

Anhidrotic Ectodermal Dysplasia: The Dental Perspective: A Case Report

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Abstract: Ectodermal Dysplasia is a large, heterogeneous group of inherited disorders, the manifestations of which could be seen in more than one ectodermal derivatives. These tissues primarily are the skin, hair, nails, exocrine glands and teeth. The most common form of ectodermal dysplasia is Anhidrotic Ectodermal Dysplasia. This case report describes a method of restoring function and aesthetics in a 8-year-old boy with removable acrylic prosthesis. Ectodermal dysplasia is both physically and emotionally devastating to patients. Treatment had major impacts on self esteem, masticatory function, speech and facial aesthetics.

Keywords: Ectodermal dysplasia; Hypodontia; Hypohidrotic dysplasia; Partial denture.

Introduction

Ectodermal Dysplasia represents a group of inherited conditions in which two or more ectodermally derived anatomic structures fail to develop. Thus, depending on type of ectodermal dysplasia, hypoplasia or aplasia of tissues may be seen¹. Patients with ectodermal dysplasia are characterized by hypoplasia or aplasia of structures such as skin, hair, nails, teeth, nerve cells, sweat glands, parts of the eye and ear and other organs.² Significant oral findings include hypodontia, loss of occlusal vertical development, protuberant lips and lack of normal alveolar ridge development³.

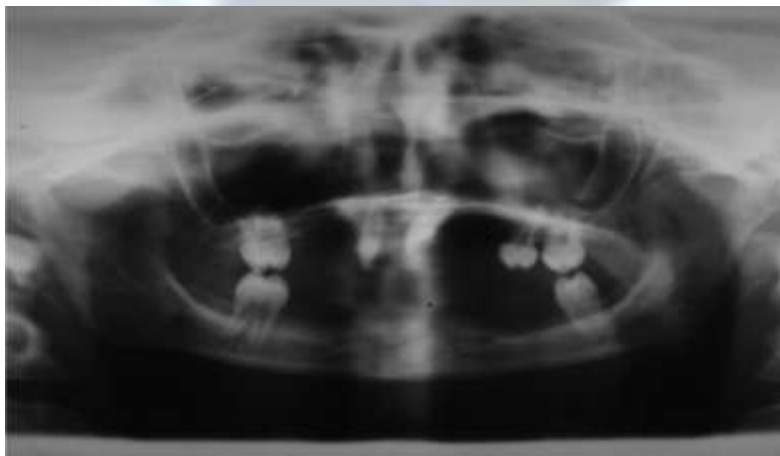


Fig 1: OPG

Thurman published the first case report in 1848 while term Ectodermal Dysplasia was coined by Weech in 1929. The frequency in a given population is highly variable with a prevalence of seven cases per 10000 live births⁴.

In cases where the salivary glands are hypoplastic or absent, varying degrees of xerostomia are expected⁵. Affected individuals typically display heat intolerance because of reduced number of exocrine sweat glands. These glands may be either absent, reduced in number, or nonfunctioning (hypohydrotic), which may result in elevated body temperature⁶. Ectodermal dysplasia might be inherited in any form of several genetic patterns including autosomal-dominant, autosomal recessive, and X-linked modes⁷. Although more than 170 different subtypes of ectodermal dysplasia have been identified, these disorders are considered to be relatively rare with an estimated incidence of 1 case per 100,000⁸.

Case report

The patient was 8 years old and the OPG showed presence of some teeth (Fig 1). Other features included pigmented nevus on right side of face with sparse hair (Fig 2), dry and scaly skin (Fig 3). Intraoral findings revealed presence of some teeth in maxillary and mandibular arch (Fig 4,5,6) The objectives of the oral treatment were to improve esthetics and correct oral functions, especially mastication, in order to finally allow the normal psychological development.

To accomplish these objectives we decided for a simple conventional prosthetic treatment. Alginate (Zelgan) was used for primary impressions due to its rapid-setting qualities and pleasant smell and taste and diagnostic casts were poured.



Fig 2: Pigmented nevus and Sparse hair

Custom trays were fabricated using self cure acrylic resin with 2 mm full wax spacer. Border moulding was done with a green stick compound. For the final impression a polysiloxane was used. The impressions were boxed to preserve the periphery created by the muscles. Occlusion rims constructed on the working casts were transferred in the mouth to establish the occlusal vertical dimension and centric relation. The vertical dimension was first determined by swallowing, checked by the rest position and associated free way space, and finally verified by means of phonetics. The vertical occlusal dimension was increased in order to establish better muscle tonus and lip support. Teeth selection was given a lot of importance for optimum aesthetic results. Acrylic teeth with reduced cusps inclines were used and the trial dentures were used. The dentures, processed with a conventional type resin, were placed into the mouth and the necessary adjustments were carried out. The patient was examined the next day (Fig 7,8). Retention and stabilization of the dentures were clinically acceptable. Discomfort areas were relieved. The management of the patient included instructions for the maintenance of oral hygiene and periodic recall visits. every 6 months to evaluate developing jaws and teeth eruption. In the delivery appointment, instructions were given to maintain proper oral hygiene. Continuous follow-ups every six months were planned for adjustment or replacement of old denture.



Fig 3: Dry and Scaly skin.

Discussion

There is no general cure for ED, but preventive treatment consisting in genetic counseling and prenatal diagnosis and many treatments addressing the symptoms are available^{9,10,11}. General treatment may require the coordinated efforts of a multi-disciplinary team of specialists who need to systematically and comprehensively plan an individualized treatment. The general treatment is symptomatic and depends on which ectodermal structures are involved^{11,12,13}. Medical care also improves esthetics and functions^{12,13}. Dental treatment is usually necessary. Optimal dental treatment requires: early and continuous dental interventions during many years, numerous phases of treatment, the involvement of a multi-disciplinary dental team (oro-maxillo facial surgeon, orthodontist, prosthodontist, pedodontist) and a marked financial effort of the patients and their families^{13,14}. The main goals of dental treatment in patients with ED are enhancement of functions (esthetics, mastication, phonetics) and psychosocial activities¹⁴. Dental treatment is directed towards the specific symptoms that are apparent in each individual^{14,15}. Treatment should be commenced as soon as possible in order to avoid possible resorption and atrophy of the alveolar ridges, and to control the vertical dimension, which can be severely affected by the total or partial lack of teeth^{16,17}. Conventional fixed prosthodontic treatment is seldom used exclusively in the treatment of ED primarily because afflicted individuals have a minimal number of teeth.



Fig 4: Intraoral View

When planning dentures in these patients, care should be taken to obtain a wide distribution of occlusal load fully extending the denture base. The remaining anterior teeth, due to their atypical conical shape, may not be suitable to stabilize

RPDs In addition, ED patients are often quite young when they are first treated, fixed partial dentures (FPDs) with rigid connectors should be avoided in young, actively growing patients. This is because rigid FPDs could interfere with jaw growth, especially if the prosthesis crosses the midline⁹ However, individual crowns may be given. The patient should use topical fluoride daily for prophylaxis against caries during the treatment. Other treatment options were described in the literature, consisting of a removable prosthesis, has shown also excellent results when the patient maintains proper oral hygiene^{16,17}. Nevertheless a fixed solution is closer to the normality and allows the young patient affected by ectodermal dysplasia to integrate more easily into the society. Oral rehabilitation of patients with ectodermal dysplasia is necessary to improve sagittal and vertical skeletal relationships during craniofacial growth and development as well as esthetics, speech, and masticatory efficiency¹⁸.



Fig 5: Maxillary Arch

Conclusion

This case report discusses management of a patient with anhidrotic ectodermal dysplasia with removable acrylic prosthesis. Dental rehabilitation aids the patient in developing proper speech, mastication, psychosocial benefits and gaining self confidence at an early age. Patients with ectodermal disorders often seek treatment from dermatologic surgeons but even an alert dentist can help diagnose the lesion and give the proper treatment for the benefit of mankind.



Fig 6: Mandibular Arch

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Fig 7: Post Operative intraoral view.



Fig 8: Post operative Frontal view

