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Restorative treatment in patient with Williams Syndrome: case report

Tratamento restaurador em paciente com Síndrome de Williams: relato de caso

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ABSTRACT

In Dentistry, any user with one or more limitations, of temporary or permanent mental nature, of physical, sensory, emotional or medical growth, is considered to be a Patient with Special Needs, preventing him from being subjected to a conventional dental situation. These patients form a group which may be considered at high risk for the development of oral diseases, according to the kind of pathogen. Among them, is the Williams-Beuren Syndrome, a rare congenital disease with cardiovascular involvement, mental retardation, dysmorphic face, idiopathic hypercalcemia, musculoskeletal problems, dental and growth anomalies. Familial and sporadic cases are thought to result from deletion of genetic material from adjacent genes located on the long arm of chromosome 7. This article reports a clinical case involving a four-year-old boy with Williams-Beuren Syndrome, referred to the clinic of the Specialization Course for Patients with Special Needs, at the São Leopoldo Mandic Dental Research Center, Campinas (SP). As the patient was resistant to dental care, in order to reduce anxiety and increase collaboration in clinical sessions, the Hixizine® medication was administered. For atraumatic restorative treatment and restorations, the following materials were used: Cleanjoy®, Futurabond DC®, Ionofil Plus® Grandioso®, Grandio® and Profluorid®. As result, it was possible to reach a level of excellence using the indicated materials and specific protocols. Based on this work and the lived experience, it can be observed that these patients can receive welcoming dental care in specialized clinics.

Indexing terms: Dental atraumatic restorative treatment. Pediatric dentistry. Williams Syndrome.

RESUMO

Na Odontologia é considerado Paciente com Necessidades Especiais todo usuário que apresenta uma ou mais limitações, temporárias ou permanentes, de ordem mental, física, sensorial, emocional, de crescimento ou médica, que o impeça de ser submetido a uma situação odontológica convencional. Esses pacientes formam um grupo que pode ser considerado de alto risco para o desenvolvimento de doenças bucais, de acordo com o tipo de patogenia. Dentre elas, tem-se a Síndrome de Williams-Beuren, uma desordem congênita rara com envolvimento cardiovascular, retardo mental, face dismórfica, hipercalcemia idiopática, problemas musculoesqueléticos, anomalias dentárias e de crescimento. Os casos são hereditários ou esporádicos e a causa está na concepção com a deleção no braço longo do cromossomo 7. Este artigo relata um caso clínico envolvendo um menino de quatro anos, portador da síndrome,

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encaminhado para atendimento na Clínica do Curso de Especialização em Pacientes com Necessidades Especiais do Centro de Pesquisas Odontológicas São Leopoldo Mandic, Campinas (SP). Como o paciente apresentava resistência ao atendimento odontológico, com o objetivo de diminuir a ansiedade e aumentar a colaboração, nas sessões de atendimento foi administrado o medicamento Hixizine®. Para o tratamento restaurador atraumático e restaurações estéticas utilizou-se os seguintes materiais: Cleanjoy®, Futurabond DC®, Ionofil Plus®, Grandioso®, Grandio® e Profluorid®. Como resultado, foi possível atingir grau de excelência utilizando os materiais indicados e protocolos específicos. Mediante o caso relatado e frente à experiência vivenciada, pode-se perceber que esses pacientes podem receber um atendimento odontológico acolhedor nas clínicas especializadas.

Termos de indexação: *Tratamento dentário restaurador atraumático. Odontopediatria. Síndrome de Williams.*

INTRODUCTION

The Williams syndrome, also known as Williams-Beuren Syndrome (SWB), was first described in 1961 by a cardiologist named John Williams and colleagues, who evaluated four children with similar faces, supravalvular aortic stenosis, cognitive impairment, reading and writing difficulties, and affinity for music. In the following year, Beuren and his collaborators added strabismus, pulmonary artery stenosis and dental abnormalities as characteristics of the syndrome [1-6].

The SWB is a rare multisystem congenital disorder determined by the deletion of genes 26-28 in the long arm of chromosome 7 in 7q11.23. It is known for marked physical and developmental abnormalities, including unique behavioral characteristics, idiopathic hypercalcemia and dental problems [2-6]. A prevalence of 1 in 7,500 births is reported, with no gender or race predominance [2,6].

Individuals with SWB have a peculiar face, described as "elf face", due to the accumulation of periorbital fat, small nose with low nasal bridge, anteverted nostrils and long filter, thick lips, and little development of the jaw. Furthermore, dental anomalies, such as generalized diastemas, hypodontia, microdontia, short roots, enamel hypoplasia, pressure from the tongue on the teeth, and altered morphology including malocclusions (Angle Class II and III) are common [1,2,4,5]. In addition to dental cleaning, a four-month recall interval for dental follow-up has been recommended to prevent the risk of caries, periodontal and endodontic infections in these patients [2,4,6]. Many of them suffer from congenital heart disease and oral problems increase the risk of subacute bacterial endocarditis [5,6].

Compared with the general population, the risk of sudden cardiac death is 25 to 100 times higher in patients with SWB [1,3]. Therefore, the dental surgeon must know the precautions and protocols to be followed when treating patients with SWB [3,7,8].

Regarding the behavior management of Patients with Special Needs (PSN) in the dental office, the main difficulty is understanding the treatment itself. In addition to implementing effective and individualized preventive protocols and a differentiated psychological approach, respecting patient autonomy is essential for health promotion [9].

Although oral diseases can cause pain, oral health care is seen as a low priority when compared to medical and social care directed to PSN [10]. This is a reality that needs to be changed. Although they are the minority of the general population, PSN should not be disregarded, or even ignored [10,11].

Several articles on different pathologies or aggravations have reported the impact of oral health on quality of life and pointed to the importance of research focusing on the subjective factors related to oral problems. However, studies on specific conditions such as SWB are still needed. Thus, it is essential that affected patients are fully assisted by the health team, with dental care being part of that assistance.

In order to promote comprehensive care, and due to the lack of investigations on the approach to oral health for patients with SWB, this article describes a clinical case of a restorative dental treatment involving a four-year-old boy and discusses the importance of access to preventive dentistry and parental counseling.

CASE REPORT

The present case report was written after obtaining authorization through the Free and Informed Consent Form (FICF) to obtain and use the image/data, signed by the person responsible for the dependent child involved.

In December 2018, the patient EGMS, with SWB, that year at the age of four, was seen at the clinic of the Specialization Course for PSN, at the São Leopoldo Mandic Dental Research Center (CPO), Campinas-SP. In the seventh month of pregnancy, the mother contracted dengue. At the birth of the baby, she was informed that her son might have a syndrome. The diagnosis of which was confirmed at two years and two months of age.

Anamnesis, clinical examination, oral hygiene instructions and brushing as prophylaxis were performed. The mother was educated on how to proceed with brushing and oral hygiene of the patient, showing willingness to collaborate with the proposed treatment. The clinical dental examination revealed a high level of active caries and generalized enamel demineralization (Figure 1).

Figure 1 – Initial appearance of the dental features.

For the Atraumatic Restorative Treatment (ART) and aesthetic restorations, the following materials from VOCO do Brasil Ltda. (Porto Alegre, RS, Brazil) were used: Cleanjoy® prophylaxis paste; Futurabond DC® self-etch dual-cured adhesive; Ionofil Plus® glass ionomer; Grandioso® composite resin; Grandio® universal nano-hybrid restorative; and Profluorid® fluoride varnish.

As the patient was resistant to dental care, the use of oral medication (benzodiazepine) for anxiety control was planned, with the mother's consent and approval (before the dental appointment), in order to reduce anxiety and increase collaboration during the interventions period. The child was underweight for his chronological age and, because he did not reach the minimum weight for the use of this medication in his age (which would be 18kg), the team decided that the use of oral benzodiazepine for sedation was not safe. Thus, Hixizine® (hydroxyzine hydrochloride syrup) was chosen due to the possibility of taking advantage of the side effect of drowsiness that the medication provides, which after all, worked very well. Then, in all sessions, three doses of Hixizine®, antihistamine of the piperazine group, were administered

in the following therapeutic regimen: 7.5mL the night before the procedure, upon waking, on the day of the procedure, by morning, and 1 hour before the procedure.

In the following consultation, pulpectomy and reconstruction of the anatomical shape of teeth 51 and 61 (figure 2) were performed with Futurabond® adhesive and Grandioso® composite resin for later making the crowns of these teeth. Then, the ART technique was performed on teeth 55, 54, 65, 64, 75, 74, 85, 84, restored with Ionofil Plus®. In the third session, cavity preparations and restorations with the composite resin Grandioso® were performed on dental elements 62, 71, and 81 (figure 3).

Figure 2 – Teeth 51 and 61 restored.

Figure 3 – Teeth 62, 71, and 81 restored.

In the fourth session, elements 51 and 61 (figure 4) were prepared for reconstruction with Grandio® universal nano-hybrid resin and polycarbonate matrices, considerably improving the patient's aesthetics (figure 5). Finally, in the fifth and last session, cavity preparations and reconstructions were performed with Grandio® nano-hybrid resin and polycarbonate matrices of teeth 52 and 62 (figure 6).

Figure 4 – Preparation of teeth 51 and 61.

Figure 5 – Making dental crowns for teeth 51 and 61.

Figure 6 – Final appearance - crowns of teeth 52 and 62 cemented.

It is worth mentioning that, in all sessions, the attendance was performed with sedation with Hixizine® as specified, in order to reduce the patient's anxiety and increase his collaboration. In the sessions, before starting any restorative work, prior prophylaxis of the dental elements was performed using Cleanjoy® and, at the end of all procedures, the fluoride varnish Profluorid® was applied.

During treatment, an excellent improvement in the patient's oral hygiene conditions was observed, demonstrating the mother's adherence to previous instructions.

DISCUSSION

PSN are considered to be those with disabilities (physical, mental, sensory, developmental, behavioral, emotional, cognitive impairment) and self-limiting conditions that need medical attention (systemic health problems), requiring specialized treatment programs or services. The pathological condition can be developmental or acquired, which can cause limitations or incapacity in activities of daily living. PSN' health care is beyond routine and requires specialized and advanced level of knowledge [9].

With this case report selected at the clinic of the Specialization Course for PSN, at the São Leopoldo Mandic Dental Research Center (CPO), Campinas (SP), the first dental visit was to approach the patient and his mother and thus obtain knowledge of pre-existing medical conditions. It is important to note that many of these patients have organic complications that can only be obtained with parents and/or doctors [2,4,6]. The mother of the patient in the case described, accompanied him on all visits, demonstrating that she was attentive and collaborating with all the information necessary for the best care.

Regarding dental findings, according to some authors [1,2,4-6], anterior open bite, tongue pressure on the teeth, excessive interdental spacing and hypoplasia were evidenced. A high degree of active caries, hypomineralization of the enamel of the incisors and generalized demineralization of the dental enamel were also observed in this child.

Dental treatment under sedation or general anesthesia is usually preferable for patients with SWB, especially for anxious patients who do not cooperate and/or need extensive dental care [8,12]. Thus, the professional must work with the due precautions and follow the protocols during the treatment of these patients [3,7,8,12]. In this case, with the mother's consent and approval before the dental appointment, the Hixizine® medication was used orally in all sessions (1 dose of 7.5mL the night before the procedure, 1 dose upon waking up, on the day of procedure, by morning and one more dose 1 hour before the procedure) to reduce anxiety and increase collaboration during the dental treatment period.

In the health area, PSN are at increased risk of developing oral diseases that can have a direct and negative impact on their overall health [13]. As patients with SWB have congenital heart problems, poor oral health increases the risk of bacterial endocarditis [5,6]. Thus, the PSN has a growing need to prevent cavities and periodontal diseases. Most of these patients do not have full capacity to perform oral care, requiring the help of other people [4,9,13].

The participation of family members or guardians in this care is essential for the success of dental treatment and the promotion of oral health. The greater the patient's degree of dependence on nursing care, the more attention the caregiver must pay to hygiene and prevention [6,9]. Guardians should be advised on individual planning of a strict diet, especially in children with severe enamel hypoplasia, high caries risk and vulnerable cardiac conditions, to reduce the chances of oral infections [6]. In this clinical case, the mother adhered to the instructions provided, contributing to the improvement of the patient's oral hygiene observed during and at the end of treatment.

CONCLUSION

The treatment of the patient has reached a high level of excellence using the materials and techniques proposed for PSN in pediatric dentistry.

Early dental evaluation and parental counseling are extremely important in cases of SWB. The dental specialist in PSN must assume that effective communication is the key factor for the success of the treatment as it consists of the way to better know the individuality of each patient.

Also, it must be understood that syndromes manifest in several different manners in each special individual. Thus, when necessary, sedation can be performed to provide satisfactory dental treatment with adequate protocols for this group of patients.

Collaborators

A Rech was responsible the clinical procedures and wrote the manuscript. IC Rinaldi provided writing assistance. T Marega served as research advisor, provided clinical support obtaining images, and writing assistance reviewing the article.

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