**Case Report**

# Ectodermal Dysplasia in Primary Dentition – A Rare Case Report

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## Abstract

Development of the primary and permanent dentition is a complex process wherein there is series of interactions between the ectoderm and ectomesenchyme. A cascade of signaling pathways occur in a spatio-temporal manner resulting in the development and eruption of the human dentition. Any developmental aberrations in shape, size, number, and position can lead to deviations from normal development of teeth. In this book, dental anomalies including gemination, fusion, concrescence, dilaceration, dens invaginatus, DE, taurodontism, enamel pearls, fluorosis, peg-shaped laterals, dentinal dysplasia, regional odontodysplasia and hypodontia etc. are discussed. Diagnosing dental abnormality needs a thorough evaluation of the patient and careful clinical and radiographical examination is required. Furthermore, more complex cases need multidisciplinary planning and treatment.

## Key words

Primary Dentition, Ectodermal Dysplasia, Partial Anodontia, Fixed Partial Denture, Crown And Bridge.

## Introduction

The term Ectodermal dysplasia was first described by Thurman in the year 1848 [1, 5]. It is a rare hereditary disorder occurring as a consequence of disturbances in the ectoderm of the developing embryo. It affects both males and females of all races and ethnic groups. Prevalence is estimated to be 7 cases in 10,000 births. An individual affected by Ectodermal Dysplasia syndromes have abnormalities of the glands, tooth buds, hair follicles, and nail development. Ectodermal Dysplasia represents a large and complex group of diseases comprising more than 170 different clinical conditions. The triad of nail dystrophy is onchodysplasia,

hypotrichosis and palmoplantar hyperkeratosis which is usually accompanied by a lack of sweat glands and a partial or complete absence of primary and/or permanent dentition. There are two major types of this condition depending on the number and functionality of the sweat glands: (1) Hypohidrotic, where sweat glands are either absent or significantly reduced in number and (2) Hidrotic, where sweat glands are normal and the condition is inherited as autosomal dominant. Dental manifestations occurring in this syndrome, which includes conical or pegged teeth, enamel hypoplasia, reduced vertical asymmetric alveolar ridge height, maxillary retrusion, high palatal arch, hypodontia or complete anodontia, and delayed eruption of permanent teeth. Absence of teeth may cause masticatory difficulties, nutritional deficiencies, speech problem, and compromised facial appearance. Therefore the objectives of oral rehabilitation in young Ectodermal Dysplasia patients are preservation of bone, early development of correct patterns of chewing, swallowing, speaking, and restoring normal facial characteristics [3]. Thus, this case report aims to present the management of Ectodermal Dysplasia in a 4 year old female patient, and purposing to help dentists to determine the general appearances, extra and intra oral manifestation of this disorder.

## Case report

A 4-year-old girl was referred to the Department of Pediatric and Preventive Dentistry in K.D. Dental College representing the lack of teeth as the chief complaint which compromise mastication, speech and esthetics.

In the medical history revealed, there was no history of birth complications during the delivery. Furthermore family history revealed that patient's father had the similar conditions as her like hypodontia, fine light hair on scalp.

On general physical examination, it was perceived a thin body structure with normal gait and posture (**Figure – 1**). On extra-oral examination, it was noted that she had a dry and dark skin, hypo plastic midface, everted lips, fizzy hair, sparse fine hair on scalp and eyebrows, mild finger deviations, saddle nose, prominent forehead and no deformity in nails, her lips were competent (**Figure – 2 to 4**).

Fig. 1 – General Examination

On Intra-oral examination, it was perceived that partially edentulous upper and lower jaw, four conical shaped anterior teeth in upper jaw and two conical shaped anterior teeth in lower jaw, thin alveolar ridge, reduced vertical bone height, loss of vestibular depth in the lower jaw, arch discrepancies and no abnormalities in salivary gland were seen with normal salivation flow rate. It was observed that in upper both the left and right primary canines were missing along with left primary first molar. In the lower arch primary anterior incisors were missing along left primary canine (**Figure – 5 to 9**).

For Investigation, Orthopantomography has been taken, on interpretation it was revealed that only four permanent 1st molars and two upper Permanent central incisors tooth buds were present. The rootless primary upper left and lateral incisors, and one lower canine were also observed. At the same time following 53, 63, 64, 71, 72, 73, 81 teeth were missing (**Figure – 10**).

So based on the observations our final diagnosis was Hidrotic Ectodermal Dysplasia (autosomal dominant) and treatment plan were made Porcelain Non Metal Crowns for maxillary

anterior teeth and Split Fixed Partial Denture for mandibular anterior teeth.

For the treatment crown cutting was done on abutment teeth (51, 52, 61, 62, 74, 82). Then Impression was taken with Putty material and sent it to laboratory for crown and bridge fabrication. After the fabrication, PNM Crowns were delivered to the respective teeth. Here Fixed Partial Denture was splited in between 71 and 81 and they are joiner lingually by ribbond elastic material. So we were trying here to provide a newer technique for oral rehabilitation of Ectodermal Dysplasia in child patient so that growth of mandible does not hinder and good esthetic & retention is maintained (**Figure – 11 to 14**).


**Fig. 2 - Extra-oral Examination**

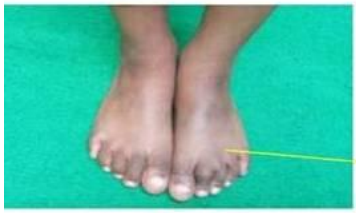

Fig. 3 - Normal toe-nail structure


Fig. 4 - Mild finger deviation

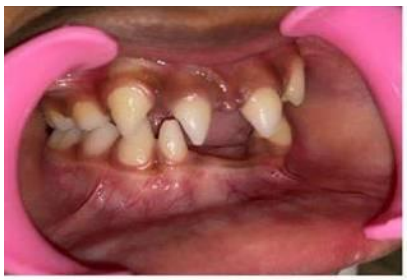

Fig. 5 - Right Posterior occlusion

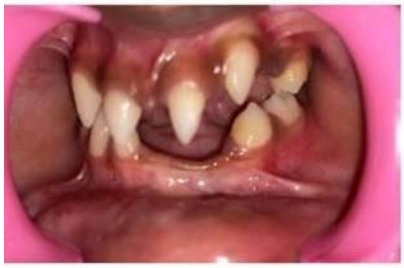

Fig. 6 - Left Posterior Occlusion

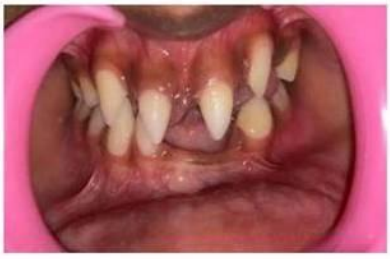

Fig. 7 - Anterior Occlusion

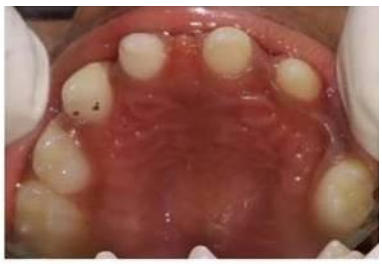

Fig. 8 - Maxillary Arch

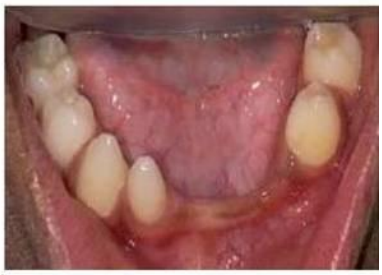

Fig. 9 - Mandibular Arch

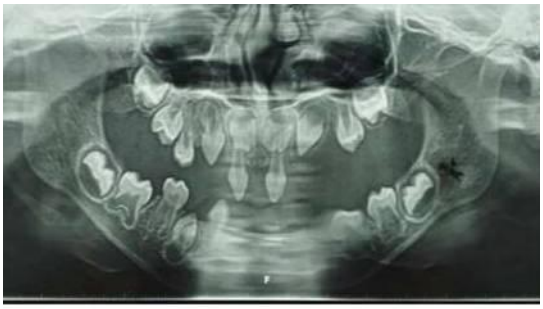

Fig. 10 - Orthopantomograph

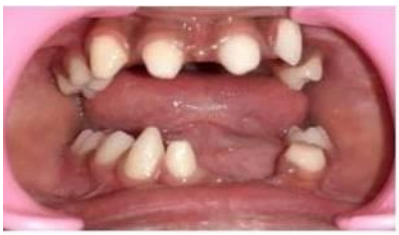

Fig. 11 - Crown Cutting Done on Abutment Teeth (51,52,61,62,74,82)

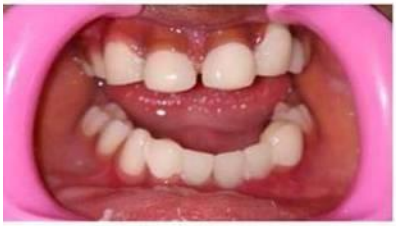

Fig. 13 -PNM Crowns on 51,52 , 61,62 and Split Fixed Partial Denture on71,72,74, 81,82

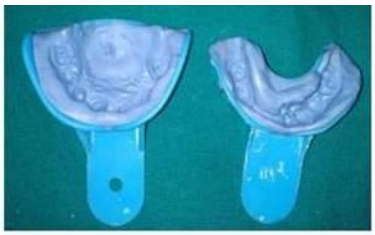

Fig. 12 - Impression Taken with Putty material and send it to laboratory for split crown and bridge fabrication

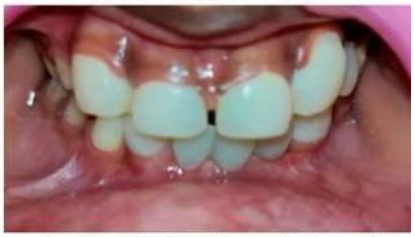

Fig. 14 - Anterior occlusion view After Treatment

## Discussion

Oral rehabilitation of the ectodermal dysplasia patient is necessary to improve both the sagittal and vertical skeletal relationship during craniofacial growth and development as well as to provide improvements in esthetics, speech, and masticatory efficiency [5]. Therefore, it is important to diagnose the associated problems in dento-alveolar complex, so that an accurate treatment plan can be established to rehabilitate the patient at optimum level [3]. 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. Early prosthodontic rehabilitation of patients with ED helps to restore and normalize function of muscles of mastication and the skeletal growth pattern. Moreover, it helps to reduce the unwanted side effects caused by absence of teeth, such as alveolar ridge resorption, loss of vertical dimension, and a tendency to class III malocclusion [2]. FPDs with sufficient abutment teeth in children are gaining popularity because of their superior aesthetics and improved retention and stability. Reasoning behind this split crown and bridge design was allowing jaw growth as it does not cross midline and also increase the occlusal vertical dimension leading

to a superior aesthetic outcome [2]. Early implant placement in a growing child may cause cosmetic problems because the implants act like ankylosed teeth. With the vertical development of the jaws, implant over-structures may not meet with the teeth of the opposite jaw, and may result in prosthetic infraocclusion [5]. Periodic dental recall of patients with ED should be done at regular intervals to be able to monitor the patient's growth and development and consequently adjust or replace the prosthesis accordingly. Vergo recommended relining/ rebasing an intraoral prosthesis in a growing patient every 2–4 years and remaking a new prosthesis every 4–6 years. In our case, this Split FPD technique is the new and beneficial technique for oral rehabilitation of Ectodermal Dysplasia in young children as it does not interfere with the growth of jaw, providing good aesthetics and retention.

## Conclusion

Management of clinical manifestations associated with ectodermal dysplasia presents a unique challenge for pediatric dentists [4]. Since oligodontia or complete anodontia leads to atrophy of the alveolar bone, prosthetic treatment is of great value to these patients from functional, psychological, and psychosocial standpoints [5]. The transitional prosthesis should be replaced by more definitive prosthesis once the skeletal growth is completed.

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