

Treatment Considerations for a Patient with Hypohidrotic Ectodermal Dysplasia: A Case Report

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Abstract

Aim: This clinical report describes the oral rehabilitation of a 6-year-old male ectodermal dysplasia (ED) patient diagnosed with hypodontia.

Background: ED is a hereditary disease characterized by a congenital dysplasia of one or more ectodermal structures and their accessory appendages. Common manifestations include defective hair follicles and eyebrows, frontal bossing with prominent supraorbital ridges, nasal bridge depression, and protuberant lips. Intraorally, most common findings are anodontia or hypodontia, conical teeth, and generalized spaces. The patient may suffer from dry skin, hyperthermia, and unexplained high fever as a result of deficiency of sweat glands.

Report: A six-year-old boy who exhibited many of the manifestations of ED as well as behavioral problems and a severe gag reflex. The treatment was designed to improve his appearance and oral functions and included the fabrication of several removable prostheses and acid-etched composite resin restorations during his growth and development.

Summary: Young patients with ED need to be evaluated early by a dental professional to determine the oral ramifications of the condition. When indicated, appropriate care needs to be rendered throughout the child's growth cycle to maintain oral functions as well as to address the esthetic needs of the patient. This clinical report demonstrates that removable partial dentures associated with direct composite restorations can be a reversible and inexpensive method of treatment for young ED patients.

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Keywords: Ectodermal dysplasia, ED, hypodontia, prosthodontic treatment, ectodermal structures

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Introduction

Ectodermal dysplasia (ED) is a hereditary disorder that can affect several ectodermal structures.¹ These structures may include: skin, hair, nails, teeth, nerve cells, sweat glands, parts of the eye and ear, and parts of other organs.² Several types of ED are known. Freire-Maia and Pinheiro described more than 100 different taxonomic groupings of ED across a wide spectrum of clinical situations.^{1,3-5} Each type of ED involves different structures and severity of the disorder varies from patient to patient.

In general, ED syndromes have been described as a group of disorders of morphogenesis displaying two or more of the following signs and symptoms:^{3,5-8}

1. Trichondysplasia (abnormal hair)
2. Abnormal dentition
3. Onchondysplasia (abnormal nails)
4. Dyshidrosis (abnormal or missing sweat glands)

The most reported ED syndrome is x-linked hypohidrotic (anhidrotic) ED (Christ-Siemens-Touraine syndrome) which affects one to seven individuals per 10,000 with males afflicted more frequently than females.^{3,4,5,7,9,10,11}

Patients with hypohidrotic ED generally have prominent supraorbital ridges, frontal bossing, and a saddle nose. The maxilla may be under developed and the lips thick and prominent. The nose may appear pinched and the alae nasi hypoplastic. The patient may resemble an edentulous old person.^{10,11} The skin is usually dry, scaly, and easily irritated as a result of poorly developed or absent oil glands. Sweat glands can be absent, few in number, or nonfunctioning which may result in a high body temperature. Scalp hair may be absent, sparse, very fine pigmented, or abnormal in texture. Eyebrows, eye lashes, and other body hair may also be sparse or absent. When hair is present, it may be fragile, dry, and generally unruly because of the lack of oil glands.² Finger and toe nails are usually normal.¹¹

Orofacial characteristics of this syndrome include anodontia or hypodontia, hypoplastic conical teeth, underdevelopment of the alveolar ridges, frontal bossing, a depressed nasal bridge, protuberant lips, and hypotrichosis.^{12,13} Teeth in the permanent dentition are frequently small, conical, tapered (peg-like), and widely spaced. Lack of alveolar growth may be associated with this condition and frequently results in increased interocclusal distance which allows optimum artificial tooth placement.¹⁴ Patients may present with a marked mandibular protrusion on closure or a deep vertical overlap. Depending on the severity of the condition, various prosthodontic treatments are available to improve appearance, mastication, and speech.¹⁵

Children with ED usually have a normal mentality and life expectancy, and their facial appearance warrants professional concern for their emotional well being and social progress.^{10,16} Tanner¹⁷ states ectodermal dysplasia with an abnormal appearance may affect normal social and psychological development in young patients. Functional needs also must be considered since the difficulty these children experience in masticating may cause nutritional problems.^{10,16} Therefore, dental care for ED patient is important. This clinical report describes the oral rehabilitation of a 6-year-old male ED patient diagnosed with hypodontia.

Case Report

Diagnosis

A 6-year-old male with hypohidrotic ectodermal dysplasia was referred by his physician to the Faculty of Dentistry at Gazi University for an examination due to the non-eruption of his permanent teeth. His parents consulted with a general dentist when the boy was four-years-old because his deciduous teeth had not erupted and he had difficulty speaking and chewing. Treatment was not possible at that time due to behavioral problems at that age. At age six, the boy became uncomfortable with the appearance

of his teeth and very reluctant to smile. He was now motivated to seek help from a dentist.

A clinical examination revealed only two deciduous teeth and one permanent tooth in the maxilla; the others were missing. The three existing teeth were extremely tapered (peg-like) and widely spaced (Figure 1).

All mandibular deciduous teeth were missing and the color of alveolar mucosa and gingiva was normal, but the alveolar ridges were rather atrophic except in the areas where teeth were present (Figure 2).

The patient exhibited typical characteristics of hypohidrotic ED, including a saddle nose, fine sparse hair, everted lips in profile, and

hypodontia. The skin of the body was dry and atrophic. However, the shape of the fingernails and toenails appeared normal. During the physical examination he showed no signs of mental problems and his vital signs were normal.

A panoramic radiograph revealed four permanent teeth (central and lateral incisors) and two deciduous anterior teeth in the maxilla. In the mandible all of the posterior and anterior permanent tooth germs were missing except for the mandibular canines (Figure 3).

Treatment

Initially, a maxillary removable partial denture and mandibular complete denture were fabricated for the patient (Figures 4 and 5).



Figure 1. Intraoral view of maxilla.



Figure 2. Intraoral view of mandible.

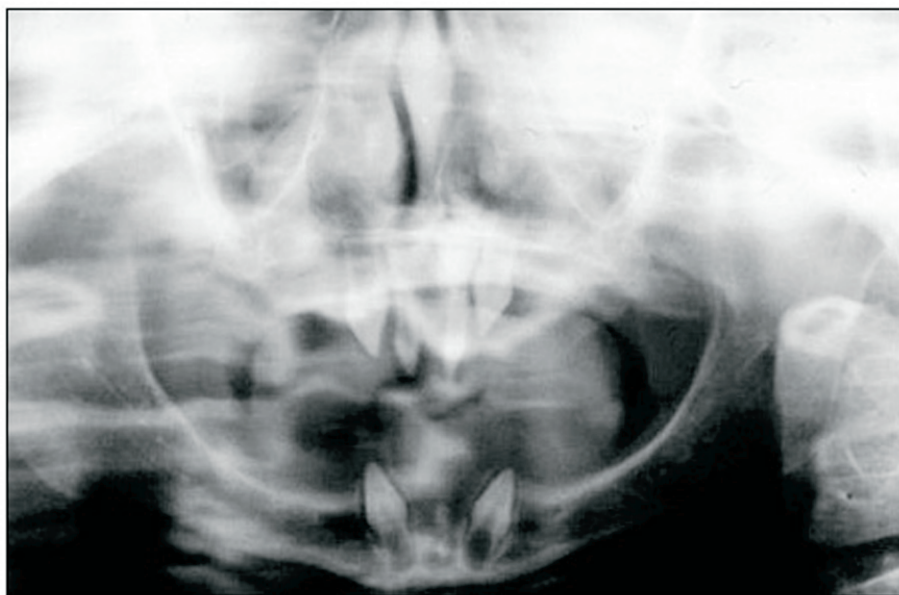


Figure 3. A panoramic radiograph revealing four permanent teeth (central and lateral incisors) and two deciduous anterior teeth in the maxilla.

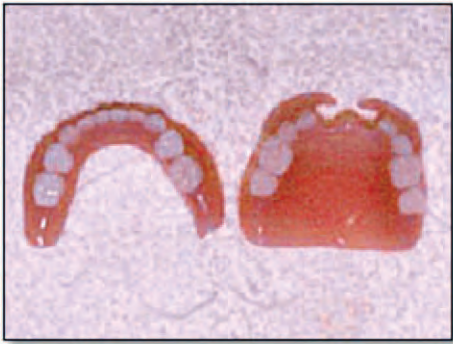


Figure 4. Maxillary partial and mandibular complete dentures.



Figure 7. Composite restorations of all erupted teeth when he was eight years old.



Figure 5. Dentition after insertion of the protheses.



Figure 8. Intraoral photograph with insertion of the acrylic partial dentures. Dentures restored oral function and provided facial and psychosocial support.



Figure 6. Intraoral photograph with restored central incisors and the new acrylic partial dentures.

The dentures greatly improved his facial appearance and jaw functions. After six months, the patient complained of his appearance created by the wide distance between his peg-shaped deciduous maxillary central incisors. Acid-etched composite resin restorations of the teeth were fabricated using a strip crown and the dentures were remade (Figure 6).

The removable partial denture was fabricated in conjunction with the composite restorations to

increase the vertical dimension and to address the patient's esthetic concerns related to the reduced number and the peg-like shape of the teeth.

At age seven, the deciduous left maxillary central incisor was lost and the eruption of his permanent maxillary lateral incisors and right mandibular canine was complete. As a result, the composite restorations were redone on his newly erupted teeth and a new prosthesis was fabricated. The dentures were adjusted and remade at intervals to allow for the eruption of the permanent teeth.

At age eight, the eruption of left mandibular canine was completed and the dentures could no longer be used. The left mandibular canine was restored with composite resin and the mandibular and maxillary arches were restored with new removable partial dentures (Figures 7 and 8).

The patient and his mother were instructed to handle the dentures carefully and to have the

patient wear them at all times except while tooth brushing and sleeping. The dentures were adjusted carefully and regular recall appointments were scheduled at two-month intervals to make necessary adjustments and monitor the oral hygiene. The patient adapted well to the dentures, and the treatment improved his masticatory and speech functions, esthetics, and established a more favorable plane of occlusion. The patient's social confidence also improved significantly as a result of treatment. The patient will continue to be evaluated regularly. When growth is complete, a more permanent treatment can be performed.

Discussion

ED is usually a difficult condition to treat with prosthodontic restorations because of the typical oral deficiencies and the young age of when they are evaluated for treatment. Therefore, when treating a child with ED, it is important to motivate both the child and his or her parents prior to treatment and to work with them to ensure compliance.⁶

In the present case the patient's increasing age made his social development and social interaction increasingly important. Reports indicate children rejected by their peer groups are more likely to become aggressive, delinquent, and are more likely to remain unaccepted by their peers and may experience mental health problems in adulthood.¹ Therefore, successful treatment of the present case can be expected to assist the patient both physically and psychologically. The boy's attitude, self-confidence, and peer group interaction showed signs of significant improvement during treatment.

The successful use of any prosthesis is dependent on the cooperation and communication between the dental team and the patient and his parents. For example, great care will need to be taken to maintain the boy's oral hygiene in order to benefit from the long-term treatment plan. In addition, the boy and his parents must be educated and motivated about the dental problems related to his genetic and psychologic conditions.^{1,6}

Clinical reports have stated the importance of prosthetic dental treatment in patients with anadontia or hypodontia for physiological and psychosocial reasons.^{13,16} Treatment of the ED

patient generally includes a removable and/or fixed partial denture, a complete denture prosthesis (overlying affected teeth when the vertical dimension of occlusion permits), and an implant retained prosthesis when indicated.² These treatment approaches can be used individually or in combination to provide an optimal result. The proper sequencing of treatment is important to achieve the desired function and esthetic results. Because of early-age intervention and the need to easily modify the intraoral prosthesis during rapid growth periods, a removable partial denture or complete denture prosthesis is indicated initially.²

Hypodontia is associated with lack of development of the alveolar ridge and results in less volume of bone for support of a conventional prosthesis. Therefore, an implant supported prosthesis can be recommended for these patients. The placement of endosseous implants in locations favorable for subsequent restorations may be difficult, require bone grafting, and the placement of implants in growing children is not recommended as a routine practice.^{18,19} The short-term survival data reported by Guckes et al.²⁰ suggested it is possible to successfully place dental implants in male and female patients of different ages with ED and congenitally missing teeth. However, they stated a careful evaluation of each patient is necessary to determine the bone volume available for implant placement.²⁰

Due to the boy's young age, on-going development of the jaws, and insufficient quantity of alveolar bone, multiple implant placements were not possible. The bone height and width was not sufficient for implant insertion without using an advanced surgical approach. Application of removable dentures may be the only restorative option for this patient. When growth is completed, treatment planning may include an implant-retained prosthesis.

As a conclusion, children and adolescents with ED often require complex prosthetic treatment to manage the hypodontia. Oral rehabilitation is important from functional, esthetic, and psychological perspectives. Oral rehabilitation involves a number of challenges which are further complicated by the growth and development of the child, variation in tooth development and eruption, the type of age-appropriate prosthesis, and the

planning and timing of treatment. Therefore, a multidisciplinary team approach is recommended for successful treatment of affected individuals. The team should include at least a pediatric dentist, a prosthodontist, orthodontist, and an oral and maxillofacial surgeon.⁶

Summary

Young patients with ED need to be evaluated early by a dental professional to determine

the oral ramifications of the condition. When indicated, appropriate care needs to be rendered throughout the child's growth cycle to maintain oral functions as well as to address the esthetic needs of the patient. This clinical report demonstrates that removable partial dentures associated with direct composite restorations can be a reversible and inexpensive method of treatment for young ED patients.

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