Osseointegrated Implants in the Oral Rehabilitation of a Patient with Cleft Lip and Palate and Ectodermal Dysplasia: A Case Report

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Hereditary ectodermal dysplasia is an inherited disorder characterized by aplasia or dysplasia of ectodermal tissues, such as hair, nails, teeth, and skin, that occurs in approximately 1 in every 100,000 live births. Dental abnormalities and abnormal facial appearance are of major concern in childhood and adolescence, since they can restrict the individual socially and affect his or her self-confidence. Oral rehabilitation in the early stages of the patient's life may provide functional and esthetic restoration as well as safeguard psychologic health. This report presents the clinical procedures involved in the rehabilitation of a 10-year-old female patient with complete bilateral cleft lip and palate and ectodermal dysplasia. INT J ORAL MAXILLOFAC IMPLANTS 2004;19:896–900

Key words: *cleft lip, cleft palate, dental implants, ectodermal dysplasia, maxillofacial development*

Hereditary ectodermal dysplasia (HED) is an inherited disorder characterized by aplasia or dysplasia of ectodermal tissues, such as hair, nails, tooth enamel, and skin, and is thought to occur in approximately 1 in every 100,000 live births.[1] Clinically, HED may be divided into 2 broad categories: *(1)* an X chromosome–linked hypohidrotic form, characterized by the classic triad—hypodontia, hypohidrosis, and hypotrichosis—and dysmorphic facial features, and *(2)* a hidrotic form that usually spares the sweat glands but affects teeth, hair, and nails and is inherited as an autosomal trait.[2]

Diagnosis of HED is more easily made in childhood and is based on the patient's history and clinical examination. Heat intolerance, inability to perspire, abnormal dentition, and sparse hair can be noticed in the affected child. Dental abnormalities and abnormal facial appearance are of major concern in childhood and adolescence,[3] since they can restrict the individual socially and affect his or her self-confidence.[4]

Oral rehabilitation in the early stages of the patient's life can provide functional and esthetic restoration as well as safeguard psychologic health. Reconstruction of the dentition can be carried out by means of fixed or removable prostheses, which may be supported by implants. Implants provide extra support for the prosthesis when the replacement of several teeth is required.

However, the patient's age may make implant-supported rehabilitation a challenge. Since osseointegrated implants are incapable of eruptive movement, they behave as ankylosed teeth.[5] Thus, it is advisable to wait for the completion of skeletal and

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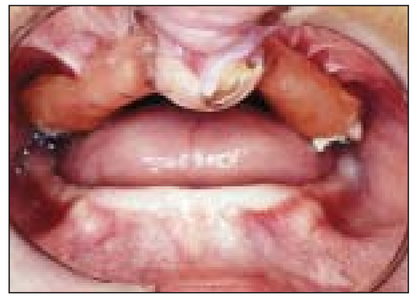

Fig 1 *(Left)* Facial and *(above)* intraoral aspects of a patient with ectodermal dysplasia and bilateral cleft lip and palate.

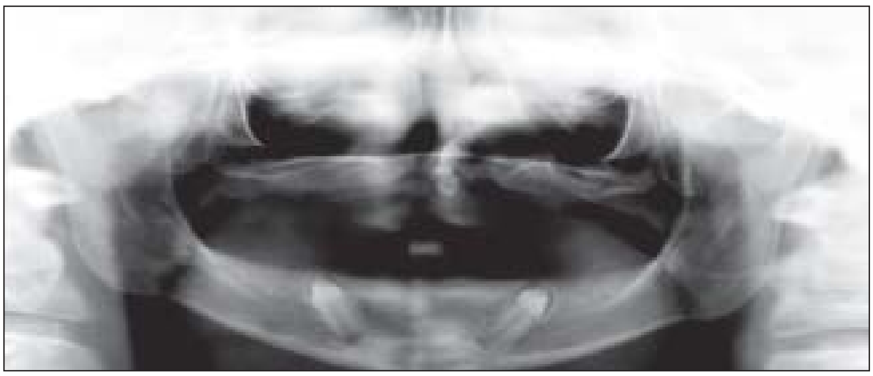

Fig 2 Orthopantomogram showing absence of teeth in both jaws, with exception of the mandibular canines, which were unerupted.

dental growth before starting an implant-based treatment plan.[6] Implants placed in growing children may present infraocclusion with time because of both the natural maxillary growth and the eruptive movement of the adjacent teeth. If it is necessary to lengthen the restoration, both the esthetics and the biomechanics of the prosthetic crown may be compromised. The probability of component fracture may increase, and treatment failure may result. The risk of peri-implantitis may also increase, because longer abutments than usual are needed in a such a case, and thus the peri-implant gingival sulcus is deeper than usual.

Psychologic and physiologic factors, however, can make it desirable to begin treatment early[7]: *(1)* the agenesis of teeth produces inhibition of alveolar bone apposition in the vertical dimension, and the implant location is affected only by sutural growth between the maxillary and cranial bones[2]; *(2)* the lack of alveolar bone in areas of anodontia decreases the tissue support for removable prostheses[8]; and *(3)* the absence of teeth and impaired esthetics and phonetics can cause social restriction.

The most effective treatment modality as determined on the basis of the scientific knowledge available in the literature should be offered. This report presents the clinical procedures involved in the rehabilitation of a patient with cleft palate and HED at the Hospital for Rehabilitation of Craniofacial Anomalies (HRAC) of the University of São Paulo (USP) in Bauru, Brazil.

CASE REPORT

A 10-year-old female patient presented with HED and bilateral complete cleft lip and palate at the Dental Implantology Department of HRAC/USP (Fig 1). Her files at the Medical Genetics Sector indicated that she had been diagnosed with ectrodactyly ectodermal dysplasia clefting syndrome (EEC) upon presentation with cleft lip and palate, dry and blond hair, bilateral lacrimal duct obstruction, wide nasal alae, dry skin, and ectrodactyly of hands and feet, with lobster-claw deformities and nail dysplasia. She also presented with complete agenesis of the teeth, with the exception of the mandibular canines, which remained unerupted (Fig 2). Surgical closure of the lip had already been performed at 2 years of age. Palatoplasty was accomplished at 9 years of age without resection of the premaxilla. Her mother reported that she had

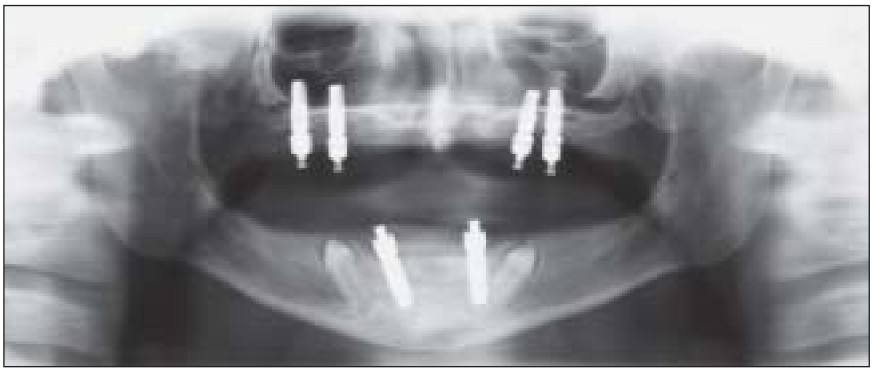

Fig 3 Radiographic aspect after abutment connection.

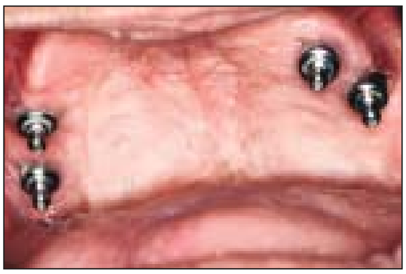

Fig 4a Clinical aspect of the maxilla after the connection of ball attachments. Notice the nonretentive alveolar ridge.

Fig 4b The mandible was provided with a bar-clip system for retention of an overdenture.

Fig 5 Facial appearance of the patient after prosthetic rehabilitation.

exhibited introverted and antisocial behavior and had started to refuse to go to school.

The evaluation for oral rehabilitation favored implant-retained overdentures in both jaws. However, radiographic examination showed very little remaining bone in the maxilla for implant placement. Bilateral sinus lifting procedures were therefore performed under general anesthesia, following a rigid protocol.[9] Particulate autogenous bone harvested from the mandible was placed into the sinus bilaterally together with 4 screw-type implants made of commercially pure titanium (Conexão; Sistemas de Prótese, São Paulo, Brazil). The implants, which were 13 mm long and 3.75 mm in diameter, were placed at the sites of the maxillary right premolars, left canine, and left first premolar. Two other identical implants were placed in the mandible in the canine areas, where the canines remained unerupted (Fig 3).

Five months later, second-stage surgery was performed under local anesthesia. To retain an overdenture, 3-mm ball abutments were connected at the sites of the maxillary right second premolar, maxillary right first premolars and maxillary left canine, and a 4-mm ball abutment was connected at the site of maxillary left first premolar (Fig 4a). The mandibular implants received 3-mm-long standard abutments, and a bar-clip–retained overdenture was fabricated (Fig 4b). Maxillary and mandibular prostheses were seated 4 months after second-stage surgery, and the patient was extremely satisfied with her new facial appearance and improved oral function (Fig 5). At the time of this reporting, the prosthetic restorations had been in function for 32 months without any signs or symptoms of failure.

DISCUSSION

Since first employed, osseointegrated dental implants have played an important role in the field of dental rehabilitation. Their use has spread to more complex cases, which requires clinicians to be aware of treatment outcomes widely accepted in the literature[2–6] to avoid potential risks to the patients.

Young patients represent a challenge in dental implant therapy, since their growth pattern is not predictable and the long-term effects of this treatment remain unknown.[10] Although the completion of skeletal and dental growth is recommended before the implementation of an implant-assisted treatment plan,[6] aspects such as esthetics and function can favor early implant rehabilitation.[11]

Guckes and colleagues[8] rehabilitated a 3-year-old child presenting with HED using implants and followed the child for 5 years. The patient was provided with implant-supported overdentures, which were relined or replaced every year to allow eruption of the maxillary teeth and facial growth. One of the implants, which was retained submerged, did not follow the forward and downward maxillary eruptive movements and remained in close proximity to the maxillary sinus. Mackie and Quayle[11] published a case report showing the applicability of implants in young patients without presenting a long-term follow-up.

One important recommendation is to avoid rigid connectors in young, actively growing patients, because they can prevent normal jaw growth, especially if the prosthesis crosses the midline. Retrievability is also recommended to allow adjustments with time.[4]

HED has not always been related to submergence of implants because of the virtual aplasia of the alveolar process that occurs in HED patients and in the presence of multiple anodontia. Ruhin and associates[12] evaluated 16 patients with HED in a retrospective study and reported that anthropometric analyses revealed facial height reduction and concavity in 8 patients compared with normal values. They also found a global growth reduction of the alveolar dimensions, especially in length. In an evaluation of 12 lateral cephalograms of 7 patients, which was conducted before and sometimes after treatment, Delaire[13] found that most cases displayed maxillary retrusion, mandibular protrusion, and Class III malocclusion, and that the worst cephalometric abnormalities occurred when dental agenesis was severe.

Moss and Salentijn[14] stated that as teeth and muscles enable mastication, they transmit tension to the maxilla and mandible. The teeth facilitate the maintenance of bone because of the resorption-apposition process elicited by dentoalveolar ligaments.[15] The consequence is that dental agenesis could impair transverse[16,17] and sagittal growth in the maxilla and mandible,[18] besides leading to reduction of the mandibular angle.[19] Vertical growth could also be affected by the alveolar height decrease associated with tooth agenesis.[17,20,21]

More recently, Bondarets and coworkers[22] longitudinally analyzed the facial growth of 61 subjects with syndromic ectodermal dysplasia. Their serial cephalometric analysis revealed that maxillary growth was slightly reduced, yet there was no detectable effect on the mandible. The most significant findings were a universal tendency toward a change in the sagittal jaw relationship, with worsening Class III malocclusion over time. Another finding was a tendency toward anterior growth rotation.

Kearns and associates[2] stated that placement of maxillary implants in patients with HED may require bone grafting and lifting of the sinus membrane, as observed in the present case. This condition is related to the unavoidable resorption of the alveolar bone that takes place in the absence of teeth. Therefore, a deficiency in the width of the alveolar crest can also be found in patients with HED, as well as an expanded maxillary sinus. Both conditions hinder the placement of screw-type dental implants without additional procedures for bone augmentation. Jansma and associates[23] concluded that maxillary augmentation with autogenous bone grafts followed by placement of endosseous implants can serve as a reliable alternative to conventional prosthetic rehabilitation in patients with clefts.

In this case, the age of the patient, combined with the complexity of and time required for surgical procedures, argued in favor of the convenience of general anesthesia. The lateral aspect of the mandibular ramus was harvested en bloc and then milled to fill the sinus cavity around the implants. As the chin would also receive implants, it was not chosen as a donor site. Moreover, the presence of impacted teeth in that area would have limited the quantity and quality of bone grafts.

Besides esthetic and functional aspects, there must be concern about psychologic support to young carriers of such syndromes, and their follow-up treatment must include helping them to accept the prosthesis and all procedures involved in its maintenance and hygiene. Neglecting these factors will undoubtedly lead to treatment failure. According to Tanner,[24] reports on the emotional aspects of HED are rare and tend to be included in the documentation of related dental work. Bhatia and Grandel[25] and Album[26] related that children became more "comfortable" with their appearance or more outgoing after dental treatment. These authors clearly suggested that failure to provide a child with dentures beyond the preschool period may result in the child developing an inadequate self-image. However, they failed to mention whether adjustment problems are likely to occur in youngsters who are already fitted with dentures, or whether other unusual aspects of their appearance lead to social difficulties.

Implants should be used in HED patients whenever possible, but even though good results can be obtained, the long-term results are still unpredictable. The combination of HED and cleft lip and palate presents a complicating factor, since the size, shape, and location of the residual maxillary alveolar bone provide little support for a complete denture.[27] Patients and their families must be aware of the limitations concerning the treatment success. Analyses of many more cases and their follow-up are needed for more conclusive data related to the treatment of this defect combination.

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