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Dental Management of Siblings with Ecto-dermal Dysplasia: A Case Report

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Abstract: *Ectodermal dysplasias are a complex group of genetically determined disorders clinically characterized by congenital alterations of the structures derived from the ectoderm. Even though ectodermal dysplasia with partial anodontia is common, this condition with total anodontia is not commonly seen. We describe features of anhidrotic ectodermal dysplasia with oligodontia in a 9 and13 year-old brothers. In order to improve esthetics, speech and mastication, the child was provided with upper and lower removable partial dentures.*

Keywords: *Anodontia, Hypodontia, Oligodontia, Oral rehabilitation, Removable prosthesis.*

I. Introduction

Ectodermal dysplasia is not a single disorder, but a group of syndromes all deriving from abnormalities of the ectodermal structures. 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 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 and have certain cranial-facial features which can be distinctive, frontal bossing is common, longer or more pronounced chins are frequent, broader noses are also very common[1-5]. Scalp and body hair may be thin, sparse, and very light in color, even though beard growth in affected males may be normal. The hair may grow very slowly or sporadically and it may be excessively fragile, curly, or even twisted[6-7]. The skin may be lightly pigmented. Skin sustaining injury may grow back permanently hypo-pigmented. In some cases, red or brown pigmentation may be present. Skin can be prone to rashes or infections and can be thick over the palms and soles. Care must be taken to prevent cracking, bleeding, and infection. Individuals affected by certain Ectodermal Dysplasia syndromes cannot perspire[9]. Their sweat glands may function abnormally or may not have developed at all because of inactive proteins in the sweat glands. Without normal sweat production, the body cannot regulate temperature properly. Therefore, overheating is a common problem, especially during hot weather. In the development of tooth buds frequently result in congenitally absent teeth (in many cases a lack of a permanent set) and/or in the growth of teeth that are peg-shaped or pointed. The enamel may also be defective. Cosmetic dental treatment is almost always necessary and children may need dentures as early as two years of age. Multiple denture replacements are often needed as the child grows, and dental implants may be an option in adolescence, once the jaw is fully grown[10-12]

II. Case Report

Case-1

A nine year old boy reported to department of pedoodontics and preventive dentistry with the complaint of missing teeth in both upper and lower region of jaws since birth and have difficulty in chewing and esthetic problems . As per familial history, a brother of the patient also suffered from oligodontia and heat intolerance.Boy have a dry skin, hypoplastic midface, prominent lips, sparse fine hair and four peg shaped teeth present in the maxillary anterior and three molars in the posterior region,with thin alveolar ridge and reduced vertical bone height and two teeth present in the mandibular region (Fig a) . Extra-oral and Intra-oral examination revealed typical features of an-hidrotic ectodermal dysplasia.Radiographic findings also confirmed the diagnosis.

Treatment plan include construction of removable partial denture for both maxillary and mandibular arches to improve function speech and appearance and vennering the peg shaped anteriors with composite .So The Primary impressions were made with irreversible hydrocolloid impression material as it is comfortable and can be easily removed from undercut area.Border Moulding was done with green stick compound ,When properly fitting custom tray was ready, final impression was made by elastomeric materials. Jaw relationship was recorded using temporary base and wax rim. The arranged teeth were verified in the mouth during the try-in appointment. After laboratory processing, the Removable partial dentures were delivered to the patient,

instructions were given to maintain proper oral hygiene. Continuous follow-ups every six months were planned for adjustment or replacement of old denture. (Fig b)

A nine year old boy with anhidrotic ectodermal dysplasia -A) Pre Treatment Front View B) Post Treatment Front View

Case-2

After watching the younger brother eating properly and looking pleasant, A 13 year old boy reported to the department with the complaint of missing teeth in both upper and lower region of the jaw.Family history revealed younger brother also sufferd from oligodontia and heat intolerance. Extraoral examination revealed sparse fine hair, dry skin, reduced vertical height of facial lower third and prominent chin and lips.Intraoral examination revealed presence of three peg shaped teeth in maxillary and one in mandibular region along with four molars in posterior area (Fig-a). These findings matched typical features of anhidrotic ectodermal dysplasia.

In order to improve aesthetics and functions,Peg shaped anterior teeths were build with composite resin and for filling the space Removable partial denture was designed. Primary impressions were made with irreversible hydrocolloid impression material as it is comfortable and can be easily removed from undercut area. Casts were prepared with type III dental stone. Custom trays were prepared, and border moulding was done with green stick compound. The final impressions of the maxillary and mandibular arches were made with medium and light body type of rubber base impression material, respectively. Maxillo-mandibular relation was recorded, and the master casts were mounted on a semiadjustable articulator. The teeth were arranged according to a balanced occlusal scheme.Try-in was done and after careful evaluation the maxillary prosthesis, and the mandibular prosthesis was fabricated in the conventional heat cure acrylic resin. The dentures were then inserted in the patient's mouth(Fig-b).The patient was instructed to maintain their oral hygiene and educated about proper insertion and removal of prosthesis. Recall was done after 2 days for final adjustments and the future visits were planned after 3 months for relining and to assess further growth.

A)  B) 

A)  B) 

A 13 year old boy with anhidrotic ectodermal dysplasia -A) Pre Treatment Front View B) Post Treatment Front View

III. Discussion

Early oral rehabilitation improves oral function, phonesis ,esthetics, reducing social impairment . It also improve saggital and vertical skeletal relationships during craniofacial growth and development.In childrens most acceptable treatment plan is removable partial denture.Implant supported denture are to be given in patients more than 15 years of age[2-4]. When implant therapy is indicated, the main problem is insufficient bone; if bone atrophy progresses in these already alveolar deficient patients, implant placement may not be possible without bone grafting[4-6]. All treated patients reported a significant improvement in their own diet because the oral rehabilitation also allowed them to eat meat, fish, fibrous vegetables and cheese[7-8]. However, removable partial or complete dentures require regular adjustments and should be replaced when a decreased vertical dimension of occlusion and an abnormal mandibular posture are detected due to growth[9]. The occlusion of removable partial denture should be in harmony with the patient's occlusion. Since oligodontia or anodontia leads to atrophy of the alveolar ridges, reduced vertical dimention, prominent chin, and class III intermaxillary relationship,so early prosthetic treatment should be performed as soon as possible. This treatment modality improves the patient's quality of life and it can be regarded as an acceptable treatment modality for functional and esthetic rehabilitation.

IV. Conclusion

It's a big challenge for a pedodontist /prosthodontist to treat the patients of ectodermal dysplsia. However Treatment of these patients with Removable partial denture or complete denture is cost-effective, improves the patient well-being and socio-psychological conditions. All the success depends on patient maintenance of oral hygiene and routine visits to the dentist.

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