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# *Prosthetic rehabilitation of a child with Rubinstein-Taybi Syndrome after dental trauma: case report*

# *Reabilitação protética de criança com Síndrome de Rubinstein Taybi após traumatismo dentário: relato de caso*

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## ABSTRACT

Rubinstein Taybi syndrome has a genetic origin in the chromosome 16. It has physical characteristics, delay in physical and mental development, and may present oral alterations. This paper aims to report a case of prosthetic rehabilitation of a patient with syndrome who, due to dental trauma, lost dental elements early. Female patient, 4 years and 7 months old, with loss of the deciduous maxillary central incisors. The data collection was carried out through anamnesis, clinical and complementary exams. After the diagnosis and the establishment of the treatment plan, we made a Denari type prosthesis. It has a tube-bar mechanism that allows maxillary growth and development. At the end of treatment, the caregiver was instructed to make periodic visits to the dentist for clinical and radiographic follow-up of the prosthesis, as well as the need for treatment with the speech therapist. The loss of anterior deciduous teeth can be associated with trauma in this region and the dental absence can affect the development and behavior of children, promoting a change in daily life and impacting the quality of life of the whole family. The Denari prosthesis is a viable treatment option for children with this syndrome, as it accompanies the maxillary growth, restores the functions of the stomatognathic system, prevents deleterious oral habits and helps with phonetics. It is necessary for the dentist to be able to offer the best treatment for these patients, as the syndrome makes it difficult, but does not impede its execution.

**Indexing terms**: Mouth Rehabilitation. Pediatric Dentistry. Syndrome.

## RESUMO

*A Síndrome de Rubinstein Taybi tem origem genética no cromossomo 16. O paciente com essas condições apresenta características físicas, atraso no desenvolvimento físico e mental podendo apresentar alterações bucais. O presente trabalho tem o objetivo de relatar um caso de reabilitação protética de um paciente com a síndrome que, devido a traumatismo dentário, perdeu precocemente elementos dentários. Paciente do sexo feminino, 4 anos e 7 meses de idade, com ausência dos incisivos centrais superiores decíduos. A coleta de dados foi realizada por meio de anamnese, exames clínico e complementares. Após o diagnóstico e estabelecimento do plano de tratamento, foi confeccionada uma prótese do tipo Denari. A mesma possui um mecanismo tubo barra que permite o crescimento e o desenvolvimento maxilar. Ao término do tratamento, a responsável foi orientada a*

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*fazer visitas periódicas ao cirurgião-dentista para acompanhamento clínico e radiográfico da prótese, bem como a necessidade do tratamento com o fonoaudiólogo. A perda de dentes decíduos anteriores pode estar associada a trauma nessa região e a ausência dentária pode afetar o desenvolvimento e o comportamento das crianças promovendo uma mudança no cotidiano e impactando a qualidade de vida de toda a família. A prótese Denari é uma opção viável de tratamento, para crianças com essa síndrome, pois não impede o crescimento maxilar, restitui as funções do sistema estomatognático, previne hábitos bucais deletérios e auxilia na fonética. É necessário que o dentista esteja capacitado para oferecer o melhor tratamento para esses pacientes, pois a síndrome dificulta, mas não impede a execução.*

***Termos de indexação***: *Reabilitação Bucal. Odontopediatria. Síndromes.*

## INTRODUCTION

Rubinstein-Taybi Syndrome (RTS) is caused by genetic conditions, and its origin is on chromosome 16 due to a mutation, translocation or chromosomal inversion [1]. Its incidence is from 1:100,000 to 300,000 births [2]. Patients with this syndrome have a characteristic face, small head, pulmonary stenosis, enlarged foramen magnum, vertebral abnormalities, thick or curved eyebrows [3,4].

Children with RTS may also have delays in physical and mental development. As for the behavior, it can be friendly and joyful [5]. People with RTS will not necessarily have all the characteristics of the syndrome, but a combination of them [6]. They may have feeding difficulties, respiratory infections, eye problems, heart anomalies, spinal and kidney abnormalities, in addition to orthopedic problems [7].

Oro-dental characteristics are significantly underreported [8]. There are reports of ogival palate, retromicrognathia, small mouth opening and bifid uvula. In rare cases, the individual has a cleft lip and palate. Most have malocclusion. Changes in dental development such as enamel hypoplasia, Hutchinson's teeth and talon cusp are described [8,9].

Dental trauma is considered a public health problem that entails high costs and consequences both for oral health and for the quality of life and well-being of the patient. The highest prevalence of dental trauma takes place in children and adolescents [10]. About 38.59% of children with special needs have dental fractures [11], and syndromic children have physical and mental conditions that may increase the rate of dental fractures due to decreased reflexes [11]. Malocclusions are oral manifestations present in syndromic patients, and this oral condition leads to a higher prevalence of dental trauma in this population [11]. The upper teeth and adjacent soft tissues, especially the upper central and lateral incisors, both deciduous and permanent, are the most affected elements [12]. Limitations in chewing and speaking, changes in the eruptive and occlusal pattern, development of deleterious oral habits, and relationship difficulties – due to embarrassment in showing teeth – are consequences of dental trauma [13].

Due to the scarcity in the literature relating RTS to the dental approach and the difficulty of caring for these patients, it becomes relevant to discuss this issue by highlighting the clinical procedures for outpatient care. The objective of this work is to inform about RTS by describing the technique of a prosthetic rehabilitation, using the Denari's prosthesis in a pediatric patient with the syndrome.

## CASE REPORT

This clinical case report was approved by the Research Ethics Committee of the Southwest Bahia State University, under opinion nº 3.560.150. The free and informed consent form was signed by the child's guardian. The report describes the treatment of a female child, 4 years and 7 months old, leucoderma who attended the pediatric dentistry clinic of the Southwest Bahia State University accompanied by her mother. The mother reported that the child had suffered dental trauma in the first year of life that resulted in root fractures of the upper central deciduous incisors. At the time, she sought dental care, and then the extractions were performed. The reason for seeking care was the desire to replace the teeth that had been lost, because she was aware of the impact it had on the child's socialization and speech.

During the anamnesis, the mother informed that her daughter is a carrier of RTS and showed medical reports that attested to this condition. She reported that no other family member had this syndrome. Among the characteristics of the syndrome, the child had a deficit between chronological and mental age, verified through a neuropsychological evaluation report, difficulty in the cognitive area, showing a developmental age of less than 2 years, and also did not reveal motor planning compatible with her age, besides reduced concentration time and verbal difficulty. In the systemic aspect, the child had Marcus Gun's ophthalmological alteration, mild facial and limb dysmorphisms. It was also reported that they were assisted by a multidisciplinary team composed of a pediatrician, geneticist, neuropediatrician, speech therapist, psychopedagogue, occupational therapist and psychologist. The child regularly attended school, did ballet, judo and therapeutic horseback riding as measures to stimulate motor and cognitive development.

At the time of the consultation, it was reported that the child was undergoing homeopathic medical treatment, did not use allopathic medication, did not have allergies and had a varied and balanced diet. Brushing was performed by her mother, since the eruption of the first teeth, three times a day with fluoridated toothpaste containing 1,100 ppm of fluorine, but there was some resistance on the part of the child, and dental floss was used.

During the clinical examination, the absence of elements 51 and 61 was confirmed, besides the fact that the other teeth were healthy and without visible biofilm. The occlusion evaluation showed a slight overjet, limited mouth opening, absence of TMJ crepitation and mandibular deviation. It was not possible to take the panoramic radiography due to the child's lack of cooperation. Therefore, the occlusal technique was conducted with adult film (AGFA Dentus E-Speed, Kulzer, São Paulo, SP, Brazil), which suggested an image of the germ of teeth 11 and 21 in Nolla stage 5 (figure 1). Due to the lack of cooperation on the part of the child, before starting the clinical examination, the mother was informed about behavior management techniques in pediatric dentistry and advised on the need to perform active protective stabilization to ensure the care and treatment of the patient. The clinical examination initiated only after written consent.

After diagnosis, the treatment chosen was the esthetic and functional rehabilitation of elements 51 and 61 with the Denari's prosthesis, since this type expands and follows the maxillary growth due to the presence of a "male and female" fitting slider (socket) between the central incisors. At the end of the service, the guardian and the child received oral hygiene instruction and dietary guidance.


**Figure 1**. Occlusal radiography showing the germ of teeth 11 and 21.

In the following session, intra-oral photographs were taken (figure 2). For shade taking, the color scale (VITA classical, Wilcos, Petrópolis, RJ, Brazil) was employed, obtaining the most adequate color B1 (figure 3A), and then the selection of the tray was made. The casting was performed with dense addition silicone (AdisilPutty Soft-Coltene, Rio de Janeiro, RJ, Brazil) of the upper and lower arches (figure 3B). After impression taking, the next procedure was casting with type IV plaster (GC Fujirock, Labaski-ku, Tokyo, Japan), in order to obtain the master cast (figure 3C).


**Figure 2**. Initial intra-oral photographs. Frontal (2A) and occlusal (2B) view.

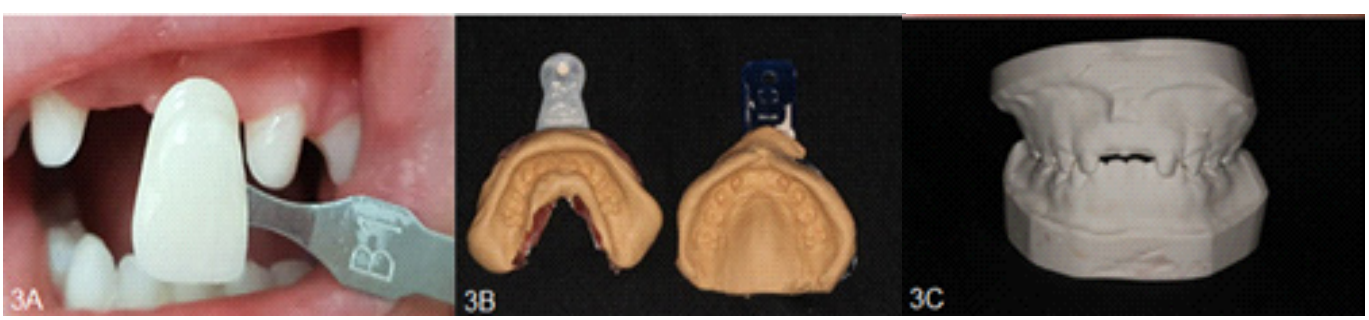


**Figure 3**. Color selection for Denari's prosthesis (3A). Casting with addition silicone (3B). Plasher model of upper and lower arches (3C).

The Denari's prosthesis was made of acrylic resin containing a mechanism of connectors in a tube-bar system (figure 4). Teeth 52 and 62 were the pillars for the prosthesis. These elements were completely covered, in the vestibular and palatal portions, by acrylic resin, thus avoiding possible wear.

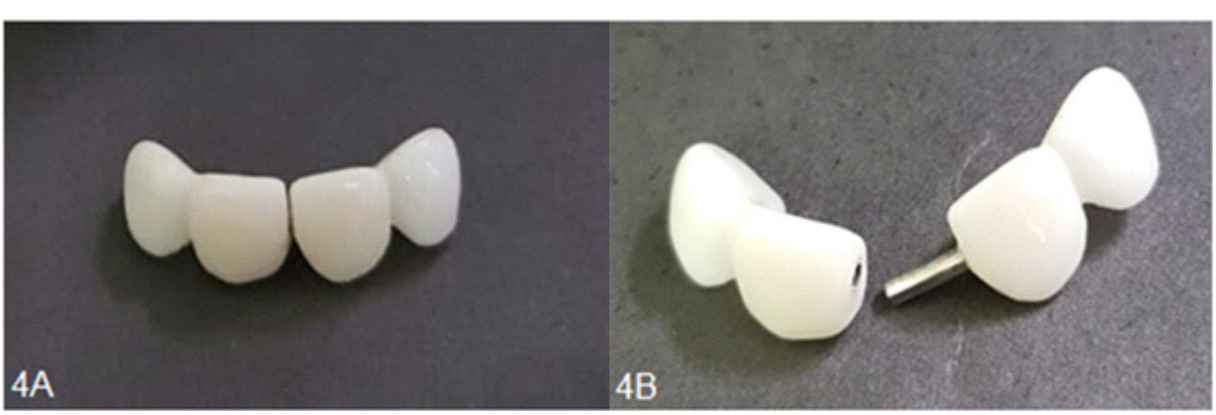


**Figure 4**. (4A) Denari's prosthesis with adapted 'male-female' socket (4B) and view of the 'male-female'socket.

For the installation of the prosthesis, initially, a prophylaxis with Robinson's brush (Microdont, São Paulo, SP, Brazil) and prophylactic paste (Technew, Rio de Janeiro, RJ, Brazil) was performed, followed by a prosthetic piece try-in (figure 5). The adaptation, size and color of the prosthesis were satisfactory. The material chosen for cementation was the typical glass ionomer for this procedure (Fuji Plus- GC, America, Alsip, IL, USA) following the manufacturer's guidelines. The cementation step was performed under relative isolation, aiming to reduce the consultation time, since this factor was essential for patient collaboration. A silicone mouth opener (Indusbello, Londrina, PR, Brazil) was also used during the entire cementation process, allowing a good and dry working field, besides contributing to the child's handling. After setting, excess cementation material was removed (Figure 6). With the use of Muller forceps (Golgran, São Caetano do Sul, SP, Brazil) and carbon (Accu Film II, Wilcos, Petrópolis, RJ, Brazil), occlusion checking was performed. It is noteworthy that protective stabilization was adopted in all care sessions.


**Figure 5**. Denari's prosthesis try-in.


**Figure 6**. Installed prosthesis (6A). Final photographs (6B) and (6C).

At the end of the service, both the mother and the child were instructed on how to proceed with the hygiene of the prosthetic piece, as well as the precautions regarding diet and oral habits that could compromise the longevity of the treatment. They were informed about the need for dental follow-up for an effective clinical and radiological control of the prosthetic piece and the permanent central incisors, as well as the need for speech therapy, in order to reestablish tongue posture and promote improvement in the child's speech.

## DISCUSSION

RTS is often identified at birth or in infancy because of the outstanding facial features and characteristic findings in the regions of the hands and feet [3,4,6], besides moderate intellectual disability [14]. The study participant had a

mild degree of RTS, showing as phenotypic characteristics: mild facial and limb dysmorphisms, as well as Marcus Gun's ophthalmological alteration, which is characterized by unilateral ptosis of the upper eyelid and low weight gain. She also had impaired cognitive and intellectual development, besides a disparity between chronological and mental age. It was clear that this discrepancy was mitigated by the family's efforts to ensure that the child received all possible stimuli for her development, while being assisted by a multidisciplinary team.

Knowing, informing, knowing how to identify and mastering methods of welcoming and conducting care, appropriate for a patient with syndrome, is one of the roles of health professionals. In addition, they must always seek a multidisciplinary performance, since it consists in the annulment of the individualistic model, expanding the team work, sharing the planning and the division of tasks, cooperating so that the whole is able to make a permanent contribution to society in the health field. It must be assumed that health problems are always interdisciplinary [15].

Dental problems related to TRS reported in the literature include crowding, malocclusion, ogival palate, retromicrognathia, thin upper lip, small mouth opening, bifid uvula, carious lesions, hypodontia, hyperdontia, natal teeth and cusp claws in the upper incisors of the permanent dentition [8,9,14]. In this work, mouth opening limitations and a slight overjet were noted. The inexistence of carious lesions is probably attributed to the maternal figure, as she showed total involvement and care with the child's health at all levels of assistance, since the diet was controlled and had low cariogenic power, brushing was supervised and performed with fluoridated toothpaste, and there was assiduity in the scheduled consultations.

The physical consequences of dental trauma can range from a small enamel fracture to tooth loss. The most prevalent etiological factor is a fall [16], and the most affected teeth are the central and lateral incisors of both dentitions [12]. In this study, the etiological factor was also confirmed, possibly associated with the precocity of the child's age at the time of the event, a period in which the child in general does not yet have the proper balance of his/her own body, potentiated by the fact that she (studied girl) is a syndromic child with motor development below that expected for her chronological age. The prevalence in relation to dental elements for trauma was reiterated.

The early loss of dental elements can lead to the acquisition of parafunctional habits, as it can interfere with the eruption and correct positioning of the corresponding successor tooth in the dental arch [16]. The absence of anterior teeth has entailed a psychological impact on children, undermining their socialization. For parents and guardians, this condition also raises concern and is one of the reasons for seeking dental care [17]. In this report, tooth loss led to a mistaken lingual posture, as well as generated a difficulty in the pronunciation of certain phonemes, showing that, in order to reestablish the functions of the entire stomatognathic system, multidisciplinary and interdisciplinary care between pediatric dentistry and speech therapy is indispensable. The literature reports this importance of care with a multidisciplinary team, especially with children with the syndrome or any other special need [18].

The esthetic rehabilitation of anterior deciduous teeth has a vital psychological impact on restoring the patient's self-esteem [16]. The manufacture of an esthetic functional space maintainer appliance is a treatment option. Among the indications for this appliance are:functional, phonetic and esthetic recovery, as well as prevention of emotional disorders in children, improvement in social interaction and preservation of the correct arch dimension, avoiding occlusal disharmonies. Denari's [19] tube-bar anterior partial prosthesis is among the main space-maintaining devices, and this was precisely the choice made for the rehabilitation of the study participant, since the Denari-type prosthesis promotes aesthetic, functional and phonetic recovery of the stomatognathic system.

Between the deciduous central incisors, the Denari-type prosthesis features a "male and female" fitting slider (socket) that opens as the maxilla grows and develops. As for this type of prosthesis, there is no need to wear out or prepare the teeth during its installation [19], resulting in less complexity for the execution of this type of prosthesis. This characteristic provides the preservation of the dental structure, reduction in the time of treatment and in the number of consultations, which are favorable and necessary conditions for the treatment of pediatric and syndromic patients.

The precautions that this type of prosthesis requires are: carry out a periodic clinical control to evaluate adaptation and possible displacements or fractures. Moreover, hygiene orientation and motivation must be intensified, with a view to avoiding the accumulation of bacterial biofilm on the prosthesis region [20]. Radiological control is essential to evaluate the

stage of formation of the permanent successor tooth and determine the appropriate time for removal of the prosthesis, which should be performed by the pediatric dentist following-up the case [21].

Besides the search for multidisciplinary services, there is the behavioral difficulty in pediatric and special care. Anxiety and lack of understanding during dental care are the biggest obstacles to patient collaboration [18]. This demonstrates the need and importance of a differentiated approach to treatment. Accordingly, in some cases, there is a need for behavior control techniques such as physical restraint, sedation, and, as a last resort, general anesthesia [18]. In the present report, protective immobilization was used with the help of the mother, ensuring that the treatment was carried out in a safe, effective and fraternal manner. It is relevant to highlight that the use of this technique does not offer psychological risks to the child or to those responsible for him/her [22].

## CONCLUSION

The use of partial anterior prostheses by the Denari's tube-bar system in patients with Rubinstein-Taybi Syndrome proved to be a viable option for space maintenance in cases of early loss of deciduous anterior teeth, as it promotes esthetic and functional rehabilitation and the reestablishment of the stomatognathic system.

It is important that the dentist is trained to offer the best treatment for these patients, since the syndrome imposes some difficulties, but does not prevent the treatment from being carried out.

### Collaborators

SKO Silva, was responsible the clinical procedures and wrote the manuscript. NCN Santos and FXPC Simões, provided writing assistance. ACDSA Maia, served as research advisor, assisted in conducting the case, provided clinical support obtaining images and writing assistance reviewing the article.

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