology and origin of beats from right ventricle (5 with inferior axis and 1 with left axis deviation) and 6 patients with right bundle block morphology (5 with left axis deviation and 1 with inferior axis). Main features of our population are shown in the table. Conclusion. In our population VT was suppressed with medical teraphy in 8 pts (66%). VT originating from right ventricle occurred at a higher heart rate. Nonetheless, right originating VT were usually sustained or incessant, thus producing tachycardiomyopathy as first presentation.

P1152

Syncope and sudden cardiac death in children with catheccolaminergic polymorphic ventricular tachycardia

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Background. Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a clearly recognized life-threatening arrhythmia that can occur in children without heart disease and with normal QT interval. We report the clinical features, treatment, and clinical follow-up of children with syncope and CPVT. Methods: We evaluated 3 children (8 to 12 years-old, 3 female) with recurrent syncope. The clinical presentation, delay before the correct diagnosis, 12-lead ECG pattern, 24-hour Holter monitoring, treadmill test (TET), isoproterenol (ISO) infusion, slit-table test, echocardiography and invasive electrophysiologic (EP) study were studied. Patients (pt) were treated with medical therapy, and clinical evaluations, Holter, TET, and ISO were performed at each appointment. Results. Syncope and near syncope were always triggered by exercise or emotion and one pt had sudden cardiac death (SCD). Structural heart disease was ruled out by echocardiogram. Pt were previously treated for epilepsy (2) and for vasovagal syncope (1), and mean time to correct diagnosis was 28 months. Polymorphic VT was induced in all pt during provocative test (TT and ISO infusion) and when sinus tachycardia achieved 170 bpm on Holter. Beta-blocker therapy was effective in abolishing syncope, as well as VT on Holter, TT and ISO. However, one pt died suddenly in the fourth year of follow-up. Conclusion. 1. CPVT is a life-threatening arrhythmia that must be individualized in children with syncope triggered by physical effort or emotion. Confusing diagnosis occurs having these pt no structural heart disease.

P1153

Radiofrequency modification for inducible and unexpected AV nodal reentry tachycardia in pediatric patients

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AV Node Reentry Tachycardia (AVNRT) is the second most common supraventricular tachycardia (SVT) undergoing radiofrequency ablation (RFA) in pediatrics. AVNRT can be difficult to induce under general anesthesia during electrophysiology study (EPS) and dual AV nodal physiology may not be demonstrated in young patients. We report our experience with radiofrequency (RF) modification of slow AV node (AVN) input fibers for inducible or suspected (non inducible) AVNRT in pediatric patients. 73 procedures were performed in 70 patients. The mean patient age was 10.7 years (4yrs 20yrs, 35M/38F). SVT was documented by ambulatory monitoring in all 73 patients. AVNRT was induced in 62/73 patients (group A) with a mean cycle length 342msec (240 -570msec). Initial AVN modification was successful in 59/62 patients (95%). During 11/73 EPS, AVNRT was non-inducible, dual AV nodal physiology was noted in 6/11 (group B), and 5/11 showed no evidence for dual AV nodal physiology (group C). These 11 patients underwent empiric AVN modification following discussion with patients' families. Freedom of recurrence from SVT at 1 year was 89% in group A (55/62), 83% (5/6) in group B and 80% (4/5 pts) for group C. 2 patients recurred following successful RFA, each has undergone repeat successful RFA with no recurrence. Complications occurred in 1 patient (CHB). In conclusion AV nodal modification for AVNRT can be performed safely and successfully in pediatric patients with good long-term results. Empiric slow pathway AVN modification for non-inducible but clinically documented SVT may result in freedom from recurrence of tachycardia.

P1154

Optimization of double-chamber physiologic pacing in children

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Physiologic double-chamber cardiac pacing is now routinely used in children but optimal programming remains to be established. Twenty-two children with atrioventricular sequential (DDD) pacemakers (mean age, 9.5 ± 4.4 years) underwent Doppler evaluation of aortic flows at rest while varying pacemaker parameters. Pacemakers were implanted for sinus node dysfunction, congenital atrioventricular block (AVB) or post-operative AVB. Cardiac index was evaluated first in VVI mode then in DDD mode with an AV delay paced or sensed varying from 50 to 250 ms. The optimal AV delay allowing the greatest cardiac index varied from one patient to the other: AV pace 100 ms in 3,150 ms in 6 and 200 ms in 7, AV sense 90 ms in 3,120 ms to 6,150 ms in 2. There was no relation between optimal and worst AV intervals and age of patients or presence of a congenital heart defect. Cardiac index increased significantly when changing pacing mode from VVI to DDD at the optimal AV delay (27.5 ± 12%). Patients with the lowest cardiac index in VVI mode had the greatest increase in cardiac index at the optimal AV delay. The increase in cardiac index when comparing worst to optimal AV interval was statistically significant for each patient. A 50 msec variation from optimal AV interval paced could change the cardiac index as much as 27%. In conclusion, Doppler echocardiography provides essential information required to optimize cardiac function in children with pacemakers.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P1155

Rhythm disturbances after correction of partial anomalous pulmonary venous connection to the superior vena cava: long term follow-up study

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Correction of partial anomalous pulmonary venous connection (PAPVC) can be complicated by obstruction of venous channels. To avoid this, we described in 1976 a new technique: derivation of the anomalous venous return towards the left atrium by plicating the SVC enlarged with the right atrial appendage. As some patients presented immediate post-op atrial arrhythmia: we were concerned about the possibility of late rhythm disturbances. Twenty-four patients (14F, 10) operated with this technique were evaluated clinically with echocardiogram, electrocardiogram, 24-hour Holter monitoring and exercise testing 9.24 years (3.7 – 19.9 years) post-op, mean age at surgery 6.66 years (2.1–13.5 years). Pre-op electrocardiograms showed sinus rhythm in all. Early post-op arrhythmia occurred in 9/24 (38%) junctional rhythm in 7, flutter in 1 and supraventricular tachycardia in 1. At follow-up all patients were asymptomatic with normal physical examination. Echocardiogram showed normal flow in venous channels. On electrocardiogram, 11 (46%) had atrial rhythm and 2 (8%) 1st degree atrioventricular block. On Holter no significant bradycardia was found, only 1 pt had advanced 2nd degree AV block and no active arrhythmia was recorded. Exercise testing showed normal chronotropic response and normal exercise tolerance. Correction of PAPVC carries an overall good prognosis, atrial rhythm is however a frequent finding. This does not appear to be of clinical significance within the period of observation studied.

P1156

Incessant polymorphic ventricular tachycardia in a patient with short-coupled variant of torsades de pointes successful treated by radiofrequency catheter ablation

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Background. Sudden cardiac death (SCD) and polymorphic ventricular tachycardia (PVT) can be related to short-coupled variant of torsades de pointes (TdP). We describe a successful management of incessant PVT in a patient with short coupled variant of TdP by radiofrequency catheter ablation (RFCA). Methods: A 16-year-old boy was referred for evaluation of an episode of SCD. Cardiac structural disease was ruled out by ECG (normal QT interval), echocardiogram, ventriculography and programmed ventricular stimulation. Holter monitoring revealed frequent short-coupled (>3000/day) ventricular premature beats (VPB) and polymorphic ventricular tachycardia (PVT). He was treated with verapamil, but 6 months later he had another episode of SCD. After a successful cardiopulmonar resuscitation he presented incessant PVT suggesting TdP unresponsive to antiarrhythmic drugs (AA). The first PVB that triggered PVT showed always right bundle branch morphology and left axis deviation. Electrophysiologic study was performed and ablation site was selected using pace-mapping and earliest activation criteria during VPB. RF energy was delivered on the left posterior fascicle, where a Purkinje potential was also observed and abolished PVBs and PVT. He received also an ICD and was discharged off AA. Two months later a new Holter showed frequent PVBs and PVT, but the first VPB showed left bundle branch morphology, which was successful ablated in the inflow right ventricle tract. During 6 months follow-up he has been asymptomatic with no AA and ICD discharges. Conclusion: 1 – Monomorphic premature ventricular beats may initiate PVT in patients with short-coupled variant of TDP. 2 – Ablation of these foci can be achieved by radiofrequency catheter ablation.

P1157

Mechanical reperfusion of right coronary artery occlusion complicating radiofrequency catheter ablation of postero-septal: acute and mid long-term follow-up

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Background: Although posteroseptal accessory pathways run close to the distal portion of right coronary artery (RCA) coronary artery injury is quite rare. However, late evaluation of the coronary circulation has not yet been reported. We report an acute RCA injury in a child due to radiofrequency catheter ablation (RFCA) treated with mechanical reperfusion and its long-term angiographic follow-up. Methods: A 12-year-old girl with Wolff-Parkinson-White syndrome refractory to medical therapy was referred for RFCA. Her ECG showed short PR interval and a negative delta wave in V1 and aVF. Electrophysiologic study confirmed posteroseptal accessory pathway. RF energy was delivered next to coronary sinus ostium with intermittent disappearance of preexcitation. After 6 RF applications (30–50s, 300, 60°C) with no impedance rise, ECG showed acute ST elevation in the inferior leads and ST depression in leads V1–V4 without changes in cardiovascular status. Coronary angiography was immediately performed and revealed total occlusion of distal RCA, which was opened mechanically with angioplasty guidewire and ST segment was normalized. There was a moderate elevation of plasma MB-CK. There was no further complication and after 1 year of follow-up, RCA angiography depicted patent RCA and normal circulation through posterior descending artery that had been opened mechanically. Conclusion: 1 – Acute RCA occlusion is a potential complication during RFCA of posteroseptal accessory pathways. 2 – Coronary angiography must be performed when ST change are noted and mechanical reperfusion should be performed.

P1158

Junctional ectopic tachycardia: a rhythm to fear!

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Junctional Ectopic Tachycardia (JET) is a potentially lethal cardiac arrhythmia that complicates the post-operative course of pediatric patients following surgical repair of congenital heart lesions. This narrow complex QRS tachycardia originates from an automatic focus in the AV node or surrounding tissue (Bundle of His) and is classically marked by AV dissociation. Unlike other well-known arrhythmias of AV dissociation, the junctional rate in JET exceeds the sinus rate by appearance, often in the first few hours to 1 days post cardiopulmonary bypass, presents the caregiver with a true clinical challenge, as hemodynamic decompensation often ensues. A complete understanding of this potentially life-threatening arrhythmia is crucial, since its management is typically resistant to usual forms of treatment. This presentation will review high risk pediatric patients, JET's etiology, pathophysiology, clinical picture, diagnostics and nursing and medical management (i.e. cooling, pacing and permissive drug therapy such as Amiodarone). Rhythm strips will clearly demonstrate JET's unique characteristics including p-wave dissociation and periodic versus regular beats. Current statistical data will be highlighted as well as a review of its management in the Critical Care Unit at The Hospital for Sick Children in Toronto, Canada.

P1159

The use of atrial anti-tachycardia device in patients with congenital heart disease, sick sinus syndrome and intra-atrial reentry tachycardia

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Introduction: Intra-atrial reentry tachycardia(IART) is associated postoperatively with Fontan operation, Senhr atrial switch operation for d-transposition of great vessels(d-TGA). Although anti-arrhythmics and RF ablations are sometimes effective therapy(Rx), proarrhythmia and recurrence remain management problems. In addition, IART remains a risk factor for sudden death from ventricular dysrhythmia. Moreover, the relative efficacy of surgical excision or anti-arrhythmic Rx as compared to defibrillator implantation for treating ventricular dysrhythmia remains unclear in these pts. A new anti-atrial tachycardia & defibrillation device, Jewel AF(JAF), has been used with success in adults without congenital heart disease(CHD) for the Rx of atrial fibrillation(AFL), atrial flutter(AT) and ventricular dysrhythmia. Method: 2 patients, S/P Fontan or atrial switch for dTGA and had at least 2 of the conditions below received JAF: 1) failure of or intolerance(thyroid toxicity) to multiple anti-arrhythmics, 2) severely limited venous access(interrupted IVC and baffled hepatic vein) for RF ablation, 3) indication for atrial pacing, 4) indication for ventricular defibrillator implantation, 5) parental choice against AV nodal ablation. Epicardial pacing leads and posterior patches were used. Pts were followed for 5 and 3 months respectively. Results: Successful conversion of IART was found after 36 of 39 (92%) atrial burst Rx delivered. No inappropriate AT/AF detection due to far-field sensing, as reported in the adult population, was observed. 2 episodes of 1:1 conducted IART were defibrillated (successfully) prior to atrial burst Rx due to shorter atrial ventricular effective refractory period than the lowest programmable R-R SVT limit(240ms) despite digoxin and amiodarone Rx. Addition of beta-blocker to

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

enhance AV nodal blockade resulted in no further ventricular defibrillation for 11 consecutive IART. One IART event was preceded by atrial burst RR, and followed by successful ventricular defibrillation. We conclude that JAT provided effective therapy for most of the IART episodes in two patients with CHD.

P1160

Transient arrhythmias after cardiac surgery in neonates and children

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Purpose: to analyze distribution and risk factors of transient arrhythmias (AR) after pediatric cardiac surgery. Patients and methods: in 392 children and 88 neonates, 24h-Holter-ECG was recorded preoperatively, from the immediate postoperative (po) period on for 72 hours, and 2 weeks po. Following AR were observed supraventricular (SV) and ventricular (V) extrasystoles (ES), SV and V tachycardia (SVT and VT), accelerated junctional rhythm (AJR) and junctional ectopic tachycardia (JET). AVB2 and AVB3. Interleukin (IL)-6, and IL8, as markers of systemic inflammation, were determined at the end of CPB, 4h and 24h po in 80 unselected patients. Results: Overall incidence of AR was 20% preoperatively, 75% on the 1st, 30% on the 2nd, 31% on the 3rd po day and 21% 2 weeks po. Incidence of AR on po day 1 was significantly lower in neonates than in children (p<0.0001) with lower incidence of SVES (18 vs 36%), VES (7 vs 19%) and VT (3 vs 13%). In contrast, SVT (7 vs 32%), AJR (15 vs 38%), JET (8 vs 54%), AVB2 and AVB3 (1% respectively) were not less frequent. In children logistic regression showed that older age and longer duration of CCA were risk factors for AR. TCPC-operation and higher IL6 and IL8 levels for AJR and closure of non isolated VSD, and TCPC for JET. In neonates, VSD-closure in addition to arterial switch operation was a risk factor for AJR and JET. Conclusions: Transient AR after cardiac surgery are common and more frequent in children than in neonates. TCPC operation in children and VSD-closure in neonates are risk factors for AR, particularly for AJR and JET. The release of inflammatory mediators is likely to be an additional risk factor for AR, as in adults.

P1161

Perinatal HIV and associated conduction abnormalities

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Background: Perinatal HIV is associated with cardiac complications particularly left ventricular (LV) dysfunction. Little is known about rhythm/conduction disturbances in this group. This study examines the ECG disturbances seen in conjunction with perinatal HIV in the pediatric population. Methods: 12-lead electrocardiograms (ECGs) and echocardiographic reports of patients with perinatal HIV were reviewed. Patients with normal cardiac anatomy and function who underwent ECG and echocardiographic evaluation served as controls. Results: Normal ECGs and echocardiograms were seen in 28/47 (59%) of the HIV patients, depressed LV function in 6 (13%) and rhythm/conduction disturbances in 15 (32%). These included first degree heart block (1° HB) (5 patients), intraventricular conduction delay (IVCD) (4), sinus bradycardia (3), ectopic atrial pacemaker (1), sinus tachycardia (1), atrial premature contractions (1) and superior QRS axis (1). None of the patients with 1° HB demonstrated LV dysfunction. Of the 38 controls, 8 (19%) had rhythm/conduction disturbances including IVCD (5 patients), ectopic atrial pacemaker (2) and left atrial enlargement (1). There was no difference in the age or sex distribution between the two groups. The number of rhythm/conduction disturbances was significantly greater in the HIV group as compared to control (p=0.03). In particular, the presence of 1°HB was significantly greater in the HIV group (p=0.02). There was no difference between the two groups regarding the other ECG disturbances. Conclusion: Conduction disturbances particularly 1°HB despite normal LV function, occur in a significantly greater proportion of the pediatric HIV population. This may be secondary to inflammatory infiltrates of the conduction tissues as seen with rejection in transplanted hearts. Close monitoring is necessary to assess for progressive disturbances that might lead to symptomatic arrhythmias, cardiomyopathy or sudden death.

P1162

Clinical significance of variability of QT dispersion in Long QT syndrome

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QT dispersion (the difference between the shortest and longest QT intervals on 12-leads surface ECG) may reflect a risk of ventricular arrythmias and cardiac morbidity. However, the clinical significance of variability of QT dispersion (QTd) in the patients with Long QT syndrome (LQTS) is unknown. Method: We studied 5 patients with LQTS (2 male and 3 female, age ranged 9.8±6.8 year) and 10 healthy subject (6 male and 4 female, 7±5.8 years) as a control (CON). LQTS patients were divided into two groups: 3 of 5 had cardiac events at rest (group A), 2 of 5 had cardiac events during exercise (group B). QT intervals of 12 leads in every one minute were automatically measured for 24 hours by computerized QT analysis system (QT guard Marquette Medical System) from which QTd and QTc dispersion (QTcd) were calculated. We also calculated QTd variability from mean QTc dispersion (MQTcd) and standard deviation of QTcd (SDQTcd), both during day time(D, 12:00–18:00) and at night (N, 0:00–6:00). Results: In group A, both D-MQTcd and N-MQTcd were longer than those in control group, and so as D-SDQTcd and N-SDQTcd, while, in group B, D-MQTcd, N-MQTcd, D-SDQTcd and N-SDQTcd were same as those in control groups. D-SDQTcd is approximately 2 times larger than N-SDQTcd in group A. Conclusion: The patients with LQTS who have cardiac events at rest, have a greater dispersion of ventricular depolarization time and QTd variability especially in day time. The variability of QT dispersion may provide an information about the difference between LQT1 and LQT2.

P1163

Clinical observations on polymorphic ventricular tachyarrhythmias in the absence of structural heart disease in the youth.

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Background: Polymorphic Ventricular tachyarrhythmias (PVT), namely catecholaminergic ventricular tachyarrhythmia (CVT) occur in young healthy children and may cause syncope or sudden cardiac death (SCD). Methods: This study was designed to evaluate distinct clinical characteristics in six patients presenting PVT with: age of first syncope, age of diagnosis, and rate of syncope and cardiac arrest. Results: The age of first syncope was 9.3 + 4.6 yrs and the diagnosis was only achieved at 14.1 + 5.4 yrs. Despite the high number of syncopal episodes of 6.0 +5.0 yrs and a 50 % rate of cardiac arrest in this population an 'epilepsy' was first misdiagnosis in 50 % of cases. The diagnosis was easily achieved during a 24 hs ambulatory Holter monitoring or treadmill testing. Conclusions: 1- The current study demonstrates that patients presenting syncope episodes should be seen by a cardiology in order to rule out VT as the cause of potential malignant symptoms. 2- The diagnosis of CVT can easily be done with a 24hs Holter monitoring.

P1164

Asymptomatic ventricular pre-excitation in children and adolescents: a 15-years follow-up study

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Background. Diagnostic assessment and treatment have been described in detail in symptomatic Wolff-Parkinson-White syndrome, but little information exists about significance and prognosis of discovering ventricular pre-excitation (VPE) on a routine ECG in asymptomatic children. Aim of the study was to examine retrospectively the follow-up of a cohort of children with asymptomatic VPE, referred during 15-years time to our Unit. Methods: Forty-one patients (18F/23M) found incidentally to have VPE on a 12-lead ECG, were referred to our Division between 1985 and 2000. Patients were considered asymptomatic if they had no documented tachycardia and no history of palpitations. All the patients underwent clinical examination, Echocardiogram, 24-hour ECG Holter and, when possible, exercise test. In the absence of symptoms, patients were seen in outpatient clinic at 6–12 month intervals after the presentation. Loss of VPE was defined as the absence of electrocardiographic signs of VPE for more than 3 consecutive controls. Results: Age at presentation varied from 1 month to 17 years. Follow-up time ranged from 6 months to 15 years. Ten patients (24%) exhibited congenital heart disease together with VPE. In 25 patients the pathway localisation, according to Fitzpatrick criteria, was right (1 postero-lateral, 8 antero-lateral, 8 antero-septal, 3 midseptal and 5 postero-septal), in 16 patients the pathway localisation was left (7 antero-lateral, 7 postero-septal and 2 postero-lateral). During

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

the follow-up 13 patients were found to have intermittent VPE and 4 to have complete fusion VPE. During follow-up 6 patients were referred to the Clinic with symptomatic supraventricular tachycardia and underwent treatment. Fourteen patients underwent electrophysiological study. 6/14 patients(40%) experienced sustained supraventricular tachycardia. Conclusion: VPE found on routine ECG in a healthy children has generally an excellent prognosis. However as changes in the electrophysiologic properties of the accessory pathway may alter the risk of arrhythmia, great attention is required in the follow-up.

P1165

QT and corrected QT dispersion in children with aortic valve anomaly.

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Increased dispersion of repolarization may contribute to an increased risk of ventricular arrhythmia. The aim of the study was to asses QT and QTc dispersion and type of arrhythmia in children with congenital aortic valve anomaly. 61 children (46 boys and 15 girls), aged 5 to 17 years, participated in this study. QT and QTc dispersion (dQT, dQTc) defined as a difference between maximal and minimal QT and QTc intervals occurring in any of 12 leads surface ECG were measured manually by two observers. The QTc interval was calculated using Bazett's formula. Arrhythmia was diagnosed in Holter ECG monitoring. 42 normal age-matched, subjects served as a control group. Patients were divided into 3 groups: First group consisted of 15 children with isolated aortic stenosis (AS), second of 46 children with aortic stenosis and insufficiency (AS+IA). Non-sustained ventricular tachycardia was diagnosed only in 1 patient with AS+IA in whom dQT was 0,06s and dQTc 0,08s. No complex arrhythmia was diagnosed in AS and control group. Comparison of dQT and dQTc values in following groups: AS (table 1) and AS+IA (table 2) versus control group and AS versus AS+IA (table 3) group, is shown below: Table 1 Table 2 Table 3 Conclusions: 1. QT and QTc dispersion were higher in children with SA and SA+IA compared to control group. 2. No differences of QT and QTc dispersion between SA and SA + IA leak were found. 3. Increased QT and QTc dispersion in children with aortic valve anomaly may indicate increased risk of ventricular arrhythmia.

P1166

Heart rate variability in children with congenital aortic valve pathology.

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The aim of the study was to analyse heart rate variability (HRV) in children with congenital pathology of the aortic valve. 25 children aged 5 to 17 years participated in this study: 9 of them had isolated aortic stenosis (AS), 16 had aortic stenosis with aortic regurgitation (AS – AR). All children were in I and II group according to NYHA. In all patients diagnosis was established by Echo-2D+Colour Doppler imagine. Ejection fraction was within normal range in all children and ranged from 60%–84%. 24 hour ECG Holter monitoring was performed in all to analyse arrhythmia and HRV. Only one child had complex arrhythmia – ventricular tachycardia. Time domain analysis of HRV was performed. Received values of HRV were compared (using Student test) with the values of the control group, which consisted 20 healthy children aged 6 to 15 years. The results are shown in table 1: Table 1 Conclusions: 1. In children with isolated aortic stenosis and with aortic stenosis coexisting with aortic regurgitation parameters of time domain analysis were significantly decreased. 2. Low heart rate variability may indicate endanger of cardiac arrhythmia.

P1167

Behavior of the evoked response for automatic pacing threshold adjustment (Autocapture) in combination with steroid eluting epicardial leads

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The efficacy and safety of beat-to-beat capture confirmation and automatic output regulation of Pacesetter's Autocapture (AC)-algorithm had been confirmed for transvenous leads in an adult patient cohort, but its application for pacing with epicardial leads in infants with congenital heart disease is limited. To gain more information about the behavior of the evoked response (ER) this animal study was initiated. Method: In 11 Göttingen minipigs (7–23kg) an epicardial steroid eluting lead (Medtronic CapSureEpi 4968) was fixed on the right ventricle and connected with a Pacesetter Microny SR pacemaker (PM) prior to transvenous high frequency ablation of the av-node. During continuous ventricular pacing (120/min) monthly PM interrogation for 1 year was performed. As a high evoked response (ER) to lead polarization (LP) ratio ensures safe AC-function the progression of ER and LP were evaluated at: 0.21/0.37/0.49/0.77/1.5 ms pulsewidth (PW). Additionally pacing threshold (THRESH) at 0.21/0.49/1.5ms PW and sensing threshold (SENS) were determined. The table summarizes the data at PW 0.49 ms. The mean ER shows a stable behavior with time but there is significant interindividual variation. LP and PW show a direct proportional correlation. Conclusion: The combination of PMs with the Autocapture-algorithm and steroid eluting epicardial leads is possible but the interindividual ER-variability warrents individual verification of the ER and LP stability to ensure safe AC-function.

P1168

Volume expansion prevents tilt-induced syncope.

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Spatially resolved near-infrared spectroscopy (NIRS) was used as an objective, continuous, non-invasive measure of perfusion in children with a history of unexplained syncope. It assessed change in regional tissue oxygen saturation (rSO2) with upright tilt. During 15-minute 70° upright tilt, gastrocnemius and frontal cerebral cortical rSO2 each decreased an average 22% in 13 patients (6 requiring an additional isoproterenol challenge) who had a positive syncopal response. In contrast, only gastrocnemius rSO2 decreased (21.1±1%, P<.001) in negative responders (n=6). After normal saline volume loading (15 mL/kg) and repeat tilt-testing, no syncope was observed. Or, despite a similar magnitude of gastrocnemius desaturation, cortical rSO2 values in previously positive responders declined from supine baseline only an average of 6±6% (P<.001). These results suggest that 1) volume expansion can prevent postural mediated syncope and 2) NIRS reliably and objectively discriminates positive syncopal responders from non-responders. The benefit of volume expansion may occur by redistribution of hemoglobin through reduction of lower body venous pooling.

P1169

An original mathematical formula for computerized generation of normal 12-leads electrocardiographs and different cardiac diseases, a near view to perfect mathematical mapping of normal and abnormal electrocardiograms

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Introduction: "Mathematics is a construct of the human mind capable of reflecting in symbols the reality of physical and physiological events" (Dr. Barnaik Yaghoobiant) Automated interpretation of electrocardiograms has been put into wide use by less-experienced physicians and other health-care professionals. The generation and interpretation of normal and abnormal electrocardiograms through mathematical methods have been major areas of research for several decades and there already exists software capable of such generation and interpretation. If we consider the electrocardiograph to be a Cartesian plane, the data from each lead can be represented by a cyclic mathematical function correlating voltage to the independent variable of time. Voltage = f (time). The ECG strip, then, is simply a two-dimensional plot of the heart's electrical activity along the axis of time. This paper presents a new mathematical function, which, for the first time, is capable of generating normal 12-lead ECG's and nearly all patterns of abnormal electrocardiograms. Materials and Methods: Using the Quick-Basic programming language on the personal computer, the mathematical functions most closely approximating the morphology of electrocardiograms was found through trial and error. When the crude features of the formula were worked out, the coefficients were refined based on the known parameters of each lead that composed the 12-lead ECG. These parameters include the width of each wave (wn), height of each wave (hn), rate of each wave (rn), the phase difference of each wave relative to the second peak(R wave) of the QRS complex (delta n), the PR, QRS, and QT intervals, and the ST and PR segments. Using these parameters, the coefficients for generating normal 12-lead ECGs, and some known patterns of abnormal ECGs, were calculated. Results and Conclusions: The twenty-eight coefficients of the formula for generating a normal 12-lead ECGs are presented in a 12.28 matrix. A similar, but larger, matrix has been partially completed representing patterns of electrical abnormality common to some cardiac diseases – hopefully, this project will be complete in the near future. The basic function is flexible enough to

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

incorporate multiple abnormalities in a single electrocardiogram. In this dynamic, two – dimensional model of electrocardiography, each electrocardiographic permutation of disease can be approximated through the equation. The parametrical approach of the function allows for near-perfect replication of the electric signatures of cardiovascular disease. By correlating a patient's electrocardiograph with the function, the resultant equation can be compared with known equations of disease and a diagnosis made by an automated system. While the technology is very promising, the complexity of the real world of electrocardiography, may be difficult to simulate in the ideal language of mathematics. The efficacy and accuracy of this system in the clinical setting still requires much more research and testing to reliably serve our healthcare providers.

P1170

Single midline approach for intramuscular abdominal implantation of

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Background: There are many approaches and techniques for epicardial pacemaker implantation in children. Development of steroid eluting epicardial lead has offered new therapeutic and surgical opportunities. Methods: Single midline approaches was used for placement of both epicardial electrodes and pacemaker generator. The platinized porous-tipped steroid eluting epicardial leads were used in all patients. There were twelve children underwent these techniques. The mean age was 1.5 years (range, 2 days to 4 years). The mean operative time was 97 minutes (range, 50 to 90 minutes). Results: This approach was successful in all patients. The pacemaker worked well without any postoperative complications. Conclusion: Placement of the platinized porous-tipped steroid eluting epicardial leads with single midline approach together with intramuscular abdominal implantation of permanent pacemaker was rapid, simple and safe in children, especially in small neonate and infant.

P1171

Pacemaker implantation in the pediatric age group: anything new ?

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Background: Pacemaker implantation in children often necessitates an epimyocardial approach. Concerns about morbidity and unsatisfactory stimulation thresholds keep being raised. A single surgeon experience was analyzed. Methods: Between 6/1995 and 9/2000, 137 pacemaker operations were performed in 106 patients (age: 1 day – 19 years, median 6 years). Indications were: postoperative AV-block/sick sinus syndrome 66, bradyarrhythmia in univentricular heart disease (Fontan patients) 24, congenital AV-block 14, cardiomyopathy 2, DDIVVI, 2 AAI, and 52 DDD systems were implanted. In 7 older patients a transvenous route was taken. All others underwent a subxiphoid approach for one-chamber and (re-)sternotomy for two-chamber systems. In epimyocardial implantations, steroid-eluting stitch on electrodes were chosen for atrial, screw-in electrodes for ventricular stimulation. Results: There was no mortality or major morbidity and one wound infection. 4 children showed chronically elevated thresholds necessitating repeated operations. In 8 patients acute problems developed (electrode loosening / damage). For the epimyocardial systems stimulation thresholds at 0.5ms averaged 1.5V (range 0.5–4.2V) for the atrial and 1.2V (range 0.1–4.0V) for the ventricular electrodes. With the newest type of ventricular screw-in electrode (n=58) this has come down to 0.75V (range 0.1–2.0V). 50/52 non-chamber systems are functioning in DDD mode. Conclusions: With the small generators and optimized electrodes available today, excellent stimulation thresholds can be achieved with epicardial access, and pacemaker placement poses no technical problem even in small infants. Epimyocardial implantation shows no morbidity, nor even after reoperatory for two-chamber-systems, and remains our preferred technique.

P1172

P wave characteristics before and after trans-catheter closure of secundum atrial septal defects

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P maximum (Pm) and dispersion (Pd) have been used as predictors of atrial dysrhythmias in adults. The purpose of this study is to assess these parameters before and after transcatheter closure of ASD. Twenty patients (9 males, 11 females; mean(SD) age at procedure 15.0(16.8) years) with isolated ASD who had undergone transcatheter device closure (10 Amplatzer septal occluders and 2 Sideflex occluders) and 20 age- and sex-matched controls were studied. Resting 12 lead ECG was used to measure P waves from which Pm and Pd (difference between maximum and minimum P wave durations) were derived. Patients had significantly longer mean Pm (107.3(10.6) vs 85.8(12.3) ms) and Pd (40.3(12.4) vs 25.1(6.4) ms) before device closure compared to controls ($p<0.0001$). Significant reduction of mean Pm (99.2(11.0) ms) and mean Pd (32.2(8.8) ms) the next day, compared to the pre-procedural values ($p<0.05$) remained roughly unchanged up to 3 years of follow-up. Prolonged atrial conduction time and in-homogeneity of atrial conduction in patients with ASD were favorably altered by transcatheter closure.

P1173

Does autocapture compensate for smaller device batteries?

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Background: Smaller pulse generators facilitate the implantation of pacing systems in pediatric patients. Substantial stimulation energy savings were demonstrated with AutoCapture (AC) devices. We compared device volume, battery capacity, stimulation output and the experienced battery service life of conventional devices with corresponding data and calculated battery life of AC devices which have been used as replacement units. Methods: In 7 children, aged 48+/-34 months a conventional pacemaker (Dash 2, Relay 4, Marathon 1, Intermedics connected to epicardial pacing leads Medtronic 10366 Capsure Epi) was replaced by AC devices (Affinity 4, Regency 2, Integrity 1, St. Jude) due to battery depletion. Battery life calculation of AC devices was based on the actual percentage pacing, pacing mode (VVIR: 1, DDD: 4 patients), Holter acquired mean heart rate, lead impedance and pacing threshold as determined during 7+/-3 months follow up. Results: are table. High pacing threshold precluded AC controlled pacing in 1 child. Conclusion: AutoCapture does not only compensate but significantly extends battery service life despite smaller device size and lower battery capacity if compared to conventional devices.

P1174

Qrs axis in isolated perimembranous ventricular septal defect and influences of morphological factors on qrs axis

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To detect the frequency of left axis deviation in isolated perimembranous ventricular septal defect we retrospectively analyzed electrocardiograms of 59 patients, aged 8 months to 15 years. Patients were grouped into those having ventricular septal aneurysm formation (n: 20) and those who did not have ventricular septal aneurysm (n: 39). Patients with ventricular septal aneurysm were then stratified into two groups according to the presence of left ventricular-to-right atrial shunt. Four hundred age and sex-matched healthy children served as control group. We found that 12 (30.3 %) of 59 patients with isolated perimembranous ventricular septal defect had a left axis deviation. Left axis deviation was more prevalent in patients with ventricular septal aneurysm (40 %) than without ventricular septal aneurysm (10.2 %) ($p < 0.05$). We also found that incomplete right bundle branch block pattern was more frequent in patients with ventricular septal aneurysm (55 %), especially who had left ventricular-to-right atrial shunt (75 %) than without ventricular septal aneurysm (10.2 %) ($p < 0.0001$). However, we could not find significant difference between patients with or without left ventricular-to-right atrial shunt for the incidence of left axis deviation and incomplete right bundle branch block pattern. Localization of perimembranous ventricular septal defect was not found to have an effect on frequency of left axis deviation and incomplete right bundle branch block pattern in this patient group. In patients with clinical findings of ventricular septal defect, the existence of left axis deviation especially if it is associated with incomplete right bundle branch block pattern should raise the possibility of perimembranous ventricular septal defect with ventricular septal aneurysm formation.

P1175

Long qt, sudden infant death syndrome and maternal alcoholism– a hypothesis

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https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Prolonged QT-interval in ECG in the newborn is considered to be a risk factor for the sudden infant death syndrome (SIDS). Suspected reasons for QT-prolongation are genetic variants either familiar or as spontaneous mutation. Newborn babys of alcohol dependent mothers might show QT-prolongation during their withdrawal syndrome after delivery. To rule out a longlasting effect on QT duration ECG's of 313 children with clinical signs of alcoholic embryopathy were analyzed. Further a case of a newborn whose mother was alcohol addict is presented: Because of tachycardia on the third day of life several ECG were obtained. On the third day of life frequency corrected QT-interval (QTc) was 0,48s. During the next days QTc normalized without any treatment. We presume an association of maternal alcoholism QT-prolongation and SIDS. As long as alcoholism is a hidden

P1176

Qt duration: beat to beat holter-analysis in healthy children

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Normal values for QT duration in routine-ECG exist, the upper limit is 0,44 s. No data was available yet for QT dynamics in holter ECG. Today software for automatic analysis of QT duration in 24-h-ECG exists. We use Marquette-Hellige system MARS 8000 to analyze QT duration in healthy children aged 6–14 years. Corrected QT interval (QTc) during sleep, awakening and physical activity are presented. The results will be discussed.

P1177

Steerable stylets for implanting pacemaker leads in congenital heart disease patients

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Children with congenital heart disease have challenges in lead implantation due to patient size, venous anatomy and capacitance, and structural heart abnormalities. Nonstandard endocardial lead positions are often necessary for optimal fixation and pacing performance and intraoperative custom shaping of lead stylets frequently becomes necessary. A steerable stylet may have value for this unique patient population. The purpose of the study was to review the subjective utility of a transvenous steerable stylet as an implantation aid. Methods: All transvenous pacemaker implants procedures in children and adults with congenital heart disease at a single institution during a 3-year period from November 1997 to November 2000 were retrospectively reviewed. The LOCATOR steerable stylet (St Jude Medical CRMD) was utilized. Each eligible lead (Tendril 1388 or Tendril SDX 1488, St. Jude Medical CRMD) took longer than 15 minutes to successfully fixate using standard techniques with manually custom-shaped stylets. Measured parameters included subjective value of each manual intercostal anatomic dissection, electrical implant characteristics, and procedural variables including lead-related complications. Results: A total of 274 leads in 232 patients (age 2–52, mean 15 years) were implanted during the period. LOCATOR-comparible (Tendril) leads were utilized 77 times in 53 patients and comprise the analysis group. Leads were fixated within 15 minutes each with acceptable pacing thresholds and intrinsic sensing in 72/77 leads (94%) using manual stylets, whereas LOCATOR steerable stylets were utilized in 5 patients (mean age 13 ± 3 yr, p=NS vs manual stylets). All 5 were active-fixation atrial lead implants in patients with repaired congenital heart disease. All LOCATOR-assisted lead implants were successful with good intraoperative pacing characteristics and no acute complications. Conclusions: Although not necessary for the vast majority of pacemaker patients, a steerable stylet may have utility as a tool for optimizing pacing lead implantation in particularly challenging procedures. This study demonstrates the feasibility of using this implant tool in children and congenital heart disease patients.

P1178

Evaluation of cardiac reserved function by dobutamine stress qt dispersion in children after anthracycline therapy.

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Purpose: To estimate the late Anthracycline (ATC) cardiotoxicity by dobutamine (DOB) stress QT dispersion (QTd) in patients with cancer. Subjects: 13 patients (aged 3–18 years) were subjected(ATC group), and 5 volunteers were control (aged 8–28 years, control group). ATC cumulative doses were 80–760 mg/m2. Methods: Standard 12 lead electrocardiograms were recorded. QTds were measured at rest, and DOB 5 and 30 mg/kg/min administration. Moreover, left ventricular ejection fraction (EF) was measured and calculated by 2-DE echo. We compared QTd and EF values at rest and DOB30 mg/kg/min administration between the two groups. Result: At rest, QTd of the two groups were similar. At DOB 5 and 30 mg/kg/min administration, as showing the figure, QTds in ATC group were significantly greater than those in control group. Conclusions: DOB stress QT dispersion will be helpful method for detection of late Anthracycline cardiotoxicity, especially patients who cannot tolerate physical ex

P1179

Estimation of late anthracycline cardiotoxicity by dobutamine stress qt dispersion in patients with cancer.

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Purpose: To estimate the late Anthracycline (ATC) cardiotoxicity by dobutamine (DOB) stress QT dispersion (QTd) in patients with cancer. Subjects: 13 patients (aged 3–18 years) were subjected(ATC group), and 5 volunteers were control (aged 8–28 years, control group). ATC cumulative doses were 80–760 mg/m2. Methods: Standard 12 lead electrocardiograms were recorded. QTds were measured at rest, and DOB 5 and 30 mg/kg/min administration. Moreover, left ventricular ejection fraction (EF) was measured and calculated by 2-DE echo. We compared QTd and EF values at rest and DOB30 mg/kg/min administration between the two groups. Result: At rest, QTd of the two groups were similar. At DOB 5 and 30 mg/kg/min administration, as showing the figure, QTds in ATC group were significantly greater than those in control group. Conclusions: DOB stress QT dispersion will be helpful method for detection of late Anthracycline cardiotoxicity, especially patients who cannot tolerate physical ex

P1180

Evaluation of permanent pacing systems in children with holter monitoring

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Background: Circadian variations in pacing and sensing thresholds as well as the appropriateness of the actual device programming may not be assessed solely with regular pacemaker follow-up. We therefore evaluated pacing system functions in children with Holter monitoring. Methods: In 70 children, aged 8 days–56 months, 92 pacing systems (22 replacement systems) were implanted. Various endocardial leads were used in 29 and epicardial leads in 63 systems. Single chamber (VVIR) systems were implanted in 45 and dual chamber systems in 47 cases. Congenital cardiac defects were present in 44 children. Indications for pacemaker implantation were complete atrioventricular block in 48, sinus node disease in 20 and long QT syndrome in 2 children. During a median follow-up of 10 months (range 0.1–110 months) 197 Holter examinations were performed (60 post pacing system implantation, 137 during regular follow-up). Holter and pacemaker follow-up data were retrospectively analyzed for pacing system dysfunctions and subsequent device reprogramming. Results: Pacing system dysfunctions were found in 51/197 (26%) Holter (post implantation 18) and consisted of atrial sensing dysfunction in 22, ventricular sensing dysfunction in 15, ventricular pacing dysfunction in 8 and atrial pacing dysfunction in 3 Holter. Pacing system dysfunctions were corrected after 37 Holter by device reprogramming. The safety of pacing was not jeopardized after 14 Holter where device reprogramming was not possible. Cardiac defects and epicardial leads were no risk factors for pacing systems dysfunctions. Conclusion: Pacing system dysfunctions in children are frequent and can be accurately analysed by Holter monitoring. Most dysfunctions can be corrected by device reprogramming. Thus, Holter monitoring is a valuable tool to assure the correct function of a pacing system.

P1181

Characteristics of atrioventricular node in atrioventricular nodal reentrant tachycardia and dual atrioventricular nodal pathway in children and adolescents

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https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

We investigated the characteristics of atrioventricular node in the electrophysiological response to programmed atrial and ventricular stimulation between common form of atrioventricular nodal reentrant tachycardia (AVNRT) and dual atrioventricular nodal pathway (DAVNP) in children and adolescents. We also examined the change of the response in AVNRT before and after radiofrequency catheter ablation (RFCA). 26 AVNRT patients (mean age 14 years, range 6–26 years) compared to 11 DAVNP patients without tachycardia (mean age 12 years, range 6–25 years). We found AVNRT patients have a longer anterograde fast pathway effective refractory period (ERP) (median 391 vs 320 ms, p=0.02) and a shorter retrograde VAERP (median 264 vs 322 ms, p=0.01). 2 DAVNP patients have no retrograde VA conduction. There was no difference in anterograde slow pathway ERP (median 268 vs 257 ms) and AH interval at anterograde fast pathway ERP (median 265 vs 256 ms). After successful RFCA in all AVNRT patients, anterograde fast pathway ERP were shortened (median 391 vs 298 ms, p=0.01). In 9 patients, anterograde slow pathway remained and one echo occurred, AH interval at anterograde fast pathway ERP didn't change. We found a difference of the electrophysiological characteristics between AVNRT and DAVNP. Electrophysiological changes of slow pathway after RFCA in AVNRT were supposed to make no influence o

P1182

Percutaneous cardiac pacing with an epicardial lead in a newborn with congenital heart block

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Complete heart block in child with structural normal heart is not very often disease. Second degree atrioventricular block is also possible to find in a newborn. But there is a big possibility to reverse it on a complete heart block after 3 month. Aim: To show how the effective cardiac pacing improve the symptoms of severe heart failure in small children. Material: Newborn 46 cm, 2700g have a second degree congenital heart block structural normal heart, min heart rate was 55 bpm, LV EF was 0,58, symptoms of heart failure as edema, liver increasing, wheeze, breath rate 45 per min. Mother and a baby have a high value of anti Ro (SSA) antibodies. We decided to implant a permanent pacemaker, because of the high risk of complete heart block formation and the presence of severe heart failure. Operating technique: We used the left thoracotomy to achieve the LV apex, where were 2 ends of the bipolar lead fixed (Medtronic capsure EPI 4968 epicardial lead) and pacemaker 'MicromySR+' from 'Pacesetter' was implanted in subpectoral pocket on the left side. Acute stimulation parameters were: sensitivity – more than 20 mV, pacing threshold – 0.5V. Results: In 6 month remotely pay concerns, and the pacing threshold became 0.9 V. There were no symptoms of heart failure. Pacing rate 120 bpm. Conclusion: Newborns and infants with a high degree congenital heart block need permanent cardiac pacing if they have a bradicardia below 60 bpm and severe heart failure.

P1183

Safety of Electrophysiologic Interventions in Pigs with an Amplatzer Septal Occluder

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The Amplatzer Septal Occluder (ASO) is constructed from a mesh of metal wire. There are concerns that the presence of such a device can confound electrophysiologic interventions such as direct current shock (DCS) or radiofrequency ablation (RFA), by inducing injurious currents or dispersing energy delivery. PURPOSE: To evaluate the effects of DCS and RFA on pigs with an ASO. METHODS: (#1) We performed DCS on 4 pigs who had undergone ASO implantation. Each pig received 2 'low-dose' shocks (mean 2.2 joules/kg) and 1 'high-dose' shock (mean 4.8 joules/kg). Immediate ECG effects were monitored; acute (2 pigs) and chronic (2 pigs) tissue effects were observed by gross and microscopic examination. (#2) We also performed RFA in 3 other minipigs, 2 of which had chronic ASO implants. Eight to 10 energy applications were made in each pig, at analogous atrial sites both remote from and close to the ASO. Half the lesions were 'low-temperature' (mean 52.1°C) and half were 'high-temperature' (mean 64.8°C). Gross and microscopic histologic examinations of the area were performed acutely (1 pig) and after 1 month (2 pigs). RESULTS: (#1) All animals tolerated DCS without developing secondary arrhythmias. All ECG tracings returned to baseline within 2 minutes of DCS. No necropsy specimen demonstrated acute burn or chronic scar. (#2) The presence of an ASO did not affect energy delivery during RFA, and did not result in altered RFA lesions. CONCLUSIONS: DCS and RFA are safe and feasible in pigs with an implanted ASO.

P1184

Outcome of Pediatric Patients with Transvenous Pacemaker or Lead Revision

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Transvenous cardiac pacing has become standard practice for children who require pacing therapy. During long-term follow-up many of these children may require revision or explantation of the pacing. Our aim was to evaluate the outcome of transvenous pacing in children who underwent pulse generator or lead revision. Between 1990–2001, 26 patients (pts) with transvenous pacing (17 male, 9 female) were enrolled in this study. The mean age of the pts at initial pacemaker implantation was 7.4±3.9. The indications for pacing were complete atrioventricular block in 22 pts and sick sinus syndrome in 4 pts. Passive fixation leads were used in 18 pts and active fixation leads in 8 pts. The leads were inserted into the heart via right subclavian vein and left subclavian vein in 21 and 5 pts, respectively. They were placed into the right ventricle (24 pts) and the right atrium (2 pts). Twenty-three pts were paced in the VVI mode, 2 in AAI mode and one in VDD mode. The mean time from implantation to revision was 52.1±26 months. In fourteen of 26 pts, pulse generators were revised due to elective replacement and lead revision was performed in four pts. Indications for lead revision included high threshold (1), lead migration (2) and infection (1). In five pts, both battery and lead were replaced because of elective replacement (3), lead migration (1) and infection (1). After lead replacement, 2 pts were paced in VVI mode, one in AAI mode and one in VDD mode. Lead revising procedure of 9 patients was made by simple traction method through subclavian vein. In 5 pts, leads were retained in the subclavian vein, which were of passive fixation type. Twice replacement of battery and lead were done electively in two pts. One of these leads was removed by open heart surgery method and the other by simple traction. Pacemaker explantation was performed in four pts. 3 of them had normal sinus rhythm and the other had recurrent pocket infection. In conclusion, 1) Pacemaker replacement in children can be performed safely. 2) Simple traction is a useful method in most of the patients.

P1185

Combined intervention and radiofrequency ablation in congenital heart disease.

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Arrhythmias can co-exist with structural congenital heart disease or they may become acquired as a result of previous surgical procedures. Arrhythmia therapy using radiofrequency ablation has evolved over the past 10 years and catheter intervention techniques have reached new heights due to advanced technology. We described combined procedures in a group of patients who had arrhythmias as well as mechanical problems both of which required treatment and were conducted during the same session. Seven patients with congenital heart disease were treated for an arrhythmia and a structural defect. Five were female and 2 male and the mean age ranged from 4–26 years (mean 14.8 years). The congenital heart disease consisted of 2 patients with Ebstein's anomaly and atrial septal defect, 2 patients with obstructed superior vena cava following Mustard operation for transposition of the great arteries, 2 patients with atrial septal defect II with concomitant patent arterial duct, and 1 patient with coronary artery fistula. The arrhythmias which were ablated consisted of atrial flutter in 2 patients, AV nodal re-entry tachycardia in 2 patients and accessory pathways in 4 patients. The interventional procedures consisted of stent implantation in 2 patients, coil closure of patent duct in 1, atrial septal defect closure with Amplatzer device in 3 and coil embolisation of coronary fistula in 1. There were no complications or deaths but 1 patient with atrial flutter experienced recurrence but with better control. Combined radiofrequency ablation and intervention is safe, feasible and desirable. It provides quality care which is also recommended to minimise vessel trauma in when vessel access is limited because of congenital anomalies or previous cannulation.

P1186

Influence of age and underlying physiology on dispersion of depolarization and repolarization in infants.

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Studies have demonstrated that alterations in ventricular loading conditions may alter the dispersion of depolarization & repolarization (DDR). Little is known about the effects of the underlying physiologic condition (UPC) and

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

age, especially in the very young, on DDR. A retrospective review of all pre-surgical electrocardiograms (ECGs) was performed between 1/92 and 1/00 from patients with Tetralogy of Fallot with Absent Pulmonary Valve (TOFAPV, eg Pressure & Volume Load). Two ECGs from patients with Tetralogy of Fallot alone (TOF, eg Pressure Load) were matched on surgical date along with two normals (NL) matched on age. Shortest and longest QRS, QT, QTc, JT & JTc intervals were measured on the 30 ECGs from patients with TOFAPV, 20 ECGs from TOF patients and 20 ECGs from normals. Results: Median age at time of ECG was 1, 55 and 2 days for TOFAPV, TOF, and NL groups respectively. Univariate analysis demonstrated no significant difference in dispersion of QRS, QT, QTc, JT or JTc between ECGs from TOFAPV and normal patients, all of whom had the same age. Dispersions of QT (p=0.04), QTc (p=0.05), JT (p=0.01) & JTc (p=0.02) were significantly greater in TOFAPV patients as compared with TOF. Only JT (p=0.02) & JTc (p=0.02), however, were significant when comparing TOF vs NL patients (See Table). Conclusions: Age may play a more significant role than LVPC in determining DDR.

P1187

Fifteen Year Experience with Endocardial Pacing in Infants and Children

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While transvenous pacing in infants and children has been described, little long term follow-up is available. Accordingly, we reviewed endocardial pacing for 1985–2000 at Babies and Children's Hospital in 186 patients 0–18 years of age at the initial operation. Thirty-nine of these were less than two years old and 17 less than six months of age. Fixed-screw 7 french unipolar leads (cephalic cutdown), and an intracardiac loop allowing growth were employed. There was one death in this series, due to anesthesia induced heart failure. Kaplan-Meier analysis of 94 right ventricular leads revealed 91% freedom from lead replacement at three years, 90% at six years, and 82% at nine years. This included elective lead replacement at the time of generator replacement because of patient growth. Stratified by age at implant, lead survival at three years was 95% for patients greater than six months of age and 59% for patients less than six months of age. Freedom from lead failure in 46 right atrial leads was 76% at 7 years. Average RV pacing threshold at reoperation was 1.1±0.3 volts and 2.2±0.8 milliamps, with an average R wave of 9.8±5.2 millivolts. We conclude that endocardial pacing in infants and children with this technique produces excellent clinical results.

P1188

Changes in pressure overload in congenital heart disease: evidence for mechano-electrical feedback in humans

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Background: Basic research and animal experiments have shown electrophysiological changes during or after changes in mechanical loading. Electrical instability, following mechanical stretch, has been observed as development of afterdepolarization and in terms of increased dispersion of refractoriness and repolarization. The aim of the study was to evaluate ventricular repolarization time indexes following acute changes in left ventricular pressure in humans. Material and methods: The study group comprised 35 pts (17M/18F, aged 2 days – 24 years) affected by severe congenital aortic stenosis and 25 pts (15M/10F, aged 6 months – 11 years) with severe coarctation of aorta who underwent respectively successful balloon valvuloplasty and angioplasty. Ventricular repolarization was evaluated before and after the procedures both in terms of absolute measures (JTc, QTc) and in terms of dispersion across the myocardium: QTc dispersion (QTcD), JTc dispersion (JTcD) and T-peak to T-end interval (Tp-Te). Results: Patients with severe aortic stenosis showed following balloon valvuloplasty a significant decrease in ventricular repolarization indexes: JTc (353.2±27.7vs341.5±25.7; p=0.002) and QTc (447.2±22.2vs417.2±19.7, p=0.01) and dispersion of ventricular repolarization indexes: QTD (44.3±16.5vs30.2±17; p=0.0002), QTcD (60.5±21.1 vs39.1±20.7, p=0.0001), JTD (48.8±25.6vs32.9±18.1, p=0.01), JTcD (64.4±25.4vs42.5±23.1; p=0.0002) and Tp-Te (114±13.4vs108.5±14.9, p=0.04). Similarly, patients with severe coarctation of aorta showed following balloon angioplasty a significant decrease in ventricular repolarization indexes: JTc (340.1±18.7vs330.9±23.6, p=0.05) and QTc (443.3±17.4vs 433.9±13.1, p=0.01) and dispersion of ventricular repolarization indexes: QTD (48.9±10.5vs33.6±10; p=0.0001), QTcD (70.3±24vs49.2±16, p=0.005), JTD (63±41.3vs38±13.2, p=0.01), JTcD (93.2±67.1vs52±15.7, p=0.008) and Tp-Te (105±19vs98.6±24.5 p=0.05). Conclusions: Changes in haemodynamic loading can produce electrophysiological effects in humans. Acute reduction in left ventricular pressure overload, following balloon valvuloplasty and angioplasty, decreases electrical instability, as expressed by ventricular repolarization time shortening both in terms of absolute measures and in terms of depolarization across the myocardium.

MAY 31 Time: 11:30–12:30

Session 7
Basic Research, Biology/ Experimental Teratology, Cellular and Molecular Biology, Vascular Biology

P1189

Age-related differences of direct cardiac effects of cisapride: a narrower safety range in the young heart

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Background: Cisapride is a prokinetic agent and is widely used to treat the gastrointestinal motility-related disorders in adults and children. However, it has been associated with QT prolongation, torsades de pointes and cardiac arrest. The cellular mechanism for these events may be related to a blockade of IKr. Methods: The direct effects of cisapride on cardiac conduction properties were assessed in the neonatal (< 7 days) and adult (> 3 months) rabbit hearts with Langendorff-perfusion. Results: Cisapride at clinically relevant dose (0.1 _M) could significantly prolong the refractoriness of the His-Purkinje system and the conduction through this system at shorter coupling interval. The recovery curve of His-Purkinje system (H2V2 versus H1H2 relation) was shifted to right dose-dependently. The corrected QT interval was also prolonged. The degree of prolongation of these parameters was significantly more in the neonates than in the adults. The ventricular refractory period was lengthened only in the neonates. At higher concentrations (0.3 and 1 _M), cisapride caused 2:1 AV block below the His bundle in the neonates but not in the adults. The AV nodal refractory period was also prolonged and its recovery curve was shifted to right in both the neonate and the adult. Conclusion: In the neonatal heart, cisapride at clinically relevant dose prolonged the refractoriness of the His-Purkinje system and the ventricular tissue, and consequently the QT interval. Such modification may even progress to pseudo-AV block at higher concentrations. Such susceptibility to cisapride may indicate a narrower therapeutic safety range in the young heart.

P1190

Expression of connexin cx 43 in children with tetralogy of Fallot.

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Gap junctions created by a family of connexin proteins play a key role in the development of human heart. The right ventricular outflow tract (RVOT) abnormalities were shown to be linked with increased or decreased level of expression of connexin Cx43. The RVOT narrowing, as well as hypertrophy of the right ventricle are observed in tetralogy of Fallot (TOF). The aim of the present study is to determine the organization and expression of connexin Cx43 on the surface of cardiomyocytes obtained during surgery for TOF. These prepared cells were compared with cardiomyocytes collected from patients without RVOT pathology. Cardiomyocytes isolated from tissue biopsy (14 patients with TOF, aged 3 weeks – 6 months, weighing 4 – 6.2 kg and 8 patients aging 2 weeks – 5 months, weighing 2.8 – 6.7 kg as controls) were cultured on collagen substratum, fixed and labelled with anti-human Cx43 antibodies for laser scanning confocal microscopy. Suspension of the cardiomyocytes was prepared for flow cytometry. The 3D images of Cx43 fluorescence revealed a three-dimensional distribution of connexons on the surface of a single cardiomyocyte. The level of expression of Cx 43 was determined by flow cytometry. Cardiomyocytes from TOF hearts and RVOT non-defected hearts differ in organization and expression of Cx 43. Cardiomyocytes from TOF hearts reveal a significant increase of Cx43 compared with the control's

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

(p<0.05). In the TOF hearts the protein is situated on the entire surface of the cell. In the control group cardiomyocytes contain less protein localized within the intercalated disks. Disturbances of distribution and expression of Cx43 in TOF hearts are observed. They may influence the development of TOF in the course of cardiac morphogenesis.

P1191

An histopathological study of pedicle and free autologous pericardial patches on pulmonary arteries

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We experimentally assessed the growth potential and histopathological behavior of pedicle and free autologous pericardial patches on pulmonary arteries. Ten beagle dogs, 3 months old, had pulmonary artery (PA) patch grafting in which an autologous pericardium was placed on half of the left PA wall. They were divided into two groups, a pedicle patch group (P group: n=5), and a free patch group (F group: n=5). The size of left PA was measured by angiography at 3 months, 6 months and 12 months after the procedure. After 12 months, the animals were sacrificed and all left PAs examined macroscopically and microscopically. Body weights increased significantly from 7.0±1.0kg to 10.6±1.6 in the P group(p<0.05) and from 6.9±0.5 to 11.8±0.4 in the F group(p<0.05). The diameter of the left PAs also increased significantly, from 6.6±0.4mm to 7.6±0.9 in the P group (p<0.05) and from 6.9±0.5 to 8.6±2.0 in the F group. In both groups, histopathological studies showed that the autologous pericardial patches consisted of an endothelium layer and a mature smooth muscle layer containing elastic fibers. Patches in the P group were significantly thicker than those in the F group, and in 3/6(50%) of the 6 in the P group, calcium deposits were observed, whereas they were not observed in the F group. These results suggest that pedicle and free autologous pericardiums have growth potential. However, the pedicle pericardium is not superior to free one as a patch material on pulmonary arteries.

P1192

Leukocyte migration induced by pediatric cardiac surgery

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Aim: The immune response after cardiac surgery with cardiopulmonary bypass (CPB) contributes to the sometimes adverse outcome with capillary leakage and migration of activated cells to sites of inflammation. This migration may be induced by attractant and repellent chemokines acting in concert. Method: We established a chemotaxis assay for peripheral blood leukocytes (PBL). PBL from healthy donors were isolated and placed into a migration chamber separated from a second lower chamber filled with patient serum by a filter (pore width 3µm). After incubation (1h, 5% CO2, 37°C) cells from top and bottom chamber were harvested and stained with a cocktail of 7 monoclonal antibodies for leuko- and lymphocyte subsets and analysed on a dual Laser FCM. Result: both chambers the total number of cells recovered was 5–15% below that of the initial cell number due to attachment of migrating cells to the point of the filter. These cells belong to the migrating compartment and were quantified by Laser Scanning Cytometers (LSC) after staining of nuclear cells and the whole filter was analysed. Results: Increased chemotactic activity started at onset of anesthesia followed by a phase of low activity immediately after surgery and a second phase of high activity at postoperative days 1–2. In the first phase mainly monocytes and NK-cells migrated. The in vitro results correlated with results obtained by immunophenotyping of circulating PBL of the same patients showing that at CPB onset monocyte and NK-cell count increases. After surgery T- and B-cell count decreased probably due to homing into lymphatic tissues. Conclusion: In chemotaxis assays with low amount of available serum and blood the combined use of FCM and LSC proved as a useful tool for analysis. During paediatric cardiac surgery the chemotactic activity of the serum changes following characteristic patterns.

P1193

Re-oxygenation does not reverse chronic hypoxia

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Previous exposure to chronic hypoxia is generally considered detrimental for myocardial performance in children with cyanotic congenital heart defects exposed to re-oxygenation at the time of intracardiac repair. We decided to compare hearts exposed to chronic hypoxia followed by re-oxygenation with normoxic hearts made acutely hypoxic and re-oxygenated. Five-week old Sprague-Dawley rats were continuously maintained either in a hypoxic (O2 content=10%) or normoxic (O2 content=21%) environment for 2 weeks (n=9 per group). At the end of this period hypoxic rats presented with lower body weight (182±5 vs 351±11g, P<0.0001), higher heart/body weight ratio (6.10±0.25 vs 3.74±0.04mg/g, P<0.0001) and higher hematocrit (69±2 vs 40±2%, P<0.0001) than normoxic rats. Hearts of both group of rats were isolated and connected to the Langendorff hypoxic perfusion (10% oxygen content, flow=15 ml/min) for 30 minutes. At the end of the hypoxic perfusion the coronary vascular resistance was lower in hypoxic than normoxic hearts (1.73±0.11 vs 2.23±0.12mmHgXminXgXml-1, P<0.005), the lactate release was lower (4.4±0.3 vs 10.5±0.4µM/min), without significant difference with regard to the LVDPXHR (46.9±3.8 vs 48.0±6.5mmHg/min/1000, =NS). All hearts then underwent 30 minutes of re-oxygenation (100% oxygen content, flow=15 ml/min). At the end of the re-oxygenation the LVEDP was higher in hypoxic than normoxic hearts (7.7±3.5 vs 5.5±0.5mmHg, P<0.01), the LVDPXHR was lower (83.9±2.0 vs 119.5±14.9mmHg/min/1000, P<0.05) and the oxygen uptake was lower (7.4±0.6 vs 9.2±0.2µM/min, P<0.01). Coronary vascular resistance was lower in hypoxic than in normoxic hearts (2.80±0.14 vs 3.68±0.26mmHgXminXgXml-1, P<0.05) although it remained higher than baseline, indicating the potential occurrence of reoxygenation injury. Our model shows that with acute uncontrolled re-oxygenation both systolic and diastolic myocardial functions are impaired by previous exposure to chronic hypoxia, despite a significant reduction of coronary vascular resistance.

P1194

Apoptosis in Adriamycin-induced cardiomyopathy in rats-comparison with pirarubicin

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We had reported that apoptosis was observed in Adriamycin-induced cardiomyopathy (ADR-CM) in rats and occured through a Fas-dependent pathway (Circulation 2000;102:572–578). In this study, we investigated whether pirarubicin (anthracycline derivative) had a lower cardiac toxicity, compared with ADR and the neutralization of anti-Fas ligand antibody (antiFas L) is effective in ADR-CM. (Methods) ADR (group A) or pirarubicin (group C) was injected weekly for 8 weeks via tail vein in young rats. In group B, antiFas L was injected with ADR at weeks 7 and 8 after first injection of ADR. In group D (as control), saline was injected instead of ADR. In every groups, the left ventricular performance was serially examined by echocardiography at weeks 8,9 and 10. At week 10 after first injection, apoptotic ratio of left ventricle was examined by TUNEL method and expression of Fas antigen of myocyte was examined by Western blotting in each group. (Results) Apoptotic ratio(apoptotic cell count/total cell count, %) were 1.29+-0.33 in group A, 0.63+-0.18 in group B, 0.29+-0.07 in group C, and 0.12+-0.04 in group D. Apoptotic ratios in group B and C were smaller than one in group A. % fractional shortening (%FS, %) were 55.7+-3.2 in group A, 51.3+-9.2 in group B, 45.6+-5.3 in group C, and 64.3+-3.7 in group D. %FS in group C is significantly better than one in group A. Fas antigen was overexpressed in the heart of group C compared with control heart, however an overexpression of Fas antigen in group C was less than one of group A and B. (Conclusion) This study showed that pirarubicin had a lower chronic cardiotoxicity compared with ADR. The neutralization of antiFas L inhibited ADR-induced apoptosis, however had no influence for cardiac function.

P1195

Cryopreservation of cardiac tissue and cardiomyocytes for cell transplantation

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Cardiomyocyte transplantation improves heart function after cardiac injury. Cryopreservation may be an effective method for cell storage for cell transplantation. We evaluated the effect of cryopreservation of cardiac tissue and cardiomyocytes by measuring contractility and proliferation. Methods: Cardiomyocyte: Fresh and passage 1, 2, 3 and 4 fetal cardiomyocytes were suspended in freeze medium and cryopreserved and stored in liquid nitrogen for 1, 2, 4, 8, 12 and 24 weeks. These cells were then rapidly thawed at 37°C and

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

cultured. A number of the cells, beating rates and percentage of beating cells were evaluated 1, 2, 4, 5, 8 and 10 days. Cardiac tissue 0.2, 2.0 and 6.0 mm3 of fetal rat myocardium were also preserved in liquid nitrogen for 1 week. Cells were then isolated from the tissue and cultured. Cell growth and contractility were measured. Results: (1) Cardiomyocytes grew and contracted after cryopreservation. Storage time did not affect cell survival rate, beating cell numbers and beating rates. Increasing cell passage prior to cryopreservation decreased the percentage of beating cells. (2) Cells isolated from cryopreserved tissue grew in vitro and contracted normally. Cell yield decreased with increased cryopreserved tissue size. (3) The subcutaneous transplant contracted regularly. The transplanted cells formed tissue in the myocardial scar. Conclusions: Cryopreserved cells survived and functioned in vitro and in vivo. Viable cells can be isolated from cryopreserved myocardium and cultured. Cryo-preservation of small pieces of myocardium is preferred for maximal cell yields.

P1196

Water Transport in Fetal Lamb Myocytes – Role of Aquaporins

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Myocardial edema contributes to contractile dysfunction post operatively and may occur as a result of ischaemia-reperfusion injury, the systemic inflammatory response to bypass, hemodilution and fluid overload. Water transport across cells occurs in response to osmotic/hydrostatic gradients; membrane permeability is enhanced by the insertion of aquaporin (AQP) molecules into the cell membrane. There are at least ten AQP isoforms (AQPs 0–9), of which seven are pure water channels. AQPs 3,7,9 are also permeable to other small molecules eg. urea, glycerol. Aquaporin 1 has been identified in the myocardium of several species. It is strongly expressed in the kidney and placenta but not AQP 3. In this study we used quantitative methodology (real time PCR) to determine the relative levels of AQP-1 in left ventricle, kidney and placenta of the ovine fetus. A preliminary study was conducted to see if AQP-3 was also present in the heart. The levels of AQP-1 in the late gestation (130 days term=150 days) left ventricle was 1.6±0.5 (mean±sem, n=7) times that of mid gestation kidney and 5.2±1.7 times that of mid gestation placenta. AQP-3 mRNA was detected in three samples at 2–4 times that of the kidney but at only ~20% of that in the placenta. Substantial expression of these two AQPs in the mammalian heart suggests they may be potential targets for the future treatment of myocyte edema which occurs postoperatively. Studies are in progress to ascertain whether other AQPs are also expressed in the heart. Ref: I Johnston et al. Placenta, 21:86–99, 2000.

P1197

Myogenesis and angiogenesis after autologous bone marrow cell transplantation improved heart function

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Background: To investigate the utility of myocardial bone marrow cell (BMC) transplants in a clinically relevant model of a myocardial infarction, we examined the survival and contractile function of infarcted hearts transplanted with chemically-induced BMCs. Methods: 35 kg pigs were studied. Sternal bone marrow was aspirated and the distal left anterior descending of adult pig heart was occluded by intracoronary artery placement of coils. The BMCs were cultured for 4 weeks and induced twice with 10uM of 5 azacytidine. Some BMCs were labeled with bromodeoxyuridine (BrdU) prior to transplantation. Four weeks after infarction, SPECT MIBI scan was performed and then 100 X 10(6) BMCs (N=5); or culture medium (N=6) were injected into the scar tissue. Four weeks after transplantation, a MIBI scan was done. Anatomical and histological studies of the hearts were performed. BrdU-labeled cells were identified in the infarcted region. Results: The BrdU-labeled cells had sarcomeres and Z-band and stained positively for troponin I. The BMC transplant sites had more capillaries than the control scars. No bone or cartilage was found. The MIBI results showed that the stroke volume, regional perfusion and regional wall motion were better (p<0.05, <0.05, <0.05) than those of the control hearts. Although the scar areas were not different (p=0.06), the scar thickness was greater (p<0.05) in the transplanted hearts than in the control hearts. The left ventricular chamber size was smaller (p<0.05) in the transplanted hearts than in the control hearts. Conclusion: Transplanted BMCs survived in the infarcted area and form new muscle tissue and capillaries. Regional perfusion was preserved and regional and global contractile functions improved.

P1198

Postreceptor defect of adenylyl-cyclase in severe failing myocardium of children with congenital heart disease

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In children with severe heart failure due to congenital heart disease beta-adrenoceptor downregulation is well known. The aim of this study was to determine, whether a postreceptor defect additionally impairs the cardiomyocytes responsiveness to beta-agonists. Methods: The severity of congestive heart failure in 38 children (9 days – 13 years) was graded by a scoring system including historical and clinical parameters. Children were divided into a group with no or mild heart failure (score < 6) and a group with severe heart failure (score > 6). Right atrial myocard biopsy was performed during cardiac surgery. The concentration of the inhibitory G-protein was measured by pertussis-toxin catalyzed ADP-ribosylation. The adenylyl-cyclase (AC) activity was measured after receptor stimulation by isoproterenol, receptor independent stimulation by forskolin and stimulation by forskolin in the presence of Mn 2+ , which uncouples the catalytic subunit from the G protein. Statistics: Mann-Whitney-test. Results: Not only beta-adrenoceptor stimulated AC activity was significantly decreased in the group with severe heart failure (65%), but also receptor independent stimulation of AC by forskolin (49%), implicating a postreceptor defect. The activity of the catalytic subunit of AC was significantly decreased in the group with score > 6. The concentration of the inhibitory G-protein was not different between groups. Conclusion: Beside the beta-receptor downregulation in children with severe heart failure a postreceptor defect located at the catalytic subunit of the adenylyl-cyclase contributes to decreased effectiveness of cyclo-AMP-increasing agents.

P1199

Alteration of heart carnitine palmitoyltransferase in adriamycin treated rat after administration of L-carnitine

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Alteration of heart carnitine palmitoyltransferase in adriamycin treated rat after administration of L-carnitine. Hong, Y.M., Yoon, H.R., Bomack, R., Bennett, M.J. Seoul and Texas Adriamycin inhibits carnitine palmitoyl transferase (CPT) system and consequently the transport of long-chain fatty acids across the mitochondrial membranes. Adriamycin-induced cardiomyopathy produces congestive heart failure. This study was devised to elucidate how adriamycin (ADR) affects heart CPT in rats given adriamycin with and without L-carnitine supplementation, and identify change of heart CPT and serum carnitine levels as a function of adriamycin concentration. Male Sprague-Dawley rats were divided into four groups as follows. The 1st group was control. The 2nd group was given intraperitoneal injection of adriamycin (5mg/kg) twice a week for 2 weeks, and the 3rd group received adriamycin (5mg/kg) with L-carnitine (200mg/kg) for 2 weeks. The 4th group was injected with L-carnitine (200mg/kg) only. Blood was collected from abdominal aorta on the 10 day, 1 week, and 2 week for determination for serum carnitine. Heart mitochondria was isolated from the frozen heart with supplying radiolabeled carnitine. The enzyme activity of CPT1 and CPT2 were measured. Enzyme activities of heart CPT1 and CPT2 significantly decreased in ADR group compared to the control group. The ratio of enzyme CPT1/CPT2 prominently decreased as a function of ADR concentration. The result suggested that the depression of enzyme CPT1 was more sensitive than that of CPT2 when exposed to ADR. The addition of L-carnitine to ADR group did not reverse the activities of CPT1 and CPT2. In conclusion, this study supports the view that adriamycin causes cardiomyopathy due to the inhibition of CPT enzyme, and L-carnitine protects from the toxicity.

P1200

Analysis of gene expression patterns in different congenital heart disease by a real time RT-PCR technique

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Severe congenital heart disease (CHD) lead to a progressive disorder with complex interaction of hemodynamic, neurohumoral and myocardial gene expression disturbances. Gene expression alteration may occur due to hemodynamic and neurohumoral changes and serve as compensatory mechanisms

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

to maintain an adequate cardiac function. However, a lot of target genes are involved in these progressive some of which may ultimately lead to cell apoptosis, fibrosis and irreversible heart failure. Myocardial gene expression patterns in infants and children with CHD have not been fully investigated so far. The aim of our studies was to identify target genes with significant expression and/or isoform shift, which may have a significant influence for the long term myocardial function of these patients. We investigated 79 myocardial biopsies from patients with CHD (age 1 week to 12 years), which were obtained during cardiac surgery. A quantitative real time RT-PCR method for small amounts of tissue (5 – 35 mg) was developed by using a commercial analyzer (ABI PRISM 7700 sequence detection system). Isoform expression patterns for beta-receptors (beta-1 and beta-2), natriuretic peptide (ANP and BNP) and connexins (Cx 40 and Cx 43) were analysed in three patients with clinically established diagnosis. Complex heart disease such as univentricular heart revealed significantly reduced expression of beta-receptors, upregulation of BNP and Connexin 43 when compared to ASD or VSD. Our data shows, that the real time RT-PCR is a suitable method for evaluation of gene expression patterns in children with CHD, where only small amounts of myocardial tissue is available. Furthermore beta-receptors, natriuretic peptides and connexines proved to be suitable markers of myocardial adaptation in serum CHD. Further studies with target genes also influencing fibrosis and apoptosis are in progress.

P1201

Changes of cardiac troponin T isoform expression

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Changes of cardiac troponin T isoform expression in the adriamycin-induced cardiac injury in rat. Troponin T (TnT) is a biomarker of cardiac injury arising from various causes such as ischemia, myocarditis, cardiomyopathy. TnT isoform switches from the embryonic form (320bp) to the adult form (250bp) during development. The fetal heart TnT isoform expression and serum TnT level increase after cardiac injury. We analyzed the expression of heart TnT isoform and serum TnT level to identify the relationship between these two markers. Sprague-Dawley rats were divided into four groups. The 1st group was control. The 2nd group was given intraperitoneal injection of adriamycin twice a week for 2 weeks and the 3rd group received adriamycin with L-carnitine for 2 weeks. The 4th group was injected with L-carnitine only. Serum and hearts were harvested at 1st day, 1 week, 2 week after adriamycin injection. Serum TnT level was measured by sandwich ELISA. Total RNA was extracted from frozen heart, and reverse transcription PCR was done by using GAPDH as a internal standard. Fetal TnT (320 bp) and adult TnT (250 bp) isoforms were identified, and the ratios of fetal/adult TnT isoforms were analyzed by imageQuant software. Serum TnT level increased significantly after 2 weeks of adriamycin injection. On the other hand the ratio of fetal/adult TnT isoforms increased prominently after only 1 day of adriamycin injection and were maximal at 1 week. Addition of L-carnitine did not reverse serum TnT level, but decreased the amounts of the fetal TnT isoform expression significantly. In conclusion, change of heart TnT isoforms was a more sensitive diagnostic test than serum TnT level. Protective effects of L-carnitine were also reflected in normalization of changes in heart TnT isoforms.

P1202

Evaluation of angiotensin converting enzyme activity in acute right ventricular hypertrophy in an experimental model of adjustable endovascular stenosis of the pulmonary artery

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The pulmonary artery banding (PAB) has been used to promote rapid left ventricular (LV) hypertrophy in patients with transposition of the great arteries (TGA) with intact septum, treated beyond the neonatal period. This procedure is followed by high morbidity and mortality rates. Genetic changes of the cardiomyocytes resulting from acute hypertrophy have not been evaluated in models of variable systolic overload of the subpulmonary ventricle. In order to evaluate the activity of angiotensin converting enzyme (ACE) in acute right ventricular (RV) hypertrophy, a balloon catheter was implanted in the PA of eleven young goats. Systolic overload was carried out throughout progressive balloon insufflations for a period of 96 hours. Hypertrophy was followed by daily hemodynamic and echocardiographic evaluations. At the end of 96 hours, the animals were killed for harvesting the hearts. The ventricles and septum were weighed separately. Samples of each cardiac muscle were collected for ACE analyses. A group of eight goats (with similar age and weight) was used as control for weight and ACE activity. At the end of the protocol, the following parameters were increased: RV/PT gradient ($p=0.001$), RV/LV ratio ($p=0.003$), thickness of the free wall of RV ($p=0.002$) and RV weight ($p=0.002$). The evaluation of ACE activity showed a significant increase only in the hypertrophied RV muscle ($p=0.002$), indicating a high correlation with the increase in the RV/LV ratio ($r=0.87$). It may be concluded that a 96-hour period of progressive systolic overload in the goat RV induces ventricular hypertrophy. This hypertrophy is related to a significant increase in ACE activity, an important molecular marker of this process.

P1203

Myocardial damage and increased apoptosis of circulating leukocytes during cardiac and vascular surgery

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Aim: Phagocytosis of apoptotic leukocytes leads to temporary anergy of monocytes/macrophages, so that it could be in part responsible for reduced immune defense during cardiac surgery. The effect of cardiac surgery on leukocyte apoptosis has not been shown yet. Cardiopulmonary bypass surgery also induces myocardial damage. This might at least in part be affected by apoptotic activity in the serum of the patients. Method: Flow-cytometric immunoglobulin-type data from 90 children (age 3–16 yr.) who underwent cardiac surgery with (65) or without (25) CPB were analyzed retrospectively for T-cell apoptosis based on light scatter and surface antigen (CD45+/CD3+) expression (Blood 83:1268). In addition, in vitro isolated leukocytes from healthy volunteers were incubated with serum obtained before, during and after surgery. Apoptosis was detected by AnnexinV staining and flow cytometry or DNA condensation analysis by laser scanning cytometry. Serum cytokine and Troponin-I levels were determined. Results: Patients undergoing surgery with CPB had elevated lymphocyte apoptosis (e.g. T-cell apoptosis increased from 3.45% (baseline) to 11.24% (4h postoperative, ANOVA $p=0.0034$). No effect was found without CPB. The results were in accordance with in vitro findings showing elevated apoptosis activity for lymphocytes and neutrophils in the serum of patients with but not without CPB, starting at reperfusion and rising up to 5d after surgery ($p<0.01$). Increase in apoptosis correlated well with the increase in Troponin-I and IL-10 levels. Conclusion: IL-10 might be involved in pre- and post-operative neutrophil apoptosis by suppressing the protective effect of IL-6. Increased apoptosis further contributes to the immune suppressive response to surgery with CPB. Elevated apoptotic activity in the blood of patients during CPB might also contribute to the destruction of cardiomyocytes during and after pediatric cardiac surgery.

P1204

Hyperthermia Promotes Heat Shock and Sarcomeric Protein Synthesis in Adriamycin Induced Cardiomyopathy in Rat Experimental Model.

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Adriamycin (ADR), an anthracycline antibiotic is among the most important of antitumor agents. The clinical value of ADR is limited by a toxic cardiomyopathy. Hyperthermic treatment, which induces the heat shock response, could reduce the severity of late ADR cardiomyopathy. In order to evaluate hyperthermia protective strategy and its relationship with heat shock (hsp) and sarcomeric protein induction an experimental model of ADR-induced very late cardiotoxicity was developed. Female Sprague-Dawley neonatal rats, body weight 40g were randomized into four groups: control, ADR, temperature, temperature ADR. ADR was injected i.v. as a dose of 4mg/Kg (0.1ml/every third day for a total of three administrations). Thermal stress was produced by wrapping the animals with an electric heating pad until they reached a core body temperature of 45°C. After 30' of stress animals were allowed to recover at room temperature for 30'. After the third ADR subdose hearts were removed and left ventricular walls were analysed by Western blotting with use of a monoclonal antibody specific for hsp 25 and a specific monoclonal antibody for myosin. Western blotting analysis of hyperthermia-ADR hearts have demonstrated an increased hsp25 and myosin expression compared to control, temperature and ADR groups of rats. In conclusion the pattern of hsp25 and myosin expression in ADR treated heart with hyperthermia protective strategy is similar suggesting that heat shock protection may be related to myosin increased synthesis.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P1205

Possible influence of the immune system on protein-losing enteropathy after fontan-operation

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Aim. Children following Fontan operation might develop a post-operative protein losing enteropathy (PLE) with a substancial mortality (Eur Heart J:19:514–520). Single case examinations from other authors show an influence of the immunesystem on these symptoms. Its role and amount have not been clarified yet. It was investigated if in patients following Fontan operations immunological changes are found that might account for PLE development. Method. In a follow up study blood samples from 12 children were drawn 3 month to 3 years postoperatively. The serum levels of different inflammatory mediators, adhesion molecules and complement factors were determined and a large panel of cellular immunodiagnostics was performed. The resulting data were compared to preoperative values from 25 children with septal defects, 20 children with atrial corrections and data of 50 healthy children. Results: In Fontan patients compared to the control patients no significant changes of the serological parameters were found. However, some dramatic changes of the cellular immune system were observed. Among others 1. the ratio of helper/cytotoxic T-cells was 2- to 4-fold ($p<0.01$) increased, 2. the reason for the increase was the decrease of the cytotoxic T cell count from 700/ml (control) to 400/ml ($p<0.05$), 3. loss of memory T-cells (CD45RO+) by > 50% and, 4. on the other hand B cell count increased from 550/ml to 900/ml ($p<0.05$). Conclusion. Children have after Fontan operation a substantially altered immunophenotype. The loss of cytotoxic and memory T-cells might result in an impaired immune defense capability. These alterations in combination with yet undefined factors are potentially involved in the development of PLE.

P1206

Heat Shock Protein 70 expression in cardiosurgery pediatric patients

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The Heat Shock Proteins (HSPs) play a role in cytoprotection. In the heart, the expression of the inducible member of HSP70 family provides an endogenous system of protection against the ischemia-reperfusion injury. The aim of this study was to investigate the HSP70 mRNA expression in the myocardium during surgery. Fourteen pediatric patients (7 males and 7 females; age from 5 months to 12 years), affected by Tetralogy of Fallot (12), atrial septal defects (1) and ventricular septal defects (1), were analyzed. We chose patients with similar CPBP and CC time, to minimize their influence on the HSP 70 gene expression; mean cardiopulmonary bypass (CPBP) time was 128±20 minutes, mean cross-clamp (CC) time was 77±13 minutes, mean corporal temperature during surgery was 29±4°C. CPBP was established by a cold crystalloid cardioplegic solution. The first right atrium specimen has been taken soon after pericardium opening, the second after the end of the CC. A RT-PCR method was developed. First strand cDNA was synthesized by MMLV RT with oligo d(T) priming. Then HSP70 and glyceraldehyde 3 phosphate dehydrogenase (GAPDH) cDNAs were co-amplified using specific primers. PCR products have been resolved on polyacrylamide gel, stained by silver nitrate, and densitometrically analyzed by NIH Image software. After CC, a mean reduction of 53% of the HSP70 mRNA levels has been observed, except for 3 patients which had a 11%, 67% and 68% increase. According to these preliminary data, the expression of HSP70 seemed to decrease after CPBP in the most of patients in who there was a significant correlation ($R=0,71$, $p<0.02$) with the corporal temperature. We hypothesized that the surgical hypothermia could slow down all metabolic processes causing the reduction o

P1207

Capillary leak syndrome after open-heart surgery can be predicted by pre-operative serological and cellular data

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Aim. Post-operative capillary leak syndrome (CLS) can occur in children after open-heart surgery. The aim of the study was to detect preoperative differences immune status in CLS and non CLS patients and to evaluate the prognostic significance of the results. Methods: 24h pre-operative peripheral blood samples were analyzed in 30 children (age: 3–16yr) undergoing open-heart surgery. 15 of them developed CLS as found by immediate post-operative formation of edema and pericardial effusion. The serum concentrations of complement (e.g. C1 inhibitor, C3, C3c), cytokines (e.g. IL-2, IL-8, IL-10, IL-12, TNF-alpha), soluble adhesion molecules (e.g. ICAM-1), routine laboratory parameters and the immunophenotype of leukocytes was determined. Results: Between the groups some of the investigated parameters were already different before surgery. In the CLS group leukocyte and thrombocyte counts and ICAM-1 concentration were increased (all $p<0.05$). The concentration of complement was decreased. Immunophenotyping revealed in CLS patients a preoperative increase of endotoxin and MHCII receptor expression on monocytes and Fc-gammaIII receptor expression on natural killer cells by 80–100% (all $p<0.05$). Despite these initial differences none of the measured parameters was sufficient for an individual prognosis. When combined, all data by the software packages CLASSIF and SPSS in order to determine if combinations of different parameters allow the identification of CLS patients. By both programs it was possible to classify all children correctly using 10 out of 36 parameters. The classification tests selected for a (sub-clinical) inflammation (i.e. increased leukocyte count, ICAM-1 level) as predictors of CLS. Conclusion: The results suggest that preoperative differences of the immune status are important indicators of CLS development and might be suitable for risk assessment.

P1208

Hypoxia promotes production of vascular endothelial growth factor in patients

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To investigate possible role of vascular growth factors in development of collateral arteries in patients with congenital heart disease, we measured both arterial and venous plasma platelet-derived growth factor (PDGF), vascular endothelial growth factor (VEGF), basic fibroblast growth factor (bFGF), and hepatocyte growth factor (HGF) by ELISA. Seventeen patients (F/M = 5/12, aged 0.9 to 17 yrs) with congenital heart disease underwent pan-aortography. The degree of visible collateral arteries in the thorax were graded as 0 (none or minimum), 1 (mild), or 2 (moderate to severe) (collateral score). The collateral score, arterial oxygen tension and saturation, mixed venous oxygen saturation, and hemoglobin in these 17 patients were analysed in terms of correlation with their vascular growth factor levels. The average levels of PDGF, VEGF, and HGF tend to be higher in arteries than those in mixed veins. Although there was no significant correlation between severity of hypoxia and PDGF nor HGF levels, linear regression analysis revealed that VEGF was negatively correlated with venous oxygen saturation ($p=0.031$) and positively correlated with hemoglobin value ($p=0.015$). Moreover, the difference in VEGF level of venous and arterial plasma in each patient showed significant negative correlation with oxygen saturation. On the contrary, HGF level was positively correlated with the mixed venous oxygen saturation ($p=0.024$). There was no apparent correlation between the collateral score and the levels of plasma growth factors. We conclude that chronic hypoxia promote secretion of VEGF into venous blood stream, which might in turn induce well-developed collateral arteries in patients with cyanotic congenital heart disease. However, further investigation is mandatory to elucidate the role of HGF and the reason for the lack of direct correlation between VEGF level and collateral scor

P1209

Postsurgical complications following cardiac surgery in children can be predicted by antigen expression on neutrophils and monocytes

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Aim. Our initial studies indicate that children who develop post operative complications (e.g. capillary leak syndrome, CLS) following cardiac surgery with cardiopulmonary bypass (CPB) can be predicted based on their preoperative level of circulating cytokines and adhesion molecules. The determination of these values is time consuming and requires a substantial volume of peripheral blood. Therefore we tested measurement of surface antigen expression via flow cytometer (FCM) and discriminance analysis as a potential assay for individual risk assessment of CLS. Methods: 24h preoperative blood samples 49 patients were stained with cocktails of monoclonal antibodies for the adhesion molecules ICAM-1, LFA1, MAC1, beta integrin, activation markers CD25, CD54, CD69, HLA-DR, CD14 or CD4. Cells were measured by 4 color dual laser FCM calibrated with microbeads. Antigen

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

expression was detected considering mean fluorescence intensity of the respective cell population. Results The data indicate that neutrophils of CLS patients express perioperatively higher levels of LFA1 and monocytes higher levels of HLA-DR and activation markers. This could lead to combination with surgical trauma and CPB to their additional stimulation and migration into sites of inflammation and induce CLS. Using a commercial classifier (SPSS) it was possible to classify 84% of the patients correctly. Conclusion: FCM with its low sample requirement and rapid access of the results could be a powerful tool for risk assessment prior to paediatric cardiac surgery. It is planned to set up a 'Flow Classification program (CLASSIF1)' for individual risk assessment and this would allow for an individual prophylaxis of post-surgical complications.

P1210

Role of the Fibrillins in Postnatal Pulmonary Arterial Wall Remodelling

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The composition of the extracellular matrix changes rapidly after birth, with extensive deposition of fibrillar collagen and elastin. Fibrillin is the principle component of elastin associated microfibrils. Fibrillin 1 is thought to provide tissue bearing structural support while fibrillin 2 guides elastogenesis and we hypothesised that the fibrillins would play an important part in postnatal remodelling together with the integrin sub-units with which they are associated. Using porcine intrapulmonary arteries from fetal life onwards (16 antenatal and immunocytochemical in situ hybridisation and biochemical techniques, we demonstrated age specific and temporal differences in expression of both the fibrillins and integrins. By immunofluorescence, at all ages fibrillin 1 colocalised with the beta 3 integrin sub-unit and fibrillin 2 with the beta 1 sub unit. In the fetal and newborn the fibrillin 1 and its associated integrin was heavily expressed in the inner media and fibrillin 2 and its integrin in the outer media. Expression of both fibrillins and their integrins spread across the media after birth. In situ hybridisation showed that the two genes were differentially expressed, their temporal and spatial expression paralleling the protein expression. Biochemical analysis indicated that the proportion of insoluble fibrillin 1 increased with age. In conclusion, a postnatal increase in fibrillin 1 suggests increased force bearing capacity while the increase in fibrillin 2 and its beta 1 sub-unit were spatially and temporally associated with the known postnatal increase in elastin deposition. This supports the contention that the fibrillins may play an important role in elastin deposition during postnatal pulmonary arterial remodelling. Supported by The British Heart Foundation.

P1211

The preliminary observations after a experimental study to evaluate cardiomyoplasty after fontan type procedure.

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Late failure of Fontan operation is a rare but serious complication. In the present study, the concept of cardiomyoplasty was extended to improve the negligible contraction of the right atrium in a model of Fontan operation failure. Material and Method: The effect of cardiomyoplasty over pulmonary artery pressure and flow was investigated in eight healthy swine, breed Large White, weighing between 35 and 42 kg. In cardiopulmonary bypass, the circulation was arrested at 20ºC and it was performed the resection of free right ventricular walls and of the tricuspid valve. Two biological valves were then implanted in each cava vein. The right ventricle was reconstructed by a patch of bovine pericardium. Before the animals were weaned from cardiopulmonary bypass, the left latissimus dorsi muscle was inserted into the pericardium sac allowing the muscle flap to cover exclusively the right side of the heart. The heart rate, the systemic and pulmonary artery pressures, and the flow in the pulmonary trunk were recorded before(S1), during(S2), and after (S3) the skeletal muscle asynchronous activation. Results: The heart rate and the systemic arterial pressure presents no significant changes at the three situations (p=0.877 and p=0.304 respectively). Otherwise, the activation of the muscle flap induced significant increments in the pressure and flow of the pulmonary artery. The pressure raised from 33.2 mmHg to 42.5 mmHg during S2, and decreased to the same initial value in S3 (p=0.003). Pulmonary artery flow was 2.78 l/min in S1, increased to 4.48 l/min in S2, and decreased to 2.73 in S3 (p=0.002). Conclusion: The results of the present study demonstrated that cardiomyoplasty activation increased significantly the pulmonary artery pressure and flow.

P1213

Nitric oxide: a vasodilator, and inhibitor of matrix remodeling by suppressing AML1B-elastase cascade.

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Nitric oxide (NO), an endogenous vasodilator, inhibits pulmonary vascular remodeling in rats via inhibition of vascular elastase. A 20 kD smooth muscle cell (SMC) serine elastase, which is induced by serum-treated elastin (STE), appears critical to the progression of pulmonary vascular disease. In our previous study using differential display to identify transcripts expressed coincident with elastase activation, we identified AML1B, a transcription factor for neutrophil elastase, in SMC. However, the direct interaction of NO with AML1B-elastase cascade remains unknown. To uncover the signaling pathway for elastase activation and interaction with nitric oxide (NO), we found STE-induced increase in phosphorylated extracellular signal regulated kinase (ERK). Inhibition of ERK activation with PD98059 inhibited AML1-DNA binding and elastase. NO donors (SNAP and DETA NONOate) inhibited elastase as did a cGMP mimetic (8-pCPT-cGMP). SNAP inhibition of elastase was reversed by co-administration of a PKG inhibitor (Rp 8 pCPT cGMP). The increase in phospho-ERK was suppressed by NO donors and the cGMP mimetic, and reversed by co-administration of the PKG inhibitor, as was nuclear expression and DNA binding of AML1B. Taken together, the present study uniquely links NO/cGMP-generating vasodilators with inhibition of elastase-dependent matrix remodeling in vascular diseases by influencing AML1B-mediated gene expression.

P1214

Immune alterations following protein losing-enteropathy (ple) after glenn/fontan surgery are similar to those after systemic lupus erythematosus (sle) and celiac disease (cd): indications for autoimmune disease

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Aim: PLE is feared late (5–10 yrs.) complication after Glenn/ Fontan type of cardiac surgery with 5–15% of the patients exhibiting a substantial decrease of serum protein and an increased excretion of protein in the stool. The mortality among patients with a manifest PLE is up to 60% but the etiology of this disease is yet completely unknown. Method: 26 patients after Glenn/Fontan surgery were immunologically analyzed over a period of up to five years (flow cytometry, serology). One of the patients developed PLE about 6 months after surgery. This patient is the first with PLE closely followed and documented immunologically in the literature before and after PLE. The immune sequel of this patient was compared to those of seven patients with a manifest PLE after Fontan. Results: After PLE (Fontan) the cellular and humoral immune system composition changed dramatically with the selective loss in particular of T-helper cells. These immune changes were very similar to those reported after SLE or CD in the literature. For both autoimmune diseases also PLE has been reported. The following significant changes after PLE/Fontan were in agreement with SLE and/or CD: Decrease of serum protein, serum albumin and CD3+4+ cell count: PLE, SLE and CD; decrease of CD8+ cells: PLE and CD, decrease of T4/T8 ratio: PLE and SLE, increase of HLA-DR and CD45RA expression on T-cells and increased serum TNFa, IL-10, IL-8 and CRP: PLE, SLE and CD; increase of serum IL2Ra: PLE and CD. Conclusion: Changes of the cellular and humoral immune system following PLE after Fontan and during SLE and/or CD are in an unexpected parallel. These similarities might indicate that PLE after Fontan is an autoimmune response or is associated with autoreactivity.

P1215

Acute Chlamydia pneumoniae infection causes persistent endothelial dysfunction of coronary arteries in piglets

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Background – Chlamydia pneumoniae is a common cause of respiratory tract infection in children. Our earlier in vitro findings suggested that acute C. pneumoniae infection is associated in young apoE-KO mice with aortic

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

endothelial dysfunction. Methods: Ten piglets, 9–10 kg weight, were infected intratracheally with C pneumoniae and six were used as controls. The coronary flow velocity (CFV) was measured at 3 days and 2 weeks, using a Doppler flow wire placed in the left anterior descending coronary artery (LAD) in response to bradykinin, an endothelium-dependent vasodilator, before and after infusion with L-arginine, a substrate for nitric oxide synthesis, and nitroprusside, a NO donor. At 3 days, the relaxation of prostaglandin F2alpha (PGF2a)- precontracted LAD rings with bradykinin was additionally investigated in vitro, in the presence and absence of L-NAME and diclofenac, inhibitors of NO synthesis and cyclooxygenase, respectively. Results: CFV in response to bradykinin was attenuated in the infected piglets at 3 days and 2 weeks, more marked at 2 weeks ($p<0.05$), and was improved by L-arginine infusion at 2 weeks. At this time point, unnecessary bolus of physiological saline resulted in severe coronary spasm in two of the infected animals. CFV in response to nitroprusside did not differ between infected and noninfected animals. In vitro, L-NAME pretreatment of coronary rings from infected animals resulted in less relaxation to bradykinin, as compared to noninfected animals ($p<0.05$). Diclofenac pretreatment resulted in significantly decreased contraction to PGF2a in the infected animals. Conclusion: Acute C pneumoniae infection causes profound endothelial dysfunction of both conduit and resistance coronary vessels. The dysfunction may be at least in part reversed by cyclooxygenase inhibitors or L-arginine, and may contribute to the early development of atherosclerosis in children or even trigger the onset of acute coronary events in later life.

P1215

Human and rabbit hearts adapt to chronic hypoxia by activation of protein kinase signal transduction pathways

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Previously we showed that chronic hypoxia in immature rabbits increases resistance to myocardial ischemia. However, the signaling pathways mediating cardioprotection by chronic hypoxia remain unknown. To examine these pathways we measured the activation and translocation of protein kinases in children with cyanotic (SaO2<85%, n=4) and acyanotic (SaO2>95%, n=4) heart defects undergoing surgical repair and in rabbits raised from birth in a hypoxic (SaO2<85% n=6) and normoxic (SaO2>95%, n=6) environment. Right atrial samples from children and left ventricular samples from isolated perfused rabbit hearts were processed for Western analysis. In children with cyanotic heart defects protein kinase C epsilon (PKCe), mitogen activated protein kinase (p38 MAPK) and Jun N-terminal Kinase (JNK) were activated and translocated 2–3 fold from the cytosolic to the particulate fraction compared with acyanotic heart defects. p42/44 MAPK was not activated in cyanotic and acyanotic hearts. In rabbits there was a parallel response in activation and translocation for PKCe, p38 MAPK and JNK similar to data from children. p42/44 MAPK was not activated in rabbit hearts. Perfusion of isolated hearts with chelerythrine (1 uM) prevented translocation of PKCe, p38 MAPK and JNK in chronically hypoxic rabbits but had no effect in normoxic rabbits. Perfusion with SB-203580 (15 nM) prevented translocation of p38 MAPK but not PKCe or JNK in chronically hypoxic hearts. SB-203580 had no effect on normoxic hearts. Thus PKCe activates the p38 MAPK and JNK pathway in chronically hypoxic rabbit hearts. These data show that protein kinase signaling mechanisms activated by chronic hypoxia from birth in rabbits are identical to those activated by cyanotic heart defects in children. Exploration of one or more of these protein kinase signaling pathways may afford cardioprotection to children undergoing repair of congenital heart defects.

P1216

Bradykinin improves endothelial dysfunction caused by chronic Chlamydia pneumoniae infection

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Background. Chronic Chlamydia pneumoniae (C pneumoniae) infection in apoE-KO mice results in impaired and progressive aortic endothelial dysfunction to muscarinic agonists by the age of 16 weeks. This dysfunction, consisting of diminished availability of nitric oxide (NO) and increased production of constrictor prostanoids, precedes the formation of arterial intimal thickening. We investigated the endothelium-dependent relaxation response to bradykinin in apoE-KO mice with chronic C pneumoniae infection. Method: 24 apoE-KO mice, 8 weeks old, were infected with C pneumoniae every 2nd week over a 10- week period. 24 apoE-KO mice were sham-inoculated with PBS. From each group, 8 mice were sacrificed at 2, 6 and 10 weeks, respectively. The precontracted aorta rings were exposed to bradykinin, in the absence and presence of NG-nitro-L-arginine methyl ester (L-NAME) and diclofenac, inhibitors of nitric oxide synthase and cyclooxygenase, respectively. ANOVA was used for statistical analysis. Results: Bradykinin-induced relaxation was significantly enhanced in infected mice at 6 weeks and 10 weeks as compared to noninfected mice ($p<0.05$). A progressive enhancement in relaxation was noted between 2 weeks and 10 weeks in infected mice ($p<0.05$) whereas no change at all in this respect was observed in noninfected mice. In infected mice, L-NAME and diclofenac impaired the bradykinin-induced relaxation at 6 weeks ($p<0.1$) and 10 weeks ($p<0.05$), respectively. Conclusion: In contrast to muscarinic stimulation, endothelium-dependent relaxation on bradykinin is augmented in young animals with chronic C pneumoniae infection. Increased vasodilating prostanoid production and increased NO availability in response to bradykinin stimulation appear to contribute to this effect. Bradykinin may therefore play a protective role by improving the endothelial dysfunction associated with chronic C pneumoniae infection. The potential beneficial effects of ACE inhibitors, which increase the availability of endogenous bradykinin on endothelial receptors, are under investigation.

P1217

Chlamydia pneumoniae infection and Helicobacter pylori infection act synergistically in the atherosclerosis development in young apoE-knockout mice

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Background: The upregulation of vascular cell adhesion molecule (VCAM)-1 by the endothelium and its abnormal production of nitric oxide (NO) are accepted mechanisms in atherogenesis. While the role of salivary Helicobacter pylori infection in atherogenesis is controversial, several epidemiological studies suggest that H pylori infection is likely to interfere with Chlamydia pneumoniae infection in this process. These 2 infections may be diagnosed in up to 50% of children and young adults. Methods: Sixteen apoE- KO mice, 8 weeks old, were equally divided in four groups. First group was infected with C pneumoniae, the 2nd group was infected with H pylori, and the 3rd group was infected with both C pneumoniae and H pylori. Mice from the 4th group and nine wild type mice served as controls. Samples from the abdominal aortas were obtained from all mice after 10 weeks and processed for immunohistochemistry for VCAM-1. The methacholine-induced, endothelium-dependent relaxation of precontracted rings from thoracic aorta was investigated in organ chambers, in the absence and presence of NG-nitro-L-arginine methyl ester (L-NAME), an inhibitor of NO synthesis. Results: The endothelium-dependent relaxation was significantly less inhibited by L-NAME in the coinfected group as compared to the other groups ($p<0.01$). Staining for VCAM-1 was more intense at the branching sites of abdominal aortas in mice with coinfection (2.1±0.3, $p<0.05$) than in mice (0.5±0.2) or noninfected (0.7±0.3) apoE-KO mice, while no staining for VCAM-1 was observed in wild-type mice. Conclusion: When aged with C pneumoniae infection, H pylori infection decreases the endothelial NO production of aorta and enhances VCAM-1 upregulation by endothelial cells at atherosclerosis-prone sites in young animals. These pathological processes support a synergism between H pylori infection and C pneumoniae infection in the development of atherosclerosis.

P1218

Age at Operation as a Predictor for Reduced Peripheral Artery Distensibility After Successful Repair of Aortic Coarctation

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Background: Aortic coarctation (CoA) is associated with late hypertension even after optimal surgical repair. Systemic hypertension may be a clinical manifestation of abnormalities on the vessel wall mechanics due to long-standing overpressure insult. Objective: Evaluate arterial distensibility properties of medium-sized muscular arteries in the pre and post coarctation vessels after a successful surgical repair and its relationship with timing of operation. Methods and Results: Forty-seven (47) patients (40M / 7F, 14.7 years [9.60 to 44.58]) with surgically repaired isolated CoA were studied and compared to thirty six (36) healthy volunteers (28M / 16F, 18.09 years [9.99 to 37.80]). Patients were operated at a median age of 2.3 months (0.03 to 476.09 months) and their median follow-up time was 12.39 years (1.20 to 32.28 years). The arterial distensibility was assessed using pulse wave velocity (PWV) measurements, which are inversely related to the square root of distensibility.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

These were done on the brachial-radial and temporal-auricularis pulse arterial segments. The patient group showed higher PWV values in the upper body than the control group (9.12 ± 1.15 m/sec vs. 7.94 ± 1.96 m/sec; p=0.04), but in the lower body not (9.44 ± 3.16 m/sec vs. 9.16 ± 2.99 m/sec; p=NS). Patients operated on after 2 months of age had higher PWV values compared to the control group (9.84 ± 3.30 m/sec vs. 7.94 ± 1.96 m/sec; p=0.02), although patients operated on before age (8.30 ± 2.85 m/sec vs. 7.94 ± 1.96 m/sec; p=NS). Multiple regression analysis identified age at operation and body mass index as predictors of PWV. Conclusions: Arterial distensibility in the upper body arteries is affected after successfully operated CoA, and is related to the age at operation. Early operation may prevent late arterial mechanical dysfunction.

P1219

The effects of tumor necrosis factor-alpha in neonatal rat cardiomyocytes:apoptosis or necrosis

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Purpose: Tumor necrosis factor-α(TNF-α) is a pro-inflammatory cytokine that has been implicated in the pathogenesis of cardiovascular disease. Serum levels of TNF-α are elevated in many human cardiac related pathogenic conditions including heart failure. It is well known that TNF-α inhibits myocardial contractility and induces apoptosis of adult rat cardiomyocytes via stimulation of TNF receptor 1. But pathophysiologically relevant(very low) levels of TNF-α can not induces apoptosis of neonatal cardiomyocytes, so we evaluated the effects of different concentration of TNF-α in cultured rat neonatal cardiomyocytes. Methods: Neonatal ventricular myocytes were isolated from 3-day-old rats by stepwise collagenase dissociation and cells were cultured for 3 days. After then cardiomyocytes were treated with low(25ng/mL) and high(250ng/mL) concentration of TNF-α for 48 hours. Apoptosis was determined by terminal deoxynucleotidyl transferase-mediated end labelling (TUNEL) staining. Cell viability was evaluated by lactate dehydrogenase (LDH) measurements using cell culture supernatants. Results: Low concentration of TNF-α did not induce apoptosis compared with controls (10.5±2.5%, 10.4±4.3%). And high concentration of TNF-α also did not induce significant apoptosis(10.1±5.6%, 10.4±4.2%). There was no detectable morphological changes of cardiomyocytes after low and high concentration of TNF-α treatment. LDH levels after TNF-α treatment was not changed compared with controls(control, low, high: 3.2±0.1%, 3.1±0.2%, 3.3±0.2%). Conclusion: Our results suggest that even though high concentration of TNF-α alone can not induce apoptosis and no significant cytotoxicity in neonatal rat cardiomyocytes.

P1220

Early disturbances of endothelial function and antioxidants in young adults with risk factors

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To investigate the effects of smoking (smk) and hyperlipidemia (hip) in otherwise healthy young adults on plasma oxidative status, endothelial function, and intimamedia thickness (IMT), 64 subjects were enrolled in a prospective study. Flow-mediated vasodilation (FMD) and IMT were determined using high resolution external ultrasound as described by Celermajer (1992) and Lee (1998) respectively. All plasma parameters were analysed by standard laboratory methods. For both groups at risk, FMD decreased while IMT was increased. FMD pathology was decreased while IMT was increased. FMD pathology was more prominent. Significant pro/antioxidant imbalance was most reliably detected by elevated oxidised lipoprotein autoantibodies and decreased levels for sulfhydril groups and alkaliphenol normalized for lipids. Results were more significant in smokers but detectable in hip patients also. The combined parameters may help to elucidate the pathophysiologic interaction of oxidative stress, endothelial dysfunction and vessel pathomorphology.

P1221

Alterations of selected neutrophil functions in children with cyanotic congenital heart disease.

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Children suffering from congenital heart disease (CHD) have been assumed to have a deficiency in their defence mechanisms. The aim of our studies was to test the respiratory burst, chemotaxis and adhesion molecules on peripheral blood neutrophils in children with cyanotic congenital heart disease. We tested 12 children with CHD (tetralogy Fallot - ToF, and transposition of the great arteries - TGA). The control group consisted of 15 healthy children. The respiratory burst of neutrophils was determined by means of the chemiluminescence (CL) of the cells. Expression of CD11b, CD18 adhesion molecules on neutrophils were determined by flow cytometry. The chemotaxis of the cells was estimated using Boyden chambers and analysed using classical assay. We have shown that: (1) peripheral blood resting and fMLP stimulated neutrophil CL in CHD children was significantly higher compared with the healthy control group. (2) Expression of CD11b and CD18 adhesion molecules were significantly higher on CHD neutrophils than those on normal cells. We have observed significant upregulation of the CD11b expression on neutrophils in CHD children after fMLP stimulation. (3) In CHD children neutrophil spontaneous chemotaxis was significantly lower than in healthy controls. The data indicate that neutrophils from children with cyanotic congenital heart disease undergo activation. Supported by KBN grant No. 4 PO5E 092 19.

P1222

Heart rate variability in a brain dead child.

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To investigate heart rate variability in a patient with complete loss of central nerve function, we studied a case who survived about 10 months after diagnosis of brain death. The patient was a 1-year-old boy, who was referred to our hospital because of convulsion after choking. The diagnosis of brain death was made on the 30-hospital day. The ECG recording was performed on the 279 and 280 days. He was expired on the 346 day. Autopsy revealed complete disappearance of the whole brain structure including the medulla oblongata. The ECG waveform was digitized by 12-bit analog/digital converter with the sampling frequency of 1kHz and stored in a personal computer. During the measurements, the patient was mechanically ventilated with the rate of 18/min. The rhythm was sinus and no ectopies were observed. Time series of R-R intervals were created from the raw data using our own software. Four time series including about 900 to 1100 beats that were recorded while the patient was in a steady state were chosen for analysis. Magnitude of heart rate variability was smaller than that of healthy children. SDNN was 5.60 to 8.78 msec and CVNN was 1.04 to 1.35%. However, almost regular fluctuation of 0.3Hz (the frequency of ventilation) and apparently random fluctuation of lower frequencies were clearly observed. The power spectra showed a peak at 0.3Hz and 1/f^2 fluctuations at the lower frequency band. From these results, we speculate that respiratory sinus arrhythmia is partly due to a direct effect of the phasic movement of the lungs, and that the fluctuation at lower frequencies of this patient is not due to autonomic control but to a random noise, the cause of which is unknown.

P1223

Improvement initiative in pediatric cardiac surgery

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To reduce cost and improve quality of care for patients undergoing pediatric cardiac surgery, a multidisciplinary team was formed. In calendar year 1999, 162 patients underwent Diagnostic Related Grouping (DRG). Of the 162, 108 underwent cardiac surgery at our institution. The specific diagnoses of this cohort of patients were as follows: AVSD(14), ASD(21), SV(13), TOF(22), TGA (ASO)(14, VSD(26), and other(52). Using Improvement Science Methodology, proposed practice changes were identified, consensus was reached among the team, and specific practice changes were implemented. These included standardization of the following items: urinary catheters, intravenous fluid, and respiratory recruitment maneuvers. In addition, generalized guidelines were developed for appropriate use of albumin, chest percussion and postural pulmonary drainage. Clinical pathways and standing orders for simple, complex, and closed cardiac surgical procedures were developed and put into practice. Implementation of these practice changes (total of 22 changes) resulted in savings of approximately
2000 of charges per case. In addition, vasopressor drug and nitric oxide use protocols were developed and resulted in additional savings. The vasopressor protocol resulted in savings of 24 to 368 of charges per case, with 88% compliance. The nitric oxide protocol realized a cost savings of approximately 3000 per case for a projected total cost savings of 250 000 to 300,000 per year.

P1224

The effect of oxygenated custiod solution on improvement of hypothermic preservation of rat hearts

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

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Objective: To study the effect of oxygenated Stanford solution on hypothermic preservation of rat hearts. Methods. Fourteen rats were randomly divided into two groups. The control group, isolated rat hearts were preserved for 10 hours in Stanford solution, and the experimental group,where isolated rat hearts were preserved for 10 hours in oxygenated Stanford solution at a temperature of 4^{o}C. High-performance liquid chromatography (HPLC) was used to measure myocardial adenosine triphosphate and total adenine nucleotides, while the left ventricular end-diastolic pressure (LVEDP), left ventricular developed pressure(LVDP) and the rate of change of left ventricular pressure (+dp/dt) were determined before and after preservation with an isolated working heart model. Results: The mechanical recovery of the left cardiac function was significantly improved after 10 hours of preservation in the experimental group (LVDP 35.6±7.0, +dp/dt 1043±230), as compared to the control group (LVDP 23.7±5.2, +dp/dt 651±209,p<0.05). Adenosine triphosphate and total adenine nucleotides in the experimental group were significantly increased 12.05±0.02 versus 1.68±0.04 nmol/L for the control group, p<0.05). Conclusion: Oxygenated Stanford Solution improved the preservation of rat hearts.

P1225

The effect of donor heart ischemia on coronary vascular endothelium after preservation in rats

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Background: Although myocardial preservation of donor heart plays a very important role in the short-term therapeutic effect following heart transplantation, it remains unclear whether the preservation influences the long-term post-operative results, especially whether it affects the coronary vascular endothelium. We designed this study to observe the histological changes in coronary vascular endothelium following donor heart ischemia. Methods: A modified Langendorff model for functional parameter measurement was used. Wister rats were divided into 4 groups. The rat hearts were preserved for ten minutes, 4 hours, 8 hours or 16 hours in 4^{o}C physiological saline after cardioplegic infusion with St. Thomas solution and then reperfused for 30 minutes with KHB solution. The left main trunk of the coronary artery and the contracting myocardium were resected and the tissue specimens were analyzed by transelectronmicroscopy. Results: The endothelial cells of the coronary artery were damaged more seriously following a prolonged preservation time. Increased preservation time also injured the myocardium. Conclusions: The donor heart ischemia caused injury to coronary vascular endothelium, which may induce coronary artery disease after heart transplantation.

P1226

Increased sensitivity of neonate aerial myocyte to adenosine A1 receptor stimulation in regulation of the L-type Ca2+ current

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Background: The antiadrenergic effects of adenosine (Ado) via A1 purinoceptor on newborn (NB) hearts and the tolerance of NB hearts to ischemia have been reported to be higher than that of the adult (AD). We have recently found that ATP inhibits L-type Ca2+ current (ICa) not only via A1, but also by P2 purinoceptors. These effects may contribute to the ischemic tolerance of NB hearts. We examined the effects of Ado and ATP on ICa of NB and AD rabbit aerial cells. Methods and Results: The membrane-perforated clamp technique was used to record ICa in enzymatically isolated myocytes. Ado (30uM) inhibited isoproterenol (ISO, 30nM)-stimulated ICa more potently in NB cells (66.5±2.9 % of ISO-stimulated ICa) than in AD (38.2±4.5 %). Dose-response curves showed an higher sensitivity of the NB myocyte to Ado. This was accompanied by an increased maximum response and a lower IC50 concentration than in AD. In NB, the effect of ATP was equivalent to that of Ado at high concentrations (30–100uM), but was significantly weaker at lower concentrations. The effect of Ado was antagonized by the A1 purinoceptor blocker, 1,3 dipropyl 8 cyclopentylxanthine (DPCPX, 300nM). Co-addition of DPCPX and the P2 purinoceptor blocker suramin (100uM) abolished 30uM ATP inhibition completely. Both the effects of Ado and ATP were eliminated by pretreatment with pertussis toxin or by superfusing with forskolin plus 3-isobutyl-1-methylxanthine (IBMX), indicating the involvement of the pertussis toxin-sensitive, cyclic AMP-dependent pathway. Conclusions: These results suggest that NB atrial cells are highly sensitive to A1 purinoceptor stimulation, and less sensitive to P2 stimulation. This may be explained not only by increased receptor density but by a higher receptor-response coupling, and may contribute to the higher resistance of the NB myocardium to ischemia.

P1227

Improving Communication with Referring Physicians

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The ability to keep referring physicians (our customers) updated on patient status after admission can be problematic. Efforts to improve satisfaction included implementing a one page brief to FAX to referring physicians within 24 hours of admission into our 38 bed CICU. Included are patients admitted to the CICU: direct transfers from within the hospital, post surgery or those post cardiac catheterization where ICU observation is required. A FAX form is placed into the admission packet, a brief description is written by the Intensivist and then sent by FAX to the designated neonatologist, pediatrician and cardiologist. To assess the benefit of this project, a survey was designed to obtain feedback on adequacy of information, helpfulness, what additional information would be of benefit, and how we can improve on communications. The form, maximum of 1 minute to fill out, asks the physician his/her specialty as well as how timely they read the FAX. That form is coupled with the original data sheet when sent out. A review of the last 20 responses to the survey showed favorable responses for helpfulness and adequacy of information 20/20 (100%). Most physicians 17/20 (85%) read the FAX on the day it arrives. Responses to specialty showed 11/20 (55%) pediatrician, neonatologist 2/20 (10%) and cardiologist 7/20 (35%). Additional information that would be of benefit included: weekly follow-up, when transferred from CICU when discharged, and to include diagrams and cath data. Healthcare professionals know how crucial it is to partner with our referring physicians providing continuity of care across the spectrum. Communications assist in keeping this link.

P1228

p55sR is a predictor marker for cytokinaemia and development of systemic inflammatory response syndrome (IRS) after pediatric open-heart surgery

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Objective: Cardiopulmonary bypass (CPB) can cause many post-operative pathophysiological changes that may lead to systemic inflammatory response syndrome (SIRS) and multiple organ dysfunction syndrome (MODS). We investigated the cytokine network in response to CPB prospectively in 22 pediatric patients undergoing various open-heart procedures. Methods: We measured plasma levels of endotoxin, cytokines (TNF-a, IL-1, IL-6, IL-8) and cytokines specific soluble inhibitors (IL-1ra, IL-6sR, p55 and p75sR) before, then 2 hours and one day after the CPB. Clinical data including durations of CPB and cross-indicators of pathophysiological complications of SIRS/MODS were collected. Results: The cytokine inhibitors except IL-6sR increased significantly from pre-bypass levels to after bypass levels. After bypass elevations of most of the cytokines specific inhibitors were quantitatively dependent on their pre-bypass levels and on the duration of CPB. The results showed that a complex response of cytokine network is induced in response to CPB and is related to duration of the CPB surgery and all the plasma pre-bypass level of the cytokine network. Pre-operative level of p55sR (and to a lesser extent p75sR) remarkably predicted the post-operative rise of the other components of the cytokine network and the development of SIRS/MODS (p=0.005). Conclusion: This finding yields an important prognostic value that may help in the future to develop therapeutic or preventive modalities after pediatric open-heart surgery.

P1229

Sox4 is essential for migration and transformation of neural crest cells participating into conotruncal separation

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Sox4 is a member of SRY-related high-mobility-group transcriptional factors and exclusively expressed in the cardiac ridge of the outflow tract and atrioventricular canal of embryonic hearts. Targeted disruption in mice causes abnormal development of semilunar valve, resulting in cardiac anomalies such as large interventricular septal defect and persistent truncus arteriosus. In order to determine a role of Sox4 in conotruncal separation, we investigated Sox4 expression in embryos treated with lin-dumasin, which influences neural crest cells critical to cranial

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

separation and, subsequently, induces cardiac malformations including persistent truncus arteriosus and tetralogy of Fallot. Semi-quantitative RT-PCR studies revealed that Sox4 expression in the control mouse hearts was first detected on 10.5 embryonic day (ED), reaching the maximum level on 11.5 ED, then diminished on 13.5 ED. Sox4 expression in the embryos treated with bis-diamine was transiently detected on 11.5 ED, which was significantly down-regulated. In the brains, Sox4 expression of control embryos was first detected on 8.5 ED and reached the maximum level on 11.5 ED. However, Sox4 expression in the embryos treated with bis-diamine was reduced in comparison with that in the control embryos on 11.5 ED. These results indicated that bis-diamine administration to the pregnant mouse decreased Sox4 expression in the embryonic heart and neural tissue. Sox4 may play an important role in conotruncal septation, affecting migration of neural crest cells into heart and transformation to the smooth muscle cells in the vascular wall.

Cardiac Anesthesia, ICU Care/ Neonatal/Respiratory Management

P1230

Remifentanil blood concentrations in infants undergoing congenital heart surgery

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Introduction: Remifentanil's ultrashort duration of action provides an ability to rapidly titrate anesthetic depth with minimal hemodynamic impact. We measured remifentanil blood concentrations in infants undergoing cardiopulmonary bypass with deep hypothermia and compared them to those predicted by hemodilution and to those reported to provide analgesia. Methods: Thirteen infants (aged 2 days to 10 months) undergoing heart surgery received a remifentanil infusion (0.3 mcg/kg/min) as part of a balanced anesthetic. Blood was sampled at 0, 2, 5, 10, 15, 20, 21 (CPB) and 28 (DH) minutes after starting the infusion. Blood remifentanil concentrations were determined by gas chromatography and high resolution mass spectrometry. Total circulating volume for the distribution of remifentanil was calculated to be patient blood volume plus pump prime volume. Results: The mean remifentanil concentration 20 minutes after initiation of the infusion was 3.98 (±0.34) ng/ml, which decreased to 2.32 (±1.09) ng/ml 1 minute after starting CPB and increased to 3.67 (±0.91) ng/ml after 7 minutes of deep hypothermia. The predicted mean remifentanil concentration, given the dilutional effect of CPB, was 2.44 (±0.59) ng/ml. Conclusions: Measured concentrations of remifentanil on cardiopulmonary bypass decrease to values consistent with that predicted by hemodilution, implying that no adsorption occurred onto the circuit tubing. The dilution that occurs from pump priming fluid does not reduce remifentanil concentrations below that which provide analgesia for surgical procedures. The increase in remifentanil levels during hypothermia may in part be related to temperature-dependent plasma esterase activity.

P1231

Fast track anesthesia for glenn shunts and Fontan procedures. Does anesthetic dose affect outcome?

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To assess the effects of 'fast track' anesthesia on outcome of children undergoing twenty-one bi-directional Glenn shunts, two classic Glenn shunts and seventeen Fontan procedures utilizing variable doses of a narcotic-based technique, a review of the time to extubation, and length of stay in PICU/hospital was completed. Incidence of hypercarbia, airway obstruction, and reintubation were also noted. Suitable candidates for 'fast-tracking' can be successfully managed utilizing a moderate dose narcotic-based anesthetic technique without adversely affecting outcome variables.

P1232

Procedural risk-factors associated with early postoperative arrhythmias after repair of congenital heart disease: procedural factors associated with early postoperative arrhythmias after repair of congenital heart disease

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The study evaluated the influence of procedural risk-factors for the occurrence of arrhythmias early after pediatric open-heart surgery. Prospective study between July 1996 and October 2000 in children undergoing repair for tetralogy (n=50), complete atrioventricular canal (CAVC, n=43) and transatrial VSD-closure (n=75). Continuous ECG-monitoring in the intensive care unit. For each group of patients, occurrence of arrhythmias was related to maximum postoperative Troponin T (T) serum levels, aortic cross-clamp time (AT), bypass time (BT) and the hemodynamic result. 20/75 VSD-patients (26%) had arrhythmias, which were clearly associated with longer AT and BT and higher T levels (p<0.01 in each). In CAVC patients, arrhythmias occurred in 20/43 cases (47%) and were significantly associated with longer BT (p<0.05), longer AT (p<0.01) and higher T levels (p<0.01). Additionally, abnormal postoperative hemodynamics (10/43 patients) predisposed to arrhythmias (p<0.01). In 17/50 tetralogy patients (34%) arrhythmias occurred which were associated with increasing BT (p<0.05) and AT (p<0.01) as well as higher T levels (p<0.01). In groups of children with identical surgical approach, longer bypass and aortic cross-clamp times as well as higher Troponin levels were associated with early postoperative arrhythmias.

P1233

Echocardiographic assessment of preload conditions at the neonatal intensive care unit.

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Labile hemodynamics of preterm neonates requires proper assessment of loading conditions. In adults, echocardiographic measurements of the diameter of inferior vena cava (IVC) and its respiratory variations (vena cava index, VCI) has been used. To assess the value of echocardiography in preload assessment, 37 neonates (35 preterm; gest. age 31.5±4.3 w, weight 1.813±0.94 g) with known central venous pressure (CVP) were studied at the neonatal ICU with echocardiography. CVP in group 1 (no ventilation) was significantly lower than in group 2 (conventional ventilation) and 3 (HFO) (p<0.001). Vena cava index (%) reached 50.0 both in group 1 and 2, but 6.9 in group 3 (p<0.001). The CVP correlated inversely with VCI (r=-0.336, p=0.046) in the whole group, better correlation being in group 1 (r=-0.631, p=0.012). VCI differed significantly in patients with low vs. normal CVP (62.6 vs. 37.6, p=0.004). Positive correlation between VCI and area of right atrium (r=0.492, p=0.003) indicates possible effect of atrial compliance on VCI. In conclusion, echocardiography may be helpful in assessing of adequate preload in spontaneously breathing preterm and full term neonates. However, its value is limited in artificially ventilated patients.

P1234

Estimation of systemic oxygen delivery in neonates treated with hypoxic gas with nitrogen.

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To evaluate the clinical efficacy of hypoxic gas therapy using nitrogen in newborns with a univentricular parallel circulation, we studied whether systemic oxygen delivery may be improved with this therapy. Methods: We studied seven patients aged 1–6 days with hypoplastic left heart syndrome. All patients were treated by prostaglandin E1 (5–10 ng/kg/min) to maintain the patency of ductus arteriosus. Supplemental nitrogen was delivered by continuous-flow respiratory support. The fraction of inspired oxygen (FIO2) was maintained 16–18% and the peripheral oxygen saturation was kept to be more than 70%. Urine volume (UV) was measured as an indicator of systemic flow volume. Both arterial and central venous blood oxygen saturation (SaO2 and SvO2, respectively), and arterial blood pH were measured before and 6–12hr after nitrogen insufflation. The oxygen excess factor, SaO2/Sa-vO2 (OEF), was calculated as an indicator of systemic oxygen delivery. Some patients were examined the cerebral blood flow (CBF) by the near infrared spectrophotometry. Abstract Text: To evaluate the clinical efficacy of hypoxic gas therapy using nitrogen in newborns with a univentricular parallel circulation, we studied whether systemic oxygen delivery may be improved with this therapy. Results: The hypoxic gas therapy by supplemental nitrogen did not alter either systemic blood pressure nor blood pH. The UV increased from 6.8±1.1 ml/hr/kg to 7.2±4.8 ml/hr/kg (p=0.012). Although SaO2 decreased, the increase of SvO2 resulted in significant improvement of the

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

DEF from 3.72±0.89 to 5.33±1.16 (p=0.009). These findings showed the systemic oxygen delivery improved. No apparent decrease of the CBF was seen during the hypoxic gas therapy. Abstract Text: To evaluate the clinical efficacy of hypoxic gas therapy using nitrogen in newborns with a congenital cardiac parallel circulation, we studied whether systemic oxygen delivery may be improved with this therapy. Conclusions: Hypoxic gas therapy with nitrogen increases systemic blood flow volume and improves the systemic oxygen delivery in neonatal patients with intramyocardial parallel circulation. This therapy is safe and effective. Ideally if the FIO2 may be maintained 16–18%.

P1235

The severely polycythemic tetralogy : can surgical outcome be improved?

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Older age(> 4yrs) has been considered a risk factor for surgical correction of tetralogy of Fallot (TOF). Between 1995 and 1998 hospital mortality in 59 children (4–18years) who underwent surgery for TOF was high - 3/59 . Causes were - 1) Major intracranial bleed 2) severe low output 3) multiorgan failure syndrome(MOFS) All 3 had severe polycythemia (Hb> 23 gm/dl) Significant polycythemia was common (35/59) and was associated with increased bleeding problems, low output states, pleural effusions and right ventricular(RV) dysfunction. Aim -To assess the outcome of various strategies evolved to diminish postoperative mortality and morbidity in older severely polycythemic children with TOF. Methods: Strategies evolved targeted at problems encountered - 1) Preop: Staged phlebotomy, optimization of coagulopathy and aggressive coil occlusion of collaterals 2) Intraop: Echo to optimize RV resection 3) Post-op: Elective extended ventilatory and inotropic support on potential low output states (extensive RV resection, large transannular patch, elevated LA pressures, multiple collaterals). Results: Early mortality reduced to 1 in the subsequent cohort of 54 children (4–18years) operated between 1998–2000. More patients had significant polycythemia (26/54) but bleeding related morbidity was reduced from 41% to 7% and vital organ bleeds eliminated. Incidence of low output states, pleural effusions, RV dysfunction could not be reduced, though consequent deaths were minimized. Conclusion - An aggressive protective management strategy approach helped reduce mortality in this high risk group of older, polycythemic children with tetralogy from 5/59 to 1/54. Morbidity due to bleeding problems was minimized. The older child with TOF continues to be at risk for: a) RV dysfunction secondary to a fibrotic and hypertrophied RV b) MOFS & left ventricular dysfunction due to chronic hypoxemia & c) collateral induced pulmonary problems.

P1236

Failed extubation following congenital heart surgery in young children: incidence, etiology, risk factors.

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Purpose: Most children who undergo congenital heart surgery(CHS) require post-operative mechanical ventilation. We sought to identify pre- and intra-operative factors associated with failed extubation (FE). Methods: We performed a retrospective chart review of children y"yl 36 months of age who underwent CHS from 1/1998 to 7/1999. A modified version of logistic regression, which accounts for lack of independence in data with multiple records per subject, was used to assess the impact of risk factors for FE. A forward selection process was used with p< .05 as the criteria for entry into the model. Estimated odds ratios(EOR) are reported with 95% confidence limits. The predictive ability of the final model was assessed using area under the receiver operating characteristic(ROC) curve. Results: 203 children underwent 219 surgeries. 21 children, during 22 separate surgeries, experienced a total of 26 FE's. Median ventilator time(days) was 9.5(3.5–18) for successful extubation versus 5.4(1.91–35) for FE. FE #1 occurred on median POD 2.8(0.1–14). Etiologies of FE #1 included heart failure(n=6), pulmonary(n=6), airway edema(n=3), accidental(n=2), paralyzed diaphragm(n=2), hemothorax (n=1), and epistaxis(n=1). Reintubation FE (n=5) occurred on median POD 5.4(0.9–35). Etiologies of recurrent FE included heart failure(n=1), pulmonary(n=2), airway edema(n=1), and paralyzed diaphragm(n=1). Our multivariate model identified pre-operative pulmonary hypertension (EOR=21.4, 4.5–217), presence of a congenital syndrome (EOR=4 7.1.)–16.6), and intra-operative circulatory arrest (EOR=4.2,1.1–15.7) as risk factors for FE. Area under the ROC curve = 0.857. Presence of all 3 risk factors was associated with 100% specificity. Conclusion: Extubation fails after approximately 10% of CHS in young patients. Etiologies of FE are diverse. In our population, pre-operative pulmonary hypertension, presence of a congenital syndrome, and intra-operative circulatory arrest are risk factors for FE. Prospective validation with larger numbers and at multiple institutions would improve the model.

P1237

Delayed sternal closure in pediatric intensive care unit after cardiac surgery.

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Objective: To assess morbidity and mortality associated with delayed sternal closure performed in pediatric intensive care unit. Type of research: Retrospective descriptive epidemiological study. Site: Multidisciplinary pediatric intensive care unit (PICU) in a tertiary care university-affiliated pediatric hospital. Methods: Review of clinical data of all patients with delayed sternal closure admitted in the PICU after cardiac surgery. Results: Between January 1992 and January 2000, 53 patients of 3061 (2%) open heart operations for congenital heart defects had prolonged open sternotomy. Thirty-seven of these children (70%) were newborns and the sternum was electively left open to avoid postoperative hemodynamic and respiratory compromise secondary to myocardial edema. Successful sternal closure was achieved in 48 patients (92.3%): 21 (58%) were done in the operating room and 22 (42%) in the PICU at the mean of 5.1 ± 2.6 and 4.9 ± 3.0 days respectively (NS). Sternal closure in the PICU were realized by cardiac surgeon under general anesthesia administered by pediatric intensivist and assisted by fellows, respiratory therapist, intensive care and operating room nurses. The overall operative mortality rate was 5/53 (9.4%) and 3/22 (13.6%) in the PICU subgroup. One of these three patients was on extracorporeal membrane oxygenation (ECMO) after surgery and died from aspergillus and haemophilus influenzae infections. No other infectious complications were observed in the PICU group. Conclusions: Delayed sternal closure can be made safely in pediatric intensive care unit without increasing morbidity and mortality.

P1238

ECMO for the treatment of malignant arrhythmia

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The use of ECMO for the support of patients with malignant arrhythmias have rarely been reported in the paediatric population. A 9-1/2-year old boy was admitted for acute myocarditis complicated by a complete heart block. Although aggressive treatment was instituted, 12 hours after admission the patient was an intractable heart failure with an ejection fraction of 9% and cardiac index of 2.1 l/m2.1lr. He was carried on ECMO. With support a ventricular conduction immediately resumed and pacing could be stopped. After 5 days, still in sinus rhythm, the cardiac function was improved (ejection fraction 40%) and he was successfully weaned from ECMO. The patient has now been followed over a 16 month period, is currently taking ACE inhibitor his electrocardiogram shows sinus rhythm, the echocardiogram is normal with and ejection fraction of 56%. The cause of the myocarditis is still unknown with negative viral cultures and serology. A 6-month old boy ventricular septal defect and aortic valve stenosis repair. Previously, he presented at 10 days of life in cardiac arrest and underwent repair of coarctation of the aorta. After post-operative low cardiac output requiring inotropic support, as the patient started to recover he presented junctional ectopic tachycardia with relapsing hemodynamic instability and numerous episodes of ventricular fibrillation and torsades de pointe necessitating defibrillation and aggressive anti-arrhythmic treatment without success. ECMO was started and 5 hours later the arrhythmia stopped and the antiarrhythmic drugs could be weaned. After 4 days ECMO could be stopped and the arrhythmia did not relapse. cardiac function was normal. On follow-up 28 months later, the patient is doing well with normal growth. In conclusion, ECMO has been useful and indeed life-saving for the two patients presented. Its use should be considered to support patients with malignant arrhythmias refractory to conventional therapy.

P1239

Postoperative care of patients undergoing unifocalization for pulmonary atresia with ventricular septal defect.

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A subgroup of patients with pulmonary atresia (PA), ventricular septal defect (VSD), absent pulmonary arteries and multiple aorto-pulmonary collaterals

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

(MAPCA's) who benefit from multiple stage approach, are at risk of high morbidity. After precise identification of pulmonary blood supply and MAPCA's unifocalisation (U) and creation of central pulmonary arteries is done through separate thoracotomies, prior to complete repair. Since January 1997, 5 patients (5–15 years old) underwent 9 U. The postoperative management consisted of adequate pain control, including regional anaesthesia, in favour fast and aggressive physiotherapy. Patients were extubated in the first 24 hours and aerosol therapy with salbutamol and corticosteroids was instituted immediately and maintained until discharge. Anticoagulation with warfarin was instituted until complete repair (6–12 months) aiming INR of 1.4–1.8 to prevent clot formation in the 16mm Gore-Tex graft exposed to low flow used to repair the pulmonary arteries. All patients eventually presented significant changes on chest X-ray despite normal parenchymal aspect on immediate postop film. Even near total absence lung pneumatisation on CT Scan recovered with aggressive physical and aerosol therapy. All patients responded to this therapy without reintubation or bronchoscopy or lobectomy. This lung insult is thought to be secondary to reperfusion injury. If important Gore-Tex stenosis was seen on CT Scan, anticoagulation was withheld for few days. Conventional postoperative antibioprophylaxis with cefalosporin was continued until lungs were clear. Four patients underwent uneventful complete repair and one is waiting with favorable angiographic control. In conclusion, early introduction of aggressive postop management of possible pulmonary complications is essential to control the morbidity of the multiple stage.

P1240

Correlation between serum lactate levels and complications following heart surgery: does it predict outcome?

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Introduction: There is a controversy regarding the role of serum lactate(L) as a predictor of outcome in the post-operative (PO) period of congenital heart disease. Objective: To evaluate L levels in the immediate PO (IPO), 3hs PO, 6hs PO, first PO, second PO and associate with outcome in the first 14 days following cardiac surgery for congenital heart disease. Methods: We performed a retrospective study to analyse age, weight, perfusion time, cross-clamp time and outcome (discharge home, discharge with complicated course and death). For statistical analyses we used non parametric analyses of variance with specific correlation (Mann-Whitney and Wilcoxon) and values were considered statistically significant at $p<0.05$. Results: We reviewed 104 patients. The median age was 12 months for patients who had a good course, 5 months for patients who had complicated course and 2 months for the ones who died. The median weight was respectively: 8.2 kg, 4.6 kg and 3.6 kg. One-hundred and nine (66.8%) patients were submitted to cardiopulmonary bypass (CPB). The mean CPB time was 75 minutes in the survivors and 109 minutes in the non-survivors. Twenty-three (14%) patients died, 6 (23%) of these died during surgery, 11 (48%) of these had early death (defined as death in the first 14 days) and 6 (25%) of these perished late death. One-hundred and two (62.2%) had good outcome, without complications, and 39 (24%) were discharged home, however they presented complications during the post-operative period. The higher lactate level in all patients was in the IPO and 3hs PO. The mean lactate was 1.63mmol/L in the patients discharged home, 3.49mmol/L in the ones with complicated course and 8.41mmol/L in the non survivors. There was a positive association between high lactate levels and poor outcome ($p<0.001$). Conclusion: High serum lactate can be an important predictor of outcome during the post-operative period in patients undergoing heart surgery.

P1241

Effects of prostaglandin E1 infusion and balloon atrial septostomy on the outcome of the neonatal arterial switch operation

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Purpose: Preoperative management of patients with transposition of the great arteries typically includes prostaglandin E1 (PGE) infusion and balloon atrial septostomy (BAS). The aim of this study was to determine whether these 2 interventions have any effect on the perioperative course after neonatal arterial switch operation (ASO). Methods: From November 1995 to November 2000, 34 patients underwent neonatal ASO at our institution. Pre-operative, intra-operative and post-operative variables were retrospectively reviewed. Results: Twenty-one of the 34 patients underwent pre operative BAS for a small interatrial communication (mean ± SD 2.7±1.4 mm) or low systemic oxygen saturation (70±13%). Mean PGE infusion time was shorter in patients undergoing BAS (53±38 hr vs 85±41 hr, $p<0.05$). Twelve patients (9/21 who underwent BAS, and 3/13 who did not undergo BAS) had PGE weaned off prior to ASO. BAS patients vs those who did not undergo BAS had no post-operative differences in the number of inotropes required, or the length of intubation or hospital stay. Patients still receiving PGE at the time of ASO required a higher number of inotropes postoperatively (>1 drug in 73% vs 38%, $p<0.001$), and had a greater length of postoperative intubation (median 121 hr vs 74 hr; $p<0.001$) compared to patients not receiving PGE at the time of ASO. Length of postoperative hospital stay was similar (mean±SD 11±3 vs 12±13 days, $p>0.05$, PGE vs. no PGE). Conclusions: Discontinuation of PGE prior to ASO was associated with less postoperative inotropic requirement and length of postoperative intubation. This suggests that efforts should be sought to discontinue pre-operative PGE therapy.

P1242

Echo guided pulmonary artery catheter placement in neonates with pulmonary hypertension.

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Direct measurement of PA pressure is not routinely performed in neonates with PPHN. In these patients, PA catheter placement is complicated by the presence of a patent foramen ovale and ductus arteriosus with right to left shunting. Catheters placed via the IVC tend to cross these defects rather than enter a branch PA. We evaluated the success rate, efficacy and safety of echo-guided PA catheter placement in the NICU in eight neonates with PPHN who were simultaneously enrolled in an IRB approved study of inhaled NO. A 4 Fr end-hole, balloon tipped catheter was placed via percutaneous femoral venous access. A limit of one hour was set on placement in a branch PA. Echo visualization was provided to guide anterograde manipulation of the catheter into a branch PA. After 48–60 hours the catheter was removed. In 7 of 8 patients the PA catheter was stabilized in a branch PA within one hour. In one patient, placement was unsuccessful and the catheter was removed without complication. The subcostal long-axis view was found to be optimal for guiding manipulation of the catheter from the IVC across the TV, and the parasternal short axis view optimal for crossing the PV and placement in a branch PA. In the 7 patients PA pressure was successfully monitored for the 24 hours of the inhaled NO study. In addition PA pressure was monitored for an additional 24–36 hours aiding clinical management. There were no complications associated with insertion or with the indwelling catheter. PA catheters can be safely placed in neonates with PPHN using echo guidance and PA pressure can be monitored to aid clinical management. Cardiologists experienced in catheter manipulation should perform this potentially difficult procedure.

P1243

Neurological and electroencephalographic alterations in children after cardiac surgery

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Neurological and electroencephalographic alterations in children after cardiac surgery. Introduction: Although the mortality rate associated with cardiac surgery in children has fallen in the last 25 years, a significant increase in the neurological morbidity of the patients has been observed. Objectives: To evaluate the incidence of neurological complications after cardiac surgery and factors associated with them. Patients and Methods: Prospective study of 22 children that had undergone cardiac surgery, 17 with cardiopulmonary bypass (CPB), from April to November 2000, at Hospital das Clinicas FMRP – USP. The patients were evaluated by a specific protocol and monitored through electroencephalography (EEG) postoperatively. Results: 6 children (27%) presented neurological manifestation, clinically evident in the PO: 5 seizures and 1 prolonged loss of consciousness. The EEG was abnormal in 11(50%) patients. CPB was not correlated with the occurrence of abnormal EEG: 47% of patients where CPB was performed and 60% of patients that underwent surgery without CPB presented abnormal EEG ($p=0.99$). In the group that underwent CPB, there was no significant statistical correlation between the duration of both CPB and total circulatory arrest (TCA) with the presence of alterations in the EEG. However, in patients with altered EEG, the blood flow was lower and the hematocrit was higher during the CPB ($p=0.03$). Trans-operatively, there was a higher incidence of adverse events such as cardiogenic shock, pulmonary hypertension and respiratory problems in patients presenting altered EEG (83%) than in patients having normal EEG (18%). However, this difference was not statistically significant.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

(p=0.18), probably due to the sample size. In patients with abnormal EEG, the mean arterial blood pressure was lower (p=0.003), and the duration of mechanical ventilation was longer in the postoperative period (p=0.003).

P1244

Specific properties of early postoperative rehabilitation in children with congenital heart defects

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Early postoperative cardiopulmonary rehabilitation is a must in modern cardiovascular surgery in both adult and pediatric medicine, where physiatrist actively participates in the teamwork. At the University Children's Hospital during the period of 1.1.1999–1.1.2000. 250 children with different congenital heart defects (CHD) were operated. Early postoperative rehabilitation were applied in all cases. Complete recovery were successful in 244 cases, 4 patients had neurologic complications (hemiparesis in 3 cases, hypotonia in 1 case). Mortality rate were less than 1% (2 cases). The specific properties of early postoperative rehabilitation in children with CHD refer to both problems with the preoperative protocol, where the informative interview a difficult understanding patience of the parents, and investigation and evaluation of the functional status of the respiratory and the locomotor system. The postoperative protocol involves physiotherapy – thoracic light as well as spectrum of respiratory kinesitherapy from positioning, percussion of the chest, vibrations and manual hyperinflation with aspiration to provocation of deep breathing and inducing cough. In addition to respiratory kinesitherapy early mobilization of pediatric patients is very important. It includes exercise of the distal and proximal segments, early verticalization and walking. The aim of early rehabilitation in CHD is to prevent postoperative complications and promote early recovery of pediatric patients aiming at normal psychomotor development for the age and participation in all normal activities. Cardiopulmonary postoperative rehabilitation in children with CHD requires medical skills which involve a multidisciplinary approach (physiatrist, anesthetist, cardiologist) on an individually adjusted base according to the diagnosis and accompanying therapy instead of routine, in order to serve its purpose.

P1245

Normative data for volume measurements in children estimated with a double indicator technique

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Knowledge of cardiac preload is essential for perioperative management (e.g. volume vs. inotropes). However, standard monitoring parameters as central venous pressure or wedge pressure do not reflect the volume status sufficiently. Transpulmonary double indicator dilution (TDID) with indocyanine green dye provides a method for examination of cardiac function and volume parameters. Intrathoracic blood volume (ITBV) and global end-diastolic volume (GEDV) are parameters of cardiac preload. Pulmonary blood volume (PBV) and extravascular lung water (EVLW) can also be measured as well as cardiac function index (CFI=CI/GEDV) which is a parameter of cardiac performance independent of preload. Twenty-one children (age 11–154 months) without severe heart disease were evaluated with the COLD system (Pulsion, Germany) in order to establish normative data for pediatric intensive care. In each patient 3 measurements were performed. All data are indexed to the body surface area and are expressed as mean ± standard deviation (see table). These data may serve as normative data for control of postoperative volume and catecholamine therapy after surgical intervention for congenital heart disease.

Acquired Heart Disease

P1246

Rheumatic fever in indigenous children in western Australia

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Introduction: Rheumatic fever (RF) has become very uncommon in non-indigenous children in Australia. This presentation will review information concerning the incidence of RF in Western Australian children. Two recent theses for the Bachelor of Medical Science degree have addressed different aspects of RF in Western Australia (WA). Methods and Results: Johnson's thesis studied the Hospital Morbidity Data System of WA to identify cases of RF admitted to hospital from 1981–94. Seventy percent of the cases of RF were in the 5–14 age group. Over the study period a significant downward trend in the incidence of RF in the non-indigenous childhood population (5–14 years) to <2/100,000 per year was demonstrated. There was no evidence of any decrease in the incidence of RF in the indigenous population (5–14 years) which was approx 150/100,000 per year. The incidence of RF was highest in the Kimberley region of WA. Mintcham's thesis completed in January 1999 included a review of RF cases in the Kimberley from 1982–96 and an audit of aspects of management including recurrences of RF and compliance with secondary prophylaxis. A high incidence of RF of 234/100,000 per year in the indigenous 5–14 yr age group was demonstrated over the whole period with 40% of cases being recurrences. Only 11 of 88 cases of RF in 1996 and 1997 received 100% of their monthly benzathine penicillin injections. Conclusion: There is no evidence of any decrease in the incidence of RF in the indigenous population (5–14yrs) in WA over the last twenty years. The incidence of RF in indigenous children in the Kimberley is as high as any third world country with unacceptably high recurrence levels.

P1247

Surgery of the aortic root and ascending aorta in the pediatric population: techniques and results

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Dilatation of the aortic root is a well known manifestation of connective tissue disease. Idiopathic dilatation of the aortic root and ascending aorta is extremely rare. Supravalvular aortic stenosis is usually observed in Williams-Beuren syndrome. Between 1/1995 and 10/2000 a total of 505 operations on the thoracic aorta were performed in adult and pediatric patients. We present a group of 20 patients <16 years of age, who required surgery for dilatation or stenosis of the aortic root and/or ascending aorta. 10 patients suffered from Marfan syndrome, 5 patients presented with idiopathic dilatation of the aortic root (n=2) or the ascending aorta (n=3) – three of them have had prior cardiovascular surgery (PDA ligation, aortic coarctation and atrioventricular canal) – and 5 patients with Williams-Beuren syndrome had supravalvular stenosis. Mean age was 8.5 yrs (4 to 16 years). Operative technique: aortic root repair with aortic valve preservation in Marfan patients (7), homograft repair (2) and composite-graft (1). In idiopathic root dilatation 2 patients underwent Yacoub procedure and 3 patients with isolated dilatation of the ascending aorta received supracoronary graft repair. All patients with supravalvular stenosis underwent enlargement of the ascending aorta using autopericardium. Hospital mortality occurred in two cases: a 10-year old girl died from respiratory failure due to severe intrapulmonary emphysema on the 2nd postoperative day, a 14-year old Marfan patient died from low cardiac output following composite-graft, mitral and tricuspid valve repair. There was no major perioperative morbidity and no long-term mortality. Repair of the aortic root and/or ascending aorta in children and infants can be performed with satisfactory early and late results. However, these patients present sometimes with severe comorbidity which may adversely affect the early outcome. In almost every case, a surgical technique which does not require long-term anticoagulant treatment, can be offered to these young patients.

P1248

Auscultation is still superior to detect mitral regurgitation but not aortic regurgitation in acute rheumatic fever

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Carditis is seen in nearly half of cases during the attacks of acute rheumatic fever (ARF). With the widespread use of Doppler echocardiography has made it possible to detect valvular regurgitation that are not detected by auscultation which has been called silent mitral regurgitation (MR). Hence the rate of carditis has been reported as high as 80–90 %. In this study we have attempted to investigate the sensitivity of auscultation for MR and aortic regurgitation (AR) in patients with ARF. 136 ARF patients were included to study. Patients were classified according to their clinical findings: 72, 49, and 15 of patients had carditis, polyarthritis and chorea, respectively. Two-dimensional and Doppler echocardiography was performed to all patients with carditis. 32 patients with polyarthritis and 9 patients with chorea. The echocardiographic diagnosis of pathologic MR and AR were based on the previously determined criteria. The murmur of MR was heard in 8 of 72 patients with carditis. In 67 of them MR was confirmed by Doppler echocardiography. On the other hand in one patient MR was detected only with Doppler echocardiography yielding a sensitivity of 97.1 %. In 28 of 72 patients with carditis, the diagnosis of AR was made by auscultation. In an additional 11 patients AR

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

was detected only by Doppler echocardiography. Thus, the sensitivity of auscultation for AR was found as 71.8 %. Since all patients with AR except two, accompanied with MR and concerning the high sensitivity of auscultation to detect MR, our findings suggest that the clinical examination is sufficient for diagnosis of rheumatic carditis. However, in a substantial part of patients AR is easily missed by auscultation. Thus, we suggest that an echocardiographic examination is very helpful to detect double valve involvement in rheumatic carditis.

P1249

Cardiac damage as a result of blunt chest trauma in children.

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In some cases blunt chest trauma involves cardiac lesions such as: pericardial effusion, heart block, aneurysm, valvular or septal rupture. This is very rare complication in pediatric patients. Early and correct diagnosis is important to take therapeutic decisions and to prevent fatal outcome. We present 5 children after unsuspected blunt trauma and different cardiac lesions diagnosed by echocardiography. Boy with disruption of tricuspid valve and severe insufficiency, girl after traffic accident and acquired interventricular communication and girl with rupture of papillary muscle of mitral valve. Because of worsening clinical condition surgery was undertaken in all cases. Tricuspid valve valvuloplasty (in one patch closure of VSD and prosthetic mitral valve implantation was performed. In 6 month – 5 years follow up all patients are still in good condition.

P1250

Fatal Kawasaki Disease (KD) due to early fibrous obliterative coronary artery disease

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The purpose of the study is to describe 2 fatal cases of KD due to neochromic luminal obstructive coronary artery disease. Patient 1: 4yr Caucasian male. URTI, low grade fever, eventually had 4 of 5 KD criteria. Given IVIG x 2 days 15 and 17 with no response: persistent fevers and hemolytic anaemia 1 month, small joint arthritis & arthralgias 2nd month, abdominal pains 3rd month. Initial coronary artery (CA) dilatation progressed with the development of unstable angina and low cardiac output 4 months after presentation. Died after anaesthetic induction for cardiac catheterisation. Post mortem histology showed thick walled obstructed triple vessel CA disease. Histology showed marked fibrocellular luminal proliferation. There was recent myocardial and endocardial infarction. Patient 2: 6 month Caucasian male developed fever and 4 criteria for KD but no peripheral oedema or redness. Given IVIG day 8 and day 20 due to irritability without fever. Echo day 6 and day 50 showed mild CA dilatation. He represented day 95 with congestive heart failure, cardiogenic shock, poor LV function and died. Post mortem showed minor CA aneurysmal changes only. There was marked lumina obstruction caused by replacement fibrosis of the luminal and media. There was patchy chronic inflammatory infiltration. In summary, these 2 patients showed atypical KD histology with early luminal proliferation, fibrosis and luminal obstruction. Conclusion: 1. KD may be fatal due to early (within 3–4 months from onset) obliterative CA disease. 2. New treatment strategies for KD are required for late presenters or non-responders to IVIG when there is evidence of continued disease process.

P1251

Poststreptococcal reactive arthritis in children: is it really a different entity?

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We have aimed to study the clinical and laboratory features and the course of poststreptococcal reactive arthritis (PSRA) in children. The diagnosis of PSRA was established in 20 children seen in our clinic between January 1997–August 2000. Three patients did not meet the required criteria for the diagnosis of acute rheumatic fever (ARF). Among 20 patients female/male ratio was 7/13, mean age was 9.2±3.7 (3.5–15) years. Fifteen children had a history of sore throat and the latency period ranged from 2 to 14 days with a mean of 5.5–3.4 days. The arthritis was presented in mean 3.7±2.2 joints (1–10 joints) involving small and large joints. The arthritis was monoarticular in 4, oligoarticular in 10, and polyarticular in 6 patients. The arthritis was nonmigratory in 17, and symmetric in 7 patients. The mean total duration of arthritis was 21.4±13.3 days with a range of 3 to 65 days despite the administration of salicylates. One patient developed mitral valve insufficiency which was diagnosed by echocardiography during his second attack. Throat culture was positive for β hemolytic streptococci in 9 of 20 patients. High or rising titers of antistreptolysin O antibody were found in 19 cases. All patients had elevated erythrocyte sedimentation rate (> 20 mm/h). The penicillin prophylaxis was performed in 12 patients and there was no recurrence during the mean follow-up period of 7.6±12.7 months (1–45 months). PSRA has been accepted a separate entity than ARF because of its latency period, features of arthritis, and response to salicylates. During this period we also diagnosed ARF in two patients whose arthritic features were not easily differentiated from PSRA. Seven of 20 patients had also family history of ARF. We conclude that these two conditions are actually one disease. So, we suggest that prophylactic penicillin is indicated to prevent recurrences and the development of cardiac sequelae in patients with PSRA.

P1252

Incidence of pericardial involvement during the attacks of familial mediterranean fever

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Familial Mediterranean fever (FMF) is an autosomal recessive disorder characterized by recurrent, self-limited attacks of fever accompanied by inflammation of the peritoneal, synovial, and pleural surfaces. Pericardial involvement is a rare (< 5 %) but well-known feature of the disease. We have followed two patients who had recurrent pericarditis as a sole manifestation of FMF. So, we believe that pericardial inflammation is more prevalent than the contrary of general belief. We therefore undertook an echocardiographic study to assess the frequency of pericardial inflammation during the attacks of FMF. Two dimensional and M-mode echocardiographic examination was performed during the 38 attacks of 32 clinically diagnosed and genetically proven FMF patients (15 female, 17 male) aged between 5.5 to 18 years. None had amyloidosis, congestive heart failure, uremia, or other systemic diseases known to be associated with pericardial disease. All patients had careful cardiac physical examination, 12-lead EKG, and chest X-ray. Chest pain was associated in 24 of 38 attacks. No patient had clinical findings, such as friction rub suggesting pericarditis. EKG and chest X-ray during attacks were normal in all patients. Echocardiographic study revealed minimal pericardial effusion which was resolved on control echocardiogram at the end of the attack in two patients' two attacks (one with chest pain). Although echocardiographic examination did not demonstrate effusion, two other patients had chest pain strongly suggesting pericardial inflammation. We concluded that pericardial involvement is not rare in FMF attacks and sometimes may be the only manifestation of FMF. We therefore suggest an echocardiographic study should be an integral part of clinical examination of FMF attack to detect the exact frequency of pericardial disease.

P1253

Infective endocarditis in brazilian adolescents. Analysis of risk factors for in-hospital mortality.

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Purpose: To study the epidemiological, clinical, therapeutic and evolutive aspects of endocarditis in a group of brazilian patients aging 12 to 20 years old. Methods: Forty consecutive patients (21 males, 19 females) admitted with infective endocarditis retrospectively studied. Results: Infective endocarditis mortality was 30%. Reumatic heart disease was the predominant underlying condition in 65% of patients. Congenital heart disease (28%) and cardiac prosthesis (8%) were the other affections involved. The majority of patients (68%) were in functional class III and IV NYHA, with more deaths than the 32% who were in functional class I and II ($p = 0.01$). Staphylococcus aureus was the most frequently isolated agent (52% of the positive blood cultures), followed by Streptococcus viridans in 39%). Embolic complications occurred in 48% of patients. Multivariate analysis (multiple logistic regression) identified functional class III and IV at admission (OR [CI95%]=30.6[2.67–450.9], $p=0.01$), and the occurrence of embolic complications (OR [CI95%]=7.3[1.50–35.7], $p=0.01$)as independent predictors of in-hospital mortality. Conclusion: Rheumatic heart disease in Brazil remains, as an adults, the main predisposing factor for infective endocarditis in adolescents, and

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Staphylococcus aureus. Like in children, the leading agent. Mortality is high and III/IV functional class at hospital admission and embolic complications are independent predictors of in-hospital mortality.

P1254

Echocardiographic features of cardiac complications in kawasaki disease

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Analyses of morphologic and functional features of cardiac structures in Kawasaki syndrome (KS) patients, especially the coronary arteries (CA). There were 26 KS patients registered in our institute since 1988. All patients underwent echocardiographic examination at least four times. Standards of the Japan Kawasaki Research Committee were applied. 1. Children under 5 years of age with a CA lumen diameter exceeding 3 mm; 2. Where an internal diameter of a CA segment is at least 1.5 times greater than an adjacent one. Early myocardial dysfunction was established in 2 patients with KS. Pericardial effusion was found in 5 patients. Fusiform/saccular aneurysms of the CA were detected in 9 patients (34%) in the first four weeks of disease. Both left anterior descending branch (LAD) and right coronary artery (RCA) were found in 5 cases (20%); isolated aneurysms of LAD were found in 4 children. The circumflex artery was spared in all cases. The average internal diameter of the affected CA was 5.8±0.8 mm, except in a 3 months old infant, whose diameters of LCA and RCA were 8.5 mm and 8 mm, where a giant fusiform aneurysm of both CA, with thrombus in LCA, pericardial effusion, acute anterior wall infarction and ischemic myocardiopathy developed subsequently. Echocardiography is extremely useful in assessing cardiac complication in KS. CA lesions occur in a third of KS patients. Giant aneurysms of 8 mm or more present a disproportionately higher risk of myocardial infarction compared with aneurysms of smaller dimensions.

P1255

Tolmetin and salicylate therapy of acute rheumatic fever. comparison of efficacy and side effects

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The arthritis of rheumatic fever is very responsive to salicylates but there are many adverse reactions especially hepatotoxicity due to aspirin therapy. These side effects change the course and the duration of rheumatic fever. Other non-steroidal antiinflammatory agents may be equally effective, although no report is available. We studied 52 patients with rheumatic fever who were admitted to Dr Sami Ulus Children's Hospital between 1995–1999. Twenty patients with arthritis (group I) were treated with tolmetin (25 mg/kg/day) and 32 patients with arthritis and/or mild carditis (group II) were put on salicylate therapy (75 mg/kg/day) for 4–6 weeks. Arthritis had disappeared at the same time in both aspirin and tolmetin group. No adverse effect of tolmetin therapy was observed whereas side effect of salicylate group was observed in 19 patients (36.5 %). Hepatotoxicity, salicylism and gastric irritation were observed in 16, 3 and 4 patients, respectively. Renal toxicity and Reye syndrome was not demonstrated. Because of these side effects of the salicylate group aspirin therapy had to be stopped for 10–20 days and the duration of hospitalization of this group was lengthened unnecessarily. Aspirin has long been the mainstay of the acute therapy of rheumatic fever. For patients who cannot tolerate aspirin, other non-steroidal antiinflammatory medications may be used. Our study demonstrated that tolmetin can be used in rheumatic fever. However more studies are needed to evaluate the subject.

P1256

The Ross Procedure In Children

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Introduction. The Ross procedure has been proposed as the optimum aortic valve substitute in the growing child. We have reviewed our experience, specifically addressing concerns over autograft dysfunction and the need for right ventricular outflow tract (RVOT) reintervention. Methods: A retrospective review of 31 consecutive patients between 1991–2000 was performed. Median age was 7 years (range 1 month–17 years). Five patients were < 1 year and 9 patients < 2years. 25 patients (81%) had undergone a previous aortic valve intervention (median 1.4 interventions). RESULTS: There were 3 hospital deaths (9% 4–14%, 70% CI); no deaths occurred in the recent 5 years. One patient required aortic root replacement due to autograft failure at one week. The remaining 27 cases demonstrated excellent autograft haemodynamics with peak Doppler velocities of 1.25 +/-0.14m/s, with no cases of greater than mild aortic incompetence (AI). Follow-up was 100% complete, mean 38 +/-6months. There were no late deaths. Reintervention was necessary in 3 patients (1 autograft replacement for AI, 1 RVOT conduit replacement and 1 RVOT balloon valvuloplasty) with an actuarial freedom from reintervention of 74% at 10 years. There have been no endocarditis or thrombo-embolic events. Conclusions: This study has shown that the Ross procedure provides good haemodynamic performance and that midterm clinical outcome in children. Careful follow-up is mandatory to identify and intervene particularly with progressive RVOT conduit obstruction.

P1257

Echocardiographic assessment of mitral valve apparatus in active rheumatic carditis

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Purpose – To assess the anatomy of mitral valve apparatus in pts with acute rheumatic carditis we performed between January 1998 and November 2000 echocardiographic study in 121 children with acute rheumatic fever diagnosed by modified Jones' criteria. From them, 25 (61%) had carditis and cardiac failure (II co III NYHA) with ages ranging from 4 to 15 years (mean 9.3). All of them had negative blood test for bacterial endocarditis. Methods – The echo study was done using the classical views with a 2.5D Challenger echomachine with pulsed, continuous and color flow Doppler. The mitral annular diameter in the the maximum ventricular diastole and the systolic chordal length at the end of ventricular systole were measured and compared to values obtained from matched control group. Results – 17 pts (25%)(group I) showed flail of mitral valve and 13(13%) group II mitral valve prolapse. The maximum annular diameter was significantly greater in group I, 38.4(+/-1.1)AI and 37.2(+/-5.9) in group II than matched group, 24.0(+/-2.8) mm, p<0.01. The maximum systolic chordal length was 31.4(+/-5.6) in group I, 28.5(+/-9.2) in group II and 18.1(+/-4.5)mm in the control group, p<0.01. CONCLUSION – Mitral regurgitation in acute rheumatic carditis is due to a combination of mitral annular dilatation and chordal elongation leading to mitral valve prolapse and in severely ill patients, flail of mitral valve.

P1258

Predictive values of echocardiographic findings in patients with symptomatic cardiac manifestation in HIV infected children

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Background. Cardiac abnormalities are common in HIV infected children contributing to morbidity and mortality. This study aims to determine spectrum of cardiac abnormalities in these patients using echocardiographic studies in symptomatic HIV infected children and to predict risk values regarding morbidity and mortality. Method: Twenty seven perinatal HIV infected patients with cardiac manifestation underwent cardiac evaluation by echocardiography between 1995–2000 were retrospectively studied. Result: The first cardiac evaluations were carried out at mean age of 35 months (range 11–65 months). Only one patient received antiviral drug. All patients were in categories B or C of pediatric HIV clinical classification presenting with dyspnea and cardiomegaly. Signs and symptoms included edema in 8 (30%), clubbing of fingers in 6(22%), cyanosis in 4 (15%), S3 gallop in 5(18%). Associated pulmonary infection was presented in 20(74%) and bacterial septicemia in 4(15%). Echocardiographic abnormalities noted in all patients included, pericardial effusion in 20 (74%, 5 with fibrin), diminished fractional shortening in 11(40%), hyperdynamic LV in 15 (55%), RV dilatation in 11(40%), LV or combined ventricular dilatation in 12(44%) and increase RV pressure in 19(70%). Death occurred in 21(78%) within 24 months after documented finding of cardiac manifestation. Conclusion: Echocardiographic abnormalities in symptomatic HIV-infected children with cardiac manifestation commonly present in 3 spectrums 1) infectious pericarditis with pericardial effusion 2) RV dilatation, increase RV pressure and hyperdynamic LV are very common in patient with advance HIV with repeated pulmonary infection 3) LV dilatation and dysfunction. All HIV infected children with cardiac manifestation were usually associated with high mortality within 1–2 years after diagnosis. The detail values of echocardiographic findings predicting the course of illness will be presented in details in each categories.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P1259

Acute cardiac failure in critically ill children : early Kawasaki disease ?

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The causes of acute cardiac failure are numerous. The absence of the classical criteria for Kawasaki disease (KD) in the early clinical course may delay the consideration of this diagnosis. A list of diagnostic criteria for acute cardiac failure, myocarditis, cardiomyopathy and KD were established a priori by 3 experts using a 3 round Delphi method. The charts of all patients admitted to the Pediatric Intensive Care Unit (PICU) from 1987 to 1998 with the above mentioned diagnoses were reviewed independently by 3 physicians. The diagnoses and the serology were assessed taking into account all relevant data that had been collected at entry and after PICU stay. Odds ratio (OR) and 95% confidence interval (CI) were calculated for each item. Among the 25 patients in acute cardiac failure (symptom having < 3 weeks), 7 fulfilled the diagnostic criteria for KD (24%). 16 had acute cardiomyopathy. Significant differences were obtained for the following clinical criteria at entry and for the entire hospital stay respectively. Fever more than five days: OR (1.25 and 13.34), CI (0.6–18.4) and (1.03–11.7). Skin rash: OR (7.05 and 18.9), CI (1.75–28.2) and (2.6–131); Conjunctivitis: OR (7.05 and 7.8), CI (1.75–28.2) and (1.2–46.6); Pericardial effusion: OR (7.05 and 7.8), CI (1.75–28.2) and (1.3–46.6); Hydrops of the gallbladder: OR (5.5 and 5.5), CI (1.4–22) and (1.4–22). A significant proportion of patients admitted in acute cardiac failure in PICU develop KD. The classical criteria for KD may be absent in the early evolution of these patients. KD should be part of the differential diagnosis of acute cardiac failure in children admitted in PICU. Prospective studies are needed to confirm these data.

P1260

Early peripheral vascular manifestations of heterozygous familial hypercholesterolemia

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Thirty-four children aged 13.0±3.6 years with heterozygous familial hypercholesterolemia (FH), carrying the frequent French Canadian mutation (5-10kb deletion) underwent cardiac and peripheral vascular evaluation. Twenty-nine patients (pts) (85%) had a family history of premature atherosclerosis. Physical examination was normal in all including blood pressure, body mass index (BMI) and absence of xanthoma except 2 pts with tendon xanthoma. Resting and exercise electrocardiograms as well as echocardiogram were normal. Ultrasound provided transcutaneous measurements of carotid, brachial and femoral arteries flow kinetics at rest, immediately after cuff deflation, 20 seconds and 5 minutes later (brachial and femoral arteries: cuff inflation at 200mmHg for 3 minutes). The following data were obtained: peak, diastolic and mean velocities (cm/sec) and Pourcelot resistance index (RI). These results were compared to those of 33 control subjects (ctrl) of same age, blood pressure and BMI. Cuff deflation resulted in an increase in velocities and a decrease in resistance index in all subjects. Brachial peak and mean velocities of pts were similar to those of ctrl under all conditions. Changes in velocities were also similar in both groups. However, femoral mean velocity at rest and 20 sec after cuff deflation was significantly lower in pts than ctrl. Femoral RI at rest was significantly higher in pts than ctrl (1.32±0.11 vs 1.25±0.09, p<0.01). Both brachial and femoral RI at 20 sec and 5 min after cuff deflation were significantly higher in pts than ctrl. In conclusion, although having normal cardiac evaluation and having responded in the same fashion to reactive hyperaemia, children with FH show a higher peripheral resistance index following cuff deflation. This may reflect already at this early stage of the disease the presence of an endothelial dysfunction.

P1261

Physical and psychosocial functioning in children who have had Kawasaki Disease (KD)

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The Child Health Questionnaire (CHQ) is a parent questionnaire which measures overall physical and psychosocial well being in children 5–18yrs. To study the long-term impact of KD on overall health status, we mailed the CHQ to four groups of KD patients: a sample without a history of coronary artery abnormalities (CAA) (Normal group), those with previous coronary abnormalities that regressed (Regressed Group), those with current mild or moderate CAA (8mm (Mild-Mod Group), and those with current giant aneurysms (≥8mm (Giant CAA group). Of 197 questionnaires mailed, 172 were deliverable, of which 108 (63%) were returned. Patients for whom questionnaires were not returned did not differ in gender, coronary status, age at onset or time since illness. Median age (range) at KD onset was 3.1 yrs (2–12 yrs) and at CHQ completion was 10.5 yrs (5.8–17.9). We compared mean Physical Health Summary (PHS) and Psychosocial Health Summary (PSS) scores of each KD group to normative values (see table). Among subscale scores, the Giant CAA group, compared to the normative population, had lower scores on General Health Perceptions (P< 0.01). Compared to the normative population, KD patients reported more anxiety issues (P=0.05), allergies (P=0.04) and orthopedic/bone/joint issues (P=0.004). In summary, KD pts without giant CAA were similar to the normative population on their overall physical and psychosocial health. Those with giant CAA had lower overall physical summary scores and general health perceptions.

P1262

Echocardiographic abnormalities in rheumatic fever (RF) with or without clinical evidence of carditis – a five year follow-up

Ferreira, W.F, Silva, C.M.C, Terreri, M.T, Albuquerque, P, Hangai, L., Moraes, V.R, Carvalho, A.C.C., Hilário, M.O
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Echocardiographic abnormalities in rheumatic fever (RF) with or without clinical evidence of carditis. Ferreira WF, Silva, CMC, Terreri MT, Albuquerque, P, Hangai L., Moraes VR, Carvalho, A C C , Hilário, M.O. Universidade Federal de São Paulo - EPM São Paulo-Brazil. The value of echocardiography in detecting carditis in patients(pts) with RF without clinical evidence of cardiac involvement has drawn attention and has motivated some studies. The aim of this study was to determine the follow-up of echocardiographic finding in patients with RF which during the first episode with or without clinical evidence of carditis. Two groups of children with RF, diagnosed according to the 1992 modified Jones criteria were evaluated by echocardiography. Group I: 16 pts at the acute phase (8 with clinical carditis and 8 without it) and 3 months later (14/16). Group II: 16 patients were evaluated at the first episode and re-evaluated 5 years later (6 with clinical carditis and 10 without it). In the group I, all the patients with clinical carditis and 3/8 without it presented echo abnormalities. At the 2nd evaluation, 5/7 with clinical carditis and 1/7 without it persisted with echo abnormalities. In group II, 5/6 patients with clinical carditis persisted with echo abnormalities while 3/10 without clinical carditis also persisted with echo abnormalities 5 years after the last evaluation. In conclusion this study shows the possibility of occurrence of asymptomatic carditis and importance of echo in its diagnosis and follow-up.

P1263

Is subclinical valvulitis a major diagnostic criterion in diagnosis of acute rheumatic fever?

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The diagnosis of acute rheumatic fever (ARF) is difficult when the only major manifestation is non-cardiac. Recently subclinical valvular insufficiency has been identified using Doppler echocardiography, and proposed as evidence of subclinical valvulitis. This study aims to prospectively analyse the association of subclinical valvulitis in non-cardiac rheumatic fever and to describe its prognosis. Between December 1998 and September 1999, patients without clinical signs of carditis, who were diagnosed as ARF in our institution were included in this study. The diagnosis was made according to Jones criteria (JC), except in two cases who had strong evidence of streptococcal infection and migratory arthralgia without prominent joint swelling or hyperemia. Doppler echocardiographic examinations of all patients were done at 2 week-intervals during acute attack and monthly thereafter. Six female and 17 male patients were aged between 6–16 years. Major findings of JC were arthritis in 16, chorea in 4, carditis and erythema marginatum in 1 patient. Two cases had arthralgia with equivocal arthritic signs, but definite color Doppler finding of mitral regurgitation (MR). Silent pathologic MR was found in 9 cases, and aortic regurgitation (AR) in 2 cases. After a mean follow-up of 4,47 months valvular regurgitations disappeared in 4 of them, including the patient with migratory arthralgia and no major criteria. All three patients with chorea still have MR at the end of this period. ARF without clinical carditis is not a benign entity and Doppler echocardiographic findings should be added to the existing JC for the diagnosis of ARF.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Cardiac Imaging: Angiography

P1264

Echocardiographic on-line quantification of left ventricular systolic function in children

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The aim of the study was to evaluate the clinical usefulness of an on-line automatic border detection system (accuasoc quantification AQ) for determination of left ventricular volumes and ejection fraction in children in comparison to the conventional off-line method. 107 patients were enrolled in the study. The ages ranged from 0.1 to 18.8 years (mean 8.3 ± 5.6). Left ventricular volumes obtained by AQ correlated well (r = 0.96) but were slightly underestimated compared to those determined by manual tracing. Mean ejection fraction was 61 ± 6.8 % by AQ compared with 61.5 ± 5.9 % by manual tracing (r = 0.89). The time necessary for acquisition of data was similar in both methods. AQ seems to be a promising method for real-time estimation of left ventricular volume, even in children.

P1265

Aneurysm of ductus arteriosus – normal or abnormal ?

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Ductus arteriosus aneurysm (DAA) has been considered as a rare congenital lesion. A prospective study was performed to describe the incidence, clinical manifestations and outcome of DAA in full-term neonates. A total of 360 full-term newborns received a screening echocardiography after informed consents obtained from the parents. There were 34 cases (9.4%) with DAA by echocardiography. Three-dimensional magnetic resonance angiography and aortography confirmed the diagnosis of aneurysm in 5. There were no symptoms and no significant differences in gender, gestational age, maternal age and Apgar score between the newborns with or without DAA. There were higher birth body weight, higher incidence of maternal gestational diabetes, and mothers with blood group 'A' in newborns with DAA (p<0.05). Follow up echocardiograms showed spontaneous closure of ductus aneurysm in all cases but significantly later than those without DAA (p<0.01). The DAA became progressively smaller after ductal closure in 22 cases (64.7%), and completely disappeared by 7 to 35 days of life. The other 12 cases (35.3%) of DAA had aneurysmatic evidence of progressive thrombus formation during 3rd and 30th day of life. The DAA and thrombi spontaneously disappeared in all cases after 1 month of life. In conclusion, there is a higher incidence of DAA and good outcome in our study than the previous reports. Although many theories of pathogenesis have been proposed, the mechanism of the aneurysm formation remains uncertain. We speculate that the presence of DAA may be a normal process of spontaneous ductal closure in full-term neonates.

P1266

Magnetic resonance imaging (mri) in the diagnosis of congenital right coronary artery fistula

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Closure of coronary artery fistulas is recommended at time of diagnosis since delayed assignment to treatment is associated with increased morbidity and mortality. Because detailed characterization of fistulous connections is essential for proper management, angiography has been widely used as the major diagnostic tool. We proposed that magnetic resonance imaging (MRI) could provide adequate information in this context and examined that in three cases. Patients (2 female) aged 7, 10 and 20 years, at functional class I to II (NYHA) were subjected to diagnostic procedures to establish the anatomical basis for a continuous murmur heard along the left sternal border. Chest X-ray showed moderate cardiomegaly with right atrial and right ventricular enlargement in all of them. The presence of a fistula involving the right coronary artery was suspected during echocardiographic evaluation in two patients. MRI allowed detailed delineation of blood flow besides showing the exact termination site of the fistulous connection: right ventricle, coronary sinus and right atrium respectively. Findings were very similar to those observed during angiographic examination and further confirmed at operation. We conclude that MRI provides anatomical information sufficient for proper surgical management of congenital fistulas involving coronary arteries, suggesting that diagnosis may be adequately established on a noninvasive basis.

P1267

Transesophageal echocardiography using a 4mm longitudinal plane probe.

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We tested a 4 mm Sudhir 30 clearum transesophageal probe (Aloka, Tokyo). Aim of the study was to investigate whether this probe would provide high quality imaging and Doppler information. Methods: The studies were performed in 165 patients (pts), either in the operating room (n=13), or in the catheterization laboratory (n=152). The age range was 7 hrs-14 years, the body weight range was 2.7–41 kg (18 pts were < 4 kg). All pts were intubated. Results: The probe could easily be inserted without laryngoscopy in all pts even in the smallest ones, and after surgical draping. Excellent 2D imaging and color flow mapping was obtained in the near field, till a depth of 5 cm. Conclusion: longitudinal plane transesophageal imaging using a 4 mm probe is feasible and provides excellent information without 5 cm of the esophagus. Using the probe in combination with 4 mm transverse plane probe sequentially biplane imaging in UNSELECTED very small children (< 3 kg) becomes a possibility.

P1268

Application of tee for the children suffered from tetralogy of Fallot

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Seventeen children suffered from TOF whose age was between three and fourteen years old were checked up and radically operated. For all patients TEE was applied before and after the operation. Before the operation on seven patients had defined outflow VSD, which we can see on Japanese Tetralogy of Fallot. And other children had defined membranous defects. Investigation of right ventricle outflow tract have showed that eight children have had pulmonary artery annulus hypoplasia and one has had membrane under the PA valve. TEE was applied after taking off the clamp from aorta. For all patients on patch outflow VSD residual flow was not different. On tree patients was defined inadequacy of tricuspid valve of the I-st degree, and on one the with second degree, that was the reason of repetition of plastic of the valve during the same operation. For eight children with PA annulus hypoplasia was made transannular plastic and during TEE was found inadequacy of the second degree. And other children's annulus was saved and inadequacy of the first degree was found on three children. On the other cases there was no inadequacy. So application of TEE gave the possibility to estimate the results of the correction even at operating room.

P1269

Operated aortic coarctation. A study of the aortic arch by magnetic resonance

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Although good results are usually achieved with surgical correction of aortic coarctation, the transverse arch can be hypoplastic leading to persistent gradient. We analyzed the dimensions of the aortic arch in infants and children operated of coarctation to evaluate the association of arch hypoplasia. Patients and Methods: 65 infants and children operated under the age of fourteen years were studied by magnetic resonance imaging. The mean operative age was 6.0±4.6 years (range 22 days-14 years). The mean follow-up time was 5.8±4.3 years (range 45 days-16 years). They were divided in four groups according to the operative age: [0–2], [2–5], [5–10] and >10 years. The surgical techniques were resection with end-to-end anastomosis in 42 patients, enlargement with patch in 12, subclavian flap angioplasty in 7 and other techniques in the remaining four. Aortic diameters were measured at four predetermined points and the ratios between each measure and the aortic diameter at the diaphragmatic level were calculated. Segments with ratios smaller than 0.9 were considered hypoplastic, those greater than 1.5 were assumed as dilated; recoarctation was considered when it was smaller than 0.6 at the site of coarctation. Results: The mean ratios along the arch were progressively and significantly smaller from the ascending aorta to the isthmus in all age groups (p < 0.00001). There was no significant difference between the ratios of each level analyzed from all age groups. The distal transverse aortic arch was hypoplastic in 31 (47.7%). There was no statistical association between the presence of transverse arch hypoplasia and age group or type of operative technique. Recoarctation was found in three patients and aneurysm in two.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

Conclusions: Hypoplasia of the transverse arch was highly prevalent independently of the operative technique, and may contribute to persistent post-operative gradient.

P1270

Vascular rings and their effect on tracheal geometry

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Background: The major clinical impact of vascular rings in children is tracheal compression, which presents with recurrent symptoms of stridor, noisy breathing, and wheezing. We tested the hypothesis that symptomatic patients have altered tracheal geometry with respect to non-symptomatic individuals. Methods: We retrospectively reviewed the tracheal dimensions (area, longest and shortest diameters) as evaluated by magnetic resonance imaging from the cupola of the lung to the carina of 49 patients referred for evaluation of a vascular ring. The smallest dimension relative to the largest dimension (% of maximum) as well as the coefficient of variation (CVAR = standard deviation / mean) were assessed for each parameter. Significance P<0.05. Results: In all parameters measured (area, longest and shortest diameters), patients with symptoms had significantly different values than patients without symptoms (see table). The % of maximum was significantly smaller and the CVAR was significantly greater in symptomatic individuals than in non-symptomatic individuals for all the parameters. Conclusions: Patients with vascular rings who are symptomatic have significantly altered tracheal geometry with respect to non-symptomatic individuals. Magnetic resonance imaging is a useful tool for visualizing both the cardiovascular and tracheal anatomy in patients with vascular rings and adds useful information to the management and care of these patients.

P1271

The value of magnetic resonance coronary angiography in children after arterial switch operation

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During arterial switch operations (ASO) coronary arteries are translocated to the neoaorta. Recognized long term complications of this procedure are coronary artery flow abnormalities. To investigate the value of magnetic resonance coronary angiography (MRCA) in children after arterial switch operation (ASO), thirteen children (4 healthy volunteers and 9 ASO (age 9.0–15.6 year) were studied. Two- and three-dimensional MRCA was performed at 1.5 Tesla (Vision, Siemens), using a phased-array surface coil. No sedation was used. One patient was studied pre and postoperatively. Image quality was good to excellent. In 11 children both offspring and proximal course of both coronary arteries could be identified. In 3 abnormal findings were revealed. In one an unexpected course of the RCA, in one a fragile LCA (both asymptomatic), and in one, who had ECG changes during exercise, the proximal LCA could not be identified. After reoperation of the latter patient (where LCA kinking was confirmed) a new MRCA revealed a well recognizable LCA. In children after ASO, MRCA is a useful non-invasive tool to investigate patency and proximal course of the coronary arteries. It may become a helpful adjuvant in selecting or deferring invasive angiography.

P1272

Assessment of cardiac sympathetic nerve activity in children with chronic heart failure using quantitative iodine-123 metaiodobenzylguanigine imaging

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Cardiac sympathetic nerve activity in children with chronic heart failure was examined by quantitative iodine-123 metaiodobenzylguanigine (I-123 MIBG) myocardial imaging in 33 patients aged 7.5 ± 6.1 years including 8 with cardiomyopathy, 15 with congenital heart disease, 3 with anthracycline cardiotoxicity, 3 with myocarditis, 3 with primary pulmonary hypertension and 1 with Pompe's disease. Anterior planar images were obtained 15 minutes and 3 hours after the injection of I-123 MIBG. The cardiac I-123 MIBG uptake was assessed as the heart to upper mediastinum uptake activity ratio of the delayed image (H/M) and the cardiac percentage washout rate (%WR). The severity of chronic heart failure was class I (no medication) 9 patients, class II (no symptom with medication) 9 patients, class III (symptom even with medication) 10 patients, and class IV (late cardiac death) 6 patients. H/M was chronic heart failure class I 2.33±0.22, class II 2.50±0.34, class III 1.95±0.61, and class IV 2.39±0.29, respectively ($p<0.05$). %WR was chronic heart failure class I 24.8±12.6, class II 21.2±10.2, class III 49.2±24.5, class IV 66.3±26.5 (%), respectively ($p<0.05$). The low H/M and high %WR were proportionate to the severity of chronic heart failure. By comparison of 17 patients with chronic heart failure class I or II and 8 patients with severely cardiac events (2 patients of class III and all patients of class IV), a threshold of H/M and %WR for prediction of severely cardiac events was 1.6 as H/M or 48% in %WR. In conclusion, the cardiac I-123 MIBG showed cardiac adrenergic neuronal dysfunction in children with severe chronic heart failure. Quantitative I-123 MIBG myocardial imaging is clinically useful as a predictor of therapeutic outcome and mortality in children with chronic heart failure.

P1273

Brain to lungs count ratio is a simple index to detect and quantify intrapulmonary right to left shunt

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Arteriovenous pulmonary fistulae is a serious complication in patients with cavopulmonary connection. Its detection requires invasive non-quantitative methods such as pulmonary angiography. Our aim was to assess intrapulmonary right to left shunt with brain to lungs count ratio. Ten patients (8±5 years) were prospectively enrolled. 5 pts with cavopulmonary connection were cyanotic (aortic saturation 79±10%) and 5 pts were acyanotic (pulmonary artery stenosis in 1, surgical correction of abnormal total pulmonary venous return in 4). None had intracardiac shunt. Lung perfusion scintigraphy was performed in supine position after injection of Tc-99m MAA (dose/MBq) = weight (kg) with a minimum of 37 MBq and a maximum of 74 MBq). A static acquisition including brain and lungs was performed in a posterior view (180s, 256x256 matrix size). In cyanotic patients, the mean brain to lungs count ratio value was 16±18% whereas in acyanotic patients, mean value was 0.6±0.3% ($p<0.005$). Pulmonary angiography confirmed arteriovenous pulmonary fistulae in 4 cyanotic pts. Lung perfusion scintigraphy with brain to lungs count ratio is a simple and useful technique to assess and quantify intrapulmonary right to left shunt.

P1274

The transepicardic echocardiogram during surgery of congenital heart disease in children.

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The transepicardic echocardiogram during surgery of congenital heart disease in children. Kreutzer R., Jatzky C., Di Sanio M., Solsona C., Villa A., Rozenbaum. Buenos Aires, Argentina. The purpose of this paper is to evaluate the detection of residual pathology immediately after surgery mainly in patients (p) of early age and low weight. Sixty one p between 1 month to 10 years of age (x. 24 m) weighting 3 to 27 kgs. (x. 9.0 kgs.) were investigated with transepicardial echocardiogram (TEE) immediately after disconnecting the extra corporeal circulation (ECC). An ATL Ultramark 9 equipment with a 5 MH. transducer was used. The transducer as well as the connecting cable with the equipment was wrapped with an sterile polietilene cover. The operated pathology was: 22 T of Fallot, 2 isolated mitral pathology, 7 A.V canal, 3 Switch correction of d-TGV, 1 Ebstein malformation, 5 Total anomalous pulmonary venous return (TAPVR), 11 l-TGV, 14 VSD, 2 ASD, 2 Fontan, and 1 DORV. Conventional echocardiograms were performed 1, 24, and 48 hours after surgery. Residual defects immediately after surgery were detected in 9 of the p such as one with Fallot and remanent low ventricular stenosis, one p. with TAPVR with anastomosis obstruction, another Fallot required a transanular patch, one p with mitral insufficiency, which became normal after surgery, and one p with A.V canal with incomplete closure of the VSD and 1 Switch with supravalvar stenosis in the neoaortic suture line. All these p were corrected during the same surgical procedure. No arrhithmias were recorded either during or after the operation and no infection occurred. In conclusion, the TEE was able to evaluate immediately the surgical result, it was easy to perform in small children, there was no morbidity secondary to the method in the present series and it is a cheap and easy option when a transesophageal echocardiogram is not available.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P1275

Accuracy of real-time three-dimensional echocardiography for determining left ventricular volumes and ejection fractions: a physiologic balloon model and experimental animal model with sonomicrometry measurements of lv volumes

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The purpose of this study is to analyse dynamic cavity volumes in a balloon model mimicking the left ventricle and experimental animal left ventricle using real-time three-dimensional echocardiography (RT3D). The single peat like balloon mimicking left ventricle was mounted in a water bath and filled with 10 different volumes of water to produce a known end-systolic volume. Pulsatile flow was generated by a flow pump to expand balloon rhythmically. The 3.5 MHZ matrix-array transducer of the real-time scanner Volumetrics Model 1 (Volumetrics Inc) was set under the water bath to image the balloon longitudinally from the apex. Reference EDV, ESV and EF were compared with RT3D data. In an in vivo animal model we measured LV volume by a 14 crystal endocardially implanted sonomicrometry array and compared them with RT3D data. For these numbers under 25 spacing > 1.17 (B-scan) or 1.9 mm (C-scan) there was a significant difference between B- and C-scan ($P < 0.05$). At more than 25 slices for volume measurements, B- and C-scan results became closer each other but both were still significantly smaller than reference ($P < 0.05$). EF was underestimated in B-scan and overestimated in C-scan when volumes were measured using a smaller number of slices. When implemented as a dynamic animal model, a multislice algorithm which uses all the 3D data combining C scans and B scans, RT3D LV end diastolic ($r=0.90$) and end systolic ($r=0.91$) correlated well with volumes measured by a implanted sonomicrometry array. Our in vitro and in vivo data show that capitalizing on the robust data within the RT3D images by including large numbers of B- and C-scan sections in measurement significantly improves accuracy of determining LV volumes.

P1276

Atrial septal defect with 'false cor triatriatum dextre': a clinical case

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A 27 years old female with progressive effort dyspnea was sent for transthoracic echocardiogram to our Echocardiography Laboratory. This exam showed right cavities enlargement. An atrial septal defect could not be excluded. A very mobile structure in the right atria was detected. The transesophageal echocardiogram showed an atrial septal defect with right to left flow. The very mobile 'membrane type' structure was confirmed in the right atria, with insertion in the inferior vena cava and near the coronary sinus. It interfered with tricuspid valvular motion, causing a mild tricuspid regurgitation. Pulmonary venous return was normal. By cardiac catheterization, the pulmonary to systemic output ratio was 1.9:1. The patient was referred to surgery with the diagnosis of atrial septal defect and 'false cor triatriatum dextre'.

P1277

Application of differential color image helical CT angiography for the diagnosis of complicated congenital heart disease

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Backgrounds: We have developed differential color imaging helical CT angiography technique and reported an application for neonates with TAPVD. Here we studied diagnosis, scanning conditions, and problems of all the examinations which we have done during the past two years. Patients and Methods: Of 205 helical CT angiography examinations, differential color imaging 3-D reconstruction was applied to 98 patients (2neonates, 19infants, and 19children). The diagnosis includes PA+VSD+MAPCA (8), TAPVD (7), CoA (7), TOF (7), HLHS (6), PAPVD (4), IAA (3), Asplenia+PA (3), peripheral PS (3), and others (11). Patients were scanned with 2mm collimation width and table pitch for neonates, infants, and small children, and with 3mm for older children. After arteries and veins had been determined by the shape, continuity, and CT density, each area of interest was determined. 3D image of the arteries and veins were reconstructed (3.7mm) and were displayed in red and blue, respectively. Results: All the examinations including neonates with IDCHS, TAPVD with pulmonary congestion were done safely and less-invasively. False positive findings were detected in three patients (4.2%), however, false negative was not found. Conclusions: Differential color imaging helical CT angiography is less-invasive and very useful for recognizing spatial information of arteries and veins and for surgical operation of complicated congenital heart diseases.

P1278

Vascular rings – Magnetic resonance imaging as first line imaging technique?

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Introduction: Recurrent or persistent congenital stridor leading to dyspnea requires extensive, in part invasive, diagnostic examinations as besides flabbiness of the epiglottis or weakness of the airway walls numerous causes must be considered. Magnetic resonance imaging may be useful, particularly in demonstrating vascular rings. Methods and patients: Since 1997 MRI studies were performed on 18 patients suspected for vascular rings aging 3 months to 15.8 years (median 8.5 months) using a multislice spinerho technique and at least one bright blood technique. A total of 45 diagnostic imaging procedures have been carried out prior to the MRI examination (5 bronchoscopy, 5 esophagoscopy, 6 chest x-rays, 4 esophageal contrast studies, 16 echocardiographys, 4 MR/CT and 7 heart catheterization). Results: In three patients vascular anatomy was normal, 15 patients show vascular anomalies (1 double aortic arch, 3 right sided aortic arch with left arteria lusoria arising from a diverticle of Kommerell, 2 atypical high origin of the truncus brachiocephalicus, 2 right arteria lusoria, 2 pulmonary sling, 1 coarctation and 1 stenosis of left pulmonary artery). The time interval between the appearance of symptoms and first correct diagnosis was < 3 months in 7, > 2 months < 6 months in 3, > 6 months < 1 year in 4 and > 1 year and < 15 years in 4. Out of the 45 preceeding diagnostic procedures 22 failed to established the right diagnosis. Even the MR/CT examinations failed, particularly due to inadequate acquisition technique and slice thickness. Conclusion: We conclude that MRI is an excellent technique to rule out vascular rings and should be carried out as first line imaging technique in symptomatic patients. However, the investigators should be familiar with cardiovascular MRI.

P1279

Role of echo-Doppler for evaluation of cardiac involvement in thoracoomphalopagus (conjoined) twins

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Background: Conjoined twinning is very rare and the successful separation depends on the degree of communication between the two twins. Cardiac involvement being a common limiting factor for a successful separation. A combination of diagnostic modalities have been described for clear delineation of cardiac anatomy in conjoined twins. Methods: We describe our experience with echocardiography and Doppler in seven sets of thoracoomphalopagus twins, aged 4 hours to 15 days, seen over the last 12 years at our center. Echocardiography was performed from multiple available windows including parasternal, abdominal and suprasternal to define the various chambers and outflow tracts for each twin and the extent of communication of their hearts. Results: Two separate hearts, complete with their outflow tracts were seen in one set which was successfully separated. Two sets of twins had a common pericardium but separate hearts. In both of these, the heart was normal in one twin and had complex cyanotic congenital heart defect in the other twin. Separation was performed, however only one twin with normal heart survived. Fusion of atrial and ventricular chambers of various extent was diagnosed in the other four sets of conjoined twins and separation was attempted in each of these, none of these babies survived. Preoperative cardiac catheterization and angiography was performed only in one of these four sets to delineate the anatomy although no further information was added. Echo Doppler findings could be confirmed in six of seven sets of twins either at surgery (5) and/or at autopsy (2). Conclusion: Echo-Doppler can provide an accurate assessment of anatomy of heart and outflow tracts in conjoined twins and angiography may be required only in select cases.

P1280

Dipyridamole stress ultrasonic myocardial tissue characterization in patients with Kawasaki disease

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https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

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We examined the feasibility of using dipyridamole stress integrated backscatter (IBS) for evaluation of myocardial ischemia or damage in patients with Kawasaki disease (KD). Dipyridamole stress IBS was used in 31 patients with coronary artery lesions due to KD ranging in age from 2 years 8 months to 20. All patients underwent echocardiography at rest and after dipyridamole stress at three left ventricular wall segments in the short-axis view; anterior interventricular septum (AS), posterior wall (PW), and inferior wall (INF). At rest, there was no significant difference of IBS in the regions with normal or abnormal distribution on Tl-201 imaging. After dipyridamole stress in contrast, the cyclic variation (CV) of IBS in the regions with abnormal distribution became significantly smaller than that in the regions with normal distribution in each segment, 3.6±1.2 vs 6.2±1.7 dB in AS, 3.1±1.3 vs 8.0 ±1.9 dB in PW, and 4.0±1.4 vs 7.3±1.6 dB in INF ($p< 0.001$). One hour after stress, the CV recovered to the level at rest in all patients. When values below 5 0dB during stress were defined as abnormal, the sensitivity of abnormal CVIBS was 75% in PW, 91% in INF. Specificity was 91% in PW, and 94% in INF, in comparison to Tl-201 imaging. There was no significant difference in R-R intervals or blood pressure responses between the patients with normal or abnormal distribution. No one was terminated from the study because of serious side effect. Dipyridamole stress integrated backscatter successfully demonstrated silent myocardial ischemia or damage at the time when cardiac systolic function is apparently normal. The CVIBS determination with dipyridamole stress could be a helpful new addition to current cardiac monitoring methods in long-term follow-up of the patients with KD.

P1281

Spiral CT angiography in children after stents implantation to aorta.

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Spiral CT angiography (CTA) is used for follow up after vascular interventional procedures mainly in adult patients (pts). The purpose of this study was to evaluate usefulness of this method in evaluation of anatomical results of stents implantation in children with different types of aortic narrowings. In 14 children (5 with middle aortic syndrome, 3 with native CoA, 6 with recoarctation of the aorta) 18 stents – 17 Palmaz stents (9 Extra large, 8 if ar) and 1 CP stent were evaluated. Age at the time of intervention ranged between 4–18 years (mean 11.5+/-5.4). CTA was performed 24 hours, 3 months, every next year after implantation and in any other time according to clinical indications. Thirty two spiral CT examinations were performed 0–60 (mean 29 months) after procedure. Investigations were carried out with a table feed of 3 mm/s, images reconstructed at 2 mm separations, length of the bolus ranged 6–114 ml, collimation 3–5 mm. Omnipaque 300 was injected in 2–3 ml/kg at a rate 2 ml/s. 3D reformations and maximum intensity projections were obtained. The CTA visualised stent position, migration, patency, surrounding tissues and complications. Early complications documented on CTA: deformation of the proximal edge of stent implanted for aortic recoarctation – 1 pt, dissection of abdominal aorta below renal arteries – 1 pt. Late complications: aneurysm at the level of stent – 1 pt, thrombosis of the stent – 1 pt, neointimal hyperplasia – 5 pts. CTA is very useful in follow-up of children after stent placement in aorta. It is safe, easy and can be performed several times. Our experience shows that CTA can replace follow-up angiography in selected patients after stent implantation to aorta.

P1282

Usefulness of two computer programs for analysing myocardial SPECT in children

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Purpose: To examine usefulness of two computer programs for myocardial SPECT: QGS (Quantitative Gated SPECT) and p-FAST (Perfusion and Functional Analysis for Myocardial Gated SPECT) in pediatric patients. Subjects and Methods: Myocardial SPECT was analyzed by QGS in 33 patients (6 months to 20 years-old) and by p-FAST in 40 patients (1 to 15 years-old). All patients had myocardial heart diseases or symptoms associated with ischemic heart disease. Thirty minutes after injection of 99mTc sestrobamin, ECG-gated SPECT was acquired by TOSHIBA SPECT machine. LVEDV, LVESV, and LVEF were calculated by QGS or p-FAST. These parameters were compared with those by M-mode echocardiography (Echo) or biplane ventriculography (LVG). In addition, wall motion, %thickening, and myocardial perfusion in each segment of LV were quantitively evaluated by p-FAST. Results: LVEDV by QGS or p-FAST showed the data approximately 25% smaller than that by LVG, but had excellent correlation to that by LVG (QGS vs LVG: r=0.95, p-FAST vs LVG: r=0.94). LVEF also showed excellent correlation to that by LVG (QGS vs LVG r=0.89, p-FAST vs LVG r=0.84). Cardiac function analyzed by these programs was more accurate than that by Echo. p-FAST but not QGS, could distinguish pseudo-dyskinesis from true-dyskinesis by simultaneous estimation of wall motion, %thickening, and myocardial perfusion at the same segment. Conclusion: Evaluation of cardiac function by QGS or p-FAST has high reproducibility and objectivity, and therefore is useful to calculate left ventricular volume and EF in children with various heart diseases. Moreover, p-FAST is useful for simultaneous estimation of wall motion, %thickening and myocardial perfusion in each segment of LV. However underestimation of volume must be kept in mind.

P1283

How useful is MRI in patients with pulmonary atresia and multifocal blood supply.

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Introduction: Surgical treatment of patients with pulmonary atresia, central or peripheral pulmonary stenoses requires accurate definition of the pulmonary vascular tree. Methods and patients: Magnetic resonance images of 124 patients (10 PA with IVS, 69 PA with VSD, 13 PA with single ventricle, 30 PA with tricuspid atresia, 9 aplasia of one pulmonary artery, 13 central pulmonary stenosis with subtotal stenosis of one pulmonary artery branch) were compared with echocardiographic and angiocardiographic findings. Multifocal blood supply existed in 33 patients. 72 patients had undergone palliative surgery with placement of 75 systemic to pulmonary shunts. Cardiac ECG-gated MRI studies were performed using a multislice spinecho technique and at least one "bright blood imaging technique". A 3D-gradientecho sequence with 1 mm slice thickness was applied in 47 patients. Contrast enhanced MR studies were performed in 9 patients. Results: The morphology and size of the pulmonary arteries could be accurately assessed in all patients. MRI discovered unknown hypoplastic pulmonary arteries in 20 patients, what was confirmed either by pulmonary vein wedge angiography or surgery. In contrast to angiography MRI was able to demonstrate left or right pulmocary artery in 22 patients with severe central stenoses of the pulmonary artery of native origin (3) or caused by palliative shunts (16), dislocation of a banding (3). Better definition of pulmonary artery blood supply by aortopulmonary collaterals was achieved in 12 patients. However, instead of the capabilities of high resolution MR-angiography and contrast enhanced MR-angiography the description of the communications and evaluation of the number of segmental pulmonary arteries connected to collaterals was unsuccessfull in patients with multifocal blood supply. 73 palliative shunts were visualized and could be evaluated for patency in 54 patients, as unpatent in 14 and occluded in 5 patients. Conclusion: MRI is an excellent noninvasive technique for diagnosing pulmonary atresia, particularly for follow-up studies. However, further developments of pre- and post-processing modalities is required for complete noninvasive studies, particularly in patients with multifocal blood supply.

P1284

A case report: asymptomatic huge intracardiac hydatid cyst

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The human hydatid disease caused by the Echinococcus granulosus strain is endemic in Turkey. Although it has been found in almost every organ, the most frequent locations are the liver (60–70 % of cases) and the lungs (20–30 %). Cardiac hydatid cyst is unusual and its incidence is approximately 0.5–2 % of cases. 6 year-old girl patient hospitalized at the Department of Pediatric Surgery for vomiting and abdominal pain. Her blood pressure was 115/75 mm Hg, pulse rate was 110 beat/min and her heart sounds were normal. There was hepatosplenomegaly at the abdominal examination. Multiple intrahepatic cysts were suggested by abdominal ultrasonography. A chest x-ray film showed slightly increased cardiothoracic index and electrocardiogram

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

was normal. The patient was examined with two dimensional echocardiography with the following diagnosis and revealed a intracardiac cystic mass which 40x45 mm insize. It was originating from the posterior wall of the left ventricle. Color Doppler showed 10–20 mitral regurgitation. The finding was confirmed by computed tomography and nuclear magnetic resonance imaging. Result of indirect hemagglutination test for Echinococcus was negative. The patient was diagnosed with hydatid cyst, received albendazole and open brain surgery was planned. Only 10% of patients with cardiac hydatid cysts have clinical manifestations. Sign and symptoms are extremely variable. Therefore a routine follow up with echocardiography of patients with hepatic or pulmonary hydatid disease will help early detection of cardiac involvement.

P1285

Assessment of myocardial contraction abnormality in the systemic right ventricle in patients after atrial switch procedure for complete transposition of great arteries (d-TGA) by Tissue Doppler Echocardiography (TDE).

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Introduction. The purpose of this study was to use the novel TDE to evaluate systolic and diastolic myocardial motions in the systemic right ventricle (RV) in patients with d-TGA after atrial switch operations. Methods. 20 consecutive patients with a median age of 19.5 (11.2–32) year and a median postoperative interval of 16.3 (13–26.5) year following atrial switch operations were studied by TDE and compared to age-matched 15 controls. Systolic and diastolic tricuspid valve motions were measured in the basal segments of the right ventricle using a novel software for tissue Doppler analysis (Echo-pack 0.2 GE Vingmed). In apical view systolic (SyST), early diastolic (ET) and atrial contraction (AT) myocardial tissue motion and the acceleration time of these waves were analysed in standardised position from 3 heart cycles in each patient. Results In comparison to normal subjects the SyST in systemic RV were significantly decreased (p<0.001). The ET and AT in the RV lateral wall were lower in the atrial switch group (p<0.01). The acceleration time of the systolic and diastolic wall motions were significantly higher in the RV in patients with atrial switch. Conclusion. The significantly diminished systolic and diastolic myocardial excursion in association with accelerated myocardial tissue velocities in patients with d-TGA after atrial switch procedure may indicate altered myocardial function. TDE may provide a novel reliable method for quantitative assessment of systolic and diastolic RV dysfunction in patients with d-TGA after atrial switch procedures.

P1286

Assessment of myocardial perfusion in patients with congenital heart disease by contrast echocardiography with microbubbles infusion

Gomes, L.F.G., Silva, C.M.C., Arruda, A. L., Mathias, W., Bela, P., Matos, R.P., Moises, V.A., Carvalho, A. C.C., Paola, A.A.V.
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Assessment of myocardial perfusion in patients with congenital heart disease by contrast echocardiography with microbubbles infusion. Gomes, L.F.G., Silva, C.M.C., Arruda, A. L., Mathias, W., Bela, P., Matos, R.P., Moises, V.A., Carvalho, A. C.C., Paola, A.A.V. UNIVERSIDADE FEDERAL DE SÃO PAULO/EPM– São Paulo Brazil. The aim of this study was to evaluated the use of contrast echocardiography with microbubbles for assessment of myocardial perfusion, in children neonates and adults patients with congenital heart disease (CHD). The technique used was as previous described, and the contrast agent used was PESDA (perfluorocarbon – exposed sonicated dextrose albumin). The images were recorded in videotape and revised for 2 experients reviewers. The study included 30 patients. Their age ranged from 8 days to 34 years (median age = 30 months) and their weight ranged from 3 to 57 kg (median 16,5kg). 14 were male and 16 female. Tetralogy of Fallot (12 pts) and transposition of great artery (5pts) were the most often. During. Adequate mixture of the microbubbles with blood provided excellent images and opacification of the left and right ventricles. Based in observational perfusion myocardial score 19 pts showed global hypoperfusion, and only 14 had some degree of ventricular dysfunction. These evidences can provide important knowledge about the existence of myocardial ischemia in this group of patients studied and possibly to predict which ones will develop ventricular dysfunction in the future.

P1287

Contrast echocardiography with microbubbles infusion- the use of this technique in neonate and children

Gomes, L.F.G., Silva, C.M.C., Arruda, A. L., Mathias, W., Bela, P., Matos, R.P., Moises, V.A., Carvalho, A. C.C., Paola, A.A.V.
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Contrast echocardiography with microbubbles infusion- the use of this technique in neonate and children. Gomes, L.F.G., Silva, C.M.C., Arruda, A. L., Mathias, W., Bela, P., Matos, R.P., Moises, V.A., Carvalho, A. C.C., Paola, A.A.V. Universidade Federal de São Paulo / EPM – São Paulo – Brazil. The use of contrast echocardiography with microbubbles infusion is a well established technique for adults and has not been used in children, neonates and adults patients with congenital heart disease (CHD). The aim of this study was the application of this technique in this group of patients. Methods – Preparation of PESDA (perfluorocarbon – exposed sonicated dextrose albumin) was similar as previously described briefly, 8 ml of decafluorobutano was hand agitated with a 3:1 mixture of 5% dextrose and 5% human albumin and then underwent electromechanical sonication for 80 seconds. The microbubbles was administered IV as a continuous infusion at rates of 30, 60, 90, 120 ml/h during 30 to 60 second or as bolus of 0,4 to 0,8ml/kg. Clinical assessment, peripheric pulse oximetry, arterial blood pressure, were monitorized in all cases. Images were taken at basal time, with contrast with and without using ultrasound for 3 minutes each time at all rate infusion. The best dose was considered when the cavity was completly full and visually we can see the myocardial wall perfusion. This study included 30 patients, with mean age of 30 months, ranging from 8 days to 34 years. The dose of contrast agent used ranged from 0.4 to 0.6 ml/Kg. The mean time of exam duration was 28 minutes. The bolus of <0.5ml/kg and a infusion rate yyyy 90 ml/h provided better and more consistent myocardial opacification intensity. There was not adverse effect because of the use of PESDA in the population. The technique showed to be safety in all ages with and has applications in a wide variety of clinical setting. It is an increasing valuable tool with high degree of specifity and is non-invasive.

P1288

Three-Dimensional imaging of atrial septal defects: comparison of Magnetic Resonance Imaging, Intra Cardiac Echocardiography and Trans Esophageal Echocardiography in assessment for transcatheter closure

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Background. The aim of this study was to compare Three-Dimensional anatomical reconstruction of atrial septal defects (ASDs) by Trans Esophageal Echocardiography TEE, Intra Cardiac Echocardiography (ICE) and Magnetic Resonance Imaging (MRI). Methods. Six patients aged 6 to 25 years who were undergoing transcatheter closure of ASDs were studied. With an omniplane TEE probe and ECG and respiratory gating a series of cine loops were acquired for each 5° angle between 0° and 180° in 3 minutes. Prior to ASD closure a 9F, 9MHz mechanically rotating, 360° orthogonally imaging ICE catheter was pulled back 10 cm in 0.5cm steps in the right atrium. Twenty respiratory and cardiac gated images were acquired per cardiac cycle at each pullback position in 5 minutes. Both TEE and ICE image acquisition was repeated following device implantation. Image processing and reconstruction (TomTec, Germany) took 5 minutes each for both TOE and ICE. Previously the patients underwent cardiac MRI scans on a 1 Tesla magnet (Siemens Impact Expert). A series of four-chamber view breath-hold spin echo images or Cine gradient echo images were acquired. The images were segmented in a semi-automated manner. From the segmented data set, a three-dimensional outer surface was extracted, rendered and visualised. Results. With all three imaging techniques, we could identify the ASDs and all related anatomical structures. There was considerable change in defect size during the cardiac cycle (14%–88%). There was close correlation of the area of the defects and rim to surrounding structures, although in 2 patients TEE underestimated the defect size compared with the other two methods. Conclusion. ICE and MRI measurements correlating most closely with 3-D, underestimating the size of the ASD. MRI was best at visualising the defects in relation to surrounding structures.

P1289

3D MRI of congenital cardiac lesions

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https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

The use of cardiac specimens for teaching congenital heart anatomy necessitates expert dissection with inevitable compromise of the specimen. Advances in MR technology allow imaging with high spatial resolution, with 3D reconstruction. 7 fixed hearts with differing lesions were imaged underwater. Imaging was performed in contiguous 25mm blocks with a CISS sequence of approx. 0.25mm. An SGI Infinite-reality Onyx was used for post-processing. The image grey-scale was inverted, and reduced to a range of 256, centred on cardiac tissue. All extracardiac pixels were assigned to 0, using software and hand editing where necessary. Volume-rendering was performed in real time using Volren software, with multiple arbitrary cut-planes. A variety of complex lesions were successfully identified using this 'virtual dissection', and examples will be shown. The lack of specimen damage, preservation of anatomical relationships, permanency of the images recorded, user-interactivity, plus the ability to electronically transmit the specimen to remote locations all make this a technique with considerable promise for future educational purposes.

P1290

Noninvasive assessment of defect size and topography in 50 children with a secundum atrial septal defect by magnetic resonance imaging: a comparison with transesophageal echocardiography (TEE) and measurements during surgery

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Background: In children with a secundum atrial septal defect (ASD II), noninvasive pre-interventional diagnosis of defect size and topography may be of value to avoid unnecessary procedures. We hypothesized that magnetic resonance imaging (MRI) could add relevant information for therapeutical management decisions. Methods and results: 50 children (age 1.8–13.4 years, mean 5.8) with a significant atrial level shunt underwent MRI examination to delineate defect topographical anatomy by multislice/multiphase phase-contrast MRI (PC-MRI) in 4 different imaging planes. MR defect measurements were compared to intraoperative measurements in those patients with a clear contraindication to intervention. The remainders were transferred to the cath-lab for transcatheter defect closure and MRI results compared with biplane transesophageal echocardiography (TEE). TEE was superior to MRI in assessing additional defects in the submillimeter range that were all considered irrelevant to the therapeutical management decision. PC-MRI clearly identified all 3 children with multiple fenestrated ASD, one child with a atrial septal aneurysm and all 5 children with a sinus-venosus defect. Secundum atrial septal defect size measurement and assessment of distances to adjacent structures agreed well with TEE and fairly well with surgery. Moreover, inflow MR angiography correctly identified all pulmonary and systemic venous anomalies. The ratio of PC-MRI derived through-plane flow data from the pulmonary artery (=Qp) and the ascending aorta (=Qs) agreed well with the pulmonary to systemic flow ratio (Qp/Qs) as obtained by invasive oximetry. Conclusions: MRI is safe and accurate in delineating ASD size and topography, assessing venous anomalies and quantifying left-to-right shunt and thus allows for selection of suited candidates for transcatheter defect closure. The method may be considered as a first-line.

P1291

Evaluation of airway compression by cardiovascular anomalies in infants and children: magnetic resonance imaging compared with surgical anatomy and cine-cardioangiography/bronchoscopy

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Background: Airway compression can be caused by a variety of congenital cardiovascular malformations. MRI may be of value but has not been evaluated prospectively in infants and children. Methods and results: 22 children (0.3–12.4 y, mean 3.3 y) with suspected airway compression by cardiovascular anomalies (double aorta, arch n=6, absent pulmonary valve n=5, aberrant truncus brachiocephalicus n=3, a. lusoria n=1, pulmonary sling n=1, right aortic arch/left ligament n=2, no cardiovascular cause n=4) underwent MRI examination. We used diastolic-triggered, respiratory-gated T1w-TSE and MPRA (3D-GE, multiplanar TOF) and performed 3D reconstructions using surface rendering and segmentation algorithms (Philips, EasyVision R, rel. 4). MRI was followed by cardiac catheterization/bronchoscopy and MR staff was blinded to the results. MRI was perfectly consistent with cine-cardiobronchography/bronchoscopy and predicted surgical strategy in all cases. A cardiovascular cause was safely excluded in the four children without. However, a left ligament was rather suspected than visualized (2 patients), but subsequently confirmed during surgery. Furthermore, respiratory-gated TSE overestimated severity of airway stenosis in most children as compared with bronchoscopy under spontaneous breathing conditions. Conclusions: MRI was found to be safe and highly accurate in a variety of cardiovascular malformations leading to airway compression in infants and children. However, the surgeon should be alerted to the possibility of an additional left-ligamentous sling.

P1292

Estimation of right ventricular ejection fraction in patients with chronic right ventricular pressure overload using myocardial performance index

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Objective: We examined the value of myocardial performance index (MPI) by means of echocardiography in asymptomatic patients with chronic right ventricular (RV) pressure overload. The aim of this study was to obtain from this index a measurement of RV ejection fraction (EF), which is a well-known clinical parameter, but difficult to obtain in patients with RV dysfunction. Methods: RV MPI by the classical Doppler echocardiography and RV ejection fraction by means of magnetic resonance imaging were measured in 10 consecutive (7 male and 3 female) asymptomatic or minimally symptomatic (NYHA I or II) patients (age 28.7 ± 11.6 years) with chronic RV pressure overload. The MPI was obtained through the use of formula (a-b)/b where a is the interval between the cessation and onset of the tricuspid inflow, or the duration of tricuspid regurgitation (if present), and b is the ejection time. Patients with left ventricular dysfunction were excluded from the analysis. Results: The correlation between RVEF and MPI was r = -0.86, p=0.001. For practical reasons the MPI was converted to RVEF-index. The EF index was calculated by the formula 92-62 x MPI acquired from linear regression analysis. Mean RVEF in the patient group determined by MRI was 63.5 ± 12.5 % and mean RVEF-index was 64.0 ± 14.3 (P=NS). Conclusions: Our study shows a significant inverse correlation between RV MPI and RV EF determined by the means of MR imaging. These data suggest that MPI converted to EF-index can be used in clinical practice as determinant for RV EF in patients with RV chronic pressure overload.

P1293

Evolution of densitometric three-dimensional heart reconstruction from rotational angiography

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Objectives: Superimpositions and invalid volume measurements due to incorrect geometric assumptions for differently shaped heart defects are disadvantages of angiocardiography. The aim of our study was to provide three-dimensional (3D) imaging and more valid quantitative data from angiography. Methods: Digital images from biplane, orthogonal rotational angiograms with a monitored injection in a speed of 2*85°/second and a rate of 2*25 frames/second were used for 3D reconstruction. 3D reconstruction technique based on backprojection of a cone beam perspective based on Feldkamp's algorithms expanded for simultaneous rotations and angulations of the gantry. The value of each 3D part (voxel) were computed densitometrically by summation of values of virtual rays from each pixel back to its source penetrating the distinct voxel. To fulfill practical time requirements a special paralleled software was developed. 3D volume measurements were computed by voxel flood fill algorithms and by triangulating of heart surfaces with tetrahedrons. The system was validated with phantoms and right and left ventricular casts. Results: The densitometric 3D method is feasible to reconstruct concave surfaces (Fig 1). A resolution of at least 255 voxel (ullis) in clinical sequences. The system discriminates structures as low as 2 mm (Fig 2) and needs a computing time of 138 seconds (256 voxel, 14 frames, 6 nodes). Phantoms and casts from heart specimens showing reliable and valid volume measurements (Vm) compared to true volume (Vt) ranging from 14 to 250 ml (Vt= 1.0445* Vm+3.20 ml, r=0.996, p<0.0001). Conclusions: Densitometric 3D reconstruction of heart chambers from rotational angiography is feasible and improves qualitative analysis. 3D angiography seems to be a promising new tool for cardiac imaging.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P1294

Three-dimensional reconstruction improves angiographic volume and cardiac output measurements

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Objectives: Accuracy and precision of angiographic volumetric methods for left and right ventricles have been shown to be poor under clinical conditions due to geometric assumptions of ventricular shape. The aim of our study was to compare three-dimensional (3D) volume measurements with conventional angiographic methods. Methods: Casts of 21 left ventricles (LV) and 22 right ventricles (RV) were prepared from native post-mortem human, bovine, and porcine hearts by simultaneous filling with methylmethacrylat and a hydrostatic pressure of 10cm H2O. True ventricular volume was determined with water displacement by the Archimedes principle. Dynamic measurements were performed with the Medos(r) artificial heart. Conventional volume measurements consisted of mono- and biplane area-length method (2D-AL) and Simpson's method from: 30°RAO, 0°RAO, 60°LAO, and 90°LAO. 3D volume measurements were computed by voxel flood fill method (3D-FF) and by extrapolating of heart surfaces with tetrahedrons after densitometric 3D ventricle reconstruction from biplane, orthogonal rotational angiograms by backprojection algorithms. Results: 3D reconstruction of left and right ventricular casts are in excellent conformity with the original casts. LV true volume(Vt) is best characterized with volume measurements(Vm) by 3D-FF (Vt=1.008*Vm-1.5 ml, r=0.994) versus best conventional biplane 2D-AL from 30°RAO/60°LAO (Vt=0.916*Vm+3.1 ml, r=0.954)(Fig1). RV ejection measured by 3D-FF (Vt=1.049*Vm-0.7 ml, r=0.995) versus best conventional biplane 2D-AL from 30°RAO/60°LAO (Vt=0.909*Vm+2.7 ml, r=0.912)(Fig 2). Mean values, standard deviations and significant differences to true volumes are given in Tab1. Angiographic measurement of cardiac output(Qcm), computed by the product of SV stroke volume with heart rate, differs not significantly to the output pumped(Qt) by the artificial heart (Qt=1.059*Qcm-82.8 ml/min, r=0.9263)(Fig3). Conclusions: Angiographic volume measurements with the three dimensional methods is superior to conventional methods. Static and dynamic 3D flood fill volume measurements showed reliable and valid results.

P1295

Radiation dose reduction strategy during transcatheter closure of the persistent arterial duct

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Background: X ray guided interventional procedures can be associated with relatively high radiation doses. A critical appraisal of catheter laboratory technique is appropriate for these procedures. In a recent audit, we have established that all information on duct size, morphology and device implantation strategy is provided by aortography in the lateral projection for 20/20 (100%) cases of percutaneous closure of the persistent arterial duct (PCPAD). Using a biplane angiography system and adopting this strategy of combining fluoroscopy available in either plane, with lateral projection fluorography only, we have estimated the potential reduction in effective dose (ED) and associated risk of radiation induced detriment. Methods: Using transmission ionisation chambers attached to each X-ray tube, detailed dosimetry has been made for each stage of each procedure for 50 children aged 4.0+/-2.4 years (mean+/-SD) undergoing PCPAD. Using recently published conversion factors, the total ED was estimated together with age dependant associated risk. The relative contribution for all fluorography other than the lateral projection, was subtracted from the total ED. Results: The relative contributions to ED for fluoroscopy and fluorography were 58% and 42% respectively. The total ED was 3.6+/-2.7 mSv (median+/-SD) with a median associated risk of 0.068%. After removing the contribution to ED for all non lateral projection fluorography, the relative contributions to ED for fluoroscopy and fluorography were 65% and 35% respectively. The ED and risk were reduced to 2.1 +/-2.7 mSv and 0.05% respectively. Conclusion: For PCPAD procedures, fluoroscopy available in two planes, combined with single plane lateral projection aortography results in a reduction of ED and associated risk of 42.9% and 43.5% respectively without compromising procedure outcome.

P1296

Definition of pulmonary circulation in tof with pulmonary atresia.

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Currently, TOF with pulmonary atresia and major aorto- pulmonary collaterals (MAPCA) remains one of the few indications for diagnostic angiography. This study attempts to define standard views to completely delineate MAPCAs with minimal radiation and contrast load, using 2D echo (2DE) to define intracardiac anatomy. We analyzed angiograms of 113 patients, 30 early (prior to 1997) pts had 'complete' studies in > 1 session and 83(after 1997) had limited angiograms. Angiograms of diagnostic value were: Arch and descending aorta in PA view, both Subclavian at origin and MAPCA inferiors. Ventriculograms, angled and biplane views did not add information to the 2DE. Lateral views were done in 24 patients to define dual supply to a lung zone when seeing MAPCAs. A elongation and an airway view fluoroscopy were also recorded. Results: Total contrast used was 4cc/Kg(Vs) 7.2 cc in the earlier group (P<0.04). Re arch was seen in 18.2%, abnormal branching of arch vessels in 16.3%, higher with Re arch 44.4% (p<0.01), renal anomalies 13%, airway anomalies 9.2%. Confluent PAs were seen in 42.8%, MAPCAs alone in the rest. MAPCAs arose from desc aorta (100%), RT Subclavian arteries 47.7%, Carotids 21.4%. PDA was seen in 34 pts. A median of 6 (2-9) (out of 10) lung zones were supplied by MAPCAs or PAs in these patients. Conclusion: Specific limited angiographic views to completely delineate MAPCAs are described in this study. Arch anomalies, renal abnormalities and airway abnormalities are commonly seen.

P1297

Reduced incidence of ventricular ectopy during right ventriculography with a 4F Halo catheter during a pediatric cardiac catheterization

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We sought to evaluate the influence of the helical-tip Halo catheter on ventricular ectopy during right ventriculography (RVG) in a pediatric cardiac catheterization. Few studies have focused on effects on RVG, especially in a pediatric population despite its potential advantages for reducing the incidence of ventricular ectopy. We compared the incidence of ventricular ectopy in infants and children undergoing RVG using a 4F Halo catheter (22 subjects) or a standard 5F or 6F balloon angiographic catheter (147 subjects). Ventricular ectopy was captured by a simultaneously recorded electrocardiogram during contrast injection. There was no statistical difference between the Halo and control groups (mean ± SD) for age (32.6 ± 23.4 vs 30.1 ± 29.0 mo), gender, weight (12.0 ± 5.0 vs 10.9 ± 5.3 kg), volume of contrast (1.36 ± 0.37 vs 1.40 ± 0.33 ml/kg), or injection rate (0.80 ± 0.26 vs 0.74 ± 0.26 ml/kg/sec). The overall incidence of ventricular ectopy in the Halo group was significantly less than in the control group (9 / 22 vs 76 / 147, p<0.05). The incidence of ventricular couplets or tachycardia in the Halo group was also less than in the control group (4 / 22 vs 19 / 147, p<0.05). The angiographic quality, scored with grading system being unsatisfactory or satisfactory, showed no evidence of unsatisfactory imaging for the right ventricle as well as the pulmonary arteries in the Halo group. We conclude that the 4F Halo catheter produces high-quality right ventriculographic images with significantly less ventricular ectopy compared with standard balloon angiographic catheters. The use of this catheter may bring us significant benefits during RVG in pediatric cardiac catheterization.

P1298

Reevaluation of the effects on ventricular ectopy during left ventriculography with a 4F Halo catheter during pediatric cardiac catheterization

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We sought to prove the efficacy of reducing the incidence of ventricular ectopy by the 4F helical-tip Halo catheter during left ventriculography (LVG) in a pediatric population. Only a few studies have performed in children despite its potential advantages. Since March 1999 to October 2000, the patients less than 5 years of age planned to perform retrograde LVG was randomized into two groups by utilizing a 4F Halo catheter or a 4F pigtail catheter. The study group consisted of 29 patients in Halo and 24 in pigtail, aged from 5.6 to 56.3 mo (median, 18.5 mo) and 3.9 to 53.8 mo (median, 16.8 mo), respectively. Simultaneously recorded electrocardiogram evaluated ventricular ectopy during contrast injection. There was no statistical differ

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

rence between the Halo and pigtail groups (mean ± SD) for age (23.2 ± 35.5 vs. 19.5 ± 14.6 mo), gender, weight (10.5 ± 2.2 vs. 9.5 ± 2.6 kg), volume of contrast (1.45 ± 0.34 vs. 1.52 ± 0.51 ml/kg), or injection rate (0.88 ± 0.22 vs. 0.85 ± 0.23 ml/kg/sec). The overall incidence of ventricular ectopy in the Halo group was significantly less than in the control group (0/29 vs. 13/24, p < 0.0001). The incidence of ventricular couplets or tachycardia in the Halo group was also less than in the control group (0/29 vs. 6/24, p < 0.006). There was no significant difference in diagnostic quality for the anatomy or cardiac function of LVG between the two groups by grading system being unsatisfactory or satisfactory. Our results are consistent with the previous reports, suggesting that the Halo catheter is potentially safe and useful for LVG during pediatric cardiac catheterization. The 4F Halo catheter should be considered for use when it indicated.

P1299

Usefulness of counter-current aortography in assesment of the pulmonary branches diameter in patients with HLHS syndrom after 1 stage of Norwood procedure

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The aim of study was the assesing of usefulness of counter-current aortography in determing precise morfometry of pulmonary artery in children with HLHS after 1st step of Norwood procedure before the next step of cardiac surgery treatment. The echocardiographic examination of the pulmonary vessels with coexistance stenosis or hypoplasia of left pulmonary artery enables to establish precise examination of vessels. In the past three years 12 patients with HLHS syndrom after 1 stage of Norwood underwent angiography (aged 5–8 month). The aortography with retrograde injection of contrast medium via axillary artery (DSA 1) was performed in 10 cases. The cardiac catheterisation was done in 4 pts (RESULTS: The precise visual assesment of pulmonary branches using DSA 1 were achieved in 8 pts: diameter of RPA from 5,4 to 10mm (mean-7,4mm), diameter of LPA from 3,4 to 6,2 mm (mean 4,4), LPA/RPA ratio from 0,35 to 0,9 (mean 0,6). The 5 pts had a significantly hypoplasia left pulmonary branch (LPA/RPA <0,5). The catheterisation had been performed in 4 children, because of poor image of LPA (in 2 pts with central aorto-pulmocary shunt) and cannulation problems (2 pts.) CONCLUSIONS: 1. Retrograde angiography via axillary artery is an effective method which provides the essential information of pulmonary branches in patients after 1st. stage of Norwood procedure and Blalock-Taussig shunt. 2. In our own material left pulmonary hypoplasia was observed in 92 % of pts. 3. In cases of central aorto-pulmonary shunt conventional aortography via catheterisation is indicated.

P1300

Low profile large bore hydrophilic catheters for left heart angiography in infants and toddlers

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Cardiac catheterizations in infants and toddlers risk damage to the femoral vessels. In some neonates, this may be overcome by using the umbilical vessels. However, this approach is not available beyond the first weeks of life. The purpose of this study is to examine the efficacy of a 4 Fr hydrophilic braided catheter with a 0.038 lumen. Since 1998, three designs have been evaluated. The catheter tip on type I and II is angled at 60 degrees to the shaft at 3.5 and 1.7 cm from the tip respectively. The former has six side holes and the latter five. The type III has secondary curve more proximally, and two side holes near the tip. All catheter types have an end hole. Type I and II are designed for general purpose use, and type III for cannulating aorto-pulmonary shunts such as BT shunts after stage 1 Norwood procedure. The catheters permitted angiographic contrast (10.6 cp viscosity) injection at 13 ml/sec flow rate, at 750 psi maximal pressure. All procedures were performed under general anesthesia via percutaneous puncture of the femoral vessels for ventriculography, aortogram and pulmonary angiogram. Omnipaque 300 was used in all patients. There was no vascular or myocardial damage. Angiographic visualization was excellent. Pedal pulses were present after catheter removal. In conclusion, the low profile hydrophilic catheters allowed angiography in infants and toddlers and lessened the risk of peripheral vascular damage.

Cardiac Imaging: CT, PET, MRI-MRA

P1301

An assessment of the pulmonary to systemic blood flow ratio (qp/qs) in patients with intracardiac left-to-right shunt by velocity-encoded, phase-contrast mri – a comparison with fick principle

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[Purpose] To evaluate the ability of velocity-encoded, phase-contrast magnetic resonance imaging (MRI) to quantify the pulmonary to systemic blood flow ratio (Qp/Qs) in patients with intracardiac left-to-right shunt. [Patients and Methods] We performed velocity-encoded, phase-contrast MRI measurements of flow at the proximal aorta and the main pulmonary artery to estimate the Qp/Qs ratio in 38 patients with VSD and in 12 with ASD. The age ranged from 1 month to 13 years. These values were compared with measurements of the Qp/Qs ratio obtained from oximetric data derived from cardiac catheterization. As a control study, 2 patients without left-to-right shunt underwent MRI measurements. [Results] The Qp/Qs ratio ranged from 1.2 to 4.9 on oximetry. There was a good correlation (r=0.94, p<0.01) between the oximetry and MRI assessments of shunt magnitude. The control data of MRI in 2 patients without shunt showed Qp/Qs were 1.08 and 1.01 respectively. [Conclusion] The magnitude of intracardiac left-to-right shunt measured with velocity-encoded, phase-contrast MRI of flow in the proximal great vessels can be significantly correlated with data obtained from cardiac catheterization, thereby this non-invasive method is useful for determining surgical indication for correction of intracardiac left-to-right shunt.

P1302

Flow dynamics in right aortic arches of children

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In normal adults, it has been demonstrated that flow is asymmetric in left aortic arches. It is unclear that the same fluid mechanics is present in children with right aortic arches. We studied 11 children with functional right aortic arches with through-plane phase-encoded magnetic resonance velocity mapping in the ascending and descending aorta. The aortic cross-section was divided into 4 quadrants aligned along the long axis of the aorta. In the ascending aorta, although there was no significant difference in the total flow across the entire cardiac cycle between quadrants, this was not the case at the time of maximum flow in the entire aorta, the posterior and rightward quadrant carried significantly more flow than the other 3 quadrants (29 + 3% vs. 23–25 + 2–4%, P < 0.001). In 9 / 11 patients, maximum velocity occurred in the right half of the ascending aorta. Similar to the ascending aorta, in the descending aorta, there was no significant difference in the total flow across the entire cardiac cycle between quadrants. At the time of maximum flow in the entire aorta, however, the posterior half of the descending aorta carried significantly more flow than the anterior half (50% + 11 vs. 44 + 11%, P = 0.04). No; surprisingly, in 8 / 11 patients, maximum velocity occurred in the posterior half of the descending aorta. In both ascending and descending aorta, maximum flow in each quadrant and time to maximum flow in each quadrant did not significantly differ between quadrants. We conclude that although asymmetric in some respects, flow in right aortic arches demonstrate heterogeneity at different times in the cardiac cycle. This information may lead to improved aortic reconstructions.

P1303

Blood flow patterns in the aortic arch and descending aorta visualized using magnetic resonance imaging bolus tagging

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Objective: Repair of many forms of Congenital Heart Disease (CHD) include reconstruction of the aorta. Characterization of flow in the aorta is paramount to understanding the physiology pre and post repair. We present our results using Magnetic Resonance Imaging Bolus Tagging (MRI-BT) to elucidate complex flow patterns in the ascending aorta, aortic arch, and descending aorta in healthy adults. Methods: Four healthy adult volunteers were studied using a 1.5 Tesla MRI Scanner (Siemens Sonata). We used a cardiac gated, multiple phase, single slice, 2D fast gradient echo sequence, preceded by a perpendicular saturation pulse. The following parameters were

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

used: echo time=4msec, repetition time=RR interval, flip angle=20 degrees, field of view=350mm, slice thickness=8mm, resolution=256X256, saturation thickness=5mm. The first image set collected was a ramdy cine view of the aorta with a transverse saturation pulse. Next, three images sets were collected with a sagittal saturation pulse and cross sectional imaging planes through the ascending aorta (approximately 2cm above the LV outflow tract), aortic arch, and descending aorta. Results: We found helical flow established in the ascending aorta and continuing through the descending aorta. Second, we found a differential velocity profile in the ascending aorta with blood flowing fastest on the underside of the aortic arch. Finally, the flow profile across the cross sectional area of the aorta was flat or plug-like. Conclusions: Using MRI-BT we are able to characterize flow patterns in the aorta. Helical, plug-like flow with a velocity differential was consistently seen in all volunteers. This pattern of flow may confer conservation of energy. This technique can be used in children with CHD. Further characterization of flow patterns in the aorta may be useful for planning vascular reconstructive procedures.

P1304

The role of magnetic resonance imaging in the long-term follow-up of the adult with fontan circulation.

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Introduction: Imaging the heart after the Fontan operation is a vital part of long-term follow up. There is now a large population of older patients for whom echocardiographic imaging may be sub-optimal and angiography is the traditional method of imaging the Fontan pathway. Methods: We reviewed 36 MRI scans on 26 patients (11 female) with atrio-pulmonary Fontans over a 7 year period. Median age at MRI was 25 years, median time since Fontan operation 12 years. We documented: 1. Information obtained from MRI 2. Whether this information was available from other non invasive studies 3. Important information missed by MRI. 4. Cost-effectiveness of MRI. Results: MRI showed right atrial dilatation in all cases. Fontan pathway obstruction was seen in 8. There was branch pulmonary artery stenosis in 6 and hypoplasia in 3. There was pulmonary vein compression in 12. Ventricular function was abnormal in 10, severely so in 2. Trans-thoracic echocardiography provided information on ventricular function and AV valve regurgitation in all, however, only 2 of the 29 other abnormalities were detected. Thirteen patients underwent cardiac catheterisation. This revealed new anatomical information in 3: a left to right atrial shunt in 1 and stenosis of the atrio-pulmonary anastomosis in 2 (one had MRI signal loss due to an ASD device). The cost of MRI at our institution is £575, trans-thoracic echocardiography £225 and cardiac catheterisation £566. Conclusions: MRI provides information on the Fontan circulation's size, ventricular function, pulmonary arterial and venous anatomy, much of which cannot be obtained with trans-thoracic echocardiography. It provides similar anatomical information to cardiac catheterisation, at the same cost, but is non-invasive and involves no radiation exposure. MRI should be considered as a routine non-invasive means of assessing the adult Fontan patient.

P1305

Ventricular dimensions, mass, and function late after Fontan like palliation – quantification by magnetic resonance imaging

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Background: Reliable data by noninvasive means regarding ventricular function, mass, and dimensions in patients with Fontan-like palliated single ventricles is not available. The aim of the study was to examine the feasibility of measuring these variables by magnetic resonance imaging (MRI) and to determine expected values in this particular patient group. Methods: 32 patients (age: 18.3±7.0 yrs) 9.4±3.9 years after Fontan like palliation and 10 healthy volunteers (age: 28.0±6.3 yrs) were examined by MRI. End-diastolic (EDV) and end-systolic (ESV) volumes, ejection fraction (EF), and ES mass (ESM) were determined by short axis cuts under breathhold using volumetric analysis software (Mass(r)). Ventricular volumes and mass were normalized for body surface area. All patients in the Fontan group had a morphologically left ventricle which was compared to the left ventricle of the volunteers. Patients eligible to the study underwent echocardiography excluding AV valve insufficiency. Results: Preliminary data of 18 patients are available. 14 patients were s/p Fontan palliation, five s/p TCPC. Volumes, EF, and mass did not differ significantly in these two patient groups. Five patients were s/p pulmonary artery banding (PAB). ESM was 95.2±47.6 g/m2 after PAB and 73.4±23.8 g/m2 without PAB (n=ns). In patients and volunteers mean ESM (78.9±31.84 g/m2 vs 80.6±18.0 g/m2) and mean EDV (73.3±36.5 ml/m2 vs 67.4±4.7 ml/m2) did not differ significantly. In three Fontan patients, however, ESM and EDV were markedly elevated. EDV correlated well with ESM in the Fontan group (r=0.78, p<0.01). Mean EF of the single ventricle (46.6±14.0 %) was significantly smaller than in normal left ventricles (62.7±8.2 %). Conclusion: Ventricular mass and EDV are not elevated in most patients late after Fontan like palliation with left ventricular morphology. EF of the single left ventricle is diminished as compared to normal left ventricles.

P1306

Cardiac magnetic resonance in genotype positive and phenotype-negative or -positive patients with hypertrophic cardiomyopathy.

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Purpose: Sudden death resulting from hypertrophic cardiomyopathy (HCM) has been reported to be directly related to ventricular thickening with particular reference to septal thickness. We compared cardiac-MR (CMR) and echocardiography in eleven patients (pts) with HCM (obstructive n=5, non-obstructive n=6, including one who was genotype-positive/phenotype-negative) regarding the extent and location of left ventricular (LV) hypertrophy as well as symptoms. Methods: All pts were imaged with dedicated 1.5T MR Scanners (SIEMENS Sonata, Germany) for quantification of global myocardial function as well as segmental wall thickness. Additionally two-to-three slices with a focal myocardial hypertrophy were selected for assessment of myocardial tissue tagging. MR tagging images were obtained using SPAMM (TR=9.7ms, TE=5ms, flip angle=10 degrees, FOV=260mm, pixel dimension=1.015mm). The data were analyzed for global and regional function measurements using MASS software (Version 4.0, Leiden/Netherlands). LV and RV end systolic and end diastolic mass, stroke volumes, and ejection fraction were determined. Centerline method was used to assess wall thickening. SPAMM-tagged images were evaluated using SPAMMVU-Software. Echo: Standard M-mode and cross-sectional views of the left ventricle were obtained. Results: In our group of HCM patients, septal hypertrophy (>12 mm) was detected in 4 pts by echo and in 7 by CMR. The pt who was described as phenotype negative by echo standards had a regional septal hypertrophy (15 mm) as detected by CMR. With respect to the detection of the thickest LV-wall section, there was a good agreement (<2 mm difference) in 6 pts and no concordance between CMR and echocardiography in 5 pts. All pts with abnormal LV-mass (mean 346±102.92g, range 100.7 to 450.9) had symptoms but septal wall thickness was not correlated to symptoms. Conclusions: Our findings suggest that CMR in selected patients can be superior to echocardiography in the assessment of regional wall thickness and LV-mass in HCM pts. In accordance with previous studies maximal regional wall thickness does not correlate with symptoms, but total LV-mass may be a potential prognostic parameter.

P1307

Methodology of multidimensional velocity vector mapping in systemic venous flow by phase contrast magnetic resonance angiography

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Hemodynamic efficiency of Fontan circulation is believed to be a major determinant of outcome. To assess in vivo flow dynamics in the systemic venous pathway, we performed two-dimensional phase contrast magnetic resonance angiography in a healthy volunteer. MR imaging data were acquired with Magnetom Vision 1.5 Tesla unit (Siemens). ECG-triggered phase-velocity images encoded for flow in the x-, y- and z-axis directions were then acquired in sagittal plane, and were intended to include the caval and atrial components. Phase images (repetition time=50 ms, echo time=8 ms, flip angle=11, slice thickness=10 mm, field of view=300x300 mm, velocity encoding=60 cm, image matrix size=256x256. 8 images of per cardiac cycle) obtained. To minimize motion artifacts respiratory compensation was used. Data of phase images by flow encode were used. Phase shift (PS: from -180 to 180) of flow encode were translated to pixel intensity (PI: from 0 to 4095), for example PS=-180 to PI=0, PS=0 to PI=2047 and PS=180 to PI=4095. The number of PI minus 2047 was determined as the final data (from -2047 to 2047). A single velocity vector was composed of the final scalar data of three directions. It is possible to translate pixel data and distribute multidimensional vector map by using the Application Visualization System/TM Medical ViewerTM (AVS-MGT) software

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

package. Flow image was distributed by subtraction of tissue image with magnitude image. Multidimensional velocity vector mapping demonstrated that flow pattern of superior caval vein show two peak and blood entering from superior caval veins contributes to the forward rotation of blood in the right atrium.

P1308
Cine MR imaging of right coronary cusp herniation
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The purpose of this study is to clarify the cine MRI findings of right coronary cusp herniation (RCCH). Four patients with suspected VSD type I were included in this study. Cine MRI (gradient-echo sequence) was performed to evaluate whether RCC herniate or not. The diagnosis was confirmed by cardiac catheterization and operation in all patients. At first, coronal scout image was obtained through the aortic root. Slice orientation was plane parallel to the ascending aorta through the aortic root. In all patients cine MRI revealed transformation of RCC and dislocation of RCC into RV outflow during systole. The jet was observed in the RV outflow during systole, which revealed direct shunt LV to RV. The jet was also depicted in LV outflow in three of 4 cases because of complicated aortic regurgitation. Cine MRI is useful in the diagnosis of RCCH.

P1309
Extracardiac vascular disease evaluation using Magnetic Resonance Imaging.
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Extracardiac vascular disease evaluation using Magnetic Resonance Imaging. Moreira, R., Gutierrez, A., Peirani, M., Cassina, D., Guerchicoff, M., Elies, F. Buenos Aires, Argentina. Purpose: to study the capability of Angiographic Magnetic Resonance (MRI) as a diagnostic tool in extracardiac congenital vascular pathology. Methods: between august 1999 and october 2000, 19 patients (pts) X: 2.4 years (3M-19Y) with extracardiac vascular anomalies were surveyed using a 1.5 T Siemens Vision System with body array coil. A) Aortic disease.5 . C) Pulmonary artery evaluation .6. Results: A) In 8 pts with aortic coarctation MR were done to evaluate collaterals, in 4 were acceptable and were operated on and were absent in 4 sent to balloon angioplasty. B) In 2 pts with total anomalous pulmonary veins connection (TPV) post-surgery with suspected residual pulmonary veins stenosis the hemodinamic study was not diagnostic but MR defined anatomy completely. 1 with mitral atresia was inoperable and the other with diffuse left vein hypoplasia died. In 2 pts neither the Echo2D nor the catheterism could confirm pulmonary veins connection. MR showed left innominate dexter in one and TPV in right atrium in the other. In one patient post-liver transplant MR confirmed stenosis of the inferior vena cava. C) MR was indicated to delineate the anatomy and size of pulmonary branch. 2pts with branch stenosis were sent to stent placement. The other 4 were done before corrective surgery. 3 with previous anastomosis and one with pulmonary banding, before correction. Conclusions: 1) Collaterals in patients with aortic coarctation could be accurately evaluated by MR. 2) MR is a good method to delineate pulmonary veins features. 3) Visualization of pulmonary artery and branch anatomy by MR prior surgery could avoid catheterism.

P1310
Magnetic resonance imaging in the diagnosis and follow-up of takayasu arteritis in children
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BACKGROUND: Takayasu arteritis (TA) carries up to a 40% mortality rate in children. Because the clinical presentation of TA is often nonspecific, accurate and prompt diagnosis depends on a high index of suspicion and appropriate laboratory and imaging studies. We report the use of advanced magnetic resonance imaging (MRI) in the evaluation of TA, its activity, and its complications. METHODS and RESULTS: We obtained T1-weighted, T2-weighted, contrast-enhanced MR images and MR angiograms of the chest and abdomen in three children (age range 11–14 years). The MRI studies confirmed the diagnosis of active TA and were repeated to evaluate response to treatment. Two patients showed complete resolution of lesions found on MRI at 6th and 12th month follow-up while the third patient showed no significant improvement. CONCLUSION: We have shown that MRI can help establish the initial diagnosis of TA in children and help monitor disease activity to guide therapy.

P1311
Decreased aortic elasticity in operated versus non-operated Marfan patients
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Background: Following aortic root replacement Marfan patients may develop complications in the aortic tract beyond the aortic root, even without severe aortic dilation. Aortic stiffness parameters are related to aortic rupture behaviour and may serve as additional risk factors for aortic complications before the aorta is dilated. Purpose: To compare aortic elasticity between electively operated and non-operated Marfan patients. Methods: 30 Marfan patients with elective aortic root replacement (mean age 35±13 years, 21 Bentall, 9 David) and 63 non-operated Marfan patients (mean age 32±11 years) underwent magnetic resonance imaging of the entire aorta. Aortic diameters and distensibility (D) at 3 levels of the descending aorta were assessed (level 1: descending thoracic, level 2: diaphragm, level 3: above the aortic bifurcation). Furthermore flow wave velocity (FWV) between level 1 and 3 was assessed. Results: Aortic diameters were normal at all levels in the operated and non-operated group. However, the operated patients had a significantly decreased local distensibility (D) at the level of the descending thoracic aorta compared to the non-operated patients (2.5 ± 1.5 vs. 3.6 ± 2.0 x 10-3mmHg-1, respectively, P=0.011). No significant difference was found in aortic flow wave velocity (FWV) between the operated and non-operated group (5.6 ± 1.5 vs. 5.7 ±1.3 m·s-1). Conclusion: Following elective aortic root replacement Marfan patients show decreased local elasticity in the descending thoracic aorta compared to non-operated Marfan patients. This might be of clinical importance in the follow up of operated Marfan patients.

P1312
Prognostic value of aortic elasticity on aortic complications in patients with Marfan Syndrome
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Background. In Marfan patients survival is mainly determined by aortic complications at a relatively young age. The occurrence of aortic dissection and rupture in Marfan patients is difficult to predict by mere aortic dimensions. Assessment of aortic elasticity may be of additional value for risk stratification. Methods: To assess the prognostic value of aortic elasticity on aortic complications (defined as: 1. aortic root diameter increase > 2 mm/year, 2. aortic dissection or 3. death), 73 Marfan patients (aged 31 ± 8 years, 41 men and 32 women) underwent magnetic resonance imaging of the entire aorta in 1997 and were followed up for 3 years. Aortic diameter and ascending aortic distensibility were assessed. MR velocity mapping was used to assess flow wave velocity along the descending aorta as an additional index of elasticity. Results: 10 patients (13.6%) of the 73 patients examined in 1997 reached one of the endpoints (7 patients with an increase in aortic root diameter > 2 mm/year, 3 acute dissections) after 3 years (3.1 ± 0.2 years). The patients were divided in a complicated and a non complicated group. There was no significant difference in baseline characteristics and in aortic root diameter (47 ± 4.4 vs. 43 ± 7.3 mm, respectively) between the two groups. However, the 10 complicated patients had a significantly decreased local ascending aortic distensibility (2 ± 1 vs. 3 ± 1 x 10-3 mmHg-1, respectively) and significantly higher descending aorta flow wave velocity (6.6 ± 1.6 vs. 5.8 ± 1.3 m·s-1, respectively) compared to the non-complicated Marfan patients. Conclusion: Descending aorta flow wave velocity and ascending aorta distensibility are related to the occurrence of aortic complications in Marfan patients, and could be of additional prognostic value for risk stratification.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

P1313

The quality and usefulness of spiral CT and 3-D images in patients with central airway disease associated with congenital heart disease.

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We analyzed the quality and usefulness of the spiral CT and three-dimensional images in 56 patients with central airway disease associated with congenital heart disease. Forty-nine patients (88%) were less than 5 years old, including 5 neonates and 31 infants. Their median age was 9.7 months and median body weight was 7.0 kg. Spiral scanning was performed after sedation with chloral hydrate (n=47) and administration of contrast media (n=56) via the pedal route (n=48). It was performed with as thinner collimation as possible from 1 to 3 mm and overlapped reconstruction of 50–20%. The airway stenoses were located at: trachea in 2b and bronchus in 31. Their causes were aortic arch anomalies (n=8), preterminally malposed aortic arch (n=7) and ascending aorta (n=5), innominate artery compression (n=6), aortic arch anomaly (n=8), aberrt pulmonary valve (n=6), displaced or dilated cardiovascular structure (n=20), and pulmonary artery sling (n=2). Motion artifact caused mild or negligible image degradation in most patients except 7. The quality was graded as good in 32 and excellent in 15. Non-sedated children with breath-holding scan present more severe motion artifact by cardiovascular pulsation. In the evaluation of the airway disease associated with congenital heart disease, the motion artifact caused the image degradation, but diagnostic three-dimensional images could be obtained. The spiral CT and three-dimensional reconstruction may be served as primary diagnostic modality in the uncooperative children with congenital heart disease and suspected airway stenosis.

P1314

Total cavopulmonary connection. Respiratory variations in blood flow (real time MRI) at rest and during exercise

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After total cavopulmonary connection (TCPC), the quantitative importance of respiratory fluctuations on pulmonary blood flow during exercise was studied in 8 patients, 11.9±4.5 years old, 5.6±2.5 years after TCPC. During supine bicycle exercise (resting = 0 watt (W)/kg, 0.5 W/kg, 1.0 W/kg) real time blood flow was measured during entire respiratory cycles in the aorta, the superior vena cava and the inferior vena cava tunnel using a 1.5T MRI scanner. Results. *Significantly different from 0W, p<0.05. Mean of N=11. Caval flow = superior + inferior vena cava flow. Conclusion. In TCPC, caval inflow predominantly occurs during inspiration at rest and this respiratory dependency is maintained during exercise. Aortic blood flow, however, changes less with respiration.

https://doi.org/10.1017/S1047951100012142 Published online by Cambridge University Press

365

Alterações Cardíacas Fetais Relacionadas à Toxoplasmose Materna. Um Estudo Ecocardiográfico Pré-Natal

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Introdução: Toxoplasmose é uma doença infecciosa multissistêmica causada pelo *Toxoplasma gondii* que pode determinar alterações no desenvolvimento fetal. Na toxoplasmose congênita em geral aceita-se que a transmissão placentária-hematogênica só ocorra nas infecções recentes, porém há relatos de casos de mães portadoras de toxoplasmose que permaneceram com o parasita encistado nas fibras uterinas, podendo haver infecção fetal em gestações futuras. As lesões são inespecíficas como áreas de necrose que sofrem calcificação precoce.

Objetivo: Este trabalho tem como objetivo descrever os achados ecocardiográficos em fetos cujas mães tinham diagnóstico de toxoplasmose gestacional.

Delineamento: Estudo prospectivo de série de casos

Métodos: A amostra consistiu de 23 fetos examinados consecutivamente no período de outubro de 1998 a abril de 1999 através de ecocardiograma fetal a cores, cujo motivo de encaminhamento havia sido toxoplasmose materna. A idade gestacional variou de 20 a 37 semanas (média 30 semanas)

e a idade materna variou de 16 a 44 anos (média 26 anos).O diagnóstico de toxoplasmose baseou-se na presença de títulos elevados de anticorpos anti-toxoplasma (IgG representando infecção crônica e IgM, infecção aguda).

Resultados: Em 21(91,4%) fetos foram observadas áreas de hiperecogenicidade localizadas nos aparelhos valvares e subvalvares, bem como na superfície septal endocárdica de ambos os ventrículos, algumas vezes com aspecto hiperecóico ("golf ball"). Não ocorreram modificações destes achados no decorrer da gestação.

Conclusão: Embora não haja relatos prévios de lesões endocárdicas fetais relacionadas a toxoplasmose materna, é possível que os achados descritos representem uma reação inflamatória tecidual muito prevalente.

366

Anormalidades Cardíacas em Crianças com a Síndrome da Imunodeficiência Adquirida (SIDA) com Transmissão Perinatal: um Estudo Clínico e Eletro-Ecodopplercardiográfico.

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Com o objetivo de determinar a prevalência das anormalidades cardíacas em crianças com a síndrome da imunodeficiência adquirida (SIDA) com transmissão perinatal, foram estudadas prospectivamente 87 crianças, sendo 39 do sexo feminino e 48 do sexo masculino, com idades variando de dois meses a doze anos. Foram realizados exames clínico, eletro e ecodopplercardiográficos seriados durante um período de quatro anos. Utilizou-se a classificação revisada de 1994 para crianças abaixo de 13 anos infectadas pelo vírus da imunodeficiência humana (HIV). As crianças foram classificadas em dois grupos: 1) grupo controle – 53 crianças (61%) que soroconverteram (não-infectadas); 2) grupo infectado – 34 crianças (39%). Este último foi subdividido em: assintomático (grupo N – 4,6%) e sintomáticos (grupos A, B e C – 34,4%). As crianças do grupo controle não apresentaram anormalidades de nenhuma espécie do ponto de vista clínico-eletro-ecodopplercardiográfico. Obtivemos os seguintes resultados no grupo infectado: 1) clínicos – fadiga (41%), hepatoesplenomegalia (38,8%), ICC (23,5%) e derrame pericárdico com tamponamento cardíaco (2,9%); a maioria pertencia ao grupo C (severamente sintomático). Houve seis óbitos (20,5% - todos do grupo C); 2) eletrocardiográficos – taquicardia (32,4%) e bradicardia sinusal (2,9%), desvio do eixo elétrico do QRS para direita (14,7%) e para esquerda (5,8%), distúrbio de condução do ramo direito (20,5%), hemibloqueio anterior esquerdo (2,9%), anormalidades da onda T em V1 (5,8%), distúrbio difuso da repolarização ventricular (8,8%) e sobrecarga ventricular esquerda (11,8%); 3) ecodopplercardiográficos – dilatação e disfunção ventricular esquerda (20,5%), derrame pericárdico grande (2,9%), insuficiência mitral leve(5,8%) e importante (2,9%), insuficiência tricúspide leve (8,8%), prolapso de válvula mitral (5,8%) e tricúspide (2,9%), e hipertensão pulmonar (2,9%).

Concluímos que mais da metade das crianças nascidas de mães HIV-positivas soroconverteram. O envolvimento cardíaco é via de regra uma manifestação tardia nas crianças com SIDA em fase avançada. A anormalidade cardíaca mais freqüente é a miocardiopatia dilatada.

367

Análise do Emprego da Operação de Blalock-Taussig em Cardiopatias Congênitas Cianogênicas: Experiência em 834 Pacientes.

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Objetivo - Mostrar a experiência com a operação de Blalock-Taussig (BT) no tratamento de diferentes cardiopatias congênitas.

Material - De nov/86 a dez/98, 834 crianças (467 masculinas) foram submetidas à operação de BT. Foi realizado no 1º ano de vida em 72% (neonatos – 36%). Foi realizada em caráter eletivo em 58% e em 42% como emergência. Os diagnósticos mais freqüentes foram AP (283 casos), Tetralogia de Fallot e IPIV (291 casos) e AT (118 casos). A operação foi do tipo modificada com tubo de PTFE em 797(95,6%) casos, com tubos de 5mm em 580 casos. O BT foi isolado em 686 casos e associado a outros procedimentos em 148 casos, como ligadura de colaterais sistêmico pulmonares em 35, unifocação ou ampliação de artérias pulmonares em 51.

Resultados – A mortalidade hospitalar foi de 12,5% e a tardia 8,7%. Variáveis como sexo, caráter da indicação, diagnósticos pré-operatório e tipo de operação não demonstraram relação com maior mortalidade ($p>0,05$). Em neonatos observou-se maior mortalidade ($p=0,008$), assim como em pacientes operados com tubos de diâmetro menor que 5mm ($p<0,001$). Em 116 casos se realizou o tratamento definitivo da cardiopatia, estando 180 casos aguardando correção. Trombose do BT e quadros infecciosos foram os fatores mais prevalentes de morbi-mortalidade após o BT.

Conclusão - A operação de BT, apesar de apresentar morbi-mortalidade maior em alguns grupos, é procedimento fundamental no manuseio das cardiopatias congênitas cianogênicas, devendo ter seu uso continuado, reforçando a necessidade de cuidados técnico cirúrgicos e acompanhamento adequado para melhor evolução.

368

Papel da perfuração da valva pulmonar com radiofreqüência seguida da dilatação por balão no tratamento da atresia pulmonar com septo ventricular íntegro.

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Introdução: O tratamento dos pacientes (pts) com atresia pulmonar (AP) e septo ventricular íntegro (SVI) continua desapontador. A perfuração da valva pulmonar com radiofreqüência seguida de dilatação por balão tem sido uma alternativa para a valvotomia cirúrgica da AP com SVI.

Objetivo: Este estudo visa apresentar nossa experiência com a perfuração da valva pulmonar (VP) com radiofreqüência seguida de valvuloplastia com balão em pacientes (pts) portadores de AP com SVI.

Material e Métodos: No período de maio de 1996 a julho de 1998, 7 pts portadores de AP com SVI que foram submetidos a perfuração valvar pulmonar por radiofreqüência, em nosso serviço. Todos os casos apresentavam ventrículo direito (VD) tripartite, e ausência de sinusóides importantes com circulação coronária VD não dependente, infundíbulo pérvio. Todos os pts exceto o caso 7 eram do sexo feminino. A idade dos pts variou de 3 dias a 4 anos e 2 meses com média de 1ano1m. Cinco crianças tiveram cirurgia prévia (Shunt Blalock-Taussig em cinco, e aortosseptostomia em um caso). O peso variou de 2,36 à 13,4 kg, média de 6,03kg.

Resultados: Todos apresentavam pressão do VD supra sistêmica. O anel da valva tricúspide variou de 9,8 a19mm, média de 13,4mm, o valor Z variou de -0,42 a +0,03. A perfuração valvar foi possível em todos os casos exceto no caso 6, a dilatação da valva pulmonar só foi possível em 3 pts (42,8%) casos 1,5 e 7. A duração do procedimento variou de 2hs 40min a 6hs 40min. Três casos apresentaram complicações (1 óbito, 1 derrame pericárdico, 1 trombose venosa profunda).

Conclusão: A perfuração por radiofreqüência pode ser considerada como uma alternativa promissora ao tratamento cirúrgico para os casos mais favoráveis de atresia pulmonar com septo íntegro (VD tripartite, infundíbulo pérvio).

284

Ecocardiografia de Contraste com Microbolhas em Cardiopatias Congênitas

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Objetivos: A ecocardiografia de contraste com microbolhas prima pelo fornecimento de informação funcional. Desenvolvemos a técnica em crianças com doenças cardíacas congênitas para tentar definir perfusão, contratilidade e viabilidade miocárdica.

Casuística e Método: Trata-se de estudo prospectivo com 29 pts de 8dias a 34 anos, 3kg a 74 kg. As cardiopatias estudadas têm predomínio de Tetralogia de Fallot, Dupla Via e Transposição de Grandes Artérias. Sob sedação com hidrato de cloral a 10%, quando indicado, foi realizado a infusão de contraste ecocardiográfico – PESDA (perfluorcarbono sonicado com solução coloidal) e em seguida aplicado o protocolo de estudo desenvolvido com monitorização clínica. Foi simultaneamente avaliada ecocardiografia convencional acoplada a imagem harmônica contínua, intermitente em sístole, nos tempos de 1,3,5,7 batimentos.

Resultados: Foi observado completo enchimento das cavidades em todos os pts sem ocorrência de efeitos colaterais, com boa difusão das microbolhas. Na maioria a perfusão se mostrou comprometida porém sem prejuízo importante da contratilidade global. Obteve-se imagem anatômica mais detalhada bem como melhor caracterização da dinâmica do fluxo sanguíneo. Ainda há limitações como falta de padrão de normalidade para a idade e para cardiopatias complexas.

Conclusão: Apesar dos problemas esta técnica se mostra promissora, especialmente pela capacidade de contribuir com informações sobre microcirculação, perfusão, viabilidade e função miocárdica, não só de VE mas também de VD.

SOCESP

Maio/Junho — 1999
Volume 9 — Nº 3

Suplemento da **Revista da Sociedade de Cardiologia do Estado de São Paulo**

ESPECIAL

Temas Livres e Posters do XX Congresso da SOCESP

ISSN 0103-8567

Atriosseptostomia por Cateter Balão à Beira do Leito

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Introdução: Algumas crianças com cardiopatia congênita (CC) necessitam de uma comunicação interatrial (CIA) para a sua sobrevivência, seja para se obter uma melhor mistura sangüínea ao nível dos átrios e conseqüente aumento da PO2 sistêmica ou para descompressão atrial, aumentando o débito sistêmico e reduzindo a pressão venosa pulmonar. A criação de uma CIA pode ser realizada através do Procedimento de Rashkind (atriosseptostomia por balão). Classicamente este procedimento é realizado com a utilização da radioscopia, podendo ser realizado apenas com o uso da ecocardiografia bidimensional.

Objetivo: Avaliação dos resultados da realização do procedimento de Rashkind guiado apenas pela ecocardiografia à beira do leito (UTI, berçário, sala de cardiologia invasiva) em pacientes com CC.

Material e Métodos: O procedimento foi realizado em 26 pacientes: TGA (19), DVSVD, com CIV subpulmonar (2), atresia tricúspide (1), DATVP (1), hipoplasia do ventrículo esquerdo (1). 73,6% eram do sexo masc. e 26,3% do sexo fem. A idade variou de 1 a 60 dias (média de 9,7 dias) e o peso de 2620 a 3990 g (média de 3199g). O critério de sucesso do procedimento foi a elevação da Sat O2 sistêmica para níveis acima de 60%, o tamanho da CIA, a redução do gradiente pressórico entre os átrios e a melhora clínica.

Resultados: Obtivemos sucesso em 18 casos (87,5%). A SatO2 pré variou de 16 a 81% e pós de 40 a 95%. O tamanho médio da CIA após o procedimento foi de 6,7mm. O tempo médio de execução foi de 1,30h. Uma criança apresentou taquiarritmia supraventricular, sendo cardioversão com sucesso. Não houve óbito relacionado ao procedimento. A imagem ecocardiográfica foi adequada para o procedimento em todos os casos.

Conclusão: A atriosseptostomia com cateter balão guiada apenas pelo ecocardiograma bidimensional a beira do leito se mostrou segura e eficaz na grande maioria dos nossos casos, com pouco risco para a criança e evitando muitas vezes a transferência da mesma para o setor de cardiologia invasiva, servindo de ponte para posterior correção cirúrgica.

The Second World Congress

of

Pediatric Cardiology and Cardiac Surgery

Honolulu, Hawaii
May 11 - 15, 1997

ABSTRACTS

P541

Head-up tilt test for the diagnosis of unexplained syncope in children

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To investigate the efficacy of the head-up tilt test in the workup of syncope of unknown origin, 30 cases of unexplained syncope and 13 health children were studied by the head-up tilt to 60° for 45 minutes. The results showed that the head-up tilt test was positive in 73% of children with unexplained syncope, but none of control subjects. The sensitivity, specificity and diagnostic value of the tilt test were 73%, 100% and 81%, respectively. The average time to the onset of symptoms was 23±12 minutes during the tilt test. Three patterns of response to head-up tilt test were observed in positive responders: vasodepressor pattern with an abrupt fall in blood pressure in 12/22 of responders; cardioinhibitory pattern with profound bradycardia in 3/22, and mixed pattern with both blood pressure and heart rate decrease in 7/22. β-adrenoceptor was effective for the treatment of children with syncope of cardioinhibitory or mixed pattern proved by the test. The result of this study suggests that the head-up tilt test is a useful and an objectively diagnostic tool for evaluating unexplained syncope in children.

P542

Body composition as a determinant for exercise tolerance in children with heart disease

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In many determinants for exercise tolerance during long term follow-up in children with heart disease, nutritional state, especially body composition, has not been elucidated. For this purpose we performed dual-energy X-ray absorptiometry (DXA) and expiratory gas analysis during treadmill exercise test in 50 children with various heart diseases (congenital heart diseases: 27, Kawasaki disease: 22, hypertrophic cardiomyopathy: 1). 35 males and 15 females and age at investigation ranged 3 to 19 yrs (median: 11.3 yrs). Body composition indexes, fat (F) and lean body mass (LBM), were measured separately in body parts, trunk (T), upper and lower [illegible] and expressed as the percentage to total body mass. Exercise tolerance was evaluated by peak oxygen uptake (pVO_2) during exercise test. 11 children (22%) showed obese by body mass index (BMI) more than 20 kg/m^2 but 20 children (40%) have higher %F than that of 90 percentile in normal children. %F was correlated with BMI ($p<0.0001$). There was no significant difference in pVO_2 between obese and nonobese children. Neither, %F nor %LBM were correlated with pVO_2. In segmental body composition analysis, %F-T and %LBM-L were correlated with pVO_2 ($p=0.031$, <0.0001, respectively). The present study reveals that significant number of children with heart diseases have excessive fat tissue even under appropriate managements. It also has shown that the fat tissue in body trunk and leg muscle are determinants for exercise tolerance. These results suggest that children with heart diseases may require more positive exercise management.

P543

Immunologic evaluation of children with Congenital Heart Diseases

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This study was performed to evaluate the immuno system of 21 children with Congenital Heart Disease (CHD), ten cases were acyanotic and eleven of cyanotic CHD with ages ranging from 1 to 140 months (median = 8 months). Tetralogy of Fallot and Ventricular Spetal Defect were the most frequent congenital lesions. Eighteen children had malnutrition and six had recurrent infections.

Polimorphonuclear leukocyte phagocytic function of children with CHD (cyanotic and acyanotic) was impaired with decreased capacity to digest microorganisms (phagocytic index). However destruction of ingested bacteries occured normally (bactenolysis index).

The analysis of total T - lymphocytes subpopulations (CD2), T - helo (CD4), T suppressers / cytotoxics (CD8) and B lymphocytes through the immunobeads technique, did not show any significant difference between acyanotic CHD and control children. In cyanotic CHD all these cells populations were deeply reduced. These results are not similar to the literature and will have to be reconfirmed later on.

In more than 50% of children with CHD, we observed that the serum the immunoglobulins, IgG and IgG, were above that of the normal populations. C3 and C4 levels were adequate, with the complement system showing appropriate function of these patients.

P544

The thrombolytic treatment with Actylise (t-PA) in children with congenital heart disease and other cardiac pathologies.

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The purpose of the study was to analyse the results of thrombolytic treatment with (t-PA) Actylise in pediatric cardiac patients. 21 children aged 14 days to 17 yrs (mean 3.5 + 0.5 yrs) with congenital heart disease and other cardiovascular pathology were treated with Actylise. There were following indications for thrombolytic therapy: 1. Peripheral veins or arteries thromboses after baloon valvuloplasty of pulmonary stenosis (N=1), and aortic stenosis (N=1), reCoA (N=1), coil occlusion of PDA (N=1). 2. Thromboses in left ventricle or left atrium (N=3) and on the disc of mechanical prosthetic valves (St. Jude) in the mitral position (N=2). 3. Coronary arteries thromboses in Kawasaki disease (N=2). 4. Pulmonary arteries thromboses (N=3) 5. Renal vein thrombosis (N=1) and vena cava superior thrombosis (N=1) 6. Stent thrombosis in the middle thoracic aorta stenosis (N=1). Does of Actylise ranged between 0.03 to 0.1 mg/kg per hour given i.v. from 1 to 10 days (mean 5.2 days). Actylise was given to systemic vein or locally directly to occluded vessel. The PT, PTT, Fibrynogen, FDP levels and complete blood cell count were monitored during thrombolytic therapy. After Actylise treatment was completed heparine was given for 1-2 days. Very good results--complete resolving of thrombi were achieved in 14 pts, partial in 4 pts, no resolving in 3 pts. Bleeding from gastrointestinal tract as side effect of treatment with Actylise was observed only in 2 pts.

Conclusions:

1. Actylise in very effective and save thrombolytic drug in children with various cardiac pathologies.
2. Bleeding from the gastrointestinal tract was the only complication observed in 2 pts.
3. Actylise in the does 0.03-0.1 mg/kg/h was given for longer time than recommended by producer and did not cause serious side effects.

687

ANEL VASCULAR

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O Anel vascular é uma anomalia congênita rara, sendo formado pelo arco aortico e seus ramos que quando em posições anômalas, ocasionam compressões de grau variável da traquéia e do esofago. Este trabalho tem por objetivo avaliar retrospectivamente os sintomas, o diagnóstico, a evolução e os tipos de anel vascular tratados em nosso serviço, em um periodo de doze anos.

Foram analisados 31 pacientes com idades entre 15 dias e 5 anos. Os sinais e sintomas mais comuns foram: estridor, sibilos, hipersecreção de vias aéreas, dispneia, pneumonias de repetição e disfagia. O RX de esôfago contrastado foi sugestivo de anel vascular em vinte e sete pacientes. Nos demais não foi possivel realizar o exame. O ecocardiograma, o estudo hemodinâmico e a broncografia confirmaram o diagnóstico. Foram identificados 13 pacientes com subclavia direita anômala, 11 com duplo arco aortico, 5 com arco Ao à direita c/ subclavia E anomala, 1 com artéria pulmonar E saindo da art pulmonar D, e 1 com artéria inominada D anomala.

A descompressão cirúrgica foi feita em dezessete pacientes. Dos quatorze não operados dez não formavam anel verdadeiro não havendo necessidade de ressecção. Dos quatro restantes, três faleceram no pre operatório e um aguarda a cirurgia.

O anel vascular pode ser identificado com o RX contrastado do esôfago. Os sinais e sintomas são os tipicos da compressão do esôfago ou da traquéia. O ecocardiograma, a hemodinâmica e a broncografia confirmam o diagnóstico.

699

COARCTAÇÃO DA AORTA: RESULTADOS DA CORREÇÃO CIRÚRGICA

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Ainda existe muita controvérsia sobre a melhor técnica para correção cirúrgica de coarctação da aorta (CoAo), principalmente em lactentes. O objetivo deste estudo foi de avaliar os resultados cirúrgicos a médio e longo prazo de pacientes submetidos a cirurgia de CoAo, comparando as técnicas cirúrgicas empregadas.

Através de uma coorte histórica de 155 pacientes operados de CoAo nativa entre janeiro de 1987 e janeiro de 1997, foi analisada a evolução tardia destes pacientes, relacionando a incidência de recoarctação (re-CoAo) com a técnica cirúrgica empregada.

A idade dos pacientes variou entre 0,1 e 216 meses (média 44 meses) e o peso entre 1,9 e 82 Kg (média 15 Kg). O tempo de seguimento foi de 1 a 127 meses. Os pacientes foram divididos em três grupos conforme a técnica cirúrgica empregada. O grupo I consistiu de 87 casos corrigidos pela técnica término-terminal (T-T), o grupo II de 27 pacientes corrigidos por istmoplastia com flap de subclávia (SUB) e o grupo III de 38 pacientes onde foi realizada istmoplastia com patch de material sintético ou pericardio bovino ou interposição de tubo (ISTM). A incidência de re-CoAo foi de 35% para o grupo I, 27% para o grupo II e 26% para o grupo III (NS). Entretanto, 60 casos do grupo I eram pacientes abaixo de 6 meses nos quais a incidência de re-CoAo foi de 57%. Em 39% dos pacientes entre 6 e 12 meses foi empregada a técnica T-T, e nestes a incidência de recoarctação (re-CoAo) foi de 6%, enquanto que a ISTM foi mais utilizada em pacientes acima de 24 meses (79%), e a incidência de re-CoAo nesta faixa foi de 33%.

Em conclusão, a incidência de re-CoAo é maior em lactentes pequenos, independente da técnica cirúrgica empregada.

700

EVOLUÇÃO TARDIA DE PACIENTES SUBMETIDOS A CORREÇÃO CIRÚRGICA DE COARTAÇÃO DA AORTA

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O objetivo deste estudo é avaliar os resultados a médio e longo prazo de crianças e adolescentes submetidos a correção cirúrgica de coarctação da aorta (CoAo).

Foi realizado um coorte histórico através de revisão de prontuários de 155 pacientes operados de CoAo nativa entre janeiro de 1987 e janeiro de 1997, com preenchimento de protocolo especifico. As variáveis analisadas foram hipertensão arterial sistêmica (HAS) no pós-operatório (PO) imediato e tardio, mortalidade e a incidência de recoarctação (re-CoAo).

A idade dos pacientes variou entre 0,1 e 216 meses (média 44 meses) e o peso entre 1,9 e 82 Kg (média 15 Kg). O tempo de seguimento variou entre 1 e 127 meses. Os pacientes foram divididos em 2 grupos conforme a faixa etária na época da cirurgia. O grupo I consistiu de 80 pacientes menores de 12 meses e o grupo II de 75 pacientes maiores de 12 meses. A mortalidade imediata foi de 11,3% para o grupo I e 2,7% para o grupo II ($p< 0.05$). A incidência de re-CoAo foi 28% para todos os pacientes. No grupo I, reCoAo foi documentada em 34% e no grupo II em 22% (NS). Porém, em lactentes menores de 6 meses esta incidência foi de 57%. HAS no PO imediato ocorreu em 81% no grupo I e 92% no grupo II (NS), porém no PO tardio, 31% dos pacientes do grupo I mantiveram-se hipertensos, enquanto que no grupo II HAS foi observada em 15% ($p<0.05$).

Em conclusão, a correção cirúrgica da coarctação da aorta oferece adequado resultado cirúrgico na grande maioria dos pacientes, porem a incidência de coarctação residual ou re-CoAo é mais elevada quando a cirurgia é realizada em lactentes pequenos, aumentando assim a incidência de HAS tardia neste grupo de pacientes.

742

HIPERCOLESTEROLEMIA FAMILIAR HOMOZIGOTA - RELATO DE UM CASO

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Esta forma e de interesse pela grave e precoce lesao coronariana. Enfatizamos as dificuldades terapeuticas. SFS, fem. Negra, 8 anos, 20Kg, irmao com morte subita aos 8 anos. Assintomatica ate os 4. Apos observou-se xantomas nos cotovelos, fossas poplíteas. Cansaco aos grande esforços e dor precordial desde os 7. Sopro sistolico ++/4+ em foco aortico irradiando para carotidas, e deposito de colesterol em retina. ECG: com alteracoes primarias de repolarizacao na parede anero lateral. Colesterol = 1115mg/dl; HDL = 1076mg/dl; HDL = 23mg/dl; VLDL=16mg/dl e TG=96mg/dl. Ecocardiograma (ECO) de Repouso: leve do ventriculo esquerdo com insuficiencia mitral e aortica discretas, calcificacao da valvula aortica. Teste ergometrico: positivo para isquemia miocardica. ECO de estresse: Hipocinesia posterior e acinesia infero-basal, isquemia infero-posterior. Cinecoronariografia: Lesao triarterial grave e disfuncao do VE. Com terapeutica combinada (dieta, lovastatina e questran, obteve-se 30% de reducao dos niveis de colesterol. Aferese de LDL apesar das limitacoes tecnicas oferecem melhores resultados. Revascularizacao miocardica estaria indicada somente apos controle proximo do normal dos niveis de colesterol.

743

DIALISE PERITONEAL (DP) NO POS-OPERATORIO IMEDIATO (POI) DE CIRURGIA CARDIACA PEDIATRICA

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Dialise Peritoneal no POI de cirurgia cardiaca e pouco documentada. Em

adultos ha mortalidade media de 50%. Analisamos as indicacoes e perfil de apresentacao pre e pos cirurgia com avaliacao da mortalidade. Dezesseis criancas de 2 dias a 10 anos, submeteram-se a DP no POI de cirurgia cardiaca entre 1992 - 1996. A mortalidade foi de 41% (7) das 4 criancas com insuficiencia cardiaca (ICC) no pre-op, 5 morreram. Tempo de circulacao extracorporea foi maior no grupo que faleceu (112, 28' x 108') nao observamos diferencas para o K+ serico e diurese nas 48h de PO (3,6ml/Kg/h x 3,2ml/Kg/h), hipervolemia em todos foi a principal indicacao de DP e determinou ICC com 100% de obito. Insuficiencia renal (IR) foi observada em 3 dos que morreram e 2 que sobreviveram. Causa de obito foi choque cardiogenico em 3 e insuficiencia respiratoria em 4. A taxa de mortalidade com indicacao de DP e alta, basicamente pela condicao hemodinamica e complicacao extracardiaca. ICC no pre-operatorio parece associar a maior mortalidade. Nao se deve esperar instalacao de IR ou ICC com congestao pulmonar para indicar DP.

744

BALOES DESTACAVEIS E A OCLUSAO DE FISTULAS CORONARIANAS E PULMONARES DE GRANDE PORTE

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Fistulas de grande porte sao dificeis de serem tratadas com cateterismo intervencionista. A utilizacao de baloes destacaveis (BD) poderia permitir a oportunidade de fechamento de fistulas que de outro modo teriam que ser encaminhadas a cirurgia. Descrevemos 3 casos de intervencao em criancas, utilizando BD. 1) menino de 12 anos, cianotico, com sopro continuo, teve multiplas fistulas pulmonares diagnosticadas em seu pulmao direito, tres baloes e varias molas foram utilizadas para oclusao. A SO2 subiu de 87 a 94 e o hematocrito diminuiu de 62 para 47 com desaparecimento do sopro continuo na evolucao de 8 meses. 2) menino de 3 anos com fistula aorta-ventriculo direito residual voltou a apresentar ICC e cansaco facil. A fistula propiciava um grande fluxo E e D com Qp/Qs de 2,5:1. Tres baloes e varias molas foram colocadas com trombose da fistula, desaparecimento do sopro e da ICC. A isquemia miocardica desapareceu. 3) um neonato de duas semanas foi visto em ICC grave, com sopro continuo e uma fistula de coronaria direita para VD medindo 8mm de diametro. O eco demonstrou hipocinesia severa do VE e VD. Um BD unico no 16 foi colocado junto ao "colo" da fistula. Houve desaparecimento do sopro e da ICC. Um eco controle mostrou funcao normal do VE e VD em repouso. Em casos selecionados, o uso de BD e uma excelente alternativa tecnica para a oclusao de fistulas grandes do territorio pulmonar ou coronariano.

745

CATETERISMO INTERVENCIONISTA PEDIATRICO NAO USUAL

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O cateterismo (cat) intervencionista e possivel em criancas cardiopatas em grave instabilidade hemodinamica. Apresentamos 8 casos: 1o estenose pulmonar e insuficiencia tricuspide com ICC direita, submetido a 2 intervencoes sem sucesso via femoral. Optou-se por acesso jugular D para dilatacao pulmonar e queda do gradiente transvalvar de 83 r 15mmHg. 2o Sindr. Down e Tetralogia de Fallot com trombose aguda no POI de Blalock-Taussig modificado, submetido a trombolise, no cat, com estreptoquinase (200UI/Kg), com sucesso e potencia a posteriori. 3o PO de bicavo-pulmonar com persistencia de fluxo residual importante para o tronco pulmonar e ICC grave. O tronco pulmonar foi ocluido com umbrella por meio de cat com sucesso. Com queda imediata de pressao de TP de 19,0 r 7,4mmHg. 4o transposicao de grandes arterias submetido a cirurgia de Senning. No POI evoluiu com ICC direita, severa. Submetido a dilatacao de estenose na juncao cava inferior atrio-venoso com sucesso. 5o fistula arteriovenosa pulmonar. Realizada embolizacao da fistula em 2 cats com salto de oximetria de 65% r 91%. 6o fistula de seio de Valsalva para VD. Cirurgia previa com fluxo residual. Embolizacao com baloes e molas com sucesso. 7o e 8o casos com protese tricuspide pos Ebstein e estenose aortica severa, ambos em baixo debito. Feito dilatacao da protese e da valvula aortica, com posterior encaminhamento efetivo para cirurgia. O cat intervencionista constitui eficiente recurso terapeutico, evitando reop. em pacientes inoperaveis e servindo de "ponte" para cirurgias eletivas.

746

PROPANOLOL NAO AUMENTA A PREVALENCIA DE PREMATUROS EM GESTANTES CARDIOPATAS

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Analisar o uso de propanolol em gestacoes de cardiopatas, suas indicacoes e repercussoes no concepto. Analisando 66 gestacoes, de um total de 674 gestacoes acompanhadas de 1981 a 1995, nas quais foi usado propranolol na dose de 30 a 80 mg, analisamos o grau funcional (NYHA) inicial, diagnostico materno e as repercussoes no concepto. Observamos uma incidencia de 14% de prematuridade, 14% de RN pequenos para idade gestacional e 21% de RN com peso ao nascer menor que 2.500g (valor na populacao geral da EPM respectivamente sem 22%, 8,3% e 10%). Os diagnosticos maternos eram: estenose mitral = 25, dupla lesao mitral com predominio de estenose = 10; protese biologica mitral = 5, reestenose mitral = 14; cardiopatia isquemica = 1; prolapso de valva mitral = 8; CIV = 1; TPSV = 2. No 1o trimestre, 18,3% das gestantes apresentavam ICC, 66% no 2o trimestre e 25,7% no 3o trimestre. Duas pacientes fizeram uso de digoxina associada; 3 receberam diuretico associados. Nenhuma recebeu anticoagulante oral. Neste grupo, 4 pacientes foram submetidas a comissurotomia e 15 a valvuloplastia mitral. Propanolol da dose utilizada nao aumentou a incidencia de prematuridade num grupo de gestantes cardiopatas com alta incidencia de ICC.

751

CATETERISMO CARDIACO COM PUNCAO ARTERIAL EM CRIANCAS ABAIXO DE 5 ANOS

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Verificar as complicacoes decorrentes da puncao arterial em criancas menores de 5 anos. Estudamos 309 criancas submetidas a cateterismo cardiaco através de puncao arterial para verificarmos as complicacoes decorrentes da tecnica. A idade media foi de 22,4 meses e o peso medio de 9,3 kg. O tempo medio do exame foi de 121 min. O cat foi através da artéria femoral direita em 98% sendo que em 2% houve necessidade de cat arterial bilateral. Os introdutores foram do no 4 ao 7 com media de 5,7 French. O tempo de puncao variou de instantaneo a 18s. Hematomas ocorreram em 12%. Houve necessidade de transfusao e a perda de pulso em 5 pacientes respectivamente. Insuficiencia vascular com graves complicacoes em 2 casos. O cat arterial em criancas com baixo peso pode ser realizado por puncao sem maiores complicacoes, nao havendo razoes para ser evitado. O material disponivel atualmente permite resultados semelhantes aos de criancas maiores.

752

ALTERACOES HORMONAIS EM CRIANCAS COM CARDIOPATIAS SUBMETIDAS A CIRURGIA CARDIACA.

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Criancas cardiopatas sofrem alteracoes hormonais na cirurgia cardiaca. O papel da circulacao extracorporea (CEC) e da hipotermia moderada (HM) nao estao ainda bem definidas. Foram 27 ceac (6 m e 12 a), divididas em 3 grupos. I) cirurgias com CEC sem HM: 11; II) cirurgias com CEC com HM: 21 e III) cirurgias sem CEC: 5. Sangue foi colhido no pre-op., apos inducao, abertura do torax, a cada 30 min. em CEC e com 24hs de PO. Usamos testes de Wilcoxon e Mann-Whitney na estatistica. As alteracoes foram dos niveis de cortisol, T4, TSH e - de ACTH, glicemia e insulinemia. As analises de cada hormonio foram significantemente diferentes na cirurgia em relacao ao pre em todos os grupos. No mesmo intervalo de tempo, comparando-os, nao houve em geral diferenca significante entre eles. Em nosso trabalho nao houve diferencas significantes, entre os grupos estudados. Parece que a intervencao cirurgica per se e mais importante que a CEC ou HM em provocar alteracoes hormonais.

753

COMPARACAO ENTRE OXIGENIO E DUAS CONCENTRACOES DE OXIDO NITRICO NA AVALIACAO DE RESISTENCIA VASCULAR PULMONAR, DURANTE O CATETERISMO CARDIACO, EM CRIANCAS COM LESOES DE SHUNT

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A utilizacao de oxido nitrico (NO) resultou em grande evolucao na compreensao da fisiopatologia vascular. Ha pouca informacao comparando diversas dosagens de NO e oxigenio (O2) durante o cateterismo (cat). Comparar o efeito de 2 dosagens de NO vs teste de O2 no cat de criancas com shunt e hipertensao pulmonar (HP) severa. Sete ceac (6m e 3a), com shunt e HP realizaram medidas hemodinamicas basais, apos 10min = 20ppm de NO, apos 10min. = 40 ppm de NO e apos 10 a 15min. de O2. A queda de resistencia vascular pulmonar e o aumento do Qp/Qs observados nao foram maiores com o NO em nenhuma das 2 dosagens utilizadas (20 e 40ppm) quando comparadas a prova tradicional com O2.

754

AVALIACAO DO TRATAMENTO CIRURGICO NA COARCTACAO DE AORTA EM NEONATOS E LACTENTES

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A cirurgia procura aliviar a obstrucao, eliminar a recoarctacao e a hipertensao arterial. Precocidade cirurgica favorece melhores resultados. Apresentamos nossa casuistica de anastomose termino-terminal (ATT) com enfase especial nos resultados e "follow up". Estudo observacional no periodo de 1981 a 96, retrospectivo. Avaliamos 13 ceac com coarctacao de aorta (CoAo), mediana 21 dias (5d a 7m), sendo 7 no periodo neonatal. CoAo isolada em 2/13 casos, CoAo com obstrucao da via de saida de VE: 4, sendo 2 com VAo bicuspide (gradiente (grad.) medio 45 mmHg), 1 VAo tricuspide com grad. 55 mmHg e 1 estenose sub Ao. A comunicacao interventricular (CIV) esteve presente em 4/13 casos; estenose mitral em 1; comunicacao interatrial em 3; hipoplasia istmica em 2 e transposicao corrigida das grandes arterias em 1 caso. A tecnica foi a ATT. O grad. pre-op. pelo eco foi de 18-80mmHg (mediana=50mmHg), enquanto o pos-op. variou de 11-31mmHg (mediana=25mmHg). Nao houve complicacoes da tecnica cirurgica. Bandagem pulmonar foi realizada em 2 com CIV. Ocorreram 2 obitos; o primeiro no POI, por hemorragia intracraniana em um neonato com estenose Ao. associada. No seguimento (mediana=60d) nao se detectou grad. significativos, hipertensao arterial ou recoarctacao. A ATT foi metodo seguro, sem complicacoes imediatas e com significante reducao do grad. a curto e medio prazo.

747

ANTICOAGULACAO, GRAVIDEZ E CONCEPTO

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Avaliar as complicacoes no concepto e recem-nascido (RN) e em gestantes com uso de anticoagulante oral (ACO). Fizeram uso de ACO; 69 gestantes. Diagnosticos: protese (prot.) biologica mitral: 8; prot. metalica mitral: 32; prot. biologica aortica (Ao), Chagas, estenose mitral: 1; prot. mecanica Ao: 11; dupla prot. metalica mitro Ao: 4; dupla prot. biologica mitro Ao: 3; comissurotomia mitral + FA: 6 e dupla lesao mitral: 2. 47 usaram fenprocumon e 22 fenindiona. 1 recebeu heparina EV no 1o trim. A maioria chegou aos o 2o trim. No 3o trim o anticoagulante oral foi suspenso em torno da 33a sem. e substituido por heparina EV (32) e SC (7). Complicacoes maternas foram: ICC, endocardite, hemoptise, AVC embolico, embolia arterial, intoxic. digitalica: 1; TPSV: 2; TEP, FA e flutter com cardioversao eletrica, choque cardiogenico: 3; disfuncao de protese; edema agudo de pulmao; obito materno: 4. No concepto observamos abortamentos: 16 (23%); natimortos: 3. Dos 49 Rns vivos 22 (44,9%) eram prematuros, 26 (53%) apresentavam baixo peso e 5 (10,2%) eram pequenos para a idade gestacional, 7 com sind. Warfarinica (10,2%). Pacientes que necessitam de anticoagulacao sao de alto risco materno fetal. O uso de anticoagulante oral predispoe o - de incidencia de prematuridade de baixo peso do RN. A incidencia de sindr. Warfarinica foi 10%.

748

COEFICIENTE DE MORTALIDADE MATERNA DECORRENTE DE HIPERTENSAO ARTERIAL E SUAS PRINCIPAIS CAUSAS 1991 A 1993

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A hipertensao da gestacao vem se firmando como a principal causa de mortalidade materna (MM) em nosso meio. Registramos os coeficientes verificados e os principais eventos fisiopatologicos envolvidos nos obitos maternos. A partir dos obitos maternos ocorridos no Hosp. Sao Paulo entre 91-93 calculamos os coeficientes por 100.000 nascidos vivos e enumeramos suas principais causas. As principais causas foram: AVC Hemorragico 38%, Aneurisma Dissecante da Aorta 12,4%, Edema Cerebral 6,2%. Os resultados evidenciam indices alarmantes de MM, sendo que pode ter alguma correlacao com a qualidade da assistencia prestada, uma vez que estes casos sao admitidos em situacao critica. Note-se que o comprometimento cerebral ocorreu em 44,2%, sendo que o controle da pressao arterial nestas situacoes de emergencia, tem papel decisivo na reducao do risco de mortalidade materna.

749

INDICACOES DA ECOCARDIOGRAFIA DE STRESS NO ACOMPANHAMENTO CLINICO DE CRIANCAS COM SUSPEITA DE ISQUEMIA MIOCARDICA.

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A isquemia miocardica (IM) induzida farmacologicamente resulta em alteracoes da contratilidade avaliadas mediante o ecocardiograma de stress (ECO ST). A continuacao relatamos nossa experiencia em criancas com suspeita de IM. De 1994 a 96 estudamos 6 pacientes com provavel IM. 3 fem. e 3 masc., e idade de 2 a 12 anos. O protocolo seguiu as recomendacoes da Sociedade Americana de Ecocardiografia. Um paciente com coronaria anomala positivo para isquemia no ECO ST pre-op. negativou apos reimplante da coronaria. Outro com Takayasu foi negativo ate o nivel de 30 mg/kg/min. Um com hipercolesterolemia secundaria resultou negativo no ECO ST. Uma crianca de 8 anos com hipercolesterolemia familiar homozigota, colesterol de 1140 mg/dl teve teste positivo no ECO ST. Um PO tardio de fistula coronaria e fistula residual teve resultado positivo para isquemia. Outro paciente positivo para isquemia pre-oclusao de fistula coronariana negativou o teste apos embolizacao no cateterismo. O ECO ST e importante recurso diagnostico e de acompanhamento clinico em criancas com variadas formas de IM.

750

ATRIOSEPTOSTOMIA POR CATETER BALAO EM CRIANCAS A BEIRA DO LEITO.

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A seguir relatamos nossa experiencia com atrioseptostomia por balao (ASB) a beira do leito, entre os anos de 1990 a 97. A ASB foi utilizada a principio em criancas portadoras de transposicao das grandes arterias para "rasgar" o septo atrial e melhorar a mistura arterio venosa. Atualmente este procedimento pode ser expandido a patologias mais complexas. Admitimos no estudo 19 casos. 73,6% masc.; 26,3% fem.; com idades de 1 a 60 dias (media 9,7); a saturacao de oxigenio (SO2) pre variou de 16% a 81% e a pos de 40% a 95%; o tempo medio durou 1h30min. Em um caso nao houve melhora da SO2. Uma crianca apresentou taquicardia supraventricular revertida com sucesso. A imagem ecocardiografica, para visualizacao do balao foi adequada em todos os casos e nao houve complicacoes. A ASB e uma tecnica paliativa que pode ser realizada a beira do leito guiada apenas por ecocardiograma bidimensional.

755

ALTERACAO DO PH INTRAMUCOSO, MEDIDO ATRAVES DE TONOMETRIA GASTRICA, NO POS OPERATORIO DE CIRURGIA CARDIACA EM CRIANCAS.

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A oxigenacao tecidual pode ser avaliada pela tonometria gastrica (pHi). O objetivo foi avaliar as alteracoes do pHi de acordo com o tempo de circulacao extracorporea (CEC) e o comportamento apos transporte e durante a internacao na UTI pediatrica.

Foram estudadas 15 criancas com media de idade de 3 anos e 8 meses, media de peso de 12,3kg, submetidas a cirurgia cardiaca com CEC para correcao de defeitos congenitos ou adquiridos. Foi utilizado tonometro gastrico com solucao salina e coleta de sangue de arteria radial cateterizada. As medidas foram realizadas no final da cirurgia e imediatamente apos admissao na UTI em intervalos de 4, 8, 12 e 24 hs. Os pacientes receberam reanimacao fluidica com coloides, mesmo modo de ventilacao pulmonar mecanica, dobutamina e ranitidina. Para correlacionar o tempo de CEC e a estabilizacao do pHi os pacientes foram divididos em dois grupos (GI: CEC< 60' e GII: CEC > 60'). Testes nao parametricos foram realizados para a estatistica.

O tempo em media de estabilizacao do pHi no GI foi de 13.22+/-11,45 h e o tempo de estabilizacao do GII foi de 9,22+/-7.32 h (p=0,9399). Houve diminuicao do pHi em 12 pacientes (92,3%), a alteracao do pHi foi de 7.26+/-0,08 no centro cirurgico apos termino da cirurgia para 7,15+/-0.01 logo apos a chegada na UTI (p=0,0046). A media do pHi evolutivamente na UTI variou ate 7,38+/-0,08 com 24h. Nenhum paciente desenvolveu disfuncao de multiplos orgaos ou obito.

Nao houve correlacao significante entre o tempo de CEC e o tempo de estabilizacao do pHi. O transporte associou com alteracao do fluxo sanguineo esplenico. A normalizacao do pHi ocorreu apos 12 horas da admissao e apesar do pHi baixo nao houve maiores complicacoes.

346

CALAZAR NO HIAS 1995-96. 2. TRATAMENTO, EVOLUCÃO CLÍNICA E PROGNÓSTICO. Luis C. Rey, Ana C. C. Pompeu*, Vania M. de Oliveira e Maria H. L. Cavalcante. Hospital Infantil Albert Sabin, Fortaleza.

O calazar apresenta extensa distribuição geográfica nacional, especialmente no meio rural e periurbano do Nordeste. Em muitas regiões a moléstia não é diagnosticada localmente, mas referida a centros terciários, o que retarda o tratamento. Este estudo visou analisar as condutas e prognóstico dos pacientes do H. Infantil Albert Sabin (HIAS) de Fortaleza.

Estudo retrospectivo e prospectivo de 138 crianças com calazar hospitalizadas em 1995-96. Foram entrevistadas as acompanhantes e revistos os prontuários.

O mielograma foi diagnóstico em 89/133 pacientes (67%). Em 24 casos houve nova punção com 14 positivos. A imunofluorescência indireta foi realizada em 56 casos sendo 53 (95%) positivos. Aspirado esplênico foi diagnóstico em 12/17 crianças (71%). O tratamento mais utilizado foi antimonisto de meglumina (138 casos), em média por 26 dias (variação: 20-49). O alopurinol foi associado em 41 casos, em média por 18 dias. Em apenas 5/138 pacientes foi necessário introduzir anfotericina B. Antibióticos foram utilizados em 65% (90/138) dos casos e transfusões de sangue 36% (50/138). Infecção secundária ocorreu em 58 casos: 38 pneumonias, infecções muco-cutâneas (7), otite, diarréia e sepse (4). A letalidade foi de 8/138 internamentos (6%). A ocorrência de sangramento prévio esteve significativamente associada à maior letalidade (risco relativo 15.9, IC=3.0-36.1, p=0.001). A permanência hospitalar média foi de 26 dias e a maioria completou o tratamento IM a domicílio. No seguimento ambulatorial observou-se regressão do quadro clínico e laboratorial até 6 meses após tratamento. Ocorreu recidiva em apenas 2 casos.

O prognóstico dos pacientes com calazar no HIAS é bom, apesar da desnutrição, a neutropenia e infecções intercorrentes iniciais. Observamos ótima resposta ao antimônio pentavalente associado ao controle das infecções secundárias.

Doenças Parasitárias

345

CALAZAR NO HIAS 1995-96: I. ESTUDO CLÍNICO-EPIDEMIOLÓGICO. Luis C. Rey, Ana C. C. Pompeu* e Vania M. de Oliveira, H. Infantil Albert Sabin (HIAS), Fortaleza.

O calazar é uma enfermidade endemo-epidêmica no Ceará, com 510 e 213 casos em 1995 e 1996, respectivamente. O HIAS hospitaliza anualmente 15 a 20% dos pacientes do estado, muitos referidos sem diagnóstico.

Identificar características clínico-epidemiológicas de pacientes internados com calazar no HIAS em 1995-96.

Estudo retrospectivo-prospectivo de 138 casos hospitalizados. Foram entrevistadas as mães e revistos os prontuários. Considerou-se pacientes com diagnóstico firmado por achado parasitológico e/ou sorologia.

Os pacientes tinham em média 52 meses (1-153 meses); 76 (55%) do sexo masculino; 69 (50%) provinham do campo; 107 apresentaram sintomas até 30 dias da internação; 18 procuraram diretamente o HIAS, 62 após consulta e 41 após internamento prévio. As principais queixas foram: febre prolongada (134/138 casos), palidez (117), aumento abdominal (100), anorexia (67), astenia (52) e emagrecimento (51). Houve queixa de sangramento em 15/138 (11%) dos casos. A mediana da esplenomegalia era de 8 cm abaixo do rebordo costal esquerdo (RCE) e da hepatomegalia de 4 cm do RCD. Havia déficit ponderal superior a 40% (DII segundo Gomez) em metade dos pacientes. Todos apresentavam hemoglobina <10 g/L (2,8-9,5g/L), sendo 55/134 <6,0 g/L. A hemossedimentação (1ª hora) estava >100 mm em 41% (47/115) dos casos; 60% das crianças (77/136) tinham leucócitos <3.000 cel/mm³ e 83% (81/131) neutropenia (<1500 cel/mm³). Havia plaquetopenia severa (<50.000/mm³) em 32% dos casos (40/127).

O calazar é uma doença endêmica no Ceará. No HIAS as crianças apresentam alterações nutricionais e hematológicas severas. O reconhecimento clínico desta parasitose é de suma importância para que os clínicos possam tratá-la precocemente a nível ambulatorial e local, quando a duração do tratamento é menor e as chances de cura são excelentes.

453

SECNIDAZOL SUSPENSÃO (DOSE ÚNICA) VS. TINIDAZOL SUSPENSÃO NO TRATAMENTO DA AMEBÍASE INFANTIL.
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A amebíase é uma protozoose que, na sua forma invasiva, atinge cerca de 50 milhões de pessoas em todo o mundo, ocasionando entre 40-100 mil mortes anuais. Este estudo multicêntrico, aberto, comparativo, entre secnidazol e tinidazol teve como objetivo comparar a eficácia e segurança desses medicamentos, no tratamento de 303 crianças com amebíase.

Crianças de 2 a 14 anos, de ambos os sexos, procedentes de 5 centros brasileiros, foram admitidas no estudo após a verificação dos critérios de inclusão e de exclusão. Os pacientes foram randomizados para receber secnidazol suspensão em dose única de 1 ml/Kg (SEC) ou tinidazol suspensão na dose de 0,5 ml/Kg, por dois dias (TIN). Foram avaliados clínica e laboratorialmente (método: direto, de Katz e de Faust) na consulta basal e no 7º, 14º e 21º dia após o tratamento.

Foram admitidos 156 pacientes no grupo SEC e 147 no grupo TIN. Foram excluídos da análise de eficácia clínica os pacientes assintomáticos na avaliação basal e aqueles com perda de seguimento. Para a análise laboratorial, foram excluídos somente os casos com perda de seguimento. Para a avaliação da tolerabilidade, todos os casos foram incluídos. Sucesso laboratorial no grupo SEC foi superior ao grupo TIN: 119/154 (77%) e 92/146 (63%) pacientes respectivamente, p=0,007. Cura ou melhora dos sintomas clínicos ocorreram em 128/138 (93%) no grupo SEC e em 125 (91%) no grupo TIN (p=0,317). A ocorrência de eventos adversos foi semelhante nos dois grupos: 12/156 (8%) SEC e 15/147 (10%) TIN, p=0,443, sendo na maioria de intensidade leve a moderada e relacionados ao sistema gastrointestinal.

Ambos os medicamentos mostraram-se eficazes no tratamento da amebíase, entretanto os índices de cura parasitológica para o grupo SEC foram significativamente superiores ao grupo TIN (77% vs 63%). Quanto à tolerabilidade geral, não houve diferença significativa, sendo ambos os medicamentos bem tolerados pelas crianças.

157

TITULO: DILATAÇÃO DO SEPTO ATRIAL COM CATETER BALÃO EM LACTENTES: EXPERIÊNCIA COM TRÊS CASOS.

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Em inúmeras situações a septostomia atrial com o balão de Rashkind é um procedimento que melhora a mistura arterioveno sa, dilatual a pressão do átrio esquerdo, corriga hipóxias graves e salva a vida de inúmeras crianças. Infelizmente há sucesso deste procedimento terapêutico apenas nos quatro pri meiros meses de vida, em média. As outras opções para septostomia após o período neonatal incluem a lâmina de Park ainda as hemodinâmica ou a cirurgia de Blalock-Hanlon, ambos associados com morbidade e mortalidade potenciais bem maiores que o Rashkind. O cateter balão nos pareceu uma excelente alternativa tática para dilatação do septo atrial em lactentes e foi utilizado em situações de emergência absoluta descritas a seguir: três crianças com idade respectivamente de 54, 87 e 47 dias e portadoras de crise refratária de hipóxia, ICC direita grave e hipóxia grave foram levadas à hemodinâmica. Seus diagnósticos eram atresia tricúspide IR, atresia tricúspide IC já com cerclagem e PG imediato de istmo - plastia e cerclagem em transposição completa, grande CIV e CoA severa. As duas primeiras apresentavam CIA restritiva e o terceiro caso septo atrial íntegro. Foram utilizados balões de 8, 10 e 12mm sendo realizadas diversas insuflações no septo com desaparecimento do entalhe inicial. A primeira criança melhorou a SO2 de 72 para 65, não precisou cirurgia e tem 17 meses de seguimento; o segundo diminuiu a pressão atrial direita de 24 para 8, evoluiu bem sem cirurgia e tem 13 meses pós dilatação. A terceira melhorou a saturação de 29 para 38 porém apresentou hemorragia pelo dreno torácico após o procedimento e foi a óbito. A necrópsia não revelou perfurações ou tamponamento.

Conclusões: A dilatação do septo atrial com cateter balão pode ser uma alternativa tática interessante em casos com septo atrial restritivo e com hipóxia ou ICC graves. No momento atual seu uso não deve se restringir a situações de emergência e a casos que não podem fazer Rashkind.

158

TITULO: EXPERIÊNCIA DA EPM COM CATETERISMO CARDÍACO EM CARDIOPATIAS CONGÊNITAS.

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Foram realizados no Setor de Hemodinâmica do Hospital São Paulo da Escola Paulista de Medicina, no período compreendido entre Janeiro de 1986 a Dezembro de 1991, um total de 868 cateterismos cardíacos, sendo apenas como cateterismo diagnóstico em 860 (88.8%) e intervencionista em 108 (11.15%) (Balão Rashkind). A faixa etária dos pacientes variou de um dia de vida a 48 anos. 517 eram homens e 451 eram mulheres. Havia 707 cardiopatias acianóticas e 161 cianóticas. Dentre as cardiopatias cianóticas a Tetralogia de Fallot foi a mais frequente, com 80 casos (9.3%), seguida por TGA com 51 casos (5.9%) e drenagem anômala total de veias pulmonares com 20 casos (2.3%). Dentre as cardiopatias acianóticas, a mais frequente foi a CIV com 126 casos (14.6%), seguida por estenoses pulmonar em 98 casos (11.4%), CIA em 70 casos (8.14%), DSAV em 60 casos (7.6%) e PCA em 28 casos (3.6%).

No período neonatal foram realizados 36 cateterismos cardíacos, sendo a patologia mais frequente a TGA com 14 casos (38.9%). Já no grupo dos lactentes 399 cateterismos foram realizados sendo o CIV a patologia mais frequentemente encontrada - 81 casos (20.3%) nos acianóticos e o Fallot nos cianóticos. Nos pacientes maiores de 16 anos a patologia dominante foi o CIA (27/52). Na análise ano a ano não houve variações significantes nos subgrupos neonatal, lactentes e adolescente/adultos.

Concluímos que hoje, com indicações de cirurgia cardíaca em neonatos apenas com o ecocardiograma, diminuiu muito o número de cateterismos em neonatos.

A análise dos subgrupos confirma que transposição é a doença com mais cateterismos neonatais (pela necessidade de septostomia) na Instituição. Em lactentes predomina CIV (quase só com hipertensão pulmonar) e Fallot e em adolescentes predominam as cardiopatias acianóticas mais benignas tipo CIA.

Abstracts

XXVIII REUNIÃO ANUAL DA SOCIED. LATINO-AMERICANA DE INVESTIGAÇÃO PEDIÁTRICA 28-31/out/1990 SÃO PAULO-SP, BRASIL

32

HIDROPSIA IMUNOLÓGICA PERINATAL: UMA NOVA ABORDAGEM TERAPÊUTICA.

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Nos países em desenvolvimento a hidropsia imunológica ainda é causa importante de morbimortalidade perinatal, devido ao uso não rotineiro da imunoglobulina anti-D.

No período de Jan/85 a Dez/89 no Hospital São Paulo(EPM), ocorreram 84 nascidos vivos(1,2%) e 6 natimortos de gestantes Rh sensibilizadas, dos quais 16 foram hidrópicos: 4 natimortos e 12 vivos. Destes,a hidropsia foi leve em 2 (16%), moderada em 5(42%) e grave em 5(42%) segundo a classificação de PHIBBS.

Em relação aos hidrópicos vivos observou-se mediana(Mi) da idade materna de 30 anos(22-39), antecedentes gestacionais com Mi do número de gestações, abortos e natimortos, respectivamente de 5(4-11), 0,5(0-3) e 2(0-3). Realizada amniocentese em 7 e transfusão intra-uterina peritoneal em 5 casos. A Mi da idade gestacional foi 32 sem.(28-38.7), sendo realizada cesariana por sofrimento fetal em 9 casos. A Mi do peso ao nascer foi 2050g (1220-3700).

Quanto à morbidade neonatal evidenciou-se: asfixia perinatal grave em 12 (100%), síndrome do desconforto respiratório(SDR) em 4(33%), SDR e hipoplasia pulmonar em 4(33%) e aspiração de mecônio em 2(17%). Punção de ascite e/ou hidrotórax foi realizada em 7 hidrópicos, ventilação mecânica em 12, drogas vasoativas em 9, exsanguíneo transfusão(EST) parcial com papa de hemácias em 10 e EST total em 8.

Nove hidrópicos evoluíram para óbito com tempo de sobrevida de 2 a 46 horas. A mortalidade relacionou-se ao insucesso na estabilização hemodinâmica e a barotrauma. Os sobreviventes eram pré-termo: 1 hidrópico leve com boa evolução neurológica, outro hidrópico moderado com hemorragia intra-ventricular e parenquimatosa e sequela neurológica e, o terceiro hidrópico grave com bom desenvolvimento neuro-psico-motor até a idade de 6 meses.

Acreditamos que esta taxa de sobrevida deveu-se à introdução em 1986 de um novo protocolo de atendimento ao hidrópico, com ênfase na estabilização cárdio-respiratória e hemodinâmica, assim como, na correção da anemia através de EST parciais com papa de hemácias, antes da abordagem convencional ao recém-nascido isoimunizado.

32

IMMUNE HYDROPS FETALIS: A NEW THERAPEUTIC APPROACH.

Miyoshi, MH; Feijó, LEH; Gomes, LFG; Freire, MFML; Lindsey, PC; Guinsburg, R; Miyasaki, CH.
Escola Paulista de Medicina (EPM), São Paulo, SP, Brazil.

In developing countries, immune hydrops fetalis is still an important cause of perinatal morbidity and mortality. The lack of anti-D immunoglobulin routine use is partially responsible for this fact.

Rh sensitized mothers delivered 84 newborn(NB) infants and 6 stillborn(SB) during Jan/85 to Dec/89 at São Paulo Hospital(EPM). Of these, 16 were hydropic: 4 SB and 12 NB. The hydrops degree (PHIBBS classification) was mild in 2(16%) cases, moderate in 5(42%) and severe in 5(42%).

Regarding the 12 hydropic NB, median maternal age was 30 yrs(22-39), and median number of previous pregnancies, abortions and SB was respectively 5(4-11), 0.5(0-3) and 2(0-3). Amniocentesis was performed in 7 cases, and peritoneal intra-uterine transfusion in 5. Median gestational age was 32 wks (28-38,7). Nine NB were delivered by C-section due to fetal distress. Median birthweight was 2050 gms(1220-3770).

Perinatal asphyxia was present in 12(100%) cases, respiratory distress syndrome(RDS) in 4(33%), RDS and pulmonary hypoplasia in 4(33%) and meconium aspiration in 2(17%). Ascites and/or hydrotorax were relieved by paracentesis in 7 NB; assisted ventilation was necessary in 12, vasopressors in 9, partial exchange transfusion(EXT) with packed red blood cells(PRBC) in 10 and total EXT in 8.

Nine hydropic infants died between 2 and 46 hours. The mortality was related to hemodynamic instability and barotrauma. Three infants remained alive: 1 mild hydrops with good neurological outcome, 1 moderate hydrops with grade IV intra-ventricular hemorrhage, and the third one with severe hydrops and good neuromotor development at 6 months of age.

It seems that a new clinical approach started in 1986 made a real difference in the survival of these infants. This protocol emphasizes cardiorespiratory and hemodynamic stabilization, as well as anemic correction by partial EXT with PRBC, before the conventional approach to the Rh hemolytic disease.

CONGRESSO INTERNACIONAL MULTIPROFISSIONAL EM

PEDIATRIA E CARDIOLOGIA

JUNTOS CONTRA A COVID-19 .ONLINE

CERTIFICADO

[redacted]

Gisele Correia Pacheco Leite
Gisele Correia Pacheco Leite
Presidente do CIMPEC

Cybelle Dutra da Silva
Cybelle Dutra da Silva
Vice-presidente do CIMPEC

Kátia Correia Lima
Kátia Correia Lima
Presidente da SOPERN

Maria Sanali M de Oliveira Paiva
Maria Sanali M de Oliveira Paiva
Presidente da SBC-RN

UFRN UNIVERSIDADE FEDERAL DO RIO GRANDE DO NORTE

HUOL Hospital Universitário Onofre Lopes

CARDIOPEDIATRIA UNIVERSIDADE FEDERAL DO RIO GRANDE DO NORTE

SOCIEDADE DE PEDIATRIA DO RIO GRANDE DO NORTE

SBC-RN SOCIEDADE BRASILEIRA DE CARDIOLOGIA - RIO GRANDE DO NORTE -

SOCIEDADE BRASILEIRA DE CARDIOLOGIA SBC 1943

CONGRESSO INTERNACIONAL MULTIPROFISSIONAL EM

PEDIATRIA E CARDIOLOGIA

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CONGRESSO INTERNACIONAL MULTIPROFISSIONAL EM

PEDIATRIA E CARDIOLOGIA

JUNTOS CONTRA A COVID-19 .ONLINE

CERTIFICADO

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UFRN UNIVERSIDADE FEDERAL DO RIO GRANDE DO NORTE

HUOL Hospital Universitário Onofre Lopes

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SOCIEDADE DE PEDIATRIA DO RIO GRANDE DO NORTE

SBC-RN SOCIEDADE BRASILEIRA DE CARDIOLOGIA - RIO GRANDE DO NORTE -

SOCIEDADE BRASILEIRA DE CARDIOLOGIA SBC 1943

Universidade Federal de Uberlândia

PROEXC
Pró - reitoria de Extensão e Cultura

Certificado

Certificamos que o trabalho

'Coarctaªo de Aorta Atþica e Complexa em criançà escolar e a importñncia da precisªo do diagnºstico precoce '

de autoria de

LOURDES DE F`TIMA GON˙ALVES GOMES

foi apresentado na Modalidade Comunicaªo Oral, no(s) dia(s) 19/09/2018, como parte das atividades do(a) **XXV Semana Cientfica do Curso de Medicina da UFU: Caminhos do Egresso**, promovido(a) pelo(a) Faculdade de Medicina (FAMED) da Universidade Federal de Uberlñndia, realizado(a) no perðdo de 17/09/2018 a 19/09/2018, sob a coordenaªo do(a) BEN HUR BRAGA TALIBERTI.

Prof. Dr. HØlder Eterno da Silveira
PrÁReitor de Extensªo e Cultura

Uberlñndia (MG), 21 de Novembro de 2018.

XXV Semana Científica do Curso de Medicina da UFU: Caminhos do Egresso

PROPOSTA

A XXV Semana Científica da Medicina - UFU é um evento cujo objetivo é difundir e construir, de maneira crítica, conhecimento em diferentes áreas e, dessa maneira, realizar a associação da investigação científica com a prática clínico-cirúrgica. Contando com a presença de palestrantes e convidados do cenário médico nacional e internacional, o evento busca transmitir saberes provindos de experiências e vivências dos específicos e diferentes campos profissionais envolvidos. O evento, além de palestras, conta com minicursos de aperfeiçoamento, mesas redondas, apresentações culturais e espaços para diálogos e networking com profissionais de referência nas esferas médicas. Ainda, a Semana Científica da Medicina, de maneira geral, atua como mediadora na construção da consciência a respeito de variados temas, ao fornecer informações valiosas aos participantes, auxiliando concomitantemente na escolha de um rumo dentro das possibilidades de carreiras profissionais após o egresso da graduação.

OBJETIVO GERAL

Preparar o estudante e o próprio médico para os diferentes mercados e áreas de atuação oferecidos na área de saúde.

OBJETIVOS ESPECÍFICOS

Introduzir o estudante e médico à carreira de pesquisa em saúde; Introduzir o estudante e médico à carreira de docência universitária; Introduzir o estudante e médico à carreira de desenvolvimento de drogas; Introduzir o estudante e médico à carreira militar; Introduzir o estudante e médico à prática de serviços humanitários; Introduzir o estudante e médico às possibilidades de estágios internacionais; Introduzir o estudante e médico a técnicas de planejamento financeiro; Introduzir o estudante e médico à Medicina de Saúde Complementar; Introduzir o estudante e médico à carreira empreendedora na área da saúde; Destacar pontos importantes sobre a escolha da residência médica; Discursar sobre o dilema do estudante e recém formado: clínica ou cirurgia; Destacar pontos importantes sobre a saúde mental do médico e estudante de Medicina; Atualizar os médicos e estudantes sobre situações importantes e frequentes na prática médica: depressão, neoplasias, diabetes, hipertensão, IAM e AVC.

PÚBLICO ALMEJADO

Estudantes e profissionais da área da saúde das universidades brasileiras e qualquer um que se interesse pelos temas.

LOCAL DE EXECUÇÃO

Universidade Federal de Uberlândia – Campus Umuarama

Realização:
Faculdade de Medicina

Coordenador:
Prof. Ben Hur Braga Taliberti

Pró-Reitor de Extensão e Cultura:
Prof. Dr. Helder Eterno da Silveira

Diretora de Extensão:
Profª. Drª. Vânia Aparecida Martins Bernardes

UNIVERSIDADE FEDERAL DE UBERLÂNDIA

PROEXC

Universidade Federal de Uberlândia

Pró-Reitoria de Extensão e Cultura

Diretoria de Extensão / Divisão de Registro e Informação de Extensão

Conforme Estatuto e Regimento Geral Universidade Título IV Capítulo I – Seção IV – Art. 138 § 2º

Data: **16/11/2018** Cadastro SIEX/UFU: **17784/18**

Responsável: (Cadastro – Emissão – Registro)

Divisão de Registro e Informação de Extensão

Universidade Federal de Uberlândia

PROEXC
Pró - reitoria de Extensão e Cultura

Certificado

Certificamos que

Loudes de FÆtima Gonâlves Gomes

atuou como Ministrante do(a) Palestra, **Atestados para atividades fîsicas: reconhecimento clînico da crianâ cardiopata**, no(s) dia(s) 11/11/2017, como parte das atividades do(a) I SIMPSIO DE PEDIATRIA ^LAPED-UFU, promovido(a) pelo(a) Faculdade de Medicina (FAMED) da Universidade Federal de Uberlîndia, realizado(a) no perîodo de 10/11/2017 a 11/11/2017, sob a coordenaªo do(a) Cristina Palmer Barros, com carga horÆria de 2 horas.

Prof. Dr. HØlder Eterno da Silveira
PrÂReitor de Extensªo e Cultura

Uberlîndia (MG), 13 de Fevereiro de 2019.

I SIMPÓSIO DE PEDIATRIA – LAPED-UFU

PROPOSTA

A Liga de Pediatria - LAPED do Curso de Medicina da FAMED tem como um de seus objetivos organizar e promover atividades de extensão para a comunidade científica. A I JORNADA DE PEDIATRIA – UFU conta com a participação de professores do Departamento de Pediatria UFU e professores convidados de outras Instituições. Com a realização de mesas redondas e mini-conferências o público terá a oportunidade de atualização e discussão de temas relevantes para a atenção da saúde da criança e adolescente nos campos de promoção da saúde e do diagnóstico, tratamento e prevenção de doenças. A divulgação do evento será realizada nas mídias sociais e através de cartazes afixados no Campus e Unidades de saúde periféricas pelos alunos da LAPED.

OBJETIVO GERAL

Apresentar conhecimentos de atualização em Pediatria de relevância para a promoção da saúde da criança e do adolescente;
Promover a oportunidade de discussão de temas pediátricos entre professores, acadêmicos e profissionais ligados à assistência da criança e do adolescente.

OBJETIVOS ESPECÍFICOS

Atender a necessidade da LAPED-UFU de realizar atividade de extensão acadêmica.

PÚBLICO ALMEJADO

A I Jornada da Liga Acadêmica de Pediatria - UFU tem como público-alvo acadêmicos do curso de Medicina e dos demais Cursos da área da Saúde, bem como docentes, médicos e demais profissionais da saúde.

LOCAL DE EXECUÇÃO

Universidade Federal de Uberlândia – Campus Umuarama - Auditório do bloco 2A

Realização:
Faculdade de Medicina

Coordenadora:
Profª. Cristina Palmer Barros

Pró-Reitor de Extensão e Cultura:
Prof. Dr. Helder Eterno da Silveira

Diretora de Extensão:
Profª. Drª. Vânia Aparecida Martins Bernardes

UNIVERSIDADE FEDERAL DE UBERLÂNDIA

PROEXC Pró-reitoria de Extensão e Cultura

Universidade Federal de Uberlândia
Pró-Reitoria de Extensão e Cultura
Diretoria de Extensão / Divisão de Registro e Informação de Extensão
Conforme Estatuto e Regimento Geral Universidade Título IV Capítulo I – Seção IV – Art. 138 § 2º
Data: **18/10/2018** Cadastro SIEX/UFU: **16403/17**
Responsável: (Cadastro – Emissão – Registro)
Divisão de Registro e Informação de Extensão

Universidade Federal de Uberlândia

PROEXC
Pró - reitoria de Extensão e Cultura

Certificado

Certificamos que

Lourdes de FÆtima Gonâlves Gomes

atuou como Ministrante do(a) Palestra, **Afecíes Cardiolâgicas na Sîndrome e Down**, no(s) dia(s) 31/05/2014, como parte das atividades do(a) Caracterîsticas e Peculiaridades da Sîndrome de Down: uma visªo integrada, promovido(a) pelo(a) Faculdade de Medicina (FAMED) da Universidade Federal de Uberlîndia, realizado(a) no perîodo de 31/05/2014, sob a coordenaªo do(a) Prof.Dr. Carlos Henrique Alves de Rezende, com carga horÆria de 2 horas.

Prof. Dr. HØlder Eterno da Silveira
PrÂReitor de Extensªo e Cultura

Uberlîndia (MG), 14 de Janeiro de 2015.

Características e Peculiaridades da Síndrome de Down: uma visão integrada

PROPOSTA

Acredita-se que a frequência da S. Down na população em geral seja de 1 para cada 600 nascidos vivos. Vários fatores têm interferido no aumento da incidência desta condição, sendo um dos principais a idade materna mais avançada, o que reflete as alterações culturais no papel social da mulher, que agora atrasa a maternidade em prol da construção de uma carreira profissional bem consolidada. Além disso, tem sido observado aumento na sobrevida dos pacientes portadores da Síndrome, devido em grande parte aos avanços no tratamento e manejo das várias condições clínicas associadas. Isso faz com que esta doença, antes de manejo quase exclusivo de pediatras, se torne de responsabilidade de outras especialidades médicas, como a clínica médica e subespecialidades, além da necessidade de apoio das outras áreas não médicas que abarcam o cuidado integrado ao paciente portador da Síndrome. Isso torna a preparação acadêmica de fundamental importância para a oferta de serviços de saúde de alta qualidade para estes pacientes. Como ainda existem falhas em relação ao currículo de vários cursos da área da saúde, eventos como o proposto se tornam de grande valia, tanto para a preparação dos profissionais, quanto para os pacientes que receberão o fruto desta preparação.

OBJETIVO GERAL

Promover um espaço de troca de conhecimentos e de atualização sobre a Síndrome de Down, de maneira que será realizada uma abordagem multifacetada, incluindo várias especialidades médicas, bem como outras especialidades da área da saúde. Tem também o intuito de aproximar o profissional da saúde em geral da realidade de vida do paciente portador da Síndrome, fornecendo informações importantes para o entendimento da doença, bem como do seu manejo na prática clínica.

PÚBLICO ALMEJADO

Acadêmicos dos cursos de Medicina, Enfermagem, Fisioterapia, Fonoaudiologia e demais áreas afins á área da saúde; profissionais médicos em residência de Pediatria e profissionais das demais áreas da saúde em residência multiprofissional; profissionais da área da saúde em geral.

LOCAL DE EXECUÇÃO

Bloco 8C - Campus Umuarama.

Realização:
Faculdade de Medicina

UNIVERSIDADE FEDERAL DE UBERLÂNDIA

Coordenador:
Prof. Dr. Carlos Henrique Alves de Rezende

Pró-Reitora de Extensão, Cultura e Assuntos Estudantis:
Profª. Drª. Dalva Maria De Oliveira Silva

Diretora de Extensão:
Profª. Drª. Glaucia Carvalho Gomes

PROEX

Universidade Federal de Uberlândia

Pró-Reitoria de Extensão, Cultura e Assuntos Estudantis

Diretoria de Extensão / Assessoria de Extensão

Conforme Estatuto e Regimento Geral Universidade Título IV Capítulo I – Seção IV – Art. 138 § 2º

Data: **08/01/2015** Cadastro SIEX/UFU: **11948/14**

Responsável: (Cadastro – Emissão – Registro)

Assessoria de Extensão

P067

CERTIFICADO

PONTUAÇÃO CNA
Protocolo 78988

XXII CONGRESSO BRASILEIRO DE CARDIOLOGIA PEDIÁTRICA
IV CONGRESSO BRASILEIRO DE CIRURGIA CARDIOVASCULAR PEDIÁTRICA
V FÓRUM DE CARDIOPATIAS CONGÊNITAS NO ADULTO
I FÓRUM DE CARDIOLOGIA PEDIÁTRICA INTERVENCIONISTA
28 DE NOVEMBRO A 01 DE DEZEMBRO DE 2012
HOTEL BOURBON | FOZ DO IGUAÇU - PR

Certificamos que

AUGUSTO C O TRIGUEIRO; LOURDES F G GOMES; RANULFO P MATOS; ANA L P MELLO; PALOMA C F D NAPOLI; ERICKA CAVALHEIRO; RALPH B COUTINHO; ELIO V DUARTE; LUCIANA FONSECA; CELIA M C SILVA

Participou do XXII CONGRESSO BRASILEIRO DE CARDIOLOGIA PEDIÁTRICA, IV Congresso Brasileiro de Cirurgia Cardiovascular Pediátrica,V Fórum de Cardiopatias Congênitas no Adulto e I Fórum de Cardiologia Pediátrica Intervencionista, realizados no período de 28 de novembro a 01 de dezembro de 2012 em Foz do Iguaçu - PR.

na qualidade de autores do Pôster: Pericardite constrictiva - Diagnóstico pouco explorado e tardio em pediatria - Relato de dois casos.

Foz do Iguaçu, 01 de dezembro de 2012

REALIZAÇÃO

Sociedade Brasileira De Cirurgia Cardiovascular

DCC/CP
Departamento de Cardiopatias Congênitas e Cardiologia Pediátrica

DCCVPed

PATROCÍNIO

ACTELION

Dr. Nelson Itiro Miyague
Presidente do Congresso Brasileiro de Cardiologia Pediátrica

Dra. Estela Suzana Kleiman Horowitz
Presidente do DCC/CP

Dr. Marcelo Biscegli Jatene
Presidente do DCCVPed

P068

C E R T I F I C A D O

PONTUAÇÃO CNA
Protocolo 78988

XXII CONGRESSO BRASILEIRO DE CARDIOLOGIA PEDIÁTRICA

Certificamos que

LOURDES F G GOMES; RANULFO P MATOS; ANTONIO C MOREIRA; ANA L P MELLO; ELIO V DUARTE; PALOMA C F D NAPOLI; RALPH B COUTINHO; CESAR A ESTEVES; ANTONIO C C CARVALHO; CELIA M C SILVA

Participou do XXII CONGRESSO BRASILEIRO DE CARDIOLOGIA PEDIÁTRICA, IV Congresso Brasileiro de Cirurgia Cardiovascular Pediátrica,V Fórum de Cardiopatias Congênitas no Adulto e I Fórum de Cardiologia Pediátrica Intervencionista, realizados no período de 28 de novembro a 01 de dezembro de 2012 em Foz do Iguaçu - PR.

na qualidade de autores do Pôster: Controle da dose pela técnica produto dose-área aumenta a proteção e segurança em crianças e com doença cardíaca congênita no laboratório de cateterismo

Foz do Iguaçu, 01 de dezembro de 2012

Dr. Nelson Itiro Miyague
Presidente do Congresso Brasileiro de Cardiologia Pediátrica

Dra. Estela Suzana Kleiman Horowitz
Presidente do DCC/CP

Dr. Marcelo Biscegli Jatene
Presidente do DCCVPed

UNIVERSIDADE FEDERAL DE UBERLÂNDIA
Faculdade de Educação Física
Curso de Fisioterapia

CERTIFICADO

Certificamos que **Lourdes Fátima Gonçalves Gomes** ministrou a aula sobre "Cardiopatias congênitas: Abordagem clinica" na III Jornada de Fisioterapia da Universidade Federal de Uberlândia, no dia 18 de novembro de 2011.

Eliane maria de Carvalho

Profa. Dra. Eliane Maria de Carvalho
Comissão Organizadora

Universidade Federal de Uberlândia

PROEXC
Pró - reitoria de Extensão e Cultura

Certificado

Certificamos que

LOURDES GOMES

atuou como Ministrante do(a) Palestra, **Cardiopatias congŒnitas: Abordagem clˆnica**, no(s) dia(s) 18/11/2011, como parte das atividades do(a) III Jornada de Fisioterapia da Universidade Federal de Uberlˆndia, promovido(a) pelo(a) Faculdade de Educa ªo F‰sica e Fisioterapia (FAEFI) da Universidade Federal de Uberlˆndia, realizado(a) no perÍodo de 17/11/2011 a 19/11/2011, sob a coordena ªo do(a) Eliane Maria de Carvalho Silva, com carga horÆria de 2 horas.

Prof. Dr. HØlder Eterno da Silveira
PrÂReitor de Extens ªo e Cultura

Uberlˆndia (MG), 26 de Abril de 2012.

III Jornada de Fisioterapia da Universidade Federal de Uberlândia

PROPOSTA

A III JORNADA DE FISIOTERAPIA será realizada nos dias 17, 18 e 19 de novembro de 2011, no Anfiteatro do CENESP – Campus Rondon, Contaremos com profissionais de diversas áreas de atuação fisioterapêutica. Teremos neste evento apresentação oral de trabalhos pré selecionados e também comissão avaliadora dos trabalhos. A III Jornada de Fisioterapia da Universidade Federal de Uberlândia irá abordar várias áreas da fisioterapia, com profissionais experientes, consagrados na história da Fisioterapia, para que os alunos possam conhecer estes profissionais não apenas por livros e artigos, e também ampliar as oportunidades de inter-relacionamento na troca de conhecimento científico. A III Jornada de Fisioterapia tem por objetivo contribuir para a qualidade da formação acadêmica dos alunos de graduação em Fisioterapia estimulando a formação de profissionais de excelência neste campo de atuação. A possibilidade da realização da jornada, bem como a presença de profissionais externos da Instituição irá proporcionar uma possibilidade de atualização para os acadêmicos de fisioterapia e para os profissionais. A fisioterapia ocupa um importante local de destaque no cenário hospitalar, bem como nas várias faces da Fisioterapia, tanto pela autonomia profissional assegurada pela sua excelente legislação, como pela competência técnica e científica dos seus profissionais.

OBJETIVO GERAL

Apresentar e discutir a atuação da fisioterapia nos diversos níveis de atenção a saúde.

OBJETIVOS ESPECÍFICOS

- Apresentar algumas áreas de atuação da fisioterapia que possam estimular docentes e discentes da Universidade Federal de Uberlândia, bem como a integração de outras Universidades, sejam elas públicas ou privadas.
- Apresentar trabalhos de grande relevância científica nas suas diversas áreas assim como apresentação de importantes inovações da prática fisioterapêutica.
- Discussão de eixos temáticos relativos a assistência, educação, gestão, política e pesquisa em Fisioterapia.

PÚBLICO ALMEJADO

Acadêmicos de Fisioterapia e profissionais da área.

LOCAL DE EXECUÇÃO

Anfiteatro do CENESP - Faculdade de Educação Física.

Realização:
Faculdade de Educação Física - FAEFI

Coordenador (a):
Prof(a). Eliane Maria de Carvalho Silva

Pró-Reitor de Extensão, Cultura e Assuntos Estudantis:
Prof. Dr. Alberto Martins da Costa

Diretora de Extensão:
Profª. Drª. Geni de Araújo Costa

UNIVERSIDADE FEDERAL DE UBERLÂNDIA

PROEX

Universidade Federal de Uberlândia

Pró-Reitoria de Extensão, Cultura e Assuntos Estudantis

Diretoria de Extensão / Assessoria de Extensão

Conforme Estatuto e Regimento Geral Universidade Título IV Capítulo I – Seção IV – Art. 138 § 2º

Data: **25/04/2012** Cadastro SIEX/UFU: **9362/11**

Responsável: (Cadastro – Emissão – Registro)

Assessoria de Extensão

XXX Congresso da Sociedade de Cardiologia do Estado de São Paulo

30 de Abril, 01 e 02 de Maio de 2009 - São Paulo/SP

XXX CONGRESSO DA SOCIEDADE DE CARDIOLOGIA DO ESTADO DE SÃO PAULO

SOCESP

CERTIFICADO

Conferido a LOURDES DE FATIMA GONCALVES GOMES

Por sua participação na qualidade de autor do trabalho: **DOENÇA DE CHAGAS NA INFÂNCIA - ASPECTOS EPIDEMIOLÓGICOS E EVOLUÇÃO CLÍNICA**, apresentado na SESSÃO DE POSTER, realizada no dia **30/04/2009**.

São Paulo, 02 de Maio de 2009

Protocolo CNA 14390/14431

Ari Timerman
Presidente da SOCESP

Fausto Feres
Presidente do XXX Congresso da SOCESP

Raul Dias dos Santos Filho
Coordenador Científico do XXX Congresso da SOCESP

Apoio

NOVARTIS

Realização

SOCESP

SÃO LUIZ

Certificado

Conferimos o presente certificado à

Drª Lourdes de Fátima G. Rodrigues

Como Palestrante no Grupo de Estudos NEOCOR com o tema:

"Transposição de Grandes Vasos"

Hospital e Maternidade São Luiz – Unidade Itaim
Carga horária de 01h

São Paulo, 17 de junho de 2009.

Maria Alice P. L. P. Lisboa
Enfª Serviço de Educação Continuada

Maria Josélia Ribeiro
Enfª Coordenadora NEOCOR

SÃO LUIZ

Certificado

Conferimos o presente certificado à

Dra. Lourdes de Fátima Gonçalves Gomes

Como Palestrante com o Tema

"Interpretação do Eletrocardiograma em Neonatologia"

Hospital e Maternidade São Luiz – Unidade Itaim
Carga horária de 02 horas.
São Paulo, 19 de setembro de 2008.

Maria Alice L.P.L. Lisboa
Enfª Serviço de Educação Continuada

Maria Lúcia A. Pereira Cardoso
Gerente de Enfermagem

IX CONGRESSO NACIONAL DE PEDIATRIA

De 08 a 12/Outubro/2007

Certificado

Certificamos que

Pereira, GR; Cunha, JA; Gomes, LFG; Rocha, A; Cunha, TB; Baraúna, BRD; Cisdeli, FCMM; Nunes, LF; Féo, MFS; Santos, RA; Santana, AM; Roma, CR.

Participaram do **IX Congresso Nacional de Pediatria Região Centro-Oeste**, realizado no período de 08 a 12 de outubro de 2007 no Centro de Convenções de Goiânia – Goiás, na qualidade de **autores** do Pôster: **DOENÇA DE CHAGAS - RARIDADE NA INFÂNCIA?**.

Goiânia, 12 de outubro de 2007

Ivate da Sociedade Brasileira de Pediatria

Presidente da Sociedade Goiana de Pediatria

Presidente do IX Congresso Nacional de Pediatria

sanofi aventis

FQM Farmoquímica

ThermaSkin

IX CONGRESSO NACIONAL DE PEDIATRIA

De 08 a 12/Outubro/2007

Certificado

Certificamos que

Féo, MFS; Gomes, LFG; Cunha, CR; Santos, PC; Baraúna, BD; Cisdeli, FCM; Pereira, VJ; Botelho, R; Silva, CHM; Santos, RA; Santana, AM; Nunes, LF; Roma, CR; Gusmman, RH; Silveira, HL; Pereira, GR; Cunha, JAB.

Participaram do **IX Congresso Nacional de Pediatria Região Centro-Oeste**, realizado no período de 08 a 12 de outubro de 2007 no Centro de Convenções de Goiânia – Goiás, na qualidade de **autores** do Relato de Caso em Pôster: **APRESENTAÇÃO NÃO USUAL DA DOENÇA DE KAWASAKI NA CRIANÇA - RELATO DE CASO.**

Goiânia, 12 de outubro de 2007

Presidente da Sociedade Brasileira de Pediatria

Presidente da Sociedade Goiana de Pediatria

Presidente do IX Congresso Nacional de Pediatria

Realização e Apoio

sanofi aventis

FQM

TheraSkin®

CAPRIS

Certificado

AMPARO MATERNAL

O CAPRIS- Centro de Aprimoramento em Saúde certifica que
Dra. Lourdes de Fátima Gonçalves Gomes
Ministrou aulas no curso de Pós Graduação Lato Sensu em
Fisioterapia Neonatal, no dia 10 de novembro, no Amparo Maternal.

São Paulo, 10 de novembro de 2007

CAPRIS	*Coordenadora do curso*	*Presidente Amparo Maternal*
Ft. Míriam R. Diniz Zanetti	*Ft. Ana Damaris Gonzaga*	*Ir. Lydia Serrachioli Gomes*

XXVII CONGRESSO DA SOCIEDADE DE CARDIOLOGIA DO ESTADO DE SÃO PAULO

XXIII JORNADA DE ENFERMAGEM
XXII SIMPÓSIO DE PSICOLOGIA
XIII SIMPÓSIO DE NUTRIÇÃO
XII SIMPÓSIO DE FISIOTERAPIA
XII SIMPÓSIO DE ODONTOLOGIA
XII SIMPÓSIO DE FARMACOLOGIA
IX SIMPÓSIO DE SERVIÇO SOCIAL
VII SIMPÓSIO DE EDUCAÇÃO FÍSICA E ESPORTE

25 A 27 DE MAIO DE 2006
CAMPOS DO JORDÃO - SP

CERTIFICADO

Conferido a: ***LOURDES DE FATIMA GONCALVES***

por sua participação na qualidade de: ***AUTOR (A)*** *do trabalho:* ***"AVALIAÇÃO DO CARVEDILOL NO TRATAMENTO DE INSUFICIÊNCIA CARDÍACA EM CARDIOPEDIATRIA"****, apresentado na* ***SESSÃO DE TEMAS LIVRES - PRÊMIO MELHOR PESQUISA APLICADA****, do dia* ***26 DE MAIO DE 2.006.***

Campos do Jordão, 27 de Maio de 2006.

Fábio Sândoli de Brito
Presidente - XXVII Congresso

Bráulio Luna Filho
Presidente - SOCESP

Beatriz B. Matsubara
Coordenadora Científica - XXVII Congresso

Fernando Nobre
Diretor Científico - SOCESP

SÃO LUIZ

Centro de Estudos

Conferimos o presente certificado a Dra. Lourdes de Fátima Gonçalves Gomes que ministrou a Palestra:

TRANSPOSIÇÃO DOS GRANDES VASOS
Apresentação de caso clínico

Reunião do Grupo de Estudos de Enfermagem em Cardiologia Neonatal - Hospital e Maternidade São Luiz - Itaim.

São Paulo, 24 de Abril de 2006

Maria Alice P. L. P. Lisboa
Enfa. Educação Continuada

Andréa Tavares Vieira
Enf. Coordenadora

SÃO LUIZ
Centro de Estudos

Certificado

Conferimos o presente certificado para

DRA. LOURDES DE FÁTIMA GONÇALVES GOMES

pela participação como Palestrante na Reunião Científica da Equipe de Neonatologia com o tema: **Reconhecimento clínico das cardiopatias congênitas e abordagem das arritmias no período neonatal**

realizada em 11 de maio de 2005,

no Hospital e Maternidade São Luiz

Dr. João Fernando Monteiro Ferreira
Diretor Executivo do Centro de Estudos

Dr. Luis Carlos B. Ferreira
Coordenador

IX CONGRESSO MINEIRO DE
TERAPIA INTENSIVA
ADULTO - PEDIÁTRICO - NEONATAL
03 A 05 DE NOVEMBRO DE 2005
MINASCENTRO - BELO HORIZONTE - MG
TEMA CENTRAL: Qualidade em medicina intensiva

CERTIFICADO

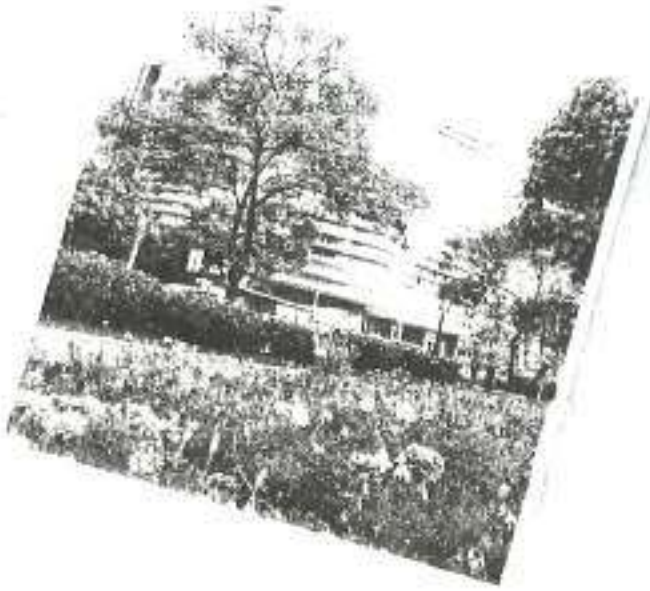
Certificamos que

LOURDES DE FÁTIMA GOMES

participou do **IX Congresso Mineiro de Terapia Intensiva** na qualidade de

PALESTRANTE DO TEMA HEMODINÂMICA NO CURSO:
MONITORIZAÇÃO INVASIVA/ NÃO INVASIVA DO PRÉ- CONGRESSO
DE PEDIATRIA/NEONATAL

Belo Horizonte, 5 de novembro de 2005.



Dra. Maria Aparecida Braga
Presidente da SOMITI e do IX Congresso Mineiro de Terapia Intensiva

Dr. José Carlos Fernandez Versiani dos Anjos
Diretor Científico da SOMITI e do IX Congresso Mineiro de Terapia Intensiva

REALIZAÇÃO SOMITI

APOIO AMMG AMIB

PATROCÍNIO MERCK SHARP & DOHME

XIII Congresso Brasileiro de Ecocardiografia

P33

28 de abril a 1 de maio de 2001 - Hotel Meliá - SP

XIII Congresso Brasileiro de Ecocardiografia
The State of the Art

Certificado

Departamento de Ecocardiografia
SBC

Certificamos que ***GOMES,L.F.G.***
participou do **XIII Congresso Brasileiro de Ecocardiografia,** do Departamento de Ecocardiografia da Sociedade Brasileira de Cardiologia, realizado em São Paulo no Hotel Meliá, de 28 de abril a 1 de maio de 2001, na qualidade de

autor apresentador - POSTER:

"ECOCARDIOGRAFIA COM CONTRASTE COM MICROBOLHAS- INFUSÃO E USO DESTA TÉCNICA NO NEONATO E CRIANÇA."

autores: GOMES,L.F.G.; SILVA,C.M.C.; OPORTO,A.V.L.; ARRUDA,A.L.; MATHIAS,W.; BELO,P.; MATOS,R.P.; MOISES,V.A.; CARVALHO,A.C.C.; PAOLA,A.A.V.; CAMPOS,O.F.

São Paulo, 01 de maio de 2001

Djair Brindeiro Filho
Presidente do Departamento de Ecocardiografia da SBC

Jorge E. Assef
Presidente do XIII Congresso Brasileiro de Ecocardiografia

Carlos Eduardo Suaide Silva
Secretário do XIII Congresso Brasileiro de Ecocardiografia

Presidente:
Jorge Assef
Vice-presidente:
Wilson Mathias Jr.
Secretário:
Carlos Eduardo Suaide Silva
Tesoureiro:
Sérgio Cunha Pontes Jr.

Comissão Científica e Organizadora:
Caio Medeiros
Ana Claudia Petisco
Claudia Gianini Monaco
José Lázaro de Andrade
Mohamed Hassan Saleh
Orlando Campos Filho
Rodrigo B. M. Barretto
Samira M. B. Leal
Valdir Ambrosio Moisés

XIII Congresso Brasileiro de Ecocardiografia

28 de abril a 1 de maio de 2001 - Hotel Meliá - SP

XIII Congresso Brasileiro de Ecocardiografia
The State of the Art

Certificado

Departamento de Ecocardiografia
SBC

Certificamos que ***GOMES,L.F.G.***
participou do **XIII Congresso Brasileiro de Ecocardiografia,** do Departamento de Ecocardiografia da Sociedade Brasileira de Cardiologia, realizado em São Paulo no Hotel Meliá, de 28 de abril a 1 de maio de 2001, na qualidade de
autor apresentador - TEMA LIVRE:

"A IMPORTÂNCIA DOS ACHADOS ECOCARDIOGRÁFICOS NO OTIMO MANEJO DA ATRESIA PULMONAR COM SEPTO VENTRICULAR INTEGRO"
autores: GOMES,L.F.G.; SILVA,C.M.C.; OPORTO,V.L.; ABUJAMRA,P.; PINHEIRO,R.P.M.N; BELO,P.; MOISES,V.A.; CAMPOS,O.F.; CARVALHO,A.C.C.; MALUF,M.; BUFFOLO,E.; LIMA,W.; PAOLA,A.A.V.

São Paulo, 01 de maio de 2001

Djair Brindeiro Filho
Presidente do Departamento de Ecocardiografia da SBC

Jorge E. Assef
Presidente do XIII Congresso Brasileiro de Ecocardiografia

Carlos Eduardo Suaide Silva
Secretário do XIII Congresso Brasileiro de Ecocardiografia

Presidente:
Jorge Assef
Vice-presidente:
Wilson Mathias Jr.
Secretário:
Carlos Eduardo Suaide Silva
Tesoureiro:
Sérgio Cunha Pontes Jr.

Comissão Científica e Organizadora:
Caio Medeiros
Ana Claudia Petisco
Claudia Gianini Monaco
José Lázaro de Andrade
Mohamed Hassan Saleh
Orlando Campos Filho
Rodrigo B. M. Barretto
Samira M. B. Leal
Valdir Ambrósio Moisés

XXI CONGRESSO DA SOCIEDADE DE CARDIOLOGIA DO ESTADO DE SÃO PAULO

XVII Jornada de Enfermagem
XVI Simpósio de Psicologia
VII Simpósio de Nutrição
VI Simpósio de Farmacologia

Campos do Jordão - SP
25, 26 e 27 de maio de 2000

VI Simpósio de Odontologia
VI Simpósio de Fisioterapia
III Simpósio de Serviço Social
I Simpósio de Educação Física e Esporte

"A EVIDÊNCIA DA NECESSIDADE DE PREVENÇÃO"

Certificado

Conferido a: ***LOURDES F. G. GOMES***

por sua participação na qualidade de: ***CO-AUTOR(A)*** *do trabalho:* ***"AVALIAÇÃO FUNCIONAL DO VENTRÍCULO DIREITO NO PÓS-OPERATÓRIO INTERMEDIÁRIO DE CORREÇÃO DE TÉTRADE DE FALLOT, COM RECONSTRUÇÃO DA VALVA PULMONAR"****,* *apresentado na Sessão de Temas Livres Orais,* ***CARDIOLOGIA PEDIÁTRICA II,*** *no dia 27 de Maio de 2000.*

Campos do Jordão, 27 de Maio de 2000.

Álvaro Avezum Júnior
Coordenador Científico

Silvia Helena G. Lage
Diretora Científica da SOCESP

José Antônio Marin-Neto
Presidente do XXI Congresso

Marcelo Chiara Bertolami
Presidente da SOCESP

SOCESP

CLEXANE Enoxaparina Sódica

Aventis

Triatec Ramipril

CERTIFICADO

CONFERIDO A

Lourdes de Fátima Gonçalves Gomes

PELA SUA PARTICIPAÇÃO NO IX CONGRESSO BRASILEIRO DE TERAPIA INTENSIVA E 4º FÓRUM LATINO-AMERICANO DE RESSUSCITAÇÃO REALIZADOS NO MINASCENTRO, NO PERÍODO DE 8 A 12 DE ABRIL DE 2000, NA QUALIDADE DE

Relatora do tema: "Acesso hemodinâmico em emergência" na Mesa Redonda:
"ATENDIMENTO EM UNIDADES DE EMERGÊNCIA"

BELO HORIZONTE, 12 DE ABRIL DE 2000

JOSÉ LUIZ DE AMORIM RATTON
PRESIDENTE DO IX CONGRESSO BRASILEIRO DE TERAPIA INTENSIVA
E 4º FÓRUM LATINO-AMERICANO DE RESSUSCITAÇÃO

MARIA APARECIDA BRAGA
COORDENADORA DA COMISSÃO CIENTÍFICA

BELO HORIZONTE
MINASCENTRO
IX CONGRESSO BRASILEIRO 8 A 12 ABRIL 2000
TERAPIA INTENSIVA
4º FÓRUM
LATINO-AMERICANO
DE RESSUSCITAÇÃO

CERTIFICADO

CONFERIDO A

Lourdes de Fátima Gonçalves Gomes

PELA SUA PARTICIPAÇÃO NO IX CONGRESSO BRASILEIRO DE TERAPIA INTENSIVA E 4° FÓRUM LATINO-AMERICANO DE RESSUSCITAÇÃO REALIZADOS NO MINASCENTRO, NO PERÍODO DE 8 A 12 DE ABRIL DE 2000, NA QUALIDADE DE

Presidente da Conferência: "AVANÇOS NA TERAPIA DA ASMA GRAVE"

BELO HORIZONTE, 12 DE ABRIL DE 2000

JOSÉ LUIZ DE AMORIM RATTON
PRESIDENTE DO IX CONGRESSO BRASILEIRO DE TERAPIA INTENSIVA E 4° FÓRUM LATINO-AMERICANO DE RESSUSCITAÇÃO

MARIA APARECIDA BRAGA
COORDENADORA DA COMISSÃO CIENTÍFICA

BELO HORIZONTE
MINASCENTRO
8 A 12 ABRIL 2000

IX CONGRESSO BRASILEIRO
TERAPIA INTENSIVA
4° FÓRUM
LATINO-AMERICANO
DE RESSUSCITAÇÃO

CERTIFICADO

Certificamos que LOURDES F. G. GOMES participou do XXI Congresso Brasileiro de Hemodinâmica e Cardiologia Intervencionista e IV Jornada Brasileira de Enfermagem em Hemodinâmica e Cardiologia Intervencionista, realizado de 15 a 17 de julho de 1999, no Centro de Convenções do Colégio Marista Santa Maria apresentando o tema "Papel da Perfuração da Valva Pulmonar com Radiofreqüência Seguida da Dilatação por Balão no Tratamento da Atresia Pulmonar com Septo Ventricular Íntegro. Experiência da Unifesp/EPM"

Curitiba - PR. 17 de julho de 1999

Enfa. Veralice Tinôs
Presidente da Jornada

Dr. Ronaldo da Rocha Loures Bueno
Presidente do Congresso

A. Iran G. Cunha

Enfa. Aparecida Irian Guidugli Cunha
Diretora do Dept. De Enfermagem em Hemod. E Cardiol. Intervencionista

CURITIBA XXI

SBHCI

Guerbet

hexabrix

II CONGRESSO
DA SOCIEDADE DE CARDIOLOGIA
DO TRIÂNGULO MINEIRO

Certificado

DRA. LOURDES FÁTIMA G. GOMES

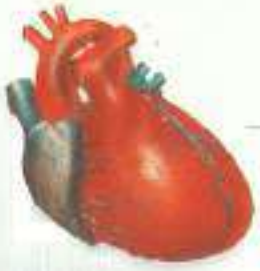
Por sua participação no

II Congresso da Sociedade de Cardiologia do Triângulo Mineiro,

na cidade de Uberlândia - MG, de 28 a 30 de outubro de 1.999,

como

EXPOSITORA DO TEMA "CRISES HIPOXÊMICAS NAS CARDIOPATIAS CONGÊNITAS" NA SESSÃO : "COMO TRATAR"



Dr.Sérgio Corrêa Prata
Pres. da Soc. de Cardiologia Do Triângulo Mineiro

Patrocínio:
NOVARTIS

Dr.Aguinaldo Coelho da Silva
Presidente do Congresso

VII CONGRESSO BRASILEIRO DE TERAPIA INTENSIVA PEDIÁTRICA

VII Encontro de Enfermagem

II Encontro de Fisioterapia

III Encontro Multidisciplinar

CERTIFICADO

Certificamos que Menezes, U.P.; Imamura, J.H.; Nakayama, P.; Oliveira, A.; Santos, J.P.C.; Fantozzi, G.V.; Gonçalves, L.F.; Cruz, N.A.; Nussenzeig, P.R.; Rubens, T.C.; Galli, R.A.; Souza, H.S.; Lopes, Jr.E.; Rosenfeld, K.G.W.

participou do VII Congresso Brasileiro de Terapia Intensiva Pediátrica, realizado em Salvador - Bahia, no período de 24 a 27 de maio de 1998, na qualidade de Autor e Co-autores de Poster HEMODIAFILTRAÇÃO VENOVENOSA CONTÍNUA EM 8 PACIENTES PEDIÁTRICOS PORTADORES DE INSUFICIÊNCIA DE MÚLTIPLOS ÓRGÃOS E SISTEMAS: EVOLUÇÃO CLÍNICA E LABORATORIAL

Salvador, 27 de maio de 1998

MFDMFreire

Maria de Fátima D. M. Freire

Presidente do Congresso

Katiací Araújo

Katiací Araújo

Presidente da Comissão Científica

Lincoln Marcelo Silveira Freire

Presidente da Sociedade Brasileira de Pediatria

XLVIII CONGRESSO DA SOCIEDADE BRASILEIRA DE CARDIOLOGIA

20 a 24 de Setembro de 1992
Centro de Convenções de Pernambuco

CERTIFICADO

Confermos a: LOURDES DE FATIMA G.GOMES

pela sua participação no XLVIII CONGRESSO DA SOCIEDADE BRASILEIRA DE CARDIOLOGIA, realizado em Recife, no período de 20 a 24 de Setembro de 1992 na qualidade de: AUTOR

DO(A) TEMA: AVALIACAO DA FUNCAO FAGOCITARIA DE POLIMORFONUCLEARES (PMN) EM PACIENTES COM CARDIOPATAS CONGENITAS

CO-AUTORES:

ANTONIO CARLOS C. CARVALHO
CHLOÉ MUSSATT
YARA JULIANO
CHARLES NASPITZ
MIGUEL MALUF
CATARINA SVIATOTOLKMIRSKY
ENIO BUFFOLLO
SIDNEY CRUZ
EULOGIO E. MARTINEZ

Recife, 24 de Setembro de 1992.

Dario C. Sobral Filho
Secretario Geral

José da Costa Rocha
Presidente Comissão Científica

Efrem de Aguiar Maranhão
Presidente

Apoio:
Ancoran Caltran
Libra Farmacêutica

XLVIII CONGRESSO DA SOCIEDADE BRASILEIRA DE CARDIOLOGIA

20 a 24 de Setembro de 1992
Centro de Convenções de Pernambuco

CERTIFICADO

Confere-se a: LOURDES FATIMA G. GOMES

pela sua participação no XLVIII CONGRESSO DA SOCIEDADE BRASILEIRA DE CARDIOLOGIA, realizado em Recife, no período de 20 a 24 de Setembro de 1992 na qualidade de: AUTOR

DO(A) TEMA: ANALISE QUANTITATIVA DAS POPULACOES DE LINFOCITOS T E B E SUBPOPULACOES DE LINFOCITOS EM CRIANCAS C/ CARDIOPATIAS.

CO-AUTORES:

ANTONIO CARLOS C. CARVALHO
CHLOÉ MUSSATI
YARA JULIANO
CHARLES NASPITZ
MIGUEL MALUF
ENIO BUFFOLLO
SILVIA DAHER
EULOGIO E. MARTINS FILHO
GLAUCIA R. VESPA
VIRGINIA AMALIA C. BELLE

Recife, 24 de Setembro de 1992.

Dário C. Sobral Filho
Secretário Geral

José da Costa Rocha
Presidente Comissão Científica

Efrem de Aguiar Maranhão
Presidente

Apoio:
Ancoron Caltron
Libbs Farmacêutica

CERTIFICADO

CONFERIDO A

Lourdes de Fátima Gonçalves Gomes

PELA SUA PARTICIPAÇÃO NO IX CONGRESSO BRASILEIRO DE TERAPIA INTENSIVA E 4º FÓRUM LATINO-AMERICANO DE RESSUSCITAÇÃO REALIZADOS NO MINASCENTRO, NO PERÍODO DE 8 A 12 DE ABRIL DE 2000, NA QUALIDADE DE

Debatedora do Painel: "PÓS-OPERATÓRIO"

BELO HORIZONTE, 12 DE ABRIL DE 2000

JOSÉ LUIZ DE AMORIM RATTON
PRESIDENTE DO IX CONGRESSO BRASILEIRO DE TERAPIA INTENSIVA
E 4º FÓRUM LATINO-AMERICANO DE RESSUSCITAÇÃO

MARIA APARECIDA BRAGA
COORDENADORA DA COMISSÃO CIENTÍFICA

BELO HORIZONTE
MINASCENTRO
IX CONGRESSO BRASILEIRO **8 A 12 ABRIL 2000**
TERAPIA INTENSIVA
4º FÓRUM
LATINO-AMERICANO
DE RESSUSCITAÇÃO

II CONGRESSO
DA SOCIEDADE DE CARDIOLOGIA
DO TRIÂNGULO MINEIRO

Certificado

DRA. LOURDES FÁTIMA G. GOMES

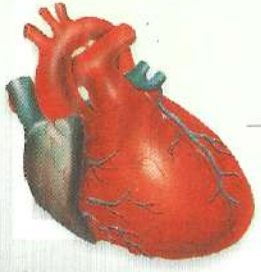
Por sua participação no

II Congresso da Sociedade de Cardiologia do Triângulo Mineiro,

na cidade de Uberlândia - MG, de 28 a 30 de outubro de 1.999,

como

COORDENADORA DO CASO CLÍNICO: "CARDIOPATIA CONGÊNITA"



Dr.Sérgio Corrêa Prata
Pres. da Soc. de Cardiologia Do Triângulo Mineiro

Patrocínio:
NOVARTIS

Dr.Aguinaldo Coelho da Silva
Presidente do Congresso

UNIVERSIDADE FEDERAL DE UBERLÂNDIA
Diretoria da Faculdade de Medicina
Avenida Para, 1720 - Bairro Umuarama, Uberlândia-MG, CEP 38400-902
Telefone: 34 3225-8604 - Bloco 2U - Sala 23

DECLARAÇÃO

Processo nº 23117.062328/2019-26

Interessado: Lourdes de Fátima Gonçalves Gomes

O DIRETOR DA FACULDADE DE MEDICINA E A COORDENADORA DO NÚCLEO DE ENSINO DA FACULDADE DE MEDICINA declaram, para os devidos fins, que Lourdes de Fátima Gonçalves Gomes participou do *Encontro de Desenvolvimento Docente: Metodologias Ativas de Ensino-Aprendizagem*, promovido pela Faculdade de Medicina, no turno da manhã do dia 13 de março de 2019, com uma carga horária total de 4 horas de atividades.

CARLOS HENRIQUE MARTINS DA SILVA
Diretor da Faculdade de Medicina
Portaria nº 1464/17

HELENA BORGES MARTINS DA SILVA PARO
Coordenadora do Núcleo de Ensino
Portaria SEI DIRFAMED Nº 19, de 01 de outubro de 2018

Documento assinado eletronicamente por **Carlos Henrique Martins da Silva**, **Diretor(a)**, em 17/07/2019, às 08:51, conforme horário oficial de Brasília, com fundamento no art. 6º, § 1º, do Decreto nº 8.539, de 8 de outubro de 2015.

Documento assinado eletronicamente por **Helena Borges Martins da Silva Paro**, **Professor(a) do Magistério Superior**, em 17/07/2019, às 19:50, conforme horário oficial de Brasília, com fundamento no art. 6º, § 1º, do Decreto nº 8.539, de 8 de outubro de 2015.

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Referência: Processo nº 23117.062328/2019-26 SEI nº 1397377

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Universidade Federal de Uberlândia

PROEXC
Pró - reitoria de Extensão e Cultura

Certificado

Certificamos que

Lourdes de FÆtima Gonâlves Gomes

participou do(a) **I SIMPSIO DE PEDIATRIA ^LAPED-UFU** promovido(a) pelo(a) Faculdade de Medicina (FAMED) da Universidade Federal de Uberlñndia, realizado(a) no perÒodo de 10/11/2017 a 11/11/2017, sob a coordenaªo do(a) Cristina Palmer Barros, com carga horÆria de 13 horas.

Prof. Dr. HØlder Eterno da Silveira
PrÁReitor de Extensªo e Cultura

Uberlñndia (MG), 13 de Fevereiro de 2019.

I SIMPÓSIO DE PEDIATRIA – LAPED-UFU

PROPOSTA

A Liga de Pediatria - LAPED do Curso de Medicina da FAMED tem como um de seus objetivos organizar e promover atividades de extensão para a comunidade científica. A I JORNADA DE PEDIATRIA – UFU conta com a participação de professores do Departamento de Pediatria UFU e professores convidados de outras Instituições. Com a realização de mesas redondas e mini-conferências o público terá a oportunidade de atualização e discussão de temas relevantes para a atenção da saúde da criança e adolescente nos campos de promoção da saúde e do diagnóstico, tratamento e prevenção de doenças. A divulgação do evento será realizada nas mídias sociais e através de cartazes afixados no Campus e Unidades de saúde periféricas pelos alunos da LAPED.

OBJETIVO GERAL

Apresentar conhecimentos de atualização em Pediatria de relevância para a promoção da saúde da criança e do adolescente;
Promover a oportunidade de discussão de temas pediátricos entre professores, acadêmicos e profissionais ligados à assistência da criança e do adolescente.

OBJETIVOS ESPECÍFICOS

Atender a necessidade da LAPED-UFU de realizar atividade de extensão acadêmica.

PÚBLICO ALMEJADO

A I Jornada da Liga Acadêmica de Pediatria - UFU tem como público-alvo acadêmicos do curso de Medicina e dos demais Cursos da área da Saúde, bem como docentes, médicos e demais profissionais da saúde.

LOCAL DE EXECUÇÃO

Universidade Federal de Uberlândia – Campus Umuarama - Auditório do bloco 2A

Realização:
Faculdade de Medicina

Coordenadora:
Profª. Cristina Palmer Barros

Pró-Reitor de Extensão e Cultura:
Prof. Dr. Helder Eterno da Silveira

Diretora de Extensão:
Profª. Drª. Vânia Aparecida Martins Bernardes

UNIVERSIDADE FEDERAL DE UBERLÂNDIA

PROEXC Pró-reitoria de Extensão e Cultura

Universidade Federal de Uberlândia
Pró-Reitoria de Extensão e Cultura
Diretoria de Extensão / Divisão de Registro e Informação de Extensão
Conforme Estatuto e Regimento Geral Universidade Título IV Capítulo I – Seção IV – Art. 138 § 2º
Data: **18/10/2018** Cadastro SIEX/UFU: **16403/17**
Responsável: (Cadastro – Emissão – Registro)
Divisão de Registro e Informação de Extensão

Universidade Federal de Uberlândia

PROEXC
Pró - reitoria de Extensão e Cultura

Certificado

Certificamos que

Lourdes de FÆtima Gonåalves Gomes

participou do(a) **Saœde Mental: seus caminhos e descaminhos** promovido(a) pelo(a) Faculdade de Medicina (FAMED) da Universidade Federal de Uberlñdia, vinculado ao programa **'PET - Programa de Educa•o Tutorial'**, realizado(a) no perÙodo de 20/06/2017 a 21/06/2017, sob a coordena•o do(a) Carlos Henrique Martins da Silva, com carga horÆria de 4 horas.

Prof. Dr. HØlder Eterno da Silveira
PrÁReitor de Extens•o e Cultura

Uberlñdia (MG), 18 de Agosto de 2017.

Saúde Mental: seus caminhos e descaminhos

PROPOSTA
O assunto de saúde mental foi escolhido de forma a trazer á comunidade acadêmica a discussão desse assunto tão presente em nosso cotidiano, mas negligenciado. A proposta é fazer um levantamento histórico cultural trabalhando os conceitos de sanidade e loucura e como foram construídos na sociedade, como mudaram com o tempo e por qual motivo houve mudanças. Queremos quebrar os estigmas sobre as pessoas, a fim de entender o processo de saúde e doença e abranger também o cuidado. Para entender como cuidar, irá se fazer um levantamento histórico-cultural do manejo com pacientes psiquiátricos, com estímulo ao debate. Haverá espaço para discutir a saúde mental também dentro da universidade. O evento ocorrerá em dois (2) dias úteis das 18h30 às 22h00, em um formato de "short-talks".

OBJETIVO GERAL
• Discutir a saúde mental.

OBJETIVOS ESPECÍFICOS.
• Definir saúde mental, sanidade e loucura;
• Trabalhar o processo histórico de cuidado ao paciente psiquiátrico com olhar crítico;
• Abordar a saúde mental dos estudantes

PÚBLICO ALMEJADO
Aberto a toda comunidade com foco para comunidade acadêmica.

LOCAL DE EXECUÇÃO
Anfiteatro do bloco 8C do Campus Umuarama.

Realização:
Faculdade de Medicina

Coordenador:
Prof. Carlos Henrique Martins Da Silva

Pró-Reitor de Extensão e Cultura:
Prof. Dr. Helder Eterno da Silveira.

Diretora de Extensão:
Profª. Drª. Vânia Aparecida Martins Bernardes

UNIVERSIDADE FEDERAL DE UBERLÂNDIA

PROEX

Universidade Federal de Uberlândia

Pró-Reitoria de Extensão e Cultura

Diretoria de Extensão / Divisão de Registro e Informação de Extensão

Conforme Estatuto e Regimento Geral Universidade Título IV Capítulo I – Seção IV – Art. 138 § 2º

Data: **31/07/2017** Cadastro SIEX/UFU: **15434/17**

Responsável: (Cadastro – Emissão – Registro)

Divisão de Registro e Informação de Extensão

XXII CONGRESSO BRASILEIRO DE CARDIOLOGIA PEDIÁTRICA

28 DE NOVEMBRO A 01 DE DEZEMBRO DE 2012
HOTEL BOURBON | FOZ DO IGUAÇU - PR

CERTIFICADO

PONTUAÇÃO CNA
Protocolo 78988

Certificamos que

LOURDES DE FÁTIMA GONÇALVES GOMES

Participou do XXII CONGRESSO BRASILEIRO DE CARDIOLOGIA PEDIÁTRICA, IV Congresso Brasileiro de Cirurgia Cardiovascular Pediátrica, IV Forum de Cardiopatias Congênitas no Adulto e I Forum de Cardiologia Pediátrica Intervencionista, realizados no período de 28 de Novembro a 01 de Dezembro de 2012 no Hotel Bourbon em Foz do Iguaçu - PR, com carga horária de 25 horas

REALIZAÇÃO

Sociedade Brasileira De Cirurgia Cardiovascular

DCC/CP

DCCVPed

PATROCÍNIO

ACTELION

Foz do Iguaçu, 01 de Dezembro de 2012

Dr. Nelson Itiro Miyague
Presidente do Congresso Brasileiro de Cardiologia Pediátrica

Dra. Estela Suzana Kleiman Horowitz
Presidente do DCC/CP

Dr. Marcelo Biscegli Jatene
Presidente do DCCVPed

XXXI
CONGRESSO DA SOCIEDADE DE CARDIOLOGIA DO ESTADO DE SÃO PAULO

SOCESP

XXXI Congresso da Sociedade de Cardiologia do Estado de São Paulo

29,30 de abril e 1º de maio de 2010 - São Paulo/SP

CERTIFICADO

A SOCIEDADE DE CARDIOLOGIA DO ESTADO DE SÃO PAULO – SOCESP CONFERE ESTE CERTIFICADO A

LOURDES DE FATIMA GONCALVES GOMES

POR SUA PARTICIPAÇÃO NO XXXI CONGRESSO DA SOCESP, NA QUALIDADE DE CONGRESSISTA.

CARGA HORÁRIA: 29H00

5438

São Paulo, 1º de maio de 2010

Realização: SOCESP

Apoio: Bayer HealthCare Bayer Schering Pharma

Dr. Luiz Antonio Machado César
Presidente da SOCESP

Dr. Carlos Vicente Serrano Junior
Presidente do XXXI Congresso

Dr. Fernando Nobre
Coordenador Científico do XXXI Congresso

Protocolo CNA 18785 Cardiologia 15 pontos / Cardiologia Pediátrica 10 pontos / Ecocardiografia 10 pontos / Ergometria 10 pontos / Hemodinâmica 10 pontos / Cardiologia Intervencionista 10 pontos

XXI Congresso Brasileiro de Ecocardiografia
I Simpósio do Departamento de Imagem da SBC
Centro de Convenções do Fortaleza - Ceará - Brasil

CERTIFICADO

ID: 13565.

Certificamos que

LOURDES DE FÁTIMA GONÇALVES GOMES

participou do XXI Congresso Brasileiro de Ecocardiografia e I Simpósio do Departamento de Imagem da SBC, realizado no período de 19 a 21 de março de 2009, Fortaleza - Ceará.

Fortaleza, 21 de março de 2009.

Realização:
Departamento de Ecocardiografia SBC

Dra. Márcia de Melo Barbosa
Presidente do DEPECO

Dr. José Sebastião de Abreu
Presidente do XXI Congresso Brasileiro de Ecocardiografia e
I Simpósio do Departamento de Imagem da SBC

CERTIFICADO

SIMPÓSIO SOCESP
Hipertensão Arterial de Difícil Controle
da Teoria à Prática Clínica

05 e 06 de Junho de 2009 - Campos do Jordão/SP

A SOCIEDADE DE CARDIOLOGIA DO ESTADO DE SÃO PAULO – SOCESP, EM PARCERIA COM O DEPARTAMENTO DE HIPERTENSÃO ARTERIAL DA SOCIEDADE BRASILEIRA DE CARDIOLOGIA – DHA SBC - CONFERE ESTE CERTIFICADO A **LOURDES DE FATIMA GONCALVES GOMES** POR SUA PARTICIPAÇÃO NO **"SIMPÓSIO SOCESP – HIPERTENSÃO ARTERIAL DE DIFÍCIL CONTROLE DA TEORIA À PRÁTICA CLÍNICA"**.

1175

Realização

SOCESP

Apoio

DEPARTAMENTO DE HIPERTENSÃO ARTERIAL

Campos do Jordão, 06 de Junho de 2009

CRM / Protocolo 14796 - Especialidade: CARDIOLOGIA 8.0 - CARDIOLOGIA PEDIÁTRICA 2.0 - ECOCARDIOGRAFIA 8.0

Ari Timerman
Presidente da SOCESP

Flavio Borelli
Presidente do Simpósio

XXIX CONGRESSO DA SOCIEDADE DE CARDIOLOGIA DO ESTADO DE SÃO PAULO

01, 02 e 03 de Maio de 2008 - São Paulo/SP

XXIX CONGRESSO DA SOCIEDADE DE CARDIOLOGIA DO ESTADO DE SÃO PAULO

SOCESP

Conferido a: **LOURDES DE FATIMA GONCALVES**

por sua participação na qualidade de: **CONGRESSISTA**

BIOLAB CÁRDIO

Carga Horária: 29 horas

São Paulo, 03 de Maio de 2008.

DR. MOACIR FERNANDES GODOY
Presidente do XXIX Congresso da SOCESP

DR. ARI TIMERMAN
Presidente da SOCESP

DR. VALTER CORREIA DE LIMA
Coordenador Científico do
XXIX Congresso da SOCESP

CNA / Protocolo 10097 / Cardiologia - 15 pontos / Cardiologia Pediátrica - 5 pontos / Ergometria - 10 pontos / Hemodinâmica e cardiologia intervencionista - 20 pontos / Ecocardiografia - 10 pontos www.cne-cap.org.br

CERTIFICADO

ID 9821

XX CONGRESSO BRASILEIRO DE ECOCARDIOGRAFIA

Certificamos que,

LOURDES DE FÁTIMA GONÇALVES GOMES

participou do XX Congresso Brasileiro de Ecocardiografia, realizado no período de 29 a 31 de maio de 2008 no Rio de Janeiro, RJ.

Rio de Janeiro, 31 de maio de 2008.

REALIZAÇÃO:

Departamento de Ecocardiografia SBC

Dra. Márcia de Melo Barbosa
Presidente do DEPECO

Dr. Luciano Herman Juaçaba Belém
Presidente do XX Congresso Brasileiro de Ecocardiografia

XXVII CONGRESSO DA SOCIEDADE DE CARDIOLOGIA DO ESTADO DE SÃO PAULO

25 A 27 DE MAIO DE 2006
CAMPOS DO JORDÃO - SP

CERTIFICADO

Conferido a: ***LOURDES DE FATIMA GONCALVES***

por sua participação na qualidade de: ***CONGRESSISTA***

Campos do Jordão, 27 de Maio de 2006.

Fábio Sândoli de Brito
Presidente - XXVII Congresso

Bráulio Luna Filho
Presidente - SOCESP

Beatriz B. Matsubara
Coordenadora Científica - XXVII Congresso

Fernando Nobre
Diretor Científico - SOCESP

Departamento de Ecocardiografia SBC

PHmed

XVIII Congresso Brasileiro de Ecocardiografia

Certificamos que

LOURDES DE FATIMA GONÇALVES GOMES

participou do **XVIII Congresso Brasileiro de Ecocardiografia**, realizado no período de 28 a 30 de Abril de 2006, em São Paulo - SP.

São Paulo, 30 de Abril de 2006.

Jorge Eduardo Assef
Presidente do Departamento de Ecocardiografia

Benedito Carlos Maciel
Presidente do XVIII Congresso de Ecocardiografia

CERTIFICADO

ID: 539

XXVI CONGRESSO DA SOCIEDADE DE CARDIOLOGIA DO ESTADO DE SÃO PAULO

12 A 14 DE MAIO DE 2005

CAMPOS DO JORDÃO - SP

CERTIFICADO

Conferido a: ***LOURDES GOMES***

por sua participação na qualidade de: ***CONGRESSISTA***

Campos do Jordão, 14 de Maio de 2005.

Ibraim M. F. Pinto
Presidente - XXVI Congresso

Otávio Rizzi Coelho
Presidente - SOCESP

Benedito Carlos Maciel
Coordenador Científico - XXVI Congresso

Rui Fernando Ramos
Diretor Científico - SOCESP

SOCESP

NOVARTIS

COACI-ID 4603/ 2005

1527

IX COPATI

Santos - 14 a 16 de abril de 2005 - Mendes Convention Center

IX Congresso Paulista de Terapia Intensiva

IX Fórum Latino-Americano de Ressuscitação Cárdio-Pulmonar e Emergências

II Fórum Latino-Americano de Neuroemergências - LABIC

Integração Multidisciplinar em UTI

CERTIFICADO

Certificamos que **LOURDES DE FATIMA G. GOMES**

participou *do IX Congresso Paulista de Terapia Intensiva, IX Fórum Latino-Americano de Ressuscitação Cárdio-Pulmonar e Emergências, II Fórum Latino-Americano de Neuroemergências – LABIC*

realizados no período de 14 a 16 de abril de 2005, em Santos - SP

na qualidade de **CONGRESSISTA**.

Santos, 16 de abril de 2005

Valter Nilton Felix
Presidente da Comissão Científica

Armando T. Guastapaglia
Presidente do Congresso

Paulo Antoniazzi
Presidente da SOPATI

IX CONGRESSO MINEIRO DE
TERAPIA INTENSIVA
ADULTO - PEDIÁTRICO - NEONATAL
03 A 05 DE NOVEMBRO DE 2005
MINASCENTRO - BELO HORIZONTE - MG
TEMA CENTRAL: Qualidade em medicina intensiva

CERTIFICADO

Certificamos que

LOURDES DE FÁTIMA GONÇALVES GOMES

participou do **IX Congresso Mineiro de Terapia Intensiva** na qualidade de

CONGRESSISTA

Belo Horizonte, 5 de novembro de 2005.

Dra. Maria Aparecida Braga
Presidente da SOMITI e do IX Congresso Mineiro de Terapia Intensiva

Dr. José Carlos Fernandez Versiani dos Anjos
Diretor Científico da SOMITI e do IX Congresso Mineiro de Terapia Intensiva

REALIZAÇÃO SOMITI

APOIO AMMG AMIB

PATROCÍNIO MERCK SHARP & DOHME

XIII Congresso Brasileiro de Ecocardiografia

215

28 de abril a 1 de maio de 2001 - Hotel Meliá - SP

XIII Congresso Brasileiro de Ecocardiografia
The State of the Art

Certificado

Departamento de Ecocardiografia
SBC

Certificamos que ***LOURDES DE FATIMA G. GOMES*** participou do **XIII Congresso Brasileiro de Ecocardiografia,** do Departamento de Ecocardiografia da Sociedade Brasileira de Cardiologia, realizado em São Paulo no Hotel Meliá, de 28 de abril a 1 de maio de 2001, na qualidade de ***CONGRESSISTA***

São Paulo, 01 de maio de 2001

Djair Brindeiro Filho
Presidente do Departamento de Ecocardiografia da SBC

Jorge E. Assef
Presidente do XIII Congresso Brasileiro de Ecocardiografia

Carlos Eduardo Suaide Silva
Secretário do XIII Congresso Brasileiro de Ecocardiografia

Presidente:
Jorge Assef
Vice-presidente:
Wilson Mathias Jr.
Secretário:
Carlos Eduardo Suaide Silva
Tesoureiro:
Sérgio Cunha Pontes Jr.

Comissão Científica e Organizadora:
Caio Medeiros
Ana Claudia Petisco
Claudia Gianini Monaco
José Lázaro de Andrade
Mohamed Hassan Saleh
Orlando Campos Filho
Rodrigo B. M. Barretto
Samira M. B. Leal
Valdir Ambrosio Moisés

2nd INTERNATIONAL SYMPOSIUM OF THE
BRAZILIAN COCHRANE COLLABORATION
CURSO INTERNACIONAL DE METODOLOGIA
EM PESQUISA CLÍNICA

UNIFESP

UNIVERSIDADE FEDERAL DE SÃO PAULO
ESCOLA PAULISTA DE MEDICINA

CERTIFICADO

Certifico que:

Lourdes de F. G. Gomes

Participou do 2nd International Symposium of the Brazilian Cochrane Collaboration - Curso Internacional de Metodologia em Pesquisa, realizado em 22 de fevereiro de 2001 na Universidade Federal de São Paulo / Escola Paulista de Medicina, teatro Marcus Limdenberg.

Prof. Dr. Álvaro Nagib Atallah
Coordenador

O Curso eqüivale a dois (02) créditos na CPG de Medicina Interna e Terapêutica da UNIFESP/EPM.

XXI CONGRESSO DA SOCIEDADE DE CARDIOLOGIA DO ESTADO DE SÃO PAULO

XVII Jornada de Enfermagem
XVI Simpósio de Psicologia
VII Simpósio de Nutrição
VI Simpósio de Farmacologia

Campos do Jordão - SP
25, 26 e 27 de maio de 2000

VI Simpósio de Odontologia
VI Simpósio de Fisioterapia
III Simpósio de Serviço Social
I Simpósio de Educação Física e Esporte

"A EVIDÊNCIA DA NECESSIDADE DE PREVENÇÃO"

Certificado

Conferido a: ***LOURDES FATIMA GONÇALVES GOMES***
por sua participação na qualidade de: ***CONGRESSISTA***

Campos do Jordão, 27 de Maio de 2000.

Álvaro Avezum Júnior
Coordenador Científico

Silvia Helena G. Lage
Diretora Científica da SOCESP

José Antônio Marin-Neto
Presidente do XXI Congresso

Marcelo Chiara Bertolami
Presidente da SOCESP

SOCESP

CERTIFICADO

CONFERIDO A

Lourdes de Fátima Gonçalves Gomes

PELA SUA PARTICIPAÇÃO NO IX CONGRESSO BRASILEIRO DE TERAPIA INTENSIVA E 4º FÓRUM LATINO-AMERICANO DE RESSUSCITAÇÃO REALIZADOS NO MINASCENTRO, NO PERÍODO DE 8 A 12 DE ABRIL DE 2000, NA QUALIDADE DE

Congressista

BELO HORIZONTE, 12 DE ABRIL DE 2000

JOSÉ LUIZ DE AMORIM RATTON
PRESIDENTE DO IX CONGRESSO BRASILEIRO DE TERAPIA INTENSIVA
E 4º FÓRUM LATINO-AMERICANO DE RESSUSCITAÇÃO

MARIA APARECIDA BRAGA
COORDENADORA DA COMISSÃO CIENTÍFICA

BELO HORIZONTE
MINASCENTRO
IX CONGRESSO BRASILEIRO 8 A 12 ABRIL 2000
TERAPIA INTENSIVA
4º FÓRUM
LATINO-AMERICANO
DE RESSUSCITAÇÃO

SÃO LUIZ

CERTIFICADO

Certificamos para os devidos fins, que

---

participou do

"II ENCONTRO DO GRUPO DE ESTUDOS EM CARDIOLOGIA NEONATAL - NEOCOR"
na qualidade de ouvinte.

TEMAS:

* Reconhecimento do Neonato Portador de Cardiopatias
* Realidades e Perspectivas da Assistência de Enfermagem na PCR em Neonatologia

São Paulo, 24 de outubro de 2000

M. Alice P. L. Ponte Lisboa
Enfa. Ed. Continuada

Juracy M.B. Galvão
Comissão Organizadora

Hospital / Maternidade Rua Dr. Alceu de Campos Rodrigues 95
Pronto Socorro 04544 000 São Paulo SP
Centro de Diagnósticos PABX (11) 3040 1100
Unidade Itaim

XVI Congresso Brasileiro de
CARDIOLOGIA PEDIÁTRICA

12 a 14 de Novembro de 1999
OURO MINAS PALACE HOTEL
Belo Horizonte - MG

REALIZAÇÃO

DEPARTAMENTO DE CARDIOLOGIA PEDIÁTRICA DA SOC. BRASILEIRA DE CARDIOLOGIA

APOIO

Baxter
DIVISÃO MACCHI

CERTIFICADO

Certificamos que

LOURDES DE FÁTIMA GONÇALVES GOMES

participou do XVI Congresso Brasileiro de Cardiologia Pediátrica na qualidade de

CONGRESSISTA e Assistente do Curso "CARDIOLOGIA PARA PEDIATRAS"

Belo Horizonte, 14 de novembro de 1999.

Cleonice de Carvalho Coelho Mota
Presidente do Congresso

Helder Machado Paupério
Presidente da Comissão Científica

XI CONGRESSO BRASILEIRO DE ECOCARDIOGRAFIA

C E R T I F I C A D O

Certificamos que Lourdes de Fátima Gonçalves Gomes
participou do XI CONGRESSO BRASILEIRO DE ECOCARDIOGRAFIA,
realizado no período de 28 de Abril a 01 de Maio de 1999, na qualidade de

CONGRESSISTA

Belo Horizonte, 01 de Maio de 1999

Helder Machado Pauperio
Presidente do Congresso

Márcia de Melo Barbosa
Presidente da Comissão Científica

Álvaro Vilella de Moraes
Presidente do Departamento de
Ecocardiografia do SBC

XII Forum de Enfermagem
XII Forum de Psicologia
IV Forum de Nutrição

Recife
19 a 22 de Setembro de 1999.

CERTIFICADO

LIV Congresso da Sociedade Brasileira de Cardiologia

Certificamos que

LOURDES GOMES

participou do LIV Congresso da Sociedade Brasileira, realizado em Recife/PE, no período de 19 a 22 de setembro de 1999
na qualidade de Congressista

Recife, 22 de setembro de 1999

Dr. Dário C. Sobral Filho
Presidente do
LIV Congresso da SBC

Dr. Hans J. F. Dohmann
Presidente da Comissão
Científica Permanente

XX CONGRESSO DA SOCIEDADE DE CARDIOLOGIA DO ESTADO DE SÃO PAULO

CAMPOS DO JORDÃO
20, 21 e 22 de maio de 1999

XVI JORNADA DE ENFERMAGEM EM CARDIOLOGIA
XV SIMPÓSIO DE PSICOLOGIA EM CARDIOLOGIA
VI SIMPÓSIO DE NUTRIÇÃO EM CARDIOLOGIA
V SIMPÓSIO DE FARMACOLOGIA EM CARDIOLOGIA
V SIMPÓSIO DE ODONTOLOGIA EM CARDIOLOGIA
V SIMPÓSIO DE FISIOTERAPIA EM CARDIOLOGIA
II SIMPÓSIO DE SERVIÇO SOCIAL EM CARDIOLOGIA

Certificado

Conferido a: ***LOURDES DE FATIMA GONCALVES GOMES***
por sua participação na qualidade de: ***CONGRESSISTA***

Campos do Jordão 20, 21 e 22 de Maio de 1999

Francisco Rafael Laurindo
Coordenador Comissão Científica

Antônio Carlos Palandri Chagas
Presidente do XX Congresso

Ari Timerman
Coordenador Científico - SOCESP

Fábio B. Jatene
Presidente da SOCESP

II CONGRESSO
DA SOCIEDADE DE CARDIOLOGIA
DO TRIÂNGULO MINEIRO

Certificado

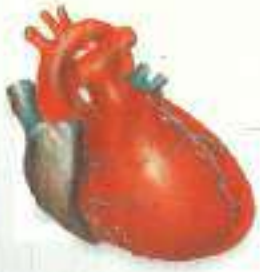
DRA. LOURDES DE FÁTIMA G. GOMES

Por sua participação no

II Congresso da Sociedade de Cardiologia do Triângulo Mineiro,

na cidade de Uberlândia - MG, de 28 a 30 de outubro de 1.999,

como Congressista.



Dr.Sérgio Corrêa Prata
Pres. da Soc. de Cardiologia Do Triângulo Mineiro

Patrocínio:
NOVARTIS

Dr.Aguinaldo Coelho da Silva
Presidente do Congresso

DEPARTAMENTO DE PEDIATRIA

SOCIEDADE MÉDICA DE UBERLÂNDIA

REGIONAL - ALTO PARANAÍBA - SMP

unimed
UBERLÂNDIA

CERTIFICADO

Certificamos que Lourdes de Fátima Gonçalves Gomes

participou I JORNADA DE EMERGÊNCIA EM PEDIATRIA

na qualidade de Congressita

Uberlândia - MG., 07 de outubro de 1994

Eduardo de Ludovade

Departamento de Pediatria
Sociedade Médica de Uberlândia
Regional - Alto Paranaíba - SMP

Orlando Cruantse

Departamento de Pediatria UFU

1.ª JORNADA DE INFECTOLOGIA PEDIÁTRICA DA ESCOLA PAULISTA DE MEDICINA

Certificado

CERTIFICAMOS que LOURDES FATIMA GONÇALVES GOMES

Participou

da **1.ª JORNADA DE INFECTOLOGIA PEDIÁTRICA,** realizada nos dias 06 e 07 de abril de 1990 no Anfiteatro Maria Tereza Nogueira Azevedo da Escola Paulista de Medicina, organizado pela Disciplina de Infectologia Pediátrica – DIPe – do Departamento de Pediatria da EPM.

São Paulo, 07 de abril de 1990

Prof. Dr. Calil Kairalla Farhat
PROF. TITULAR - CHEFE DA DIPe

Prof. Dr. João Tomás de Abreu Carvalhaes
CHEFE DO DEPARTAMENTO DE PEDIATRIA

SERVIÇO PÚBLICO FEDERAL
MINISTÉRIO DA EDUCAÇÃO
UNIVERSIDADE FEDERAL DE UBERLÂNDIA
FACULDADE DE MEDICINA
PROGRAMA DE PÓS-GRADUAÇÃO EM CÊNCIAS DA SAÚDE

DECLARAÇÃO

Declaramos que a **Profa. Dra. Lourdes de Fátima Gonçalves Gomes** desenvolve co-orientação de Mestrado do Programa de Pós-Graduação em Ciências da Saúde - Faculdade de Medicina da Universidade Federal de Uberlândia, conforme quadro abaixo:

ALUNOS	TURMA	SITUAÇÃO	PERÍODO
BRUNO FRANCO ROSSI	MESTRADO PROFISSIONAL/2016	CO-ORIENTADOR	1º e 2º semestre de 2016 1º e 2ºsemestre de 2017 1º semestre de 2018

Por ser verdade firmamos o presente.

Uberlândia, 20 de setembro de 2018.

Prof. Dr. Robinson Sabino da Silva
Coordenador do Programa de Pós-graduação em Ciências da Saúde

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ATA DE DEFESA - PÓS-GRADUAÇÃO

Programa de Pós-Graduação em:	Ciências da Saúde				
Defesa de:	Exame de Qualificação do Mestrado Acadêmico, Nº 08, PPCSA				
Data:	29.09.2022	Hora de início:	16:00	Hora de encerramento:	18:00
Matrícula do Discente:	12012CSD001				
Nome do Discente:	Bruna Zanforlin Jácome				
Título do Trabalho:	TRATAMENTO E EVOLUÇÃO DE CRIANÇAS COM CARDIOMIOPATIA DILATADA ACOMPANHADAS EM UM SERVIÇO PÚBLICO DE CARDIOLOGIA PEDIÁTRICA DE REFERÊNCIA REGIONAL				
Área de concentração:	Ciências da Saúde				
Linha de pesquisa:	2: Diagnóstico, Tratamento e Prognóstico das Doenças e Agravos à Saúde				
Projeto de Pesquisa de vinculação:	EPIDEMIOLOGIA CLÍNICA E DIAGNÓSTICO DAS DOENÇAS DEGENERATIVAS DO APARELHO CARDIOVASCULAR				

Reuniu-se em sala virtual, em web conferência pela plataforma Mconf-RNP, em conformidade com a PORTARIA Nº 36, DE 19 DE MARÇO DE 2020 da COORDENAÇÃO DE APERFEIÇOAMENTO DE PESSOAL DE NÍVEL SUPERIOR - CAPES, a Banca Examinadora, designada pelo Colegiado do Programa de Pós-graduação em Ciências da Saúde, assim composta: Professores Doutores: Lourdes de Fátima Gonçalves Gomes (UFU) e Aguinaldo Coelho da Silva e Elmiro Santos Resende (UFU) orientador(a) do(a) candidato(a).

Iniciando os trabalhos o(a) presidente da mesa, Dr(a). Elmiro Santos Resende, apresentou a Comissão Examinadora e a candidato(a), agradeceu a presença dos membros da banca, e concedeu a Discente a palavra para a exposição do seu trabalho. A duração da apresentação da Discente e o tempo de arguição e resposta foram conforme as normas do Programa.

A seguir o senhor(a) presidente concedeu a palavra, pela ordem sucessivamente, aos(às) examinadores(as), que passaram a arguir o(a) candidato(a). Ultimada a arguição, que se desenvolveu dentro dos termos regimentais, a Banca, em sessão secreta, atribuiu o resultado final, considerando o(a) candidato(a):

Aprovado(a).

Esta defesa faz parte dos requisitos necessários à obtenção do título de Mestre.

O competente diploma será expedido após cumprimento dos demais requisitos, conforme as normas do Programa, a legislação pertinente e a regulamentação interna da UFU.

Nada mais havendo a tratar foram encerrados os trabalhos. Foi lavrada a presente ata que após lida e achada conforme foi assinada pela Banca Examinadora.

Documento assinado eletronicamente por **Elmiro Santos Resende**, **Professor(a) do Magistério**

Superior, em 29/09/2022, às 18:21, conforme horário oficial de Brasília, com fundamento no art. 6º, § 1º, do Decreto nº 8.539, de 8 de outubro de 2015.

Documento assinado eletronicamente por **Aguinaldo Coelho da Silva**, **Usuário Externo**, em 30/09/2022, às 16:40, conforme horário oficial de Brasília, com fundamento no art. 6º, § 1º, do Decreto nº 8.539, de 8 de outubro de 2015.

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Referência: Processo nº 23117.070571/2022-13 SEI nº 3958084

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ATA DE DEFESA - PÓS-GRADUAÇÃO

Programa de Pós-Graduação em:	Ciências da Saúde				
Defesa de:	Dissertação de Mestrado Acadêmico Nº 07/PPCSA				
Data:	03.11.2022	Hora de início:	15:00h	Hora de encerramento:	18:00h
Matrícula do Discente:	12012CSD001				
Nome do Discente:	Bruna Zanforlin Jácome				
Título do Trabalho:	TRATAMENTO E EVOLUÇÃO DE CRIANÇAS COM CARDIOMIOPATIA DILATADA ACOMPANHADAS EM UM SERVIÇO PÚBLICO DE CARDIOLOGIA PEDIÁTRICA DE REFERÊNCIA REGIONAL				
Área de concentração:	Ciências da Saúde				
Linha de pesquisa:	3: Fisiopatologia das doenças e agravos à saúde				
Projeto de Pesquisa de vinculação:	EPIDEMIOLOGIA CLÍNICA E DIAGNÓSTICO DAS DOENÇAS DEGENERATIVAS DO APARELHO CARDIOVASCULAR				

Reuniu-se em web conferência pela plataforma Mconf-RNP, em conformidade com a PORTARIA Nº 36, DE 19 DE MARÇO DE 2020 da COORDENAÇÃO DE APERFEIÇOAMENTO DE PESSOAL DE NÍVEL SUPERIOR - CAPES, pela Universidade Federal de Uberlândia, a Banca Examinadora, designada pelo Colegiado do Programa de Pós-graduação em Ciências da Saúde, assim composta: Professores Doutores: Lourdes de Fátima Gonçalves Gomes (UFU), Claudio Ribeiro da Cunha (ICDF) e Elmiro Santos Resende (UFU) orientador do candidato.

Iniciando os trabalhos o presidente da mesa, Dr. Elmiro Santos Resende, apresentou a Comissão Examinadora e o candidato, agradeceu a presença do público, e concedeu ao Discente a palavra para a exposição do seu trabalho. A duração da apresentação do Discente e o tempo de arguição e resposta foram conforme as normas do Programa.

A seguir o senhor(a) presidente concedeu a palavra, pela ordem sucessivamente, aos(às) examinadores(as), que passaram a arguir o(a) candidato(a). Ultimada a arguição, que se desenvolveu dentro dos termos regimentais, a Banca, em sessão secreta, atribuiu o resultado final, considerando o(a) candidato(a):

Aprovado.

Esta defesa faz parte dos requisitos necessários à obtenção do título de Mestre.

O competente diploma será expedido após cumprimento dos demais requisitos, conforme as normas do Programa, a legislação pertinente e a regulamentação interna da UFU.

Nada mais havendo a tratar foram encerrados os trabalhos. Foi lavrada a presente ata que após lida e achada conforme foi assinada pela Banca Examinadora.

Documento assinado eletronicamente por **Elmiro Santos Resende**, **Professor(a) do Magistério Superior**, em 03/11/2022, às 18:08, conforme horário oficial de Brasília, com fundamento no art. 6º, § 1º, do Decreto nº 8.539, de 8 de outubro de 2015.

Documento assinado eletronicamente por **Lourdes de Fátima Gonçalves Gomes**, **Professor(a) do Magistério Superior**, em 04/11/2022, às 15:14, conforme horário oficial de Brasília, com fundamento no art. 6º, § 1º, do Decreto nº 8.539, de 8 de outubro de 2015.

Documento assinado eletronicamente por **Cláudio Ribeiro da Cunha**, **Usuário Externo**, em 08/11/2022, às 21:45, conforme horário oficial de Brasília, com fundamento no art. 6º, § 1º, do Decreto nº 8.539, de 8 de outubro de 2015.

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Referência: Processo nº 23117.082221/2022-08 SEI nº 4045185

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DECLARAÇÃO

Processo nº 23117.079533/2022-26

Interessado: Membros da banca

Declaramos para os devidos fins que a *Comissão Julgadora* da **Banca de Qualificação** da Defesa da Tese de Doutorado do aluno **Almir Fernando Loureiro Fontes**, do Programa de Pós-Graduação em Ciências da Saúde, intitulada **"Emprego da ecocardiografia de strain na identificação de comprometimento do coração em casos clínicos moderados e graves de COVID-19."*;*** realizada no dia 28 de outubro de 2022, na Faculdade de Medicina da Universidade Federal de Uberlândia, em sala virtual, em web conferência pela plataforma Microsoft Teams, em conformidade com a PORTARIA Nº 36, DE 19 DE MARÇO DE 2020 da COORDENAÇÃO DE APERFEIÇOAMENTO DE PESSOAL DE NÍVEL SUPERIOR - CAPES, foi composta pelos seguintes professores:

TITULARES:

Elmiro Santos Resende (UFU)

Lourdes de Fátima Gonçalves Gomes (UFU)

Aguinaldo Coelho Silva

Suplente:

Fernando César Veloso

Por ser verdade firmamos o presente.

YARA CRISTINA DE PAIVA MAIA
Coordenadora do Programa de Pós-graduação em Ciências da Saúde
Portaria Rnº 3020/2021

Documento assinado eletronicamente por **Yara Cristina de Paiva Maia**, **Coordenador(a)**, em 03/11/2022, às 14:12, conforme horário oficial de Brasília, com fundamento no art. 6º, § 1º, do Decreto nº 8.539, de 8 de outubro de 2015.

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Referência: Processo nº 23117.079533/2022-26 SEI nº 4033662

Criado por viviane.pires, versão 3 por viviane.pires em 27/10/2022 16:09:51.

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ATA DE DEFESA - PÓS-GRADUAÇÃO

Programa de Pós-Graduação em:	Ciências da Saúde				
Defesa de:	Tese de Doutorado Nº 012/PPCSA				
Data:	29.11.2022	Hora de início:	09:00h	Hora de encerramento:	13:00h
Matrícula do Discente:	11813CSD004				
Nome do Discente:	Almir Fernando Loureiro Fontes				
Título do Trabalho:	INJÚRIA MIOCÁRDICA AVALIADA PELA ECOCARDIGRAFIA DE STRAIN EM PACIENTES APÓS COVID-19				
Área de concentração:	Ciências da Saúde				
Linha de pesquisa:	2: DIAGNÓSTICO, TRATAMENTO E PROGNÓSTICO DAS DOENÇAS E AGRAVOS À SAÚDE				
Projeto de Pesquisa de vinculação:	EPIDEMIOLOGIA CLÍNICA E DIAGNÓSTICO DAS DOENÇAS DEGENERATIVAS DO APARELHO CARDIOVASCULAR				

Reuniu-se em web conferência pela plataforma Mconf-RNP, em conformidade com a PORTARIA Nº 36, DE 19 DE MARÇO DE 2020 da COORDENAÇÃO DE APERFEIÇOAMENTO DE PESSOAL DE NÍVEL SUPERIOR - CAPES, pela Universidade Federal de Uberlândia, a Banca Examinadora, designada pelo Colegiado do Programa de Pós-graduação em Ciências da Saúde, assim composta: Profs. Drs. José Maria Peixoto (UNIFENAS), Mohamed Hassan Saleh (Instituto Dante Pazzanese de Cardiologia), Lourdes de Fátima Gonçalves Gomes (UFU), Messias Antônio Araújo e Elmiro Santos Resende (UFU), orientador do candidato.

Iniciando os trabalhos, o presidente da mesa, Prof. Dr. Elmiro Santos Resende, apresentou a Comissão Examinadora e o candidato, agradeceu a presença do público, e concedeu ao Discente a palavra para a exposição do seu trabalho. A duração da apresentação da Discente e o tempo de arguição e resposta foram conforme as normas do Programa.

A seguir o senhor(a) presidente concedeu a palavra, pela ordem sucessivamente, aos(às) examinadores(as), que passaram a arguir o(a) candidato(a). Ultimada a arguição, que se desenvolveu dentro dos termos regimentais, a Banca, em sessão secreta, atribuiu o resultado final, considerando o(a) candidato(a):

Aprovado.

Esta defesa faz parte dos requisitos necessários à obtenção do título de Doutor.

O competente diploma será expedido após cumprimento dos demais requisitos, conforme as normas do Programa, a legislação pertinente e a regulamentação interna da UFU.

Nada mais havendo a tratar foram encerrados os trabalhos. Foi lavrada a presente ata que após lida e achada conforme foi assinada pela Banca Examinadora.

Documento assinado eletronicamente por **Elmiro Santos Resende**, **Professor(a) do Magistério Superior**, em 29/11/2022, às 11:53, conforme horário oficial de Brasília, com fundamento no art. 6º, § 1º, do Decreto nº 8.539, de 8 de outubro de 2015.

Documento assinado eletronicamente por **Lourdes de Fátima Gonçalves Gomes**, **Professor(a) do Magistério Superior**, em 02/12/2022, às 08:47, conforme horário oficial de Brasília, com fundamento no art. 6º, § 1º, do Decreto nº 8.539, de 8 de outubro de 2015.

Documento assinado eletronicamente por **José Maria Peixoto**, **Usuário Externo**, em 02/12/2022, às 13:22, conforme horário oficial de Brasília, com fundamento no art. 6º, § 1º, do Decreto nº 8.539, de 8 de outubro de 2015.

Documento assinado eletronicamente por **Messias Antônio de Araujo**, **Usuário Externo**, em 05/12/2022, às 10:46, conforme horário oficial de Brasília, com fundamento no art. 6º, § 1º, do Decreto nº 8.539, de 8 de outubro de 2015.

Documento assinado eletronicamente por **Mohamed Hassan Saleh**, **Usuário Externo**, em 07/12/2022, às 09:57, conforme horário oficial de Brasília, com fundamento no art. 6º, § 1º, do Decreto nº 8.539, de 8 de outubro de 2015.

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Referência: Processo nº 23117.087957/2022-64 SEI nº 4100801

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Declaração de Revisão Ortográfica

Eu, Luciene Teixeira, professora licenciada em Letras – pela Universidade Federal de Uberlândia, declaro para os devidos fins de direito que fiz a revisão ortográfica do memorial descritivo de Lourdes de Fátima Gonçalves Gomes intitulado, apresentado à Faculdade de Medicina da Universidade Federal de Uberlândia como requisito à promoção do Professor Integrante da Carreira do Ensino Básico, Técnico e Tecnológico do nível IV da Classe Associado IV para Classe Titular.

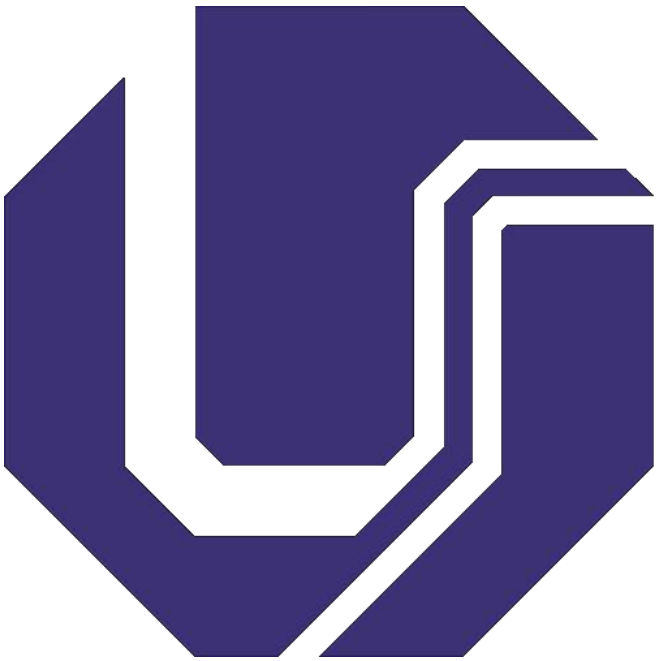
Uberlândia, 20 de março de 2023.

DECLARAÇÃO DE REVISÃO

Declara-se para os devidos fins que o projeto intitulado: **MEMORIAL DESCRITIVO**, de autoria de LOURDES DE FÁTIMA GONÇALVES GOMES, passou por revisão de Língua Portuguesa, sendo o conteúdo de responsabilidade da autora. O texto foi revisado por Ricardo Ondir, portador do RG 21.612.556-2 SSP/SP, formado em Letras pela Faculdade de Filosofia, Letras e Ciências Humanas – FFLCH, com Bacharelado em Língua e Literatura Portuguesa / Língua e Literatura Francesa e Licenciatura em ambas, pela Universidade de São Paulo – USP, que emite tal documento declarando que realizou correções ortográficas e gramaticais de Língua Portuguesa no texto acima indicado.

São Paulo, 27 de março de 2023

RICARDO ONDIR – REVISOR E TRADUTOR



FIM