Reporte de Casos
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Dental management and oral manifestations in child with Robinow syndrome: case report

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Abstract: Robinow syndrome (RS) is a rare genetic disease that may involve cardiovascular, skeletal and urogenital systems and manifest through short limb dwarfism, defects in vertebral segmentation, hypoplastic genitalia and cranial and facial abnormalities. In this article, we hereby present a patient of a 10-year-old Brazilian boy with RS who presented with a remarkable number of classical general and dental features. Physical examination revealed disproportionate short stature with shortening limbs, syndromic face with normal hair appearance, low-set ears, macrocephaly, prominent forehead, hypertelorism and ocular prominence, nose anomalies, triangular mouth with long philtrum, incompetent lip seal, midface hypoplasia, retrognathia, deficient malar prominence, among others. Clinical examination revealed gingival hyperplasia, malocclusion, crowding dental, diastemas, microdontia, enamel hypoplasia, prolonged retention, dental caries, delay of dental eruption v-shaped palate, torus palatinus, bifid tongue and ankyloglossia antecedent. Radiographically, dental agenesis, hypertaurodontism and thick roof pulp chambers and delayed tooth eruption were observed. Preventive, restorative and rehabilitative dental procedures are being carried out. Robinow syndrome presents clinical and radiographic findings that may be present from birth and require pediatric dentistry follow-up. Furthermore, the pediatric dentist plays an essential role in developing effective planning and treatment in terms of health promotion, aesthetic rehabilitation and functional interventions in the stomatognathic system.

Key words: Craniofacial anomalies; Oral manifestations; Pediatric dentistry.

Manejo odontológico y manifestaciones orales en un niño con síndrome de Robinow: reporte de caso

Resumen: El síndrome de Robinow (SR) es una enfermedad genética rara que puede involucrar los sistemas cardiovascular, esquelético y urogenital, manifestándose por enanismo de miembros cortos, defectos en la segmentación vertebral, genitales hipoplásicos y anomalías craneales y faciales. En este artículo, presentamos el caso de un paciente, un niño brasileño de 10 años con SR, que presentó un número notable de características generales y dentales clásicas. El examen físico general reveló una estatura desproporcionadamente baja con acortamiento de los miembros, rostro sindrómico con apariencia normal del pelo, orejas de implantación baja, macrocefalia, frente prominente, hipertelorismo y prominencia ocular, anomalías en la nariz, boca triangular con filtrum largo, labios incompetentes, hipoplasia de la región media de la cara, retrognatia, prominencia malar deficiente, entre otras. El examen físico intraoral reveló hiperplasia gingival, maloclusión, apiñamiento dental, diastemas, microdontia, hipoplasia del esmalte, retención prolongada, lesiones de caries dentales, retraso en la erupción dental, paladar en forma de "V", torus palatino, lengua bífida y antecedente de anquiloglosia. Radiográficamente, se observó agenesia dental, hipertaurodontismo, cámaras pulpares con techo grueso y erupción dental retardada. Se están llevando a cabo procedimientos dentales preventivos, restauradores y rehabilitadores. El síndrome de Robinow presenta hallazgos clínicos y radiográficos que pueden estar presentes desde el nacimiento y requieren seguimiento odontológico pediátrico. Además, el odontopediatra desempeña un papel esencial en el desarrollo de una planificación y tratamiento efectivos en términos de promoción de la salud, rehabilitación estética e intervenciones funcionales en el sistema estomatognático.

Palabras clave: Anomalías craneofaciales; Manifestaciones bucales; Odontología pediátrica.

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Manejo odontológico e manifestações orais em criança com síndrome de Robinow: relato de caso

Resumo: Síndrome de Robinow (SR) é uma doença genética rara que pode envolver os sistemas cardiovascular, esquelético e urogenital, manifestando-se por nanismo de membros curtos, defeitos na segmentação vertebral, genitália hipoplásica e anomalias cranianas e faciais. Neste artigo, apresentamos o caso de um paciente, um menino brasileiro de 10 anos com SR, que apresentou um número notável de características gerais e dentais clássicas. O exame físico geral revelou baixa estatura desproporcional com encurtamento dos membros, rosto sindrômico com aparência normal do cabelo, orelhas de implantação baixa, macrocefalia, testa proeminente, hipertelorismo e proeminência ocular, anomalias no nariz, boca triangular com filtrum longo, lábios incompetentes, hipoplasia da região média da face, retrognatia, proeminência malar deficiente, entre outras. O exame físico intrabucal revelou hiperplasia gengival, má oclusão, apinhamento dentário, diastemas, microdontia, hipoplasia do esmalte, retenção prolongada, lesões de cárie dentária, atraso na erupção dentária, palato em forma de "V", tórus palatino, língua bifida e antecedente de anquiloglossia. Radiograficamente, observou-se agenesia dentária, hipertaurodontismo, câmaras pulpares com teto espesso e erupção dentária tardia. Procedimentos dentários preventivos, restauradores e reabilitadores estão sendo realizados. A síndrome de Robinow apresenta achados clínicos e radiográficos que podem estar presentes desde o nascimento e exigem acompanhamento odontológico pediátrico. Além disso, o odontopediatra desempenha um papel essencial no desenvolvimento de um planejamento e tratamento eficazes no que diz respeito à promoção da saúde, reabilitação estética e intervenções funcionais no sistema estomatognático.

Palavras-chave: Anormalidades Craniofaciais; Manifestações Bucais; Odontopediatria.

Introduction

Robinow syndrome (RS) is a rare genetic disease that can present an autosomal dominant or autosomal recessive inheritance pattern.[1,2] Compromise of the cardiovascular, skeletal and urogenital systems can be present in both patterns and manifest through short limb dwarfism, defects in vertebral segmentation, hypoplastic genitalia and cranial and facial abnormalities.[1,3-5] Generally, autosomal recessive patterns present more severe dysmorphology compared to the dominant pattern and the musculoskeletal system is often minimally affected.[2,4]

Prevalence of this rare syndrome has been reported as 1:500.000, with no gender or race predilection[1,2], however, it is particularly higher in some regions of Turkey[6], Oman[7,8], Czech Republic, Brazil and Pakistan8 due to consanguineous marriage.[6,8,9] The prevalence is low, as 5%-10% of the children die in infancy because of cardiac problems.[9]

Classic oral and facial manifestations are varied and may include: bifid tongue, bifid uvula, cleft palate, dental crowding[4,10-12], malocclusion, dental agenesia, gingival hyperplasia[4,10,12,13], ankyloglossia, triangular mouth with a long philtrum[10,12,14] and highly arched palate.[11,13,14]

This article describes facial, oral and dental manifestations in a Brazilian boy diagnosed with RS. It is expected that these findings can help health professionals, especially pediatric dentists, in the care of children diagnosed with this syndrome, multidisciplinary treatment plan and dental decision making, based on the best scientific evidence. The patient and his legal representative were consulted and agreed to participate in the study, signing an informed consent form.

Case report

A 10-year-old Brazilian boy, without cognitive or motor impairment, presented to the Children's Specialty Clinic, Baby Clinic of the State University of Londrina complaining of aesthetic dental damage. Family history revealed that the patient is the second child of a non-consanguineous marriage. Mother reported no family history or characteristics of RS. The patient undergoes multidisciplinary follow-up with a speech therapist, pediatrician and endocrinologist.

Medical history

1. Gestational and childbirth history: Nuchal translucency examination at the 12th gestational week was indicative of the possibility of a syndrome. Ultrasonography at the 34th gestational week showed shortening of the bones suggestive of Down Syndrome. Cesarean delivery at 38 weeks, Apgar eight and nine, weighing and measuring, respectively, 3,720 kg and 51 cm. According to the mother, the pregnancy and delivery were uneventful and there was no exposure to radiation, alcohol or tobacco during pregnancy.

2. Neonatal period: Intensive Care Unit monitoring for seven days due to difficulty breathing and sucking. During this period, he was fed by tube. Subsequently, exclusive breastfeeding up to 6 months and supplemented up to 10 months.

General and facial characteristics:

The patient has a good communicative profile, mild speech impairment, excellent socialization and interaction. Mother reported an inadequate food diet rich in sucrose. Physical examination revealed disproportionate short stature (1,33 cm), weight 26 kg, syndromic face, normal hair appearance without compromise in quality or quantity, macrocephaly, prominent forehead, hypertelorism and ocular prominence, downslanting palpebral fissures, short nose with anteverted nares and depressed nasal bridge, triangular mouth (bottom corners face downward), long philtrum, incompetent lip seal, midface hypoplasia, retrognathia, deficient malar prominence and low-set ears (Figure 1). An examination of the hands showed clinodactyly and brachydactyly affecting both limbs (Figure 2).

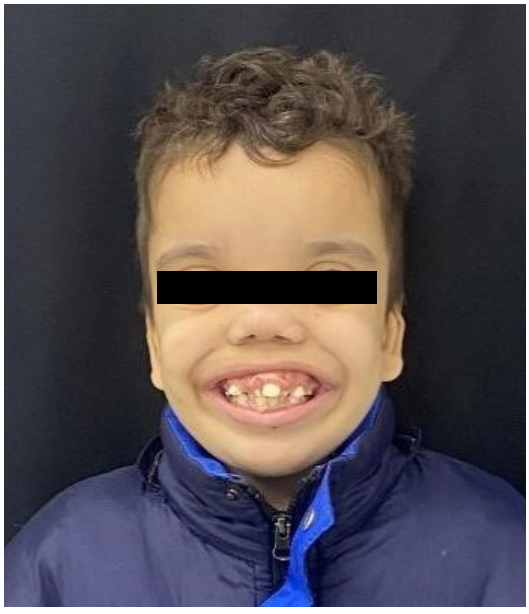


Figure 1. *Facial photograph showing general and facial characteristics of RS.*

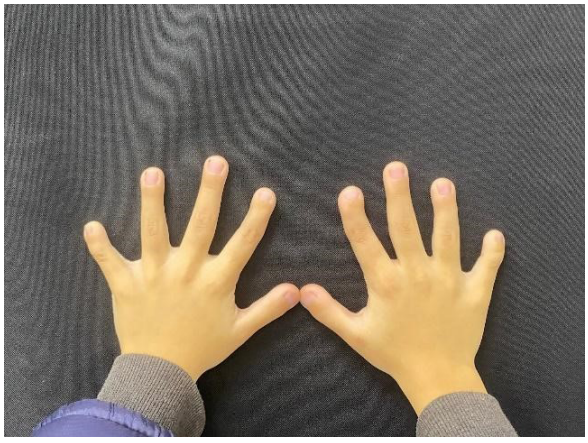


Figure 2. *Clinodactyly and brachydactyly affecting both hands.*

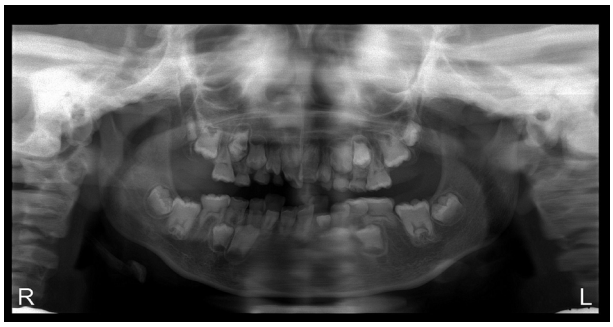


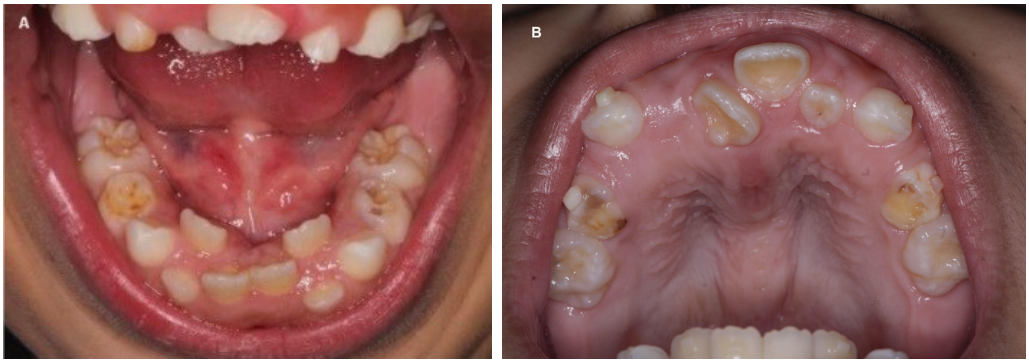
Figure 3. *Panoramic radiographic showing dental agenesis, hypertaurodontism and thick roof pulp chambers and delayed tooth eruption.*

Oral and dental characteristics

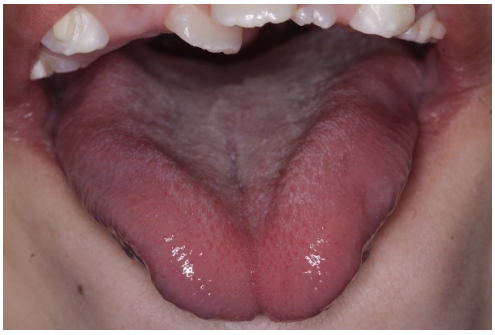
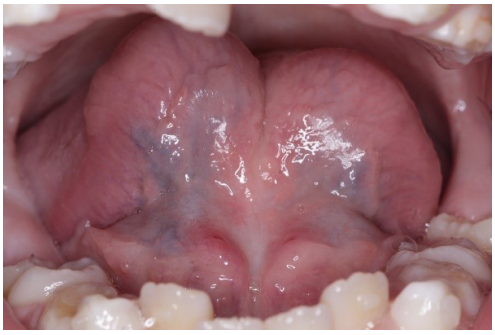
Radiographically, dental agenesis of bilateral upper and lower second premolars, hypertaurodontism in permanent molars, thick-roof pulp chambers in deciduous and permanent molars, and delayed tooth eruption were observed (Figure 3). Intraoral physical examination revealed gingival hyperplasia, malocclusion, crowding dental, multiple diastemas, enamel hypoplasia in upper incisors and prolonged retention of lower lateral incisors. In addition to these characteristics, the presence of dental caries in deciduous molars (Figures 4A and 4B) and bifid tongue (Figures 5A and 5B) was observed. The eruption of the deciduous dentition was delayed and started by 10 months of age.

Treatment plan

Consisted of dietary counseling, dental biofilm evidence, oral hygiene instructions, dental prophylaxis, topical application of fluoride varnish (Duraphat® 5000 ppm F; Colgate–Palmolive Company, Hamburg, Germany) to anterior teeth and application of silver diamine fluoride 30% (Cariestop® 30%, Biodinâmica Química e Farmacêutica LTDA, Parana, Brazil) in the deciduous molars, prolonged retention extraction,



Figures 4A and 4B. *Intraoral physical examination.*

Figures 5A y 5B. *Bifid tongue*

atraumatic restorative treatment in teeth #54, #64, #74, #84 and application of resin based sealants in teeth #55, #65, #75, #85 and #11. At this time, the patient is undergoing orthodontic treatment and reinforcement of oral hygiene guidelines, as well as biofilm control, is carried out monthly in orthodontic maintenance appointments.

Discussion

First described by Robinow and colleagues, this disorder is an extremely rare genetic disease[1,2] characterized by short limb dwarfism, hemivertebrae and genital hypoplasia and abnormalities in the head.[1,3-5] Higher prevalence may be found in some regions of the world due to consanguinity[6,8,9] and the male-female ratio is 1:1.[10] In the present report, the Brazilian boy presented normal mental development and was diagnosed as having autosomal recessive RS, based only on clinical characteristics: no genetic tests were performed. Congenital heart defects are frequently found in patients diagnosed with RS[3,4,10], but they were not present in this patient.

Characterized as either recessive Robinow syndrome (RRS) or dominant Robinow syndrome (DRS), fewer than 200 affected families have been reported with respect to RRS and fewer than 50 families with respect to DRS.[15] RRS is caused by mutation in ROR2 gene on the 9q22 chromosome, which control most basic cellular processes of chondrocytes and the resulting normal formation and ossification of the limbs, tail, vertebrae, and ribs, and it gives to the overall skeletal size. It also appears to be critical for the normal formation of the skeleton, heart, and genitals.[8,16] Loss of function of ROR2 protein during early development disrupts embryonic development, leading to the skeletal abnormalities and other phenotypic aspects of RRS such as genital abnormalities and heart defects.[17]

Common facial manifestations found in this patient corroborate those described in the literature: macrocephaly[10,12-14], forehead prominence[4,10-12,14], ocular hypertelorism[4,11], ocular prominence, downslanting palpebral fissures[10,13,14], low-set ears[10,12,13], nasal anomalies, triangular mouth with a long philtrum[10,12,14], retrognathia, midface hypoplasia[4,10-14] and deficient malar prominence.[14] Table 1 presents and compares the main facial and general manifestations of RS.

Classic oral and dental findings observed in this patient corroborate those described by several authors: bifid tongue, dental crowding[4,10-12], microdontia[3,18], delayed tooth eruption[11,19], prolonged retention of deciduous incisors[14,19], malocclusion, dental agenesia, gingival hyperplasia[4,10-13], ankyloglossia[10,12,14] and v-shaped palate.[11,13,14]

Table 1. General and facial manifestations of Robinow syndrome in the literature compared to the clinical case.

General and facial manifestations reported in the literature	General and facial manifestations in the patient
Microcephaly	Present
Forehead prominence	Present
Ocular hypertelorism	Present
Ocular prominence	Present
Iris anomalies	Not present
Palpebral fissures	Present
Low-set ears	Present
Short nose	Present
Anteverted nose	Present
Depressed nasal bridge	Present
Triangular mouth and bottom corners face downward	Present
Long/short philtrum	Present
Incompetent lip seal	Present
Retrognatia	Present
Midface hypoplasia	Present
Deficient malar prominence	Present
Genital hypoplasia	Present
Short stature	Present
Shortening of limbs	Present
Respiratory problem	Present at birth
Scoliosis	Not present
Hand anomalies	Present

However, bifid uvula, cleft palate[4,10-12] and supernumerary teeth[3,18] represent manifestations inherent to RS that were not observed. Table 2 presents and compares the main oral and dental manifestations of RS.

Table 2. Oral manifestations of Robinow syndrome in the literature compared to the clinical case.

Oral and dental manifestations reported in the literature	Oral and dental manifestations in the patient
Gingival hyperplasia	Present
Bifid tongue	Present
Macroglossia	Not present
Ankyloglossia	Present
Bifid uvula	Not present
Cleft palate	Not present
V-shaped palate	Present
Malocclusion	Present
Dental crowding	Present
Delayed tooth eruption	Present
Prolonged retention	Present
Natal or neonatal tooth	Not present
Dental agenesia	Present
Hypodontia	Not present
Microdontia	Present
Supernumerary teeth	Not present
Multiple diastemas	Present
Dental rotation	Present
Taurodontism	Present
Thick roof pulp chambers	Present
Conical teeth	Not present
Enamel hypoplasia	Present
Dental caries	Present

In this case, we presented rare dental findings such as: hypertaurodontism in permanent molars, thick-roof pulp chambers in deciduous and permanent molars and enamel hypoplasia. The present clinical case is the first report in the literature of the involvement of these manifestations associated exclusively with RS. Taurodontism is characterized by apical displacement of the pulp floor and shortening of the roots. It is believed to be caused in early fetal life due to the failure of the diaphragm of the epithelial sheath of Hertwig to invaginate at the proper horizontal level.[20,21] Presence of taurodontism is associated with several other syndromes, such as: Ellis Van Creveld[22], Klinefelter[23], Hurler syndrome[24], among others.

Another feature found in this patient associated with taurodontism is thick-roof pulp chambers. Kantaputra *et al*. (1999)[19], reported thick-floored pulp chambers in a patient diagnosed with RS. We believe that this dental alteration may be related to the presence of malocclusion, parafunctional habits, as well as the action of pulp aggressors, such as: dental caries and periodontal diseases.

In relation to the development defect of enamel, enamel hypoplasia is defined as a reduction in the amount of tissue formed, presenting as changes in the translucency or opacity of the enamel that can be diffuse or demarcated and white, yellow or brown in color.[25,26] This condition can be caused by many acquired environmental and systemic perturbations such as metabolic conditions, genetic disorders, infections, drugs and chemicals, as well as radiation and trauma.[27] Furthermore, many inherited syndromes, particularly those that involve skin, hair and nail show generalized enamel defects due to the common embryologic neural origin of the ectoderm.[28,29] Furthermore, enamel hypoplasia may represent an additional risk factor for the development of dental caries.

Measures for the prevention and control of dental caries, such as: dental biofilm evidence, oral hygiene instruction, prophylaxis, dietary counseling represent relevant strategies to promote oral health that have been implemented. The patient underwent a frenectomy procedure 4 years ago with significant improvement in speech according to his mother. At this time, the patient is undergoing orthodontic therapy through the use of invisible dental aligners with good collaboration. As the patient had difficulty effectively controlling dental biofilm, additional guidance during all control consultations regarding invisible aligners was shared. Difficulties in controlling dental biofilm, experience of tooth decay, presence of attachments, use of dental aligners and dental characteristics inherent to the syndrome, such as dental crowding, prolonged retention, represent factors that favor the development of chronic inflammatory gingival enlargement.

The present report showed varied and concomitantly remarkable general and oral manifestations. Therefore, it is essential that health professionals, especially pediatric dentists, are able to understand and identify clinical and radiographic aspects of Robinow syndrome and differentiate it from other syndromes that present similar manifestations.

Conclusion

Robinow syndrome represents an autosomal dominant or autosomal recessive disorder with varied oral, dental, facial and general manifestations that require a multidisciplinary approach. The pediatric dentist plays an essential role in diagnosing oral characteristics present since childhood. Furthermore, effective planning and treatment in terms of health promotion, aesthetic rehabilitation and functional interventions of the stomatognathic system must be carried out to improve the quality of life, self-esteem and well-being of the child and their family.

Conflict of Interest

The authors declare that there are no conflicts of interest regarding the publication of this paper.

Ethics Statement

The authors declare that the parents gave consent for images and clinical information of the case to be reported in scientific publications. Parents understand that the child's name and initials will not be published and efforts will be made to conceal the child's identity. This article complies with the protocols of the Research Ethics Committee of the State of University of Londrina.

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