UNIVERSIDADE FEDERAL DE UBERLÂNDIA

FACULDADE DE ODONTOLOGIA

JÉSSICA BRENDA RODRIGUES DE MEDEIROS

CASES REPORT:

CRI-DU-CHAT SYNDROME

UBERLÂNDIA

2017

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CRI-DU-CHAT SYNDROME

Trabalho de conclusão de curso apresentado à Faculdade de Odontologia da UFU, como requisito parcial para obtenção do título de Graduado em Odontologia.

Orientadora: Profª.Drª. Alessandra Maia de Castro Prado

Coorientadora: Me. Kesia Lara dos Santos Marques

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TRABALHO DE CONCLUSÃO DE CURSO

ATA DA COMISSÃO JULGADORA DA DEFESA DE TRABALHO DE CONCLUSÃO DE CURSO DO (A) DISCENTE **Jéssica Brenda Rodrigues de Medeiros** DA FACULDADE DE ODONTOLOGIA DA UNIVERSIDADE FEDERAL DE UBERLÂNDIA.

No dia **07 de novembro de 2017**, reuniu-se a Comissão Julgadora aprovada pelo Colegiado de Graduação da Faculdade de Odontologia da Universidade Federal de Uberlândia, para o julgamento do Trabalho de Conclusão de Curso apresentado pelo (a) aluno (a) **Jéssica Brenda Rodrigues de Medeiros, COM O TÍTULO: - "CASES REPORT: CRI-DU-CHAT SYNDROME".** O julgamento do trabalho foi realizado em sessão pública compreendendo a exposição, seguida de arguição pelos examinadores. Encerrada a arguição, cada examinador, em sessão secreta, exarou o seu parecer. A Comissão Julgadora, após análise do Trabalho, verificou que o mesmo encontra-se em condições de ser incorporado ao banco de Trabalhos de Conclusão de Curso desta Faculdade. O competente diploma será expedido após cumprimento dos demais requisitos, conforme as normas da Graduação, legislação e regulamentação da UFU. Nada mais havendo a tratar foram encerrados os trabalhos e lavrada a presente ata, que após lida e achada conforme, foi assinada pela Banca Examinadora.

Uberlândia, 07 de novembro de 2017

Profa. Dra. Alessandra Maia de Castro Prado *Universidade Federal de Uberlândia – UFU*	Aprovado/Reprovado
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Renata Pereira Georjutti *Aluno(a) de doutorado – PPGO/UFU*	Aprovado/ Reprovado

Dedico o presente trabalho aos meus pais Jerusalino e Fernanda, meu irmão Jhonatha, meu noivo Júlio, e toda minha família, que com muito carinho e apoio, não mediram esforços para que eu chegasse até esta etapa de minha vida.

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“Aqueles que passam por nós não vão sós. Deixam um pouco de si, levam um pouco de nós.”

Antoine de Saint-Exupery

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Title: Cases Report: Cri-du-Chat Syndrome

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Abstract:

The Cri-du-chat syndrome is an autosomal genetic disorder described by Lejejune in 1963, which involves the partial or total deletion of the chromosome 5 (5p). The syndrome is considered rare, with a prevalence of 1: 15,000 and 1: 50,000, with specific alterations. The aim of this study was to report three clinical cases that involve patients with Cri-du-chat syndrome emphasizing the cranio-skeletal and orofacial characteristics, that may imapair on the dental care. The cranio-skeletal and orofacial alterations in these patients were described and compared on the literature. Pathognomonic characteristics of the Cri-du-chat syndrome were found in the cases described. In one of them a mosaicism karyotype was observed as the cause of deletion chromosome 5. The knowledge of the syndrome’s characteristics, the resultante, systemic and oral problems, allows greater safety when planning and executing the dental treatment.

Keywords: Cri-du-Chat Syndrome, 5p deletion syndrome, Orofacial manifestation.

INTRODUCTION:

Cri-du-chat syndrome (CdCS) is a rare chromosomal anomaly caused by the partial or total deletion of the chromosome 5 short arm, thus the individual presents 46, XX, 5p- (female) and 46, XY, 5p- (male) karyotype. Because of this deletion, the syndrome is also known as Syndrome 5p-.[1–5] The syndrome was first described in 1963 and has an estimated incidence of 1: 15,000 to 1:50,000 live births, affecting more females, in a ratio of 2:1.[2,6–9]

CdCS occurs rarely along with mosaicism (3.7%), where the person has two distinct genetic materials, one is a normal karyotype and the other is cells showing a smaller arm deficiency in the chromosome 5.[10–12]

The syndrome's name refers to the clinical characteristic, described as an acute and distinct monochromatic cry presented by the child at the birth. This cry resembles the cat's meow and is probably related to anomalies in the larynx and epiglottis.[1,2,4,6–8,13,14] The larynx and the epiglottis has its anatomy altered, the first one being small and diamond-shaped, and the second one being small and hypotonic.[4,8,14] This cry is not present in all babies born with SCDC, however is considered a reliable and early indicator for the diagnosis.[2] The diagnosis of the syndrome is based on evident clinical signs and analysis of the karyotype.[4,9,15]

Patients with Cri-du-chat syndrome presents intellectual and psychomotor disorders along with developmental delay and mental retardation.[14,16,17] The phenotypic characteristics observed are low birth weight, microcephaly, round face, spaced eyes, low nasal bridge, low implantation ears and simian fold (only one palmar fold is present).[1–3,6,7,18–20] Through the aging process they begin to present short stature with inadequate weight, small chin, long fingers and muscular hypotonia.[1,1,2,5,18,21,22]

Most individuals diagnosed with SCDC do not develop speech, present difficulty sucking up when adults, and usually have other associated problems such as partial hearing loss, skeletal abnormalities, respiratory and hearing infections, heart diseases, kidney malformation and abdomen muscles separeted.[1,2,17,18,23,24]

The mortality rate is aproximatly of 10%, reaching 75% in the first months of life and 90% in the first year, however, after the first year of life the mortality rate reduced signifincatly.[18,25] However, there are reports of patients with SCDC over 50 years of age.[18,25]

The main orofacial alterations reported are perioral muscle hypotonia, class II occlusion, biprotrusion, micrognathia, anterior open bite, ogival palate, cleft palate, delayed tooth eruption, enamel hypoplasia, dental erosions

caused by gastroesophageal reflux, supernumerary teeth, dental transpositions, bruxism and periodontal diseases.[2,5,16,21,26,27]

Individuals with SCDC often have chronic periodontal disease associated with poor oral hygiene.[15]However, recent study comparing bacteria present in the dental biofilm of people with CdCS and of a control group showed that both had potentially pathogenic bacteria such as *Aggregatibacter actinomycetemcomitans, Porphyromonas gingivales, Prevotella intermedia, Treponema denticola* and *Tanerella forsythia.*[21,27] The amount of *T.* forsythia found in the SCDC group was statistically higher than the amount found in the control group when compared to the differences obtained in the others bacterias.[21,27]

There are few studies reporting the dental management of SCDC. This would be important to provide information that may help to tailor clinical management for care of these patients, thus improving their quality of life.

The aim of this study is to describe the cranio-skeletal and orofacial characteristics found in a case series of three patients with CdCS attending in a specialized dental service. We also compare these alteration with the most common features of the syndrome reported in the literature.

CASES REPORT:

The oral and systemic analysis of three patients diagnosed with Cri-du-chat syndrome were made. These patients are

periodically treated at a Dental Care Specialized Center for patients with disabilities.

Case 1

Case 2

Case 3

Case 1: white female, second daughter of the family nucleus, with non-consanguineous parents, born with 32 weeks by a preterm caesarean birth, weight 1.900 g, height 45 cm. Patient present few crying and significant cyanosis at birth, and was hospital admission for five days, presented difficulty of sucking and swallowing. She was diagnosed with CdCS at 2 years old, karyotype 46 XX, del (5), with hospitalization history of allergic reaction to Gardenal®, and at 3 years old of chicken pox. Cardiac arrhythmia and mild aortic insufficiency were evaluated by means of complementary exams (electrocardiogram and echocardiogram), resulting in the diagnose of the ostium secundum type interatrial communication (ASD). The continuous daily use of Acetylsalicylic acid was prescribed and a periodic follow-up with a geneticist, a cardiologist, a neurologist and a hematologist were performed. In subsequent medical visits the patient presented somatic growth retardation, global hypotonia, apparent hypothyroidism, epicanto, sialorrhea, small hypoplastic lips, psychomotor

retardation, fanned ears, ocular hypertelorism, small feet, and lack of alignment of the toes. The physical activity was recommended once a week, and physiotherapy treatment, when she was 2 years old. The relatives described that the sleep and the rest are regular. For dental treatment, interventions were performed only after prophylactic antibiotic prescrition. During the first dental treatment, at age 3, the patient had generalized active white patch caries lesions and upper incisors mobility due to dental traumatism. A subsequent fall resulted in trauma with avulsion of the permanent maxillary central incisor. The patient has been followed up the specialized dental service, however presenting a high frequency of cavity and difficulty to perform adequate oral hygiene, leading to biofilm accumulation and gingivitis. At age of 12 years old, another trauma occurred resulting in enamel fracture and mobility of the dental element left central permanent incisor. At the age 15 years old, the cardiologist recommended only local anesthesia due to the high risk of respiratory complications, cardiac arrest and neurological sequelae. For this reason all dental treatment was performed at the outpatient level. The patient is currently 23 years old and presents an open bite and micrognathia. She has been collaborating with the dental care and has been submitted to periodontal, restorative and

preventive treatments, followed by periodical dental monitoring every 4 months.

Case 2: white female, second daughter of the family nucleus, with non-consanguineous parents, was born by caesarean, delivery at 36 weeks due to reduced amniotic fluid and umbilical cord circular, birth weight 2.600 g and height 47 cm. She was diagnosed with SCDC at 1-year-old. Presented episodes of pneumonia at 3 months and at 1 year and 8 months old. Puberty was early (menarche at 9 years old) and during a nursing examination reported regular rest and sleep. Hydrotherapy, equine therapy and physiotherapy were performed once a week during the childhood period. Although she presents good understanding, she has difficulty speaking and swallowing. The presence of strabismus, severe scoliosis and difficulty in walking were observed during its growth. Due to the congenital heart disease, ostium secundum interarterial communication, the use of prophylactic antibiotic therapy was indicated through the dental treatment. At the first dental clinical examination, when she was 2 years and 10 months old, was observed diastemas on maxillar and mandibular deciduous anterior teeth, the presence of enamel hypoplasia and cavities. The treatment was performed at outpatient clinic. When she was 10 years old, after orthodontic evaluation and due to the non-cooperative behavior and dental demand observed at the clinical examination, the patient underwent general

anesthesia for multiple extraction of impacted tooth. Micrognathia, mouth breathing and gingivitis, due to inadequate oral hygiene were also observed. At the age of 19 she was submitted to periodontal, restorative and preventive treatments in an outpatient clinic, where she has been followed up periodically for oral health maintenance every 4 months.

Case 3: white female, first daughter of the family nucleus, non-consanguineous parents, born at 24 weeks due to premature eutocite, presented laryngomalacia with no need for surgical correction, weight 1.720 g and height 42 cm, hospitalized for 1 month and 29 days after birth. At 5 years old she was submitted to a bilateral inguinal hernia surgery. The patient was diagnosed with CdCS and mosaicism at age 7. She has been followed up with a geneticist since childhood, performing equine and occupational therapy. She had chicken pox at the age of 8, and had used Risperidone near her period (2mg at night), mineral oil twice a week due to constipation, and also the use of nebulizer to sleep. Physical examination showed scoliosis, kyphosis, physiological lordosis, acuity auditory without alterations, confirmed sinus arrhythmia with electrocardiogram and delayed neuropsychomotor development. She has insufficient chewing and difficulty to verbalize. During the first dental care, made when she was almost 7 years old, was observed anterior open bite and micrognathia,

cavities and face edema in the mandibular region near the first deciduous molar. The treatment was performed at the Dental Care Specialized Center for patients with disabilities, where the patient is followed up until now. At age 11 years old, she fell of her own height and had a tooth trauma with avulsion of the element 11 which was re-implanted immediately. When she was 13 years old, she suffered another fall and had a dental trauma fracturing the element 21. The patient was treated at the emergency and the semi-rigid splinting of the fractured element was made. After one month, the Rx showed oblique external root reabsorption and apical and cervical fistulas in the elements 11 and 21, followed by the indication of their extraction. Patient had collaborative during dental care, determining the treatment performed under general anesthesia. At the age of 16 years old, a new surgical procedure was performed for the extraction of the permanent second and third mandibular molar. As the posterior mandibula was fragilized, was installed a 4-hole titanium plate and a monocortical screws to stabilize the mandibular cortical cortex to prevent fracture. Currently being more collaborative, she allowed to be submit to preventive treatments in an outpatient clinic, where she has been followed up periodically to maintain oral health every 4 months. The patient need received antibiotic prophylaxis before the procedures.

Table 1- Presents the characteristics that can be found in people with Cri-du-Chat syndrome, in which are described in the literature and that was compared with those observed in the patients treated.

TABLE 1 – CHARACTERISTICS OF CRI-DU-CHAT PEOLPLE DESCRIBED IN THE LITERATURE AND THOSE OBSERVED IN THE CASE REPORTS

CHARACTERISTICS FOUND IN LITERATURE IN PEOPLE WITH CRI-DU-CHAT SYNDROME	CASE 1	CASE 2	CASE 3
GENERAL FEATURES			
- LOW BIRTH WEIGHT	+	+	+
- SHORT STATURE AT BIRTH	+	+	+
- CHARACTERISTIC CRY AT BIRTH	–	–	+
- ROUND FACE	+	+	+
- SHORT NECK	+	+	+
- SPACED EYES	+	+	+
- EPICANTHUS	+	+	+
- LOW SET EARS	+	+	+
- ENLARGED NASAL BRIDGE	+	+	+
- FACIAL ASYMMETRY	–	–	–
- SIMIAN CREASE	+	+	+
- MUSCLE HYPOTONIA	+	+	+
- MOTOR DIFFICULTY	+	+	+
- DIFFICULTY OF COMMUNICATION	+	+	+
- CONGENIC CARDIOPATICS	+	+	+

- BREATHING PROBLEMS	+	+	+
DENTAL CHARACTERISTICS			
- TEMPOROMANDIBULAR DISORDERS	not	reported	
- BRUXISM	not	reported	
- DENTAL EROSIONS	not	reported	
- PERIODONTITIS	+	+	+
- GINGIVITIS	+	+	+
- ENAMEL HYPOPLASIA	not	reported	
- DENTAL TRANSPOSITION	not	reported	
- SUPERNUMERARY TEETH	not	reported	
- MANDIBULAR REROGNATISM	+	+	+
- MACROGLOSSIA	+	+	+
- OPEN BITE	+	+	+
- DENTAL TRAUMA	+	+	+

DISCUSSION:

This study provide information on a case serie of 3 patients with CdCS. The medical and dental records were evaluated after authorization of patient's parents through the Informed Consent Form. The three individuals with CdCS were female patients, that, is most frequent, in a 2: 1 ratio.[2,6-9] The general characteristics described in the literature, the short stature and weight at birth, spaced eyes with diminished visual acuity, difficulty of communication, besides respiratory, and cardiovascular problems were observed in the

mentioned cases of this study. The characteristic cry of the syndrome was observed only in one patient of the three who was born with laryngomalacia and was not reported by the other mothers.

The three cases presented, due to the difficulty of oral hygiene, had high prevalence of cavities, and gingival inflammation, which can be explained by the prevalence of *T.forsythia* bacterium.[21,27] Regular dental follow-up is required because these patients are more susceptible to periodontal diseases.[21,27]

Individuals with CdCS diagnosis have motor difficulties, consequently, they are more susceptible of falling, as can be observed in the three cases reported in which the patients suffered dental traumatism, which requires greater attention of their caregivers.

In general, there is a certain difficulty regarding the dental care of CdCS patients due the several medical and developmental problems that are relevant for conducting the treatment. Often in specific cases is necessary to provide center-surgical care under general anesthesia. [28] The knowledge of the patient's medical history is also important, as can be observed in the three patient's cardiac alterations, being necessary the use of antibiotics prior to dental treatment.

The syndrome's characteristics, of the medical problems, as well as its association with dentistry and other areas of

health allow greater safety in the execution of the treatment, and the patient and its family remains calm. These factors collaborate to the improvement of the quality of the care given to the individuals with Cri du Chat Syndrome.

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Anexo

(NORMAS DA REVISTA)

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Cancer-Pain.org [homepage on the internet]. New York: Association of Cancer Online Resources, Inc.; c2000–01 [Cited 2015 May 11]. Available from: http://www.cancer-pain.org/.

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