MULTIDISCIPLINARY TREATMENT APPROACH OF PATIENT WITH ECTODERMAL DYSPLASIA

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Abstract

In this paper dental treatment of a 10-year-old male patient with ectodermal dysplasia was presented. A 10-year-old male patient with ectodermal dysplasia referred to Gulhane Medical Academy, Department of Pedodontics. Radiographic evaluation revealed that several teeth germs were absent. The treatment was planned by a multi-disciplinary team of pediatric dentist, orthodontist and prosthodontist. At first the conical central incisors were reshaped with direct composite restorations. Minimal orthodontic intervention was applied to close the diastema. To improve his facial esthetics, speech and oral function, removable prosthesis were applied.

The results were significant improvements in speech, masticatory function, and facial esthetics, contributing to the development of normal dietary habits, and the improved and more rapid social integration of the patient.


Keywords: Ectodermal Dysplasia, Congenitally Missing Teeth, Dental Prosthesis.

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Introduction

Ectodermal dysplasia (ED) is a hereditary disease characterized by anomalies in the structures of ectodermal origin¹. The disease effects skin, saliva, sebaceous and sweat glands (anhidrosis or hypohidrosis), hair (atrichosis or hypotrichosis), nail and teeth (anodontia or hypodontia)². In some cases, depressed immune system may cause an increased susceptibility to certain infections or allergic reactions and effected children experience respiratory infections, chronic nasal passages inflammation, eczema and asthma³. ED was firstly identified in 1860’s by Darwin⁴. A hundred years after Darwin’s definition, the disease now have more than 150 subtypes and all of them appear to be genetic in etiology⁵. ED have two different forms named Clouston syndrome (autosomal inherited form) and Christ-Siemens syndrome (hypohidrotic form)⁶. Recently a new classification has been proposed based on the alterations in the proteic molecular functions that lie behind it⁷. The disease affects both males and females. Prevalence of ED appraised to be 7 in 10.000 live births⁸. Hypohidrotic ED is the most common ED (%80) and is often inherited as an X-linked disorder⁹,¹⁰. The disorder is fully declared in man only, nevertheless women who carry a single copy of the disease gene may exhibit some of the symptoms and findings associated with the disorder¹¹. Spontaneous gene mutation is also possible¹². Most of the patients have normal life expectancy and normal intelligence. Early diagnosis is important for these patients because in early infancy, the lack of sweat glands may lead to hyperthermia, followed by brain damage or death if unrecognized².
ED patients have characteristic facial abnormalities such as prominent forehead, sunken nasal bridge (saddle nose), thick lips, large chin and darkly pigmented skin around the eyes. On most of the patients’ body, the skin may be abnormally thin, soft and dry with an abnormal lack of pigmentation. The most characteristic findings of ED are hipodontia and abnormal shape of teeth.

In addition to the absence of teeth, ED can also lead to underdevelopment of the jaws. This kind of craniofacial deformities can be effect physically and psychologically to the ED patient and may inhibit normal social interactions. The oral rehabilitation of patients needs a multidisciplinary approach. Additional considerations, such as the patient's age, stage of growth, inherent anatomic deficiencies present in conjunction with the missing teeth, soft tissue defects, existence of malformed dentition, severe diastemas and psychological status, must be considered. Prosthetic treatment is a great value to these patients from the functional standpoint as well as for psychological and psychosocial reasons.

Case Report

A 10-year-old male patient with ectodermal dysplasia referred to Gulhane Medical Academy, Pediatric Dentistry Department because of nutrition, speech and esthetic problems due to the lack of teeth. The patient exhibited the classical features of ED including abnormally thin, dry and soft body skin, sparse hair, eyelashes and eyebrows, characteristic face (so-called “old man face”), a saddle shaped noise and hypohidrosis. He was the first child of consanguineous parents. There was no family history associated with his condition. The patient was mentally normal.

A comprehensive clinical and radiographic evaluation was performed. In extraoral examination a facial physiognomy typical of ED was observed. The facial height indicated an extremely decreased vertical dimension of occlusion. The intraoral examination revealed that only six teeth (two anterior and four posterior permanent maxillary teeth) and bone atrophy of the alveolar ridges, on both the maxilla and the mandibula. Some of the existing teeth were short, conical and pointed (Figure 1). In addition oral mucosa was slightly dry and sticky. Radiographic examination confirmed the clinical findings. The treatment was planned by a multi-disciplinary team of pediatric dentist, orthodontist, prosthodontist and was clarified to both the patient and his parents.

![Figure 1. Intraoral appearance of the patient.](image1)

After the permission was received from the parents, the conical central incisors were etched with a 37% phosphoric acid gel for 20 seconds, the area was rinsed for 20 seconds then dried gently with air. An adhesive (Excite DSC, Ivoclar Vivadent AG, Schaan, Liechtenstein), was applied according to the manufacturer’s instructions. Finally, to meet esthetic requirements for the anterior segment, the dental restoration (3M ESPE, USA) was completed with composite using the incremental technique and occlusal adjustment was performed. (Figure 2,3).

![Figure 2. Conical central incisors were reshaped with direct composite restorations.](image2)

Minimal orthodontic intervention was applied to close the diastema (Figure 4,5). The fixed appliance was fitted to the upper central incisors and power chain was applied to begin...
space closure along a 0.016 x 0.022-inch stainless steel (SS) wire. Treatment phase, more root uprighting had been carried out to the central incisors using second-order bends in the archwire. Four months later, enough closure of the central incisors and good root parallelism had been achieved.

To improve his facial esthetics, speech and oral function and to limit the resorption of the alveolar ridges removable prosthesis were applied (Figure 6,7). After this application hygiene instructions for the dentures were given to the patient and his parents. Further recalls have taken place every week for 1 month and then every 3 months. The patient had no discomfort and seemed to be adapting well on the following visits.

Discussion

EDs are a heterogenous group of disorders characterized by a group of findings involving defects of two or more of the teeth, skin
and appendageal structures including hair, nails and eccrine and sebaceous glands\textsuperscript{17}. It is commonly transmitted as an X-linked recessive disorder. However rarely autosomal recessive and autosomal dominant inheritance have also been seen\textsuperscript{18}.

In most of ED patients in addition to the delay in teething, the teeth are abnormal in shape and structure\textsuperscript{19}. Not only is the shape abnormal but also the number. Some cases have congenitally anodontia\textsuperscript{14}. The main goals of dental treatment are to improve esthetics and oral functions especially mastication and phonetics\textsuperscript{20}.

Early prosthetic therapy for children with ectodermal dysplasia may require the coordinated efforts of a multi-disciplinary team of pediatric dentists, orthodontists, prosthodontists \textsuperscript{21}. Although removable prosthesis is most frequently reported treatment modality for the dental management of ED as used in the present case\textsuperscript{22}, implants should be an alternative treatment in patients with ED. Patients with ectodermal dysplasia-because of tooth absence-have reduced alveolar bone with "knife-edge" morphology, making implant reconstruction a challenge. Therefore, patients frequently require bone grafting and sinus-lift procedures\textsuperscript{23}.

Kramer et al.\textsuperscript{24} reported the case of a boy at the age of 8 years with ectodermal dysplasia who exhibited a severe hypodontia and who was treated with implants inserted into the anterior mandible and recommended the early insertion of dental implants in children with severe hypodontia. Growing implant patients present a unique age-related problem regarding implant positioning and prosthetic outcomes. Guler at al.\textsuperscript{25} reported that dental implants with or without bone grafts can be used in patients over 12 years of age. Because of the economical situation of our patient, removable prosthesis applied instead of implant therapy.

According to Vieira at al.\textsuperscript{13}, prosthetic management is important for ED patients because it provides good esthetics, phonetics, and masticatory comfort, maintains healthy supporting tissues throughout a lifetime of denture wearing experience and helps the patient develop a good psychologic self-image. The prosthesis must be periodically modified in young adults as alveolar growth, erupting teeth and rotational jaw growth change\textsuperscript{21}. In this case the prosthesis was replaced after 12 months because of skeletal growth, and new ones applied.

In some cases before the application of removable prosthesis, restorative and orthodontic treatment may be necessary to improve facial esthetic\textsuperscript{14}. In present case the conical central incisors were reshaped with direct composite restorations and minimal orthodontic intervention was applied to close the diastema.

It is important to take the motivation of the patient and parents as well as the patient's acceptance for dental treatment into consideration. In all the stages of the prosthetic treatment behavior management should be performed by “tell-show-do” method\textsuperscript{20}. After the application of removable prosthesis we gave hygiene and utilization instructions for the dentures to the patient and his parents. The patient learned to use his prosthesis quickly and usage and condition of the prosthesis were evaluated at each recall appointment.

The main difficulty consisted in dental treatment of ED patient is to keep the mandibular denture in mouth. To improve and facilitate the denture’s adaptation dentist may advice to patient to use a denture adhesive paste for the first few days\textsuperscript{16}. In our case the patient had no discomfort and adapting of the prosthesis was well on the first visit. In further recalls only discomfort areas were relieved. Retention and stabilization of the dentures were clinically acceptable.

**Conclusions**

The primary goals of dental treatment of patients with ED are enhancing esthetics and improving masticatory function. The treatment requires the cooperation of a multi-disciplinary dental team of pediatric dentist, orthodontist and prosthodontist.

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**Declaration of Interest**

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