SUMMARY

This clinical report describes the characteristics and prosthodontic restoration of 2 young male patients with anhidrotic ectodermal dysplasia. With proper care and prosthodontic treatment, the patients can enjoy a relatively normal life. It is important that the patients and their parents fully understood the dental problems related to their physiologic and psychological conditions. The need for continued dental treatment is stressed.

Keywords: Ectodermal Dysplasia; Removable Dentures

CASE REPORT (CR)

Balk J Stom, 2007; 11:141-144

Complete and Removable Partial Dentures for Patients with Ectodermal Dysplasia: 2 Case Reports*

Introduction

Ectodermal dysplasia (ED) is a hereditary disorder characterized by abnormal development of certain tissues and structures of ectodermal origin. It has been described as a group of morphogenesis displaying 2 or more of the following signs and symptoms: (1) tricho-dysplasia (abnormal hair), (2) dental anomalies, (3) onycho-dystrophy (abnormal nails), and (4) dyshidrosis (abnormal or missing sweat glands). Congenital malformation of teeth, hair, nails, or sweat glands may occur either as isolated malformations or as part of an ED syndrome. Freire-Maia and Pinheiro classified ED into 11 possible groups, based on all possible combinations of 2 or more defects. The most frequently reported ED syndrome is X-linked hypo-hidrotic dysplasia, also known as Christ-Siemens-Touraine syndrome, which affects 1 to 7 individuals per 10,000 live births. Clarke suggested that the X-link trait is transmitted with the gene being carried by the female partner and manifested in the male partner. However, an anhidrotic ectodermal syndrome may occur in a family without previous history of this disease because of gene mutation. Female carriers of this dysplasia may have a variable degree of clinical involvement, ranging from non-detectable signs to clinically significant hypodontia, hypotrichosis, and unilateral hypoplasia of the breasts.

The most common oral characteristic is hypodontia or anodontia in many cases, reflecting the complete suppression of dental ectoderm. Because of the lack of teeth and resultant loss of vertical dimension, the lips are protuberant, the vermilion border is indistinct, and the alveolar process does not develop in the absence of teeth and, hence is missing. Incisors, canines, and premolars, when present, often have conical crowns and the oral mucosa often appears dry. Histopathologic evaluation shows aplasia of the labial, buccal and lower respiratory glands. The pharyngeal and laryngeal mucosa may be atrophic, resulting in dysphonia.

Early and extensive dental treatment is needed throughout childhood because of the absence of most of the deciduous and permanent teeth. A multidisciplinary team composed of a paediatric dentist, a prosthodontist, an orthodontist, and an oral and maxillofacial surgeon has been advocated in some reports and recommended to ensure proper treatment of young ED patients. The following report describes the characteristics and prosthodontic treatment of 2 patients with anhidrotic ectodermal dysplasia.

Case Reports

A 14-year-old boy was referred to the Department of Prosthodontics (Dicle University, Faculty of Dentistry),
for prosthodontic rehabilitation due the lack of teeth. A review of the medical and dental histories revealed that the diagnosis of anhidrotic ED was established in the early childhood. The patient exhibited typical characteristics of the anhidrotic ectodermal dysplasia including protuberant lips, saddle nose, fine sparse hair, scant eyelash and eyebrow, and pigmentation around the eyes and mouth (Fig. 1). His parents did not exhibit evidence of ectodermal dysplasia, and none of his relatives were known to have a condition similar to this.

Figure 1: Facial appearance of a 14-year-old boy (Case 1)

An oral examination revealed that the patient had a loss of vertical dimension of occlusion with dry oral mucosa, protuberant lips, indistinct vermilion border, and underdeveloped alveolar ridges, which was small and thin (Fig. 2). He had the left and right permanent first molars in the maxilla. There were no teeth in the mandible; patient’s tongue occupied the space between posterior alveolar ridge (Fig. 3).

The second patient was a 9-year-old boy with anhidrotic ED. He had the same a review of medical and dental histories (Figs. 4 and 5). However, intraoral examination revealed an absence of the left and right permanent first and second molars in the maxilla. There were no teeth in the mandible. Patient’s tongue occupied the space between posterior alveolar ridge (Fig. 6).

Figure 4: Facial appearance of a 9-year-old boy (Case 2)

Figure 2: Profile appearance of a 14-year-old boy (Case 1)
Prosthodontic Treatment

Maxillary removable partial dentures and mandibular complete dentures were fabricated in the conventional manner. A silicone impression material (Lastic Xtra, Kettenbach, D-35713, Eschenburg, Germany) was used for this young patient with dry mucosa, because silicone is highly biocompatible, clean, and pleasant for the patient. It is highly elastic, and the setting time is fast and can be controlled. It is hydrophobic and requires an extremely dry field. Anterior deciduous plastic teeth were used for aesthetic reasons. Anatomic, 33-degree, plastic posterior teeth (Major Prodotti Dentari S.p.A, Moncalieri, Italy) were used to develop bilateral-balanced occlusion. Maxillary and mandibular trial dentures were verified, and protrusive record was obtained.

After the approval of the tooth arrangement by the patients and their parents, the waxed dentures were processed in a heat-polymerized denture base resin (Melliodent, Heraeus Kulzer Ltd, Germany). The dentures were inserted, and the patients and their parents were instructed on the proper maintenance of the oral hygiene (Figs. 7 and 8). The patients were then scheduled for
1-week, 2-week, 1-month, 3-month, and 6-month follow-up appointments; no major complication were observed for one and half years.

Discussion

The decision to begin treatment should be made by the treating dentist, along with the parents and the patient. Because individuals with ED are quite young when they are first treated, the treating dentist should have some knowledge and ability in the behavioural management of paediatric patients.11

Numerous clinical reports have demonstrated the importance of prosthetic dental treatment in ED patients for physiologic and psychosocial reasons.12 Prosthodontic treatment of ED can include fixed, removable, or implant-supported dentures.13,14

Fixed prosthodontic treatment is seldom used exclusively in the treatment of ED, primarily because many afflicted individuals have a minimal number of teeth. In addition, ED patients are often quite young when they are first treated, and fixed partial dentures with rigid connectors should be avoided in young, actively growing patients.8

Removable prosthodontics is the most frequently advised as a treatment modality for the dental management of patients with ED.5 Because anodontia or hypodontia is typical in individuals with this condition, complete dentures, partial dentures, or overdentures are often part of the treatment provided. When there are teeth present for the support, overdentures are a desirable treatment option for these patients.11,15

Previous studies have reported situations in which endosteal implants were successfully used in the prosthetic management of ED.7,13,16,17 A number of studies indicate an improvement in physiologic and psychosocial function of adult patients with an implant-supported denture when compared with their condition before that, or with an edentulous control group with complete dentures.18

In this clinical report, the consideration of osseointegrated implants was deferred because of the age and potential growth of this young patients. Also, overdentures were not evaluated because presented teeth were molars, available for retention of removable partial denture. Common problems of ED patients who used removable partial dentures and/or complete dentures usually related to loss of dentures retention. In this clinical report, removable partial dentures and complete dentures provided acceptable results for aesthetics, psychological support, function and phonetics. In addition, the patients were observed without a major complication for one and half years.

References


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