

UNIVERSIDADE FEDERAL DE UBERLÂNDIA
FACULDADE DE ODONTOLOGIA



ANA LUIZA RODRIGUES RIBEIRO

**Dental approach of a patient with Osteogenesis
Imperfecta Type V: case report**

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Dental approach of a patient with Osteogenesis Imperfecta Type V: case report

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

Orientadora: Prof^a. Dr^a. Danielly Cunha
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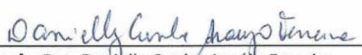


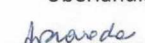
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ATA DA COMISSÃO JULGADORA DA DEFESA DE TRABALHO DE CONCLUSÃO DE CURSO DO (A) DISCENTE **Ana Luíza Rodrigues Ribeiro** DA FACULDADE DE ODONTOLOGIA DA UNIVERSIDADE FEDERAL DE UBERLÂNDIA.

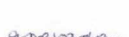
No dia seis de julho de 2017, reuniu-se a Comissão Julgadora aprovada pelo Colegiado de Graduação da Faculdade de Odontologia da Universidade Federal de Uberlândia, para o julgamento do Trabalho de Conclusão de Curso apresentado pelo (a) aluno (a) **Ana Luíza Rodrigues Ribeiro, COM O TÍTULO: "DENTAL APPROACH OF A PATIENT WITH OSTEOPENIA IMPERFECTA TYPE V: CASE REPORT"**. 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.

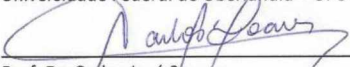
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Title: Dental approach of a patient with Osteogenesis Imperfecta Type V: case report

Short Title: Osteogenesis Imperfecta and approach

Key word: Osteogenesis Imperfecta, dental care, physical restraint

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ABSTRACT

Introduction: The Osteogenesis Imperfecta (OI) is a group of hereditary disorders that occurs due to a deficiency in the production of collagen type I. **Aim:** The aim of this study is to report a case of a male patient, four years of age diagnosed with OI type V and the care that must be taken in the dental care. **Case report:** The mother reported that the child had already suffered several bone fractures and currently was with the use of calcium carbonate, D vitamin and pamidronate. The proposed intervention was control of dental biofilm with professional prophylaxis, due to the uncooperative behavior was employed the active protective stabilization technique and mouth opener. **Conclusion:** It was concluded that due to bone fragility and risk of fractures is important to the careful positioning of the patient in the dental chair, correct use of auxiliary devices and basics and advanced techniques of behavior management.

INTRODUCTION

Osteogenesis imperfecta (OI) is a disease clinically and genetically de group of heritable disorders of connective tissue.¹ It is a rare disease, whose most common inheritance is autosomic dominant, caused by mutations in the collagen type I alpha1 (COL1A1) or collagen type I alpha2 (COL1A2) genes associated with type I collagen metabolism changes.² This change was classified into four main groups according to their clinical characteristics and pattern of genetic inheritance: type I, type II, type III and type IV.³ Subsequently, Glorieux et al (2000)⁴ expanded the classification including the types V, VI and VII. The prevalence of OI is estimated at 1 in 10.000 new births.⁵

The OI offers secondary characteristics important general that characterize the different types of the disease.¹ In addition, this aberration may also display characteristics craniofacial and oral health, such as modification of craniofacial growth, malocclusion of Class III crossbite, anterior and/or posterior and anterior open later⁶ and dentinogenesis imperfecta type I,⁷ which is reported to be higher in deciduous teeth.⁸ However the most striking feature is the bone fragility, with the tendency to fracture from minimal trauma, which leads this change to be known as Brittle Bone Disease.¹

In dental care of children and patients with disabilities the stabilization cases may be used, however observing some aspects first, such as the need of the patient receiving diagnosis and immediate treatment, ensuring the safety of the patient, professional and team.⁹ The bone fragility presented in OI can be considered a risk at the time of implementation of the protective stabilization. Therefore, the objective of this study was to report the clinical case of a patient with OI type V and describe the dental approach regarding the use of protective stabilization.

CASE DESCRIPTION

A male patient with four years of age is monitored with a multidisciplinary team composed by Pediatric Dentistry, Nurse and Nutritionist in the Special Patients, Hospital Dentistry at the Federal University of Uberlândia (SEPAC-UFU) since a month of life, having been held twelve dental consultations for follow-up.

Anamnesis

The mother revealed that there were no complications during pregnancy, but the same reports have been submitted to an ultrasound and radiographic examination, in which were highlighted some intra-uterus in the leg and ribs of the baby,

but without the confirmation of the diagnosis of OI. The child was born at 39 weeks, by means of childbirth cesarean type. The child was born weighing 2,515 g and measuring 45 centimeters.

The seven days of life was the diagnosis of OI type V, where they were found fractures of the two arms, two legs, clavicle and rib, by means of radiographic examination, the child was then immobilized only with a chamois to healing. The two months of life the patient suffered a fracture in his leg during the vaccination. At 10 months of life had a fracture of the femur being needed sedation was required for the limb to be cast, since then the patient has not suffered any bone fracture.

Currently, the patient is under treatment and medical follow-up with a pediatrician and geneticist, makes use of calcium carbonate, 7 ml per day and D vitamin, 5 drops per day, both medications are in use since the eight months of age. In addition, the patient also makes use of intravenous pamidronate a year ago, a type of bisphosphonate used in the treatment of bone mineral density. This medication is administered in a Hospital of Clinics of Federal University of Uberlândia at intervals of four months and children need to be admitted under observation during three days, because it is given in three doses.

Extraoral and Intraoral Clinical examination

The clinical examination extraoral was observed skeletal changes, such as the shape of the face triangular, frontal bone prominent, a cephalic perimeter of 30 cm and absence of bluish sclera (Figure 1). The intraoral clinical examination the child presented the deciduous dentition complete, without change in shape and number of teeth and absence of dentinogenesis imperfecta (DI). Oral hygiene was regular, with the presence of dental biofilm visible, but absence of dental caries (Figure 2).

Dental Approach

The first dental visit happened with a month of age, where information was provided regarding oral hygiene and diet. The patient is monitored at regular dental treatment every four months for the control of dental biofilm. The dental approach is strictly a preventive, acting in control of biofilm, with professional prophylaxis, using Robinson toothbrush and prophylactic folder and use of dental floss. Furthermore, the strengthening and encouraging oral hygiene at home with toothbrush and toothpaste, as well as the use of dental floss (Figure 3).

Throughout the entire monitoring dental, the techniques of managing basic behavioral were used with the objective to familiarize the child with the dental environment. Despite the

use of these practices often the patient still presents a behavior does not associate, being necessary the use of management technique advanced behavioral type active protective stabilization with the mother holding his arms and legs of the child (Figure 4). Thus, the mother received the guidelines about and justifications for the use of this technique, agreeing, in writing in its use. In addition to the active protective stabilization, it was necessary to the careful use of mouth opener for the achievement of professional prophylaxis and the diagnosis of possible dental abnormalities (Figure 5).

DISCUSSION

The OI type V is an extremely rare disease caused by a mutation in the gene IFITM5 and that presents a great phenotypic variability.¹⁰ This gene is responsible for encoding the protein BRIL, which is considered a marker of Mineralizing osteoblasts.¹¹ This study presented a case report of a child diagnosed very early with OI type V, despite its hereditary, there is a report in the family of another person with the same anomaly. OI type V does not present some classic features such as, for example, the bluish sclera.¹

The intrauterine fracture is a finding is extremely rare, but can occur in certain situations such as the OI. The

ultrasound and radiographic examination should be used for the diagnosis of possible fractures of any etiology and the choice of delivery, the biochemical tests and genetic factors may be specific to assist in prenatal diagnosis of OI. The choice of delivery is especially important when there is a risk to the mother or the baby, and the cesarean delivery type more suitable.¹² In the present report was detected intra-uterus, by means of imaging examinations. However, despite the advances in diagnosis of OI, the tests required for the early diagnosis were not performed during the prenatal period, but fortunately, the birth cesarean type was chosen, and the diagnosis occurred only after 7 days of life.

The pharmacological treatment of OI has been performed with the use of bisphosphonates, current evidence shows that this medicine orally or intravenously is able to increase bone mineral density.¹³ The patient reported here makes periodic use of intravenous pamidronate, a year ago, a type of bisphosphonate, for increasing the density. Pamidronate has been effective in the treatment of OI, and your success does not seem to be related to the change in genotype of type I collagen in patients with OI.¹⁴

Regard the oral condition, the patient had no dental alterations or dentinogenesis imperfecta, since this is not a common feature of OI type V.¹ In addition, the clinical

examination also not diagnosed with lesions of caries, which can be explained by the dental care early, before the first year of life, and the child is accompanied by specialized staff, periodically in accordance with the risk assessment with dental caries.

Although the patient receive a follow-up regular dental care, since an age very early and have their behavior managed through basic techniques, it still retains a little behavior collaborator during the dental consultations, even for the achievement of preventive procedures. Therefore, it is necessary to use the protective stabilization activates with an adult by restricting the movement of members. This method is considered to be safe and indicated in patients who need dental care and that for reasons of little development and maturity or lack of ability to understand, they cannot collaborate with the attendant.⁹

However, patients who have a bone fragility exacerbated as at OI, the method may present a high risk of bone fracture, when not performed safely and properly. That is why the participation of parents during the completion of stabilization is indispensable, since they have greater experience and possibly have already received medical information about the best way of positioning and handling of the child in specific situations.

Other arrangements necessary for the implementation of dental care in the abovementioned patient was the opener of mouth. Although this device can be used to enable and maintain the mouth opening, the use of this device in children not collaborator is not considered as protective stabilization.⁹ However, for the use of the device it is necessary to perform the mouth opening of the child, often non-voluntarily, and this procedure in a patient with OI can generate serious disorders such as the risk of occurrence of bone fractures. Therefore, further information must be disseminated in order to prevent the risk of bone fractures during dental care in patients with OI.

CONCLUSIONS

It was concluded that OI presents great bone fragility and fracture risks, so the adequate and safe positioning of the patient in the dental chair, as well as the correct use of auxiliary devices such as mouth openers should be evaluated frequently during the care dental practice. Basic behavioral management techniques should be encouraged because they have satisfactory success rates. On the other hand, advanced behavioral management techniques, such as protective stabilization, can be indicated, but performed in a cautious way with the participation of the family. Thus, they become

important preventive procedures for the maintenance of oral health, avoiding curative care that requires longer intervention time and more invasive procedures.

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Figure legends

Figure 1. **A** and **B** - Skeletal changes, such as the shape of the face triangular, frontal bone prominent, a cephalic perimeter of 30 cm. **C** - Absence of bluish sclera.

Figure 2. **A** - The present the deciduous dentition complete, without change in shape and number of teeth and absence of dentinogenesis imperfecta. **B** - Oral hygiene was regular, with the presence of dental biofilm visible, but absence of dental caries.

Figure 3. **A, B** and **C** - The strengthening and encouraging oral hygiene at home with toothbrush and toothpaste. **D** - Incentive the use of dental floss.

Figure 4. Need to use of management technique advanced behavioral type active protective stabilization with the mother holding his arms and legs of the child.

Figure 5. Need to use of mouth opener for the achievement of professional prophylaxis and the diagnosis of possible dental abnormalities.

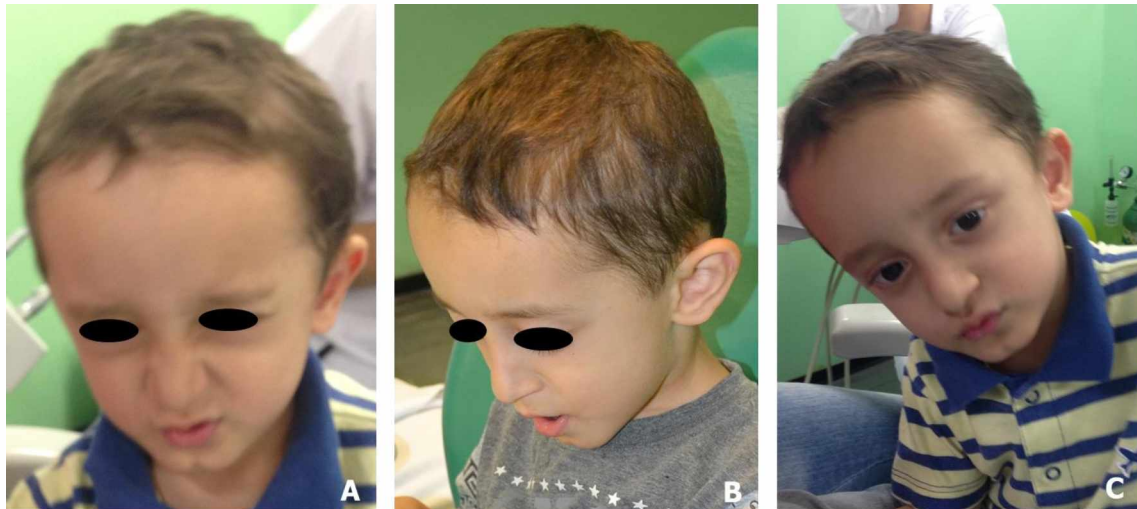
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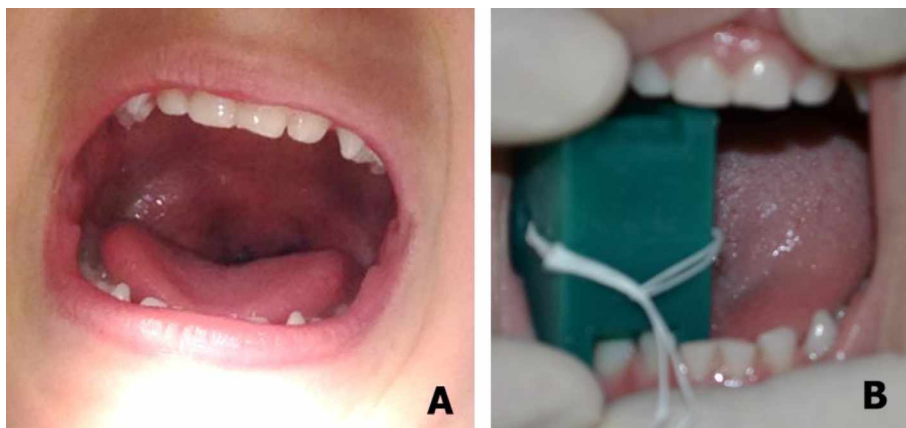
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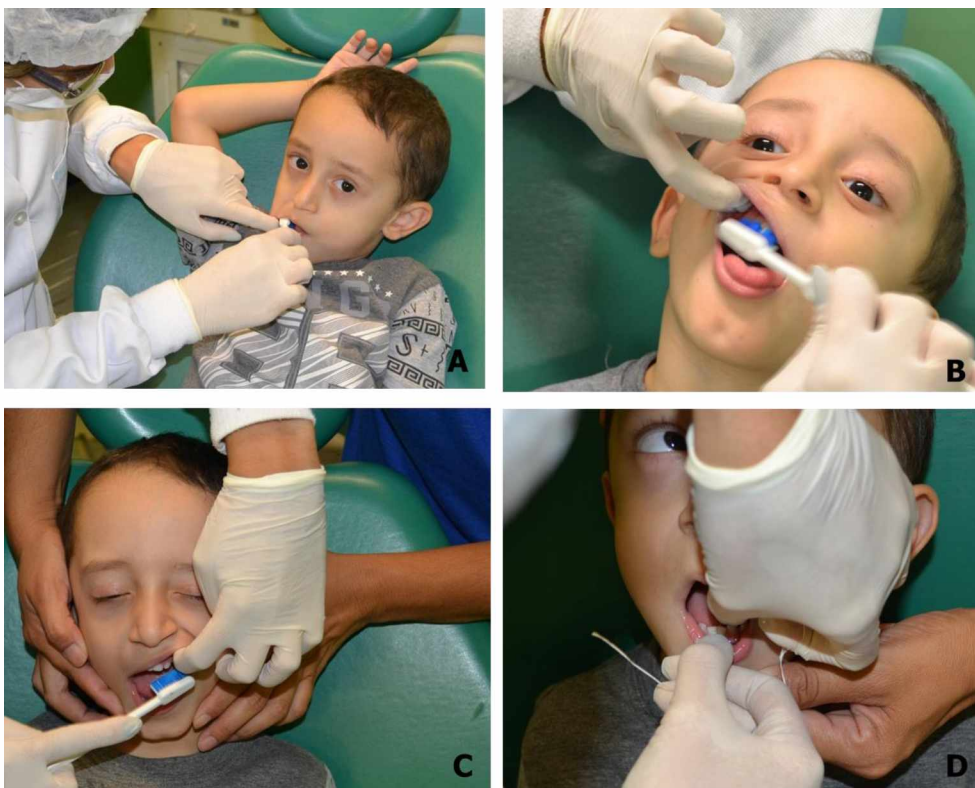
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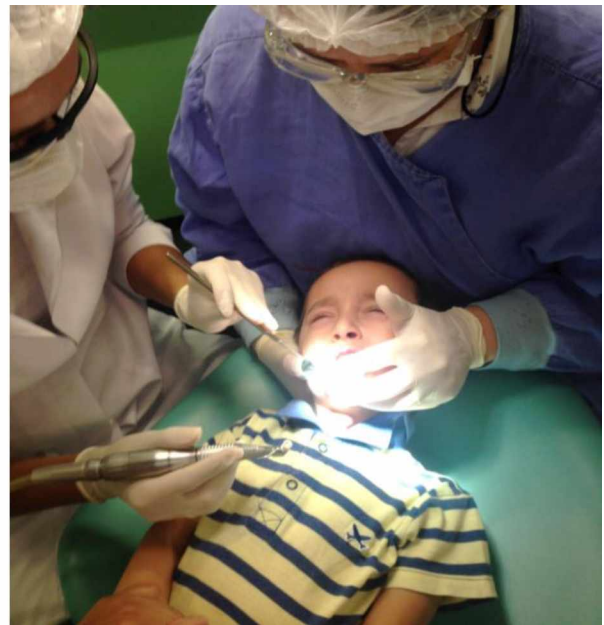
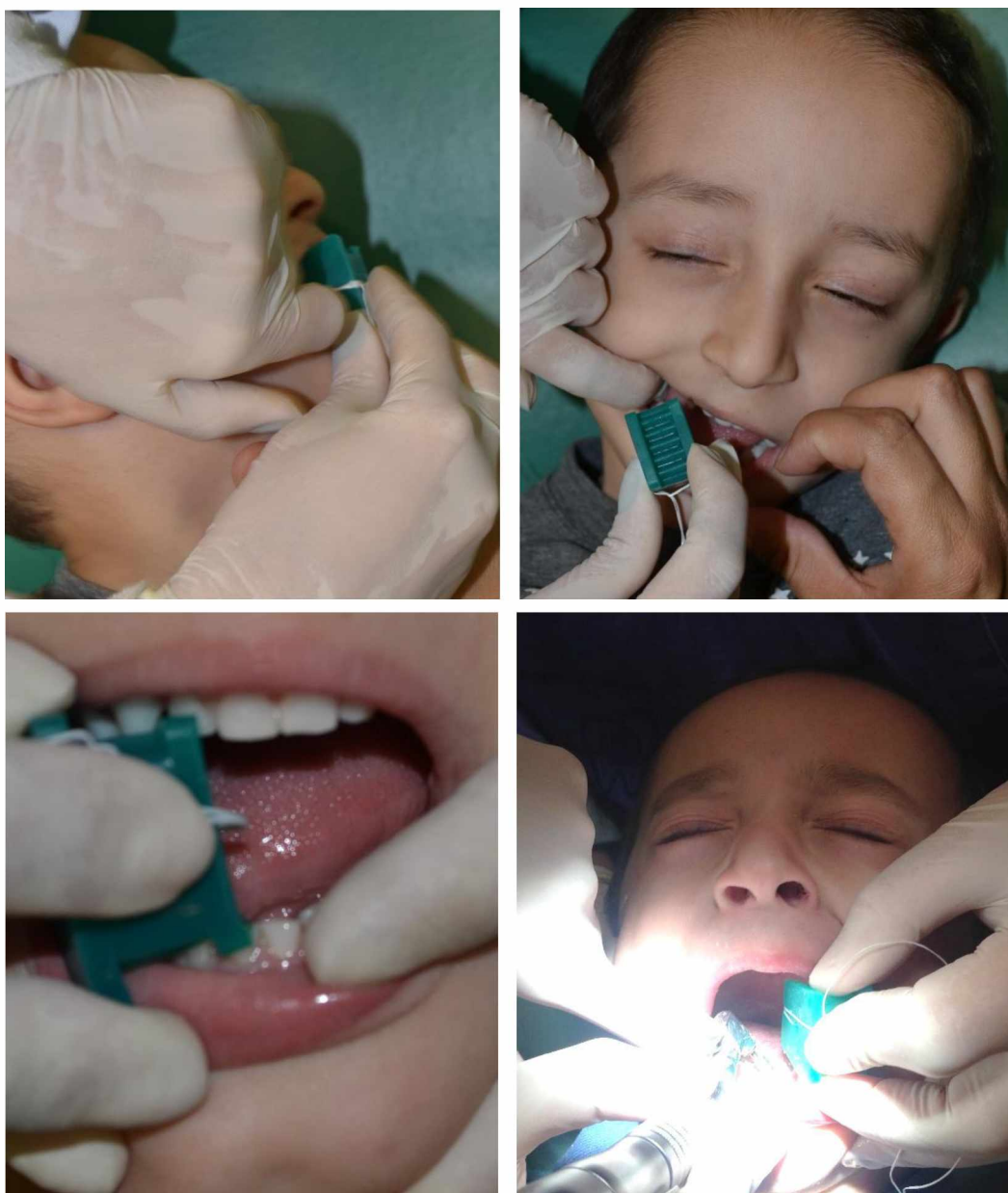


Figure 5



Appendix A

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A critical review of the manuscript topic, the rationale and significance of the study and as appropriate study aims and/or hypotheses should be presented in the introduction.

Methods: All methods used must be detailed, referenced adequately and include a description of the statistical data analysis methods.

Results: Results must be presented in a logical order with references to appropriate tables, figures and illustrations.

Discussion: Important findings from the study should be discussed and compared with the published literature on the topic. Limitations of the study and any future research implications of the study findings must be discussed.

Conclusions: Conclusions should be presented in sentence form and not as a numerical list or dot points. Conclusions should parallel those presented in the structured abstract.

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Questions? Contact: Special Care in Dentistry, 401 N. Michigan Ave., Suite 2200, Chicago, IL 60611. Call 312.527.6764, fax 312.673.6663 or send an e-mail to SCDA@SCDAonline.org.

Appendix B

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SETOR DE PACIENTES ESPECIAIS (SEPÆ)



Autorização

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