

Oral Rehabilitation of a Child with Ectodermal Dysplasia: A Case Report

Abstract:

Ectodermal dysplasia is a congenital disorder affecting multiple body parts, dominating oral cavity leading to multiple missing teeth. Dental professionals are the one who encounter the disease at early stage. They can minimize the aesthetic, functional, social and psychological impairment by simple oral rehabilitation of the patient. This case report is a story of 4 year old child suffering with Ectodermal dysplasia with missing maxillary and mandibular teeth that was rehabilitated with removable complete denture

Key-words: Ectodermal dysplasia, complete denture, oral rehabilitation, congenital disorder, child with special health care need.

Introduction:

Ectodermal dysplasia (ED) is an X-Linked autosomal disorder that may be recessive or dominant with multiple congenital conditions associated with it. On clinical representation ED have categorized into more than 200 types. ED mostly affects parts with ectodermal origin like hairs, sweat glands, nails, teeth etc. The two broadly classified ED are 1. Anhydrotic/Hypohydrotic ED represents by absence or significantly reduced number of sweats glands and 2. Hydrotic ED represent with normal sweat glands with presence of other clinical features.

General manifestation of ED mainly includes sparse hair, dry skin, intolerance to heat etc. however oral manifestation includes complete or partial anodontia, hyposalivation, abnormal shape of teeth, rudimentary alveolar ridges, shallow palatal vault etc.

Pediatric dentists are one of the health care professional that encounters these patients at early age and they can minimize the functional, aesthetic, social and psychological impairment by means of oral rehabilitation.

Case Report:

The parent of a four year old male patient reported to the OPD with chief complains of missing upper and lower teeth since birth. Complete history was taken that revealed that the patient was diagnosed with Ectodermal dysplasia at the age of one year by medical health care professional. Clinical examination revealed ectomorphic body type with dry and scaly skin. Patient was intolerable to heat as told by parent, thin and sparse hair, frontal bossing and deep mento-labial sulcus [Fig.1]. Intra oral examination showed complete edentulous mandibular arch with rudimentary alveolar ridge and partially edentulous maxillary arch with presence of only two conical shape lateral incisors. Maxillary arch was shallow with underdeveloped alveolar ridges [Fig.2].

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Investigating digital Orthopantomogram (OPG) was done that revealed the presence of only two maxillary lateral incisors which was conical in shape and complete absence of developing tooth buds in maxillary and mandibular arch. [Fig.3]

Treatment plan was discussed among group of Pediatric dentist and Prothodontist and was decided to rehabilitate with removable complete denture in maxillary and mandibular arch by considering all the factors related to patient's age, cooperative behavior, parental acceptance and socioeconomic status of parents. Extraction of maxillary teeth was avoided and planned to provide the space in complete maxillary denture as shown in [Fig.4]

All the steps of conventional complete denture procedures were followed except the primary impression was made with alginate instead of impression compound as the patient was heat intolerable. Complete maxillary and mandibular denture were made [Fig. 5 and 6] and insertion was done. Post insertion photographs were not possible due to negative behaviour of the child. Patient was followed up till one year [Fig.7] at regular intervals and parent was quite satisfied with the treatment and showed positive attitude to follow up.



Fig.1 Extra oral photograph

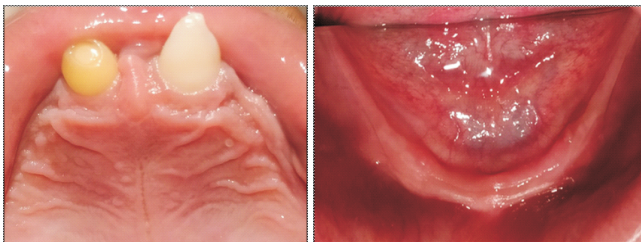


Fig.2 Intra oral image showing partial edentulous maxillary

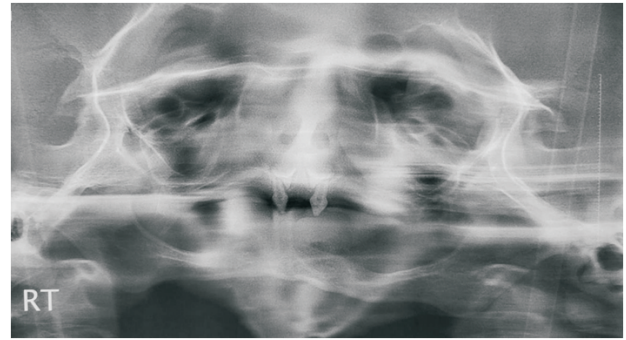


Fig.3 OPG showed complete missing of permanent tooth



Fig.4 Complete maxillary denture to accommodate



Fig.5 Complete mandibular denture



Fig.6 Maxillary and mandibular denture in occlusion



Fig.7 Follow up after one year [image received on whatsapp from parents]

Discussion:

Dental management of patients with Ectodermal dysplasia(ED) needs an interdisciplinary approach. It requires a team of specialist from the field of Pediatric dentistry, Prosthodontics, Orthodontists and Oral surgery. The team should aim to prevent, maintain and restore the form and function of the patient's teeth, which in turn help the patient to have improved functions, speech, aesthetics and overall growth and development of the child improving his or her overall quality of life. According to Nowak, "Treating the pediatric patient with ED requires the clinician to be knowledgeable in growth and development, behavioral management, techniques in the fabrication of a prosthesis, the modification of existing teeth utilizing various restorative techniques, the ability to motivate the patient and parent in the use of the prosthesis, and the long-term follow-up for the modification and/or replacement of the prosthesis"[1]

The primary choice of the prosthetic rehabilitation in ED patients is implant retained or implants supported dentures. There are several case reports showing the successful use of implant therapy in ED patients.(2,...) However there are several limitations like knife edge alveolar

bone, displacement or failure of implant due to the facial growth, implant can cause damage to the underlying developing tooth germs and further can restrict the craniofacial growth. Hence keeping these drawbacks in mind we plan to fabricate complete removable dentures to facilitate masticatory functions, phonetics, and aesthetics for improving the overall growth of the patient.

Fabrication of prosthesis and its biomechanical adaptation can be challenging in ED patients due to reduced alveolar bone height, knife edge ridges. Masticatory functions can be enhanced by achieving occlusal stability and retention in dentures. In our case maxillary teeth was used in order to enhance the retention by fabricating overdenture. Early rehabilitation of ED patients facilitate the development of lifelong dietary pattern and prevent malnutrition. Proper vertical dimension of dentures facilitate development of temporomandibular jaw relations during growth periods. Several case reports in literature shows that ED patients after rehabilitation showed larger growth than in the controls.[4,5,6]

Since the patient is in growing age periodic recall visits are required for the patient. There may be a need to adjust or replace the prosthesis accordingly depending on the growth and development of the patient. Recommend relining/rebasing an intraoral prosthesis in a growing patient every 2-4 years and remaking a new prosthesis every 4-6 years.[3] Oral hygiene instructions along with cleaning of prosthesis and the use of fluoridated toothpaste and topical fluoride applications must be follow for the age appropriate recommendations of AAPD in patients with partial edentulous arch.

Conclusion:

Oral rehabilitation in a very young patient with Ectodermal dysplasia can be done using conventional prosthetic denture with regular follow up.

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