

Prosthodontic Management of Ectodermal Dysplasia in pediatric patient

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Abstract: Ectodermal dysplasia is a large heterogenous group of inherited disorders that occurs as a consequence of disturbances in the ectoderm of the developing embryo. The tissues primarily involved are skin, nails, hairs, sweat glands and a partial or complete absence of primary and/ or permanent dentition. This clinical report highlights the imperative need of appropriate treatment strategy and application of partial acrylic dentures in a pediatric patient who suffered from congenitally oligodontia.

Key words: Partial dentures, ectodermal dysplasia, prosthetic rehabilitation, oligodontia,

I. Introduction

Ectodermal dysplasia is a well recognized syndrome which is characterized by congenital dysplasia of one or more structures derived from embryonic ectoderm. It was first reported and published by Thurnam¹ in 1848 but the term "Ectodermal Dysplasia" was coined by Weech² in 1929.

These are a heterogeneous group of disorders characterized by developmental dystrophies of ectodermal structures, which affects the development of keratinocytes and cause aberrations in the hair, sebaceous glands, eccrine and apocrine glands, nails, teeth, lenses and conjunctiva of the eyes, anterior pituitary gland, nipples and the ears.³ Although more than 170 different subtypes of ectodermal dysplasia can be defined. These disorders are considered to be relatively rare with an estimated incidence of 1 case occurring over 100,000.⁴

Due to the overlapping clinical features of many of the syndromes that involves ectodermal dysplasia (ED) A definitive classification is difficult but On the basis of sweat gland involvement ED broadly divided in to two groups. First one is Hypo-hidrotic or anhidrotic (Christ-Siemens-Touriane syndrome) in which sweat glands are either absent or significantly reduced in number. This is the most common and males are more affected because of it is X-linked inheritance pattern with the gene mapping to Xq12-q13. The second one is Hydrotic (Clouston syndrome) in which sweat glands are normal. It shows autosomal dominant inherited pattern. In both cases abnormal (hypodontia) or missing (anodontia) teeth may be found but hereditary patterns of nails and sweat glands involvement are different.^{5,6}

II. Case Report

A 8-years-old boy was referred to the Department of Pediatric Dentistry, Vananchal Dental College & Hospital Garhwa India with the chief complain of missing teeth and difficulty in chewing. He was born at 40 weeks of gestation with normal delivery. After three months of birth he had experienced recurrent attacks of high fever and crying. He was admitted to IMS BHU Varanasi, India to examine the physical condition diagnosed as a hypohidrotic ectodermal dysplasia. According to familial history, patients parent's, his elder brother and their all relatives were normal.

At first appointment patient presented a typical characteristics (Figs 1) of ectodermal dysplasia such as sparse hairs and eye brow, protuberant supraorbital ridge and chin, saddle nose, frontal bossing. The skin was dry and rough. To establish a sound treatment strategy, a meticulous and extensive evaluation was executed. An Extraoral radiography (OPG: Orthopantomogram) and cephalometric was taken to assess the status of teeth and condition of supporting structures. (Figure 2,3). The panoramic radiograph and cephalometric radiograph confirmed oligodontia with missing primary and permanent teeth except Following teeth were present 16, 55, 26,65, 36,75, 46 and 85. The tooth buds of 27,37 and 47 were also present. The colour of alveolar mucosa and other mucosa were normal.

In order to restore the oral function, the full dentures were constructed by a modified conventional approach. The child was familiarized with various materials, impressions trays using "Tell Show Do" technique. Preliminary impressions were recorded with alginate material by using selected stock tray for child. Study casts

were made and the individual trays were made with tray resin. Border moulding was done with greenstick compound by using the special trays and Final impression taking was performed with silicon rubber material (Aquasil, Dentsply, Delhi, India). Models were obtained and temporary denture bases were constructed with self cure acrylic resin (DPI Cold Cure India) then wax rims were made on the temporary denture base. At the occlusion record, maxillary rim was adjusted for lip support, phonetics and esthetics. The horizontal jaw relation was determined without using the face-bow and the proper vertical dimension and free way space were established. The vertical dimension of occlusion was first determined by measuring face height in physiological rest position and checked by means of cephalometric criteria. Wax dentures were constructed with available smaller size acrylic teeth (Cosmo, Dentsply, India). Try in was done during which the patient and parents were satisfied (Figure 4). The dentures were made of heat-cured acrylic resin (Trevelon Heat Cure, India). At 8 years and 1 months of age, the partial dentures were completed and inserted (Fig 5). Periodic check ups were scheduled after 24 hours, 1 week, 1 month, during which the patient presented with no problems. Now he was able to communicate better and was more interactive and engaging with others. Further recall appointment have been scheduled after 3 months.

III. Discussion

The ectodermal dysplasia is comprise a large heterogeneous group of inherited disorder. Among them X-linked hypohidrotic ectodermal dysplasia is more common. The gene responsible for X-linked has been reported by Monreal et al. It indicates the direct molecular diagnosis relationship.⁷ Later on it is found that ectodermal dysplasia is related with a mutation of the protein ectodysplasin-A. It is located with the EDA gene in the q12- q13 locus of the X chromosome. Mutations in one or various genes, including EDA, EDAR (EDA receptor) and are associated with hypohidrotic ectodermal dysplasia, with or without immunodeficiency.^{8,9,10}

Oral findings are of particular interest since the disease is characterized by hypodontia, oligodontia or anodontia, which can, moreover, affect both the maxilla and mandible and cause delayed eruption, malformed teeth, producing a small, pointed, conical appearance; and resorption or atrophy of the alveolar border, thus complicating the fundamental rehabilitation procedure in these patients.

As per dentist concern, an acceptable solution is sought for these young patients with complex cases of congenital lack of teeth, the possibility of fixed prosthetic and implant treatment of pediatric patients in early stage is not good because of the implant movement caused by jaw growth. Guckes et al.¹¹ recommend that this approach should be postponed until age 13 because of possible implant movement caused by jaw growth.

It is advisable to make use of such prosthesis (removable denture) which do not alter the dentition permanently during the period of assessment and accomplish patient's functional and esthetic demands. It allow the child normal a life style as possible without damaging self-esteem or psychological development and ensuring that behavior is unaffected. When the patient is in the last stage, the removable prosthesis may be replaced by a fixed type restoration using osseointegrated.

Considering the age and potential growth of our patient, it was deemed better to postpone osseointegrated implants. Prosthodontic management of this patient was challenging for all steps right from impression making to use of dentures because of the fragile mucosa, poor alveolar ridges and decreased salivation. Treatment commenced as soon as possible in order to avoid possible resorption and atrophy of the alveolar ridges, and to control the difficulties in eating and speaking.

The fabrication of partial dentures for the young patient with such a condition requires knowledge of growth and development, behavioral management and the skills of the dentist for fabricating the dentures²⁰. The effect of prosthetic management of the patient should be evaluated in long-term follow up. As the child grows, the denture will have to be modified and replaced. Replacements will be needed at least 3 times between the period of early and late mixed-dentition and permanent-dentition.¹² Unfortunately, the followup was limited in this case, so the evaluation of the treatment result could not be mentioned.

IV. Conclusion

Ectodermal dysplasia often associated with emotional consequences for affected individuals at early stage. Many dental problems are also associated thus they suffer not only from functional difficulties but also poor esthetics. Early clinical diagnosis and treatment planning is essential to encourage a normal physiological development and to improve function of stomatognathic system. From our point of view , the use of partial acrylic denture is a practical and quick alternative which provides easy, acceptable and economical solution to rehabilitate the functional and esthetic requirements.

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Figure 1: Frontal View of the patient's face



Figure 2: Panoramic radiograph revealed oligodontia



Figure 3: Cephalometric radiograph presents the oligodontia



Figure 4: Try In



Figure 5: Patient's frontal view after insertion of denture