PROSTHODONTIC MANAGEMENT OF ECTODERMAL DYSPLASIA: A CASE REPORT

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ABSTRACT
Ectodermal dysplasia is a hereditary disorder characterized by involvement of abnormal nails, abnormal hair, abnormal or missing sweat glands and also abnormal dentition. The oral manifestation of the disease depends on the severity of the ectoderm involved. The Prosthodontic management of such patients with dysplastic condition necessitates a multidisciplinary approach. However the definitive treatment can only be rendered after the completion of growth period, and till then a provisional treatment can be given to enhance the aesthetic and functional requirement of the patient. The case report discussed here explains the prosthodontic management of a 15 year old boy with anhydrotic ectodermal dysplasia.

Keywords: anhydrotic, ectodermal dysplasia, prosthodontic management, telescopic denture, cast partial denture.

INTRODUCTION
Ectodermal dysplasia is defined as an X-linked recessive disorder characterized by congenital dysplasia of the ectodermal derivatives. The syndrome is characterized by abnormal nails (Onchondysplasia), Trichondysplasia (abnormal hair), Dyshidrosis (abnormal or missing sweat glands) and abnormal dentition1. The syndrome has a male affliction than females with about 1 to 7 individuals per 10001. Two clinically distinguished forms of the syndrome are reported: hypohidrotic (anhidrotic) and Hidrotic (Clouston'syndrome)2. The extra-oral features include frontal bossing, prominent supra-orbital ridge, saddle nose, hypoplastic alae nasi, depressed midface with malar hypoplasia such that it gives a dished shaped appearance with everted chin and thick and everted lips. Presence of poorly developed or absence of sebaceous glands makes the skin dry, scaly and easily irritated. 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 hairs are present, they may be fragile, dry3.

Intraoral features include anodontia or hypodontia of deciduous and permanent dentition, hypoplastic conical teeth, underdeveloped maxilla, and mandible, with poorly developed alveolar ridges leading to reduction in vertical dimension giving an aged appearance. The oral mucosa is normal and dry with protuberant lips1. The most frequent prosthetic treatment involves construction of a removable partial denture or complete denture. The treatment of the patients is critical because of the age and also the presence of poor alveolar ridge and low self esteem. The present case report describes the prosthetic management of a patient with anhidrotic ectodermal dysplasia.

CASE REPORT
A 14 year old boy reported to the College of K.M. Shah Dental College, Piparia with the Chief complain of inability to chew. The patient also complained of burning sensation of skin while going out in the sun. (Fig: 1) Upon thorough examination the patient was diagnosed with anhidrotic ectodermal dysplasia. Clinical examination revealed presence of conical shaped central incisors and permanent molar in the lower arch. However the mucosa over the alveolus and the gingival was normal. The ridges were rather atrophic except the region where the teeth were present. (Fig 2) Oral panoramic radiograph confirmed the absence of other permanent tooth buds and underdeveloped maxilla and mandibular ridges. (Fig: 3)
Prosthodontic treatment included to improve the mastication, appearance, speech. Since the child was still in the growing age and the pulp horns were high treatment included to provide cast partial denture in the mandibular arch and telescopic denture in the maxillary arch.

**Treatment plan**

Diagnostic impressions were made in irreversible hydrocolloid impression material. (Fig: 4)  Also a Diagnostic jaw relations was done to check the amount of interarch space and the ridge relation.(Fig:5) Once the jaw relations were confirmed a conventional final impression with border moulding in green stick and zinc-oxide eugenol paste and master casts were obtained. (Fig:6) A putty index and a separate model were made only of the upper two central incisors. Pattern resin was adapted and converted into metal copings. (Fig7 (a), (b), (c)) Onto which denture bases were made and occlusal rims were made. The lower master cast was duplicated and wax pattern for cast partial denture was made on the refractory cast. Casting was done and framework was obtained. (Fig8 (a), (b), (c)) Framework was tried and denture base was adapted and occlusal rims were made on it. Jaw relations were carried in normal conventional manner. (Fig 9) Upper anterior were trimmed and only a shell of the teeth was placed in order to reduce the proclination of the lips. Teeth arrangement was then finished in Class I relationship. (Fig 10) Try was done to verify the vertical relation, centric relation, etc. Flaking, dewaxing were carried out in a normal manner. A layer of indirect composite was applied on the labial surface of the copings to mask the hue of the metal. The final packing was done and dentures were fabricated in the conventional manner. (Fig11) Finishing and polishing was done and dentures were inserted. (Fig 12) Follow up was done at 1 week and 1 month. Minor occlusal adjustments were done.

**Discussion**

Treatments of ectodermal patients vary with age, dental agenesis, malformed teeth and growth and development of the stomatognathic system. Also treating the patient requires lot of motivation to the child as well as the parents to ensure their compliance during the treatment procedure4.

Prosthodontic treatments are of great of value to these patients for the function and psychological part of the patient. Treatment with partial or complete dentures is critical during the preschool years and can be continued till adulthood. Since alveolar bone development is dependent on the presence of teeth, children with ectodermal dysplasia have little or no bony ridge upon which to construct dentures. Hence, greater than usual problems are involved in attempting to restore function and appearance. The clinical management of patients manifesting this anomaly provides a unique opportunity for cooperative effort between the pedodontist and the prostodontist5.

The first goal of prosthetic device was to meet the needs of the patient which includes mastication, aesthetics and psychological factors. Prosthodontic treatment of the patient involved fabrication of the upper telescopic denture and lower cast partial denture. The telescopic denture helps in preservation of alveolar bone around the retained teeth while periodontal sensory mechanisms guides and monitor gnathodynamic functions. Lower cast partial denture provided stability and retention to the prosthesis6.

Implant placement for an adult patient with ectodermal dysplasia is the treatment of choice. However this was not recommended because of the growing age of the patient and inadequate bone support. Yap AK, Klineberg I, have concluded that implants placed in adolescent ED patients do not have a significant effect on craniofacial growth, while implants placed in ED patients younger than 18 years have a higher risk of failure. Guckes et al. recommend that this approach should be postponed until age 13 because of possible implant movement caused by jaw growth7. In addition, he has also suggested that their use in the anterior mandible may be routinely recommended in young patients if implant-supported prostheses were shown to have positive effects on craniofacial growth, self-image and food choice.

**Fig 1: Diagnostic OPG**
Fig 2: Intra-oral picture showing conical upper incisors and lower molar

Fig 3: Diagnostic casts

Fig 4: Diagnostic Jaw Relations

Fig 5: Final impressions

Fig 6: Master Casts

Fig 7 (a): Putty index for upper central incisors
Fig 7 (b): Copings made from Pattern Resin
Fig 7 (C): Final metal copings

Fig 8 (a): Wax pattern for lower denture
Fig 8 (b): Metal framework for lower denture
Fig 9 Final Jaw relations

Fig 10 Trial Dentures

Fig 11 (a) & (b) Trial Dentures in mouth (c) Trial Dentures in Mouth

Fig 12 Final Dentures in Mouth
REFERENCES


