Treatment strategy for patients with ectodermal dysplasia: a case report

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Ectodermal dysplasia (ED) is a hereditary condition characterized by abnormal development of the skin, hair, nails, sweat glands, and the stomatognathic system. There are many different types of ectodermal dysplasia of which X-linked anhidrotic ectodermal dysplasia is the most common. Multiple genes have been discovered to cause ectodermal dysplasias. With any form of ED, children may display a range of symptoms and challenging rehabilitation. This clinical report presents the treatment plan for a young patient with ED and anodontia requiring prosthetic restoration.
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INTRODUCTION

The incidences of hereditary syndromes and congenital defects affecting the stomatognathic system are small, but the treatment challenges are great. The most common genetic causes of disturbed development of the stomatognathic system are: ectodermal dysplasia (ED), cleft lip and palate, incontinentia pigmenti, Down's syndrome, and Rieger syndrome.[1] This report focuses on the treatment challenges associated with ED.

ED occurs in approximately 1 in every 100,000 live births.[2] ED is not a single disorder, but rather a group of closely related conditions. Aproximately 132 different syndromes have been identified. The EDs are heritable conditions in which there are abnormalities of two or more ectodermal structures such as the hair, teeth, nails, sweat glands, cranial-facial structure, digits, and other parts of the body. The nature of this dysplasia requires an interdisciplinary approach for a successful treatment. Severel disciplines are involved in the study and treatment of ED: genetics, dentistry, ear-nose-throat, opthalmology, pediatric dermatology, and psychology.[3-7]

The dental phenotype of patients with ED is characterized by polymorphism. The symptoms affecting the stomatognathic system involve the number and morphology of teeth, the growth of the maxillary bones and the mandible, and the quantity of saliva. Abnormalities in the number of the teeth can be grouped in three categories: hypodontia (absence of fewer than six teeth), oligodontia (absence of six or more teeth), and anodontia (complete absence of teeth).[1]

The patient has hypodontia in 80% of all ED cases.[8] The teeth that are present appear malformed (conical in shape) and widely spaced. The alveolar bone of the edentulous ridge is knife-edged, and the growth of the midface is altered. The growth of the maxilla and the mandible appear to be connected with the number of missing teeth. The severity of oligodontia has a direct influence on bone growth. The lack of proper bone growth makes the patient appear older (e.g., reduced vertical height, increased nasolabial fold, flattened face). A cephalometric study showed that the cant of the maxilla, as defined by the angle formed by SN and ANS-PNS, and the mandibular length (Go-Pg) were decreased in patients with ED. The mandible was relatively more prognathic in the ED population, creating a pseudo-class III appearance.[9]

A study of the growth patterns of the maxillae and mandible should be performed before treatment strategies are developed. It must be explored if any of the most patient- benefiting treatments, which also happen to be the most invasive treatments, can be utilized. These treatments may involve implant placement.

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The skeletal changes in the maxilla are happening in all three dimensions: in the anterioposterior axis, the transverse axis and the vertical axis.[10] The maxilla grows anteriorposteriorly because of passive displacement, which constitutes one third of its growth and because of enlargement of the maxilla itself. In the transverse axis, the width of the maxilla increases because of the expansion of the midpalatal suture. The vertical increase occurs because of the sutural lowering, passive lowering, and bone apposition on the occlusal surface of the maxillary alveolus. Simultaneously, the nasal floor is resorbed on its nasal surface. Therefore, the growth of the maxilla is concomitant with the overall growth of the child.

The mandible grows in an anterioposterior mode as well as rotates forward. The posterior width increases by virtue of the "V" configuration. The midsymphyseal suture ceases to grow prior to eruption of the primary teeth. Intercanine distance starts increasing again 1 year before eruption of the primary central lower incisor and essentially terminates with the eruption of the permanent lateral incisors. Anterior width, therefore, stabilizes early: around age 7 for dentate patients and much earlier for anodontia patients.[10] The differences in the mode and timing of growth between the maxilla and the mandible should be contemplated when developing a treatment plan.

The dental team is composed by specialists representing pediatric dentistry, restorative dentistry, prosthodontics, orthodontics, oral surgery, or periodontics. Hypodontia and oligodontia call for a multidisciplinary approach,whereas, anodontia can be effectively treated by a prosthodontist alone or by the collaboration of a prosthodontist and an oral surgeon or periodontist.

Complete dentures,[11-14] partial dentures,[15] or overdentures[16,17] have been reportedly fitted to young ED patients with success. Although reports of the great adaptability of these patients, to dentures, have been published, the restraints of the removable design on the lifestyle of the young patient are not mentioned. Many authors recommend precautions that should be practiced when contemplating implants for the growing patient.[18-25] Cronin emphasized the use of a site-specific approach when treating young patients.[18-20] He advocated that the anterior mandible in growing patients suffering from anodontia be the only recipient site for dental implants because growth is completed early at that site for this particular group of patients. Kearns[21] and Bergandel[23] supported his recommendations. Bergandel *et al*.[23] empasized the early diagnosis of ED because many patients are proceeding with prosthodontic treatment without a prior diagnosis.

Tallgren reported that the annual mean crestal bone loss, in denture wearers, in the anterior mandible is 0.4 mm.[26] If the prosthetic rehabilitation consists of an implant –retained overdenture then the annual bone loss is 0.1 mm or 0.5 mm over 5 years.[27-29] One study reported that if instead of an overdenture, a mandibular implant supported complete denture is used, then not only the bone loss is minimized but also significant growth of the mandible can result.[30] Finally, Johnson *et al*,[9] reported on the effect of early implant treatment in stimulating halted bone growth. They suggested that more studies are necessary to evaluate the effect of implant treatment in young patients on the development of the stomatognathic system.

Implant placement for adult patients requires proactive planning. Treatment planning for young patients requires even more precautions.[21] Important factors to consider regarding implant placement in the growing patient are the possibility of implants becoming embedded or relocated because of the development of the stomatognathic system, the effect of the prosthesis on growth, and the maintenance of the prosthetic rehabilitation.[19]

Implants have proven to be effective on the treatment of edentulism in adults.[31-33] They are expected to enhance function in young edentulous patients as much as in adults while sparing them of the need to remove the prosthesis. Osseointegration success rates of implant surgery in children are higher for the mandible (91-92%) than for the maxilla (71-86%).[24] The high success rates for the mandible constitute implant surgery almost as predictable as for adults, whereas, the lower results for the maxilla require careful case selection.

In this clinical report, we explain the rationale for the treatment plan chosen for a young patient with ED and anodontia. Issues regarding implant placement are also discussed.

CLINICAL REPORT

A 5-year-old boy with ED presented to The University of Texas Pedodontic Clinic for prosthetic rehabilitation. As mentioned above, any structure originating from the ectoderm can be abnormal in ED. This particular patient presented with anodontia; xerostomia; dry, scaly skin; dark pigmentation around the eyes; sparse, very fine scalp hair; flattened facial appearance; and aged profile with increased nasolabial fold (Figure 1).

Intraoral examination revealed complete absence of teeth, xerostomia, and underdeveloped maxillary and mandibular ridges (Figures 2 and 3). Attached mucosa was missing from certain areas, especially where the frenums were attached. The panoramic radiograph confirmed the complete absence of tooth buds and the extreme underdevelopment of the maxillary and mandibular bones (Figure 4).

The objectives of treatment regarding the dental condition were the preservation of bone; the early development of normal mechanisms of chewing, speaking and swallowing; the establishment of normal facial characteristics and smile; the development of

Figure 1. Patient presented with anodontia, xerostomia, dry skin and dark pigmentation around the eyes.

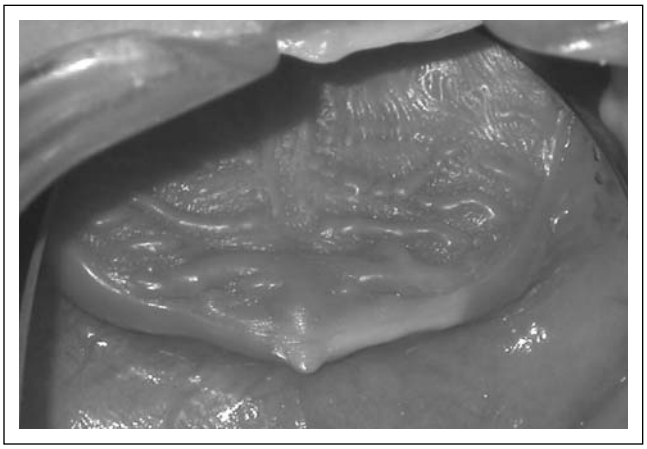

Figure 2. Intraoral examination revealed complete absence of maxillary dentition and an underdeveloped maxillary ridge.

Figure 3. The mandible was underdeveloped with anodontia.

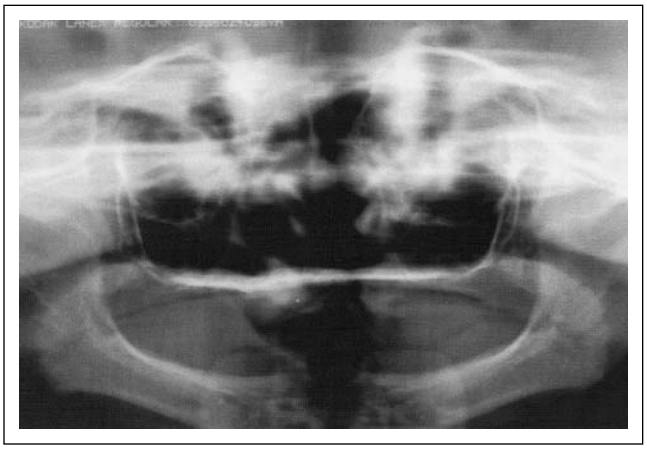

Figure 4. The Panoramic radiograph revealed absence of toothbud and abnormal osseous architecture.

Figure 5. The frontal profile with intraoral prostheses at resting position.

Figure 6. The frontal profile with patient smiling.

Figure 7. The prosthetic arrangement with appropriate lip contouring and occlusion.

Figure 8. The facial profile with prostheses.

normal emotional and psychologic profile; and the fitting of a functional prosthesis with adequate retention, stability, and support.

It was decided that a set of complete dentures would be fitted to serve the patient for an interim period of 1 year. Conventional techniques, as would have been followed for an adult patient, were followed for fabrication of the complete maxillary and mandibular dentures, with some deviations regarding the shape and shade of the teeth. More specifically, the tooth mold used was selectively ground to resemble primary teeth, and shade A1 was selected. Due to the reduced height of the alveolar ridges, denture adhesive was deemed necessary to improve retention of both maxillary and mandibular dentures. The prosthodontic intervention significantly improved the appearance of the young patient (Figures 5, 6, and 7), establishing a more normal profile (Figure 8).

The patient will be monitored for 1 year to determine the need to refit or remake his complete dentures. This will be a transitional period during which the patient's response to his rehabilitation will be evalutated. Complications of the treatment (e.g., discomfort, and sore spots) are expected and will be partially correlated with the dry mouth of the patient.

At age 6, implant placement will be re-evaluated. It is expected that by then the height of the mandible will have increased due to bone apposition on the lower border.[10] Initial evaluation of the panoramic radiograph showed enough height at the midsymphyseal region for implant placement of at least two, 8-mm fixtures. After palpation, the width seem to be marginally adequate, but any deficiency can be succesfully addressed during surgery with guided bone regeneration. Prosthetic options would be an implant-retained overdenture or an implant-supported complete denture.

DISCUSSION

After the age of 2, children begin establishing normal speaking and swallowing skills. Teeth play an important role in both functions as well as in the normal face profile and esthetics. Children are expected to have full primary dentition at 2 years of age; therefore, it is paramount that prosthetic treatment begin at that age. [22,34,35]

The young patient in this report presented 3 years after this period, at age 5. However, consequences of the delay in speech, nutrition, and growth as well as in psychological development were difficult to assess. Nevertheless, prosthetic treatment was initiated immediately.

An implant-retained overdenture or an implant-supported complete denture for that patient will significantly improve his quality of life. His extemely poorly developed mandible is not an appropriate substructure for conventional removable prosthetic rehabilitation. Also, his xerostomia is another factor guiding the treatment plan towards an implant-supported prosthesis.

There are no reports in the literature that relocation of implants placed in the anterior edentulous mandible is a problem, even if implants are placed as early as age 7. Minor lingual rotation is expected, but not to the degree that it might compromise future prosthetic rehabilitation. Selection of angulated abutments may help alleviate this problem. All other oral sites are considered to be sites at which implants might be relocated in such a degree due to growth patterns that will become unrestorable.

Implants can become embedded only if they are placed close to adjacent teeth. In cases of hypodontia and oligodontia, implants should not be placed adjacent to natural teeth in the growing patient because in the course of time, they will function as

ankylosed teeth and will become buried into the bone. The patient described here presented with anodontia. Patients with anodontia are not at risk of ankylosed teeth because there is no alveolar bone; therefore, no bone apposition is occuring on the crests of their ridges.

Diminished bone growth is another characteristic of ED. Further resorption of the supporting hard tissues will further compromise the prosthetic rehabilitation. Implants may assist in preserving alveolar bone. However, preserving alveolar bone should be achieved without compromising growth. The mandibular implant-supported complete denture does not require a split design at the midline in contrast with a maxillary fixed prosthesis, which must be split in the midline.The anterior width of the mandible stabilizes early in patients with anodontia; therefore, the growth pattern of the mandible will not be encroached.[19]

Another consideration would be the minimum dimensions of the prosthesis as the requirements for restorative materials might be incompatible with the maximum opening of a 7-year-old patient. At that age, the average maximum oral opening would be 30 to 40 mm. Also, the technical aspects of implant placement might be complicated because of the normal, but small maximum oral opening in young patients. This factor can be challenging, because the smallest height for a pilot drill would be around 30 mm. Also, bone quality and quantity justify implant placement only in the anterior mandible unless extensive bone grafting techniques are used. It is recommended that all these procedures be performed by experienced specialists.

Except for the risks involved in implant surgery, there is also an increased amount of maintenance introduced with the implant-supported complete denture design. Nevertheless, the advanced biomechanical properties of this rehabilitation offer one competitive advantage for the growing patient, that is the avoidance of a removable design that stimulates adult images and habits. It is paramount for the young patient to be as close to normal as possible, and only the fixed detouchable bridge can accomplish such normalcy. This maintenance approach is costly, but the authors believe that the cost is justified. More adjustable implant-retained overdenture designs might be appropriate in selected cases. Parents must be actively involved in the treatment decision and must agree to assume responsibility for the increased time and cost associated with the treatment.

SUMMARY

The considerations involved in determining the optimal treatment protocol for a 5-year-old boy with ED and anodontia were presented in this report. The key consideration is that the anterior mandible in edentulous, young patients completes most of growth by age 6. This provides the stable environment, necessary for considering dental implant solutions in the treatment plan. Complete dentures have been fitted, and implant placement has been deferred until the child reaches the age of 6.

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