Prosthetic Rehabilitation of a Child With Rapp–Hodgkin Syndrome

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ABSTRACT

The purpose of this paper was to report the case of a 6-year-old male patient who was clinically diagnosed as having Rapp-Hodgkin syndrome—a variant of ectodermal dysplasia characterized by dysplastic disorders of the skin, hair, and nails associated with cleft lip and/or palate. The patient exhibited only the primary maxillary and mandibular canines bilaterally. Restorative treatment was performed on all primary canines, followed by aquisition of maxillary and mandibular impressions for fabrication of acrylic removable partial dentures with circumferential clasps on the primary canines. The dentures allowed recovery of the vertical dimension and excellent esthetics. Hypodontia of several teeth associated with ectodermal dysplasia, as in this case involving Rapp-Hodgkin syndrome, causes several functional and esthetic alterations. These disorders should be diagnosed and treated as early as possible to restore the normal function, allow adequate mastication and speech, enhance esthetics, and, consequently, promote the patient's self-esteem and social integration. (J Dent Child 2012;79:115-9)

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Ectodermal dysplasia is a hereditary disease that involves dysplasia of tissues of ectodermal origin, including the hair, teeth, skin, and sebaceous glands, eventually causing also dysplasia of tissues of mesodermal origin.[1,2] The reported prevalence ranges from 1:10,000 to 1:100,000 live births, with a male to female ratio of 5:1. The condition is usually manifested in males and carried by females.[1-3] Ectodermal dysplasia was first observed by Darwin in 1838 and then by Christ in 1913, who defined it as a congenital defect of the ectoderm.[2] Nearly 120 variations of ectodermal dysplasia have been described.[3] Rapp-Hodgkin syndrome (**RHS**) was reported by Rapp and Hodgkin, who described the findings on a mother, son, and daughter.[1,2,4,5]

The syndrome is rare, presents autosomal inheritance, and may occur due to mutations involving alleles associated with the gene TP63.[6-8] Several authors have presented evidence of autosomal dominant inheritance, describing several families with 2 or more RHS cases.[2,9,10]

RHS is characterized by ectodermal dysplasia associated with cleft lip and/or palate; few cases have been described in the literature.[7,11] The clinical signs of RHS include dry, thin skin due to the deficiency of sweat glands; dystrophic and fragile nails; thin, coarse, and clear hair and eyebrows; pili torti; alopecia; high, prominent forehead; low stature; narrow nose; hypospadia in males; photophobia; prominent forehead; saddle nose; and hyperkeratosis of the hand. Patients may present intolerance to heat in more severe cases due to deficiency of the sweat glands, leading to constant fever of unknown etiology.[1,2,7]

The oral examination reveals prominent thin lips because the vertical dimension is reduced. This is due to: hypodontia; maxillary hypoplasia with high palatal vault; hypoplasia of the middle facial third with mandibular prognathism; cleft lip and palate; reduced salivary

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flow; dry oral mucosa; difficulty in swallowing; hypodontia; enamel hypoplasia; tooth crowding; abnormalities in tooth morphology; peg-shaped incisors; microdontia of molars; and delayed tooth eruption. These characteristics may occur both in the primary and permanent dentition. RHS' oral alterations, especially cleft lip and palate, may cause disorders in nutrition, speech, swallowing, and facial aspects. Enamel hypoplasia combined with reduced salivary flow increases caries risk, thus requiring fluoride therapy, dietary counseling, dental plaque control, and increased ingestion of water with food.[2,11] The development of the alveolar bone occurs according to tooth eruption; thus, it may be impaired by hypodontia.[2,9]

The neonatal diagnosis of RHS is difficult because the main characteristics of sparse hair, hypodontia, and prominent forehead are normal at this period. A diagnosis based solely on facial characteristics is not reliable up to 11 months of age.[2,9] The dental aberrations are manifested early and should be detected during childhood. After clinical and radiographic examination, the family should be screened to rule out other syndromes. The definitive diagnosis may be confirmed by the indirect molecular method (ie, by linkage analysis or biopsy, which detects the abnormal distribution of sweat glands and capillary follicles characteristic of the syndrome).[2,9]

Early diagnosis is important to prevent and control the first occurrences of hyperthermia in young children and enhance physical, emotional, and social development. In some cases, the signs of the syndrome may remain unnoticed by pediatricians; thus, pediatric dentists are often the first health professionals to diagnose this disorder due to delayed tooth eruption and other oral characteristics. Psychological follow-up is mandatory in the rehabilitation of these patients, as is utilizing the services of speech therapists, dermatologists, otolaryngologists, pediatricians, and plastic surgeons.[2,12]

Once the diagnosis is established, the patient should be informed of the immediate treatment options and be prepared for future treatment considerations (ie, to wait for growth completion before rehabilitation with removable or complete dentures, fixed dentures, orthodontic treatment, esthetic restoration of abnormal teeth with composite resin, and endosseous implants).[1,2,13-17]

The purpose of this paper was to describe the oral rehabilitation of a 6-year-old boy with Rapp-Hodgkin syndrome using removable dentures.

CASE DESCRIPTION

A 6-year, 4-month-old male patient of African descent was admitted to the Hospital for Rehabilitation of Craniofacial Anomalies, University of São Paulo, Bauru, São Paulo, Brazil (Figure 1). He was the single child of unrelated parents without a family history of malformations and was delivered normally at term, weighing 2,710 kg and measuring 47 cm.

An extraoral physical examination revealed: sparse hair; absent eyebrows; sparse eyelashes; obstruction of the lacrimal duct bilaterally; dry hair and skin; thin nails; complaint of constant fever since birth; and good neuropsychomotor development. The clinical intraoral examination revealed submucous cleft palate and hypodontia of several teeth, with presence of only the primary maxillary and mandibular canines bilaterally (Figure 2). Radiographic examination also evidenced the presence of

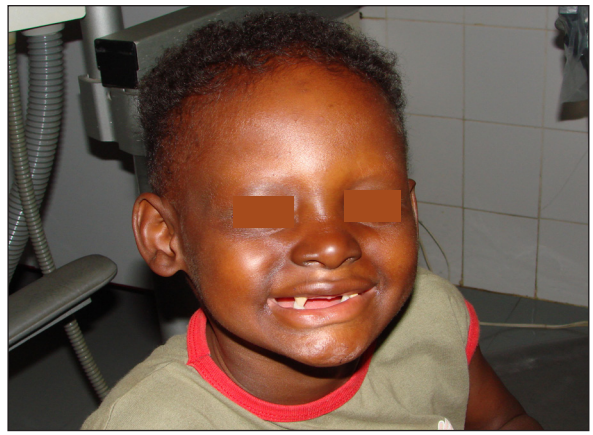

Figure 1. Facial aspect of a patient with Rapp–Hodgkin syndrome.

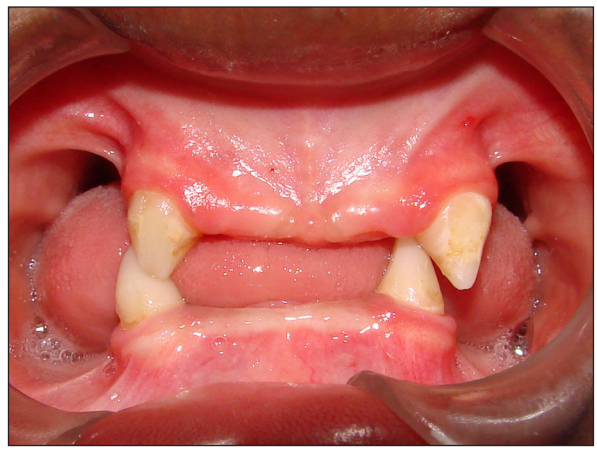

Figure 2. Intraoral aspect of primary maxillary and mandibular canines bilaterally.

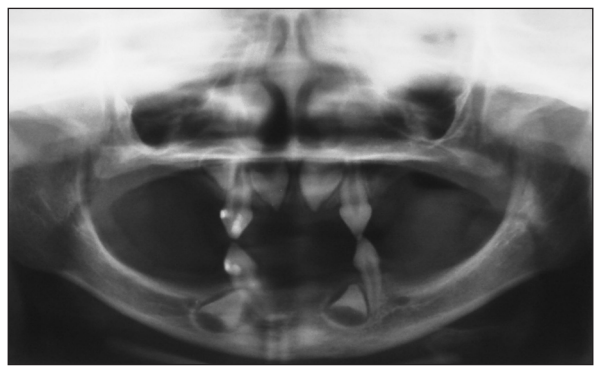

Figure 3. Panoramic radiograph showing hypodontia of several teeth, with presence of the primary and permanent maxillary and mandibular canines, bilaterally.

tooth buds of the permanent maxillary and mandibular canines bilaterally (Figure 3). The patient was evaluated by a geneticist to rule out other conditions and was diagnosed as having RHS based on the clinical findings. The mother also reported that the child was facing bullying problems at the school.

Oral rehabilitation treatment involved several short sessions, due to the patient's young age. Treatment was initiated by psychological preparation and oral health education, followed by composite resin and glass ionomer restorations on all primary canines. During these sessions, the patient was extremely interested and compliant.

After completion of restorative procedures, maxillary and mandibular impressions were taken with alginate using stock trays, and a wax bite record was obtained. Plastic teeth were mounted in wax on both maxillary and mandibular dental casts to evaluate esthetics, occlusion, speech, and swallowing (Figures 4 and 5). The teeth mounted in wax were sent to the laboratory for application of acrylic and placement of circumferential clasps fabricated with orthodontic wires on the primary canines to enhance the retention. The dentures were delivered, providing recovery of the vertical dimension and excellent esthetics (Figures 6 and 7).

At the 4-month follow-up, the maxillary denture was broken and repaired and adjusted. The patient is now being followed up with every 4 months to check the fit and adjustment of his dentures.

DISCUSSION

Children with syndromes may present psychological disorders due to their abnormal characteristics, especially those of early school age. Similarly, individuals with inherited dental aberrations may suffer bullying,[18] present psychological, esthetic and functional problems,[19] and decide not to have children for this reason.[18] Consequently, the early prosthetic treatment with removable dentures in children with hypodontia is unanimously recommended. Oral rehabilitation provides functional benefits: It enhances speech, mastication, emotional and social development, esthetics, and self-esteem.[20] When the child is compliant, treatment may be successful as early as at 2 to 3 years old. The deficient alveolar bone, however, may complicate the retention and stability of the dentures.[2,7,14,15,21]

Many dental professionals are unaware of the syndrome's characteristics and the aspects related to pediatric dental treatment using dentures. Children can usually adapt easily to removable dentures, however, provided they and their parents are well informed and motivated. The treatment should comprise short sessions to enhance patient compliance and should be initiated by oral health education.[2,7,22,23]

The dental treatment of these children should be carefully planned by a multidisciplinary treatment involving pediatric dentists, orthodontists, prosthodontists, and maxillofacial surgeons to coordinate diagnosis, treatment, and follow-up, according to the patient's age and growth period,[24,25] as well as psychological follow-up.[22] A flow chart of such multidisciplinary care was suggested by Bergendal.[24] In the present case, the rehabilitation with removable dentures enhanced the mastication, speech, and esthetics of the patient and contributed to promote his self-esteem. Craniofacial growth is normal in RHS; thus, the dentures may fit improperly and require regular replacement.

Figure 4. Dental casts in an articulator for mounting of teeth in wax.

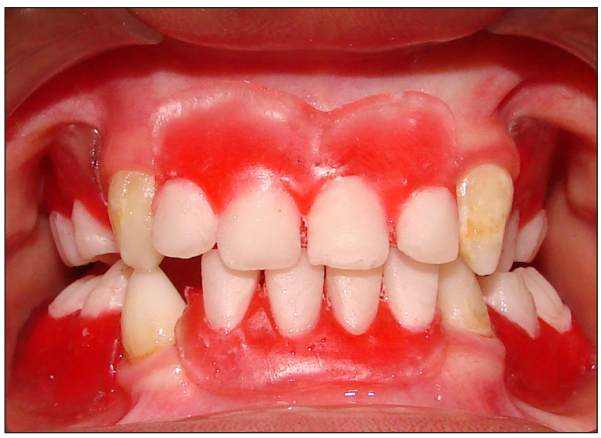

Figure 5. Analysis of teeth mounted in wax in the mouth.

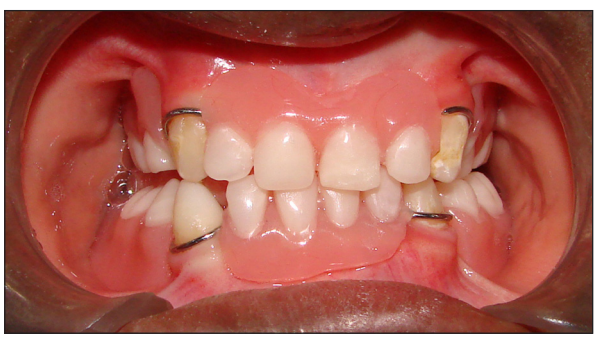

Figure 6. Intraoral aspect of a denture in place.

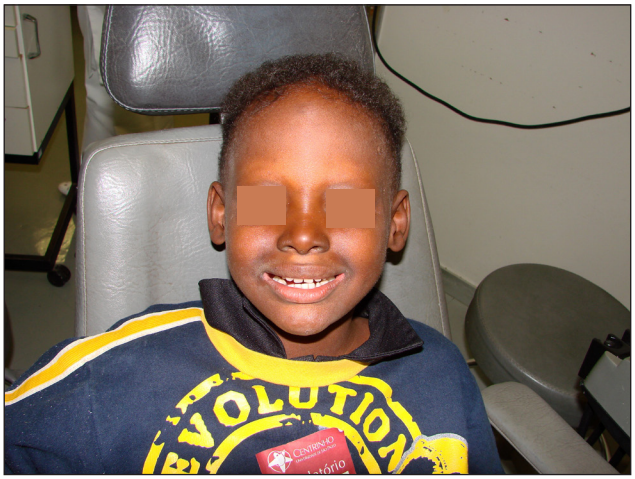

Figure 7. Excellent esthetic outcome obtained after placement of the maxillary and mandibular removable dentures.

Treatment with endosseous implants is recommended only when growth is almost complete, because it is known that implants react as ankylosed teeth when installed before completion of growth of the alveolar process.[24,26] Based on this rationale, removable dentures are indicated and should be replaced by endosseous implants in the permanent dentition stage after completion of bone growth[2,11,14,17] and when the patient can participate in the decisions and fully understand the different phases of this complicated treatment.[24] Meanwhile, in case of partial anodontia, magnetic retention of prostheses has also been suggested.[27]

No studies are available on the utilization of implants in young children, especially with the peculiarities inherent to the syndrome.[24] According to some authors, however, the possible problems related to early implant placement may be neglected considering the benefits provided by implant-supported rehabilitation in the mandibular arch of anodontic children and because the alveolar growth in the anterior mandible is relatively small when teeth are missing.[26] In any case, implant placement in the maxillary arch should be avoided until early adulthood.[26]

The utilization of orthodontic mini-implants for the anchorage of temporary prostheses until completion of alveolar growth has been suggested.[28] These devices allow good fixation and adjustment while permitting vertical growth, since they may be placed in the toothless alveolar bone. They also allow utilization with orthodontic treatment and adequate oral hygiene.[28]

No long-term studies are available on patient compliance with wearing denture at this age. Recall appointments should be scheduled as frequently as possible (eg, every 4-6 months for analysis of denture fit and child compliance with denture wear). Dentures also should be replaced as necessary according to craniofacial growth until definite rehabilitation may be performed.

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