Dental Management of Ectodermal Dysplasia

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Abstract
Ectodermal dysplasia is a hereditary disorder associated with abnormal development of embryonic ectodermally derived organs including teeth, nails, hair and sweat glands. Hypodontia of the primary and permanent dentition is the most common oral finding. Therefore, affected patients need dental prosthetic treatment. This case report presents a case of a patient affected by ectodermal dysplasia with hypodontia. Oral rehabilitation was accomplished with fixed prostheses. Treatment had major impacts on self-esteem, masticatory function, speech and facial esthetic.

Introduction
Hereditary ectodermal dysplasia represents a large group of conditions in which two or more ectodermally-derived anatomic structures fail to develop. 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. Ectodermal dysplasia might be inherited in any form of several genetic patterns including autosomal-dominant, autosomal-recessive, and X-linked modes. 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 100,000.

According to the state of sweat glands involvement, two major groups are distinguished: (1) Hypohidrotic or anhydrotic (Christ-Siemens-Touraine syndrome) in which sweat glands are either absent or significantly reduced in number; (2) Hydrotic (Clouston syndrome) in which sweat glands are normal. Dentition and hair are involved similarly in both types but hereditary patterns of nails and sweat glands involvement are different. Hypohidrotic ectodermal dysplasia as the most common type seems to show an X-linked inheritance pattern with the gene mapping to Xq12-q13; therefore, males are more susceptible than females. Hydrotic type is inherited in an autosomal dominant pattern. In general, the skin of affected children is lightly pigmented and appears thin and almost transparent; surface blood vessels are easily visible. Other manifestations include fine sparse hair, reduced density of eyebrow and eyelash hair. When hair is present, it may be fragile, dry, and generally with unruly appearance as a result of poorly developed or absent sebaceous glands. Fingernails and toenails may also show faulty development and be small, thick or thin, brittle, discolorated, cracked, and/or ridged.

The pre-ocular skin may show fine wrinkling with hyper-pigmentation and mid face hypoplasia is frequently observed resulting in protuberant lips. In cases where the salivary glands are hypoplastic or absent, varying degrees of xerostomia are expected. Affected individuals typically display heat intolerance because of reduced number of eccrine sweat glands. These glands may be either absent, reduced in number, or nonfunctioning (hypohydropic), which may result in elevated body temperature. Fever with unknown origin may lead to early diagnosis during infancy. The teeth are markedly reduced in number (oligodontia or hypodontia) and often manifest abnormal development in shape which may appear tapered, conical or pointed in incisors. Molars might be observed in reduced size. The lack of tooth bud formation causes hypoplastic alveolar bone, leading to a reduced vertical dimension of occlusion. Therefore, an old-age appearance is common in affected individuals.

This article presents the prosthetic rehabilitation of an adult with hereditary ectodermal dysplasia associated with oligodontia in permanent dentition.

Case report
A 20-year-old male was referred to the Department of Prosthodontics, Government Dental College, Srinagar with chief complaint of lack of teeth (Figure 1). According to familial history, a brother of the patient also suffered from oligodontia and heat intolerance. Intra oral examination revealed oligodontia with mal formed teeth.
In order to improve appearance and mastication, fixed partial prosthesis was established as treatment that improved the facial esthetics, speech and mastication of the patient (Figure 2).

**Treatment**
Maxillary and mandibular arch primary impressions were obtained using irreversible hydrocolloid impression material. Two sets of diagnostic casts were made by pouring the impressions twice with a type III dental stone. One cast set was used for the diagnostic wax-up and the other was saved for patient records. A centric relation record was taken and the casts were mounted on a semi adjustable articulator using face bow transfer and centric relation record. The diagnostic wax-up was made according to optimum esthetic and function. Maxillary and mandibular teeth were prepared for metal-ceramic crowns. Impressions were made using vinyl polysiloxane impression material. The metal-ceramic restorations were fabricated and were cemented with glass ionomer cement (Figure 2). The occlusion was checked and adjusted. The mutually protected occlusion scheme was preserved for this patient to allow for relatively even distribution and less stress of forces during excursive movement. Follow-up visits were scheduled at 3 months and then at 6 months. No esthetic and functional problems were seen after the 2 years of follow-up period.

**Conclusion**
Management of clinical manifestations associated with ectodermal dysplasia presents a unique challenge for Prosthodontist. Treatment has major impacts on self-esteem, masticatory function, speech and facial esthetics.

**References**
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