PROSTHETIC REHABILITATION IN ECTODERMAL DYSPLASIA

by Jana Publication & Research

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Abstract

Background:
Ectodermal dysplasia (ED) represents a group of rare, inherited disorders characterized by developmental anomalies in structures derived from the ectoderm. Hypohidrotic ED, the most common type, often presents with oligodontia or anodontia, posing significant functional and psychological challenges in children.

Case Report:
This case describes a 5-year-old boy with hypohidrotic ectodermal dysplasia presenting with oligodontia, dry skin, sparse hair, and typical facial features such as a saddle nose and thick everted lips. The child exhibited difficulty in mastication, speech, and an inability to sweat. Clinical and radiographic evaluation revealed the presence of only four primary second molars and a few developing permanent molars. A multidisciplinary treatment approach was adopted, and early prosthetic rehabilitation with removable partial dentures was undertaken to restore function, esthetics, and psychosocial confidence.

Conclusion:
Early prosthetic management in pediatric patients with ectodermal dysplasia is crucial for enhancing oral function, supporting normal craniofacial growth, and improving quality of life. Timely dental intervention addresses physical challenges and plays a vital role in the child's emotional and psychological development.

Introduction:-

Ectodermal dysplasia (ED) encompasses a diverse group of inherited disorders characterized by congenital abnormality in two or more structures originating from the embryonic ectoderm. These structures primarily include hair, teeth, nails, and sweat glands.[1]

Individuals with ED commonly present with features such as thin or absent hair (hypotrichosis), missing or irregularly shaped teeth (hypodontia or anodontia), and fragile or deformed nails.[2]

Diagnosis of ED typically involves a thorough clinical evaluation focusing on the characteristic features affecting hair, teeth, nails, and sweat glands. Genetic testing can provide definitive confirmation by identifying specific mutations responsible for the disorder.

According to the state of sweat gland involvement, two major groups are distinguished:

(1) Hypohidrotic or Anhydrotic (Christ-Siemens-Touriane syndrome) is the most common type seems to show an X-linked inheritance pattern with the gene mapping to Xq12-q13; therefore, males are more susceptible than females. In this type, sweat glands are either absent or significantly reduced in number.

(2) Hydrotic (Clouston syndrome) in which sweat glands are normal.[3]

Both types exhibit similar involvement of dentition and hair, but the hereditary patterns affecting nails and sweat glands differ.

The number of teeth is significantly reduced (oligodontia or hypodontia), often displaying abnormal shapes such as tapered, conical, or pointed incisors, while molars may appear smaller than usual. The absence of tooth bud formation results in hypoplastic alveolar bone, leading to a decreased vertical dimension of occlusion. Consequently, affected individuals often exhibit a premature aging appearance.[4]

Early dental intervention of patients with ED plays a key role in improving their functional abilities, boosting their overall wellness, and alleviating psychological stress.[2] The present case report describes the early prosthetic rehabilitation of a 5-year-old child with hereditary ectodermal dysplasia associated with severe oligodontia in the primary dentition.

Case:

A 5-year-old boy reported to the Department of Pediatric Dentistry, Government Dental College and Hospital, Nagpur, with a chief complaint of missing teeth.

The parents reported that the patient had never sweated since birth, even during hot summers. They also noted a history of delayed tooth development and frequent episodes of elevated body temperature since early infancy. The patient also complained of difficulty in mastication and speech. Family history revealed a consanguineous marriage between parents while both the parents and other family members appeared unaffected.

The extraoral examination revealed dry skin, brittle nails, and typical features of anhidrotic ectodermal dysplasia, including frontal bossing, sparse scalp and eyebrow hair, a saddled nose, a long philtrum, and unusually thick, everted lips.(Figure 1).

Intraorally, severe oligodontia was noted with only four teeth being present. These included the maxillary and mandibular deciduous second molars. The maxillary and mandibular arches exhibited severe ridge resorption (Figure 2). A thin alveolar ridge with reduced vertical bone height, and loss of vestibular depth in the lower jaw. The panoramic radiographic findings also confirmed the clinical diagnosis (Figure 3). It revealed tooth buds of four permanent molar teeth in the upper and lower jaw along with tooth buds buds of permanent second molars in lower jaw.

The treatment plan included the fabrication of removable partial dentures for both arches.

First, an impression of the upper and lower jaw was taken using an alginate material. The impressions were then poured with dental stone to obtain dental models. Custom trays were fabricated on the dental casts. Border molding was done and then the final impression was recorded with Oranwash elastomeric material. The final impression was poured with dental stone to obtain the dental cast. Occlusal wax rims were fabricated on the record bases. Orientation, vertical and centric jaw relationship was established. Acrylic teeth were used for teeth arrangement. Try-in was done to assess the aesthetics, speech, and occlusion of the patient with necessary adjustments made as required(Figure 4). Processing of the denture was done following the steps like flasking, dewaxing, packing of heat cure acrylic resin, and then polymerization. The denture was finally trimmed and polished. Final fitting and adjustments were done (Figure 5).

The patient and his parents were instructed on the correct insertion, removal, and maintenance of the dentures. The parents were reassured, and the child was guided through speaking exercises to help train oral musculature for better adaptation to the dentures. A recall was scheduled after 24 hours for necessary adjustments, with follow-up visits planned every six months to monitor bone growth and reline the dentures.

DISCUSSION-

Hypohidrotic ectodermal dysplasia (Christ-Siemens-Touraine syndrome) is the most common form among this group of disorders, with an estimated incidence of approximately 1 in 100,000 live births. Mutations in the *EDA* gene, which encodes the epithelial morphogen ectodysplasin A,

lead to the X-linked recessive form of the disease. In contrast, mutations in *EDAR* and *EDARADD* genes are associated with autosomal recessive and autosomal dominant forms.[5]

As Nowak rightly said, "successful management of pediatric patients with ED requires an understanding of growth and development, behavioral guidance, prosthesis fabrication, restorative techniques, and long-term follow-up with ongoing motivation of both patient and parents." [6]

Management of ED is multidisciplinary, involving pediatricians, dermatologists, dentists, and geneticists. From a dental perspective, early intervention is essential to address functional impairments and support emotional and psychological well-being. Dental management is a critical aspect of treating ectodermal dysplasia (ED), as patients often present with missing, malformed, or delayed tooth eruption.[7]

Studies have shown that early prosthetic treatment supports normal craniofacial growth and enhances the harmony of lip positioning in relation to the nose and chin [6]

Dental prostheses—such as complete dentures, partial dentures, or dental implants—are commonly employed to restore both function and aesthetics in patients with hypodontia, oligodontia, or anodontia.[8]

In childhood, dentures are the primary line of treatment for the dental abnormalities. These dentures are regularly evaluated by the pediatric dentist as per the child's growth and development and are modified/replaced accordingly.[9]

Future advancements in pediatric prosthodontics, including digital impression techniques, 3D printing, and bioengineered tooth regeneration, hold promising potential for improved treatment outcomes in children with ectodermal dysplasia. Long-term clinical studies and interdisciplinary research are needed to establish standardized treatment protocols and assess the effectiveness of fixed versus removable prostheses in growing children. Moreover, genetic counseling and early screening in families with a history of ED can facilitate early intervention, minimize complications, and support more personalized care strategies.[10]

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CONCLUSION-

Prosthetic rehabilitation in young patients with oligodontia due to ectodermal dysplasia is essential for promoting normal development and enhancing quality of life. A multidisciplinary approach, individualized to each patient's clinical and emotional needs, ensures optimal outcomes. Early replacement of missing teeth not only facilitates proper craniofacial growth but also restores essential functions such as mastication, speech, and aesthetics.It also contributes significantly to improving the patient's self-confidence and psychosocial well-being, underlining the critical role of timely dental intervention in the holistic management of ectodermal dysplasia.

ACKNOWLEDGEMENTS-

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FIGURE LEGENDS-

Figure 1: Saddle nose, long philtrum, thick lips, and sparse hair on eyebrows noted on extraoral examination (Frontal view).

Figure 2. Panoramic radiograph showing erupted primary second molars and unerupted permanent molars.

Figure 3. Steps in the fabrication of removable partial dentures.

Figure 4. Maxillary and Mandibular dentures in position.

FIGURES-

Figure 1. Saddle nose, long philtrum, thick lips, and sparse hair on eyebrows noted on extraoral examination (Frontal view).

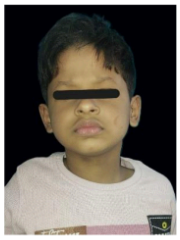


Figure 2. Panoramic radiograph showing erupted primary second molars and unerupted permanent molars.

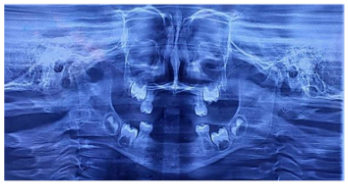


Figure 3. Steps in the fabrication of removable partial dentures.

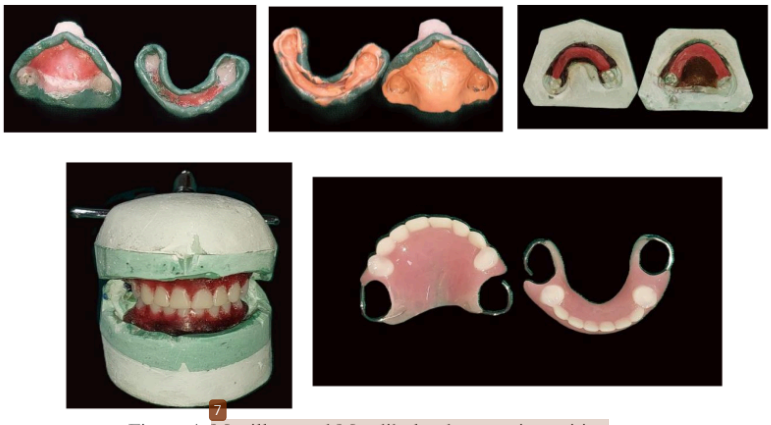
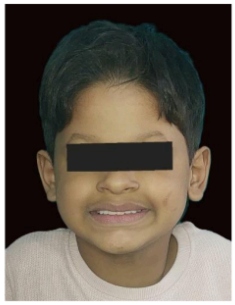


Figure 4. Maxillary and Mandibular dentures in position.



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