Premature prosthodontic intervention for two brothers afflicted by ectodermal dysplasia- A case report.

Srishti Mishra[1], Mehr Yazdani[2], Nivea Verma[2], Manesh Lahori[3].

[1]PGT, Department of prosthodontics crown & bridge and implantology, K.D Dental college and Hospital, Mathura.
[2]MDS, Private practitioners, Mathura.
[3]Prof & Head, Department of Prosthodontics, crown & bridge and Implantology, K.D Dental college and Hospital, Mathura.

Abstract

The abstract presents a case study involving two brothers afflicted by ectodermal dysplasia (ED), an uncommon genetic condition that affects structures originating from the ectoderm. The primary classification of ED discussed includes the hypo hidrosis form, characterized by features such as hypodontia, hypohidrosis, and hypotrichosis, commonly caused by ectodysplasin gene mutations. The case reports detail the clinical presentation of both brothers, including missing teeth, heat intolerance, and physical anomalies typical of ED. The treatment approach involved prosthetic intervention with complete dentures to address functional and aesthetic concerns. The procedural steps for a prosthesis fabrication are outlined, emphasizing the value of long-term monitoring and interdisciplinary cooperation in the care of pediatric patients with ED. The abstract underscores the significance of early intervention and comprehensive care customized to the specific needs of patients with ED to enhance their quality of life.

Keywords- Anodontia, complete dentures, ectodermal dysplasia, hypohidrosis, hypotrichosis.

Address of correspondence: Dr. Srishti Mishra, K.D Dental college and Hospital, Nh 2 road, Mathura, 281006, India.
Email address: - shishti778@gmail.com **Phone no:** 8770773017. **DOI:** 10.5281/zenodo.11050154
Submitted: 01-Mar-2024 **Revised**: 12-Mar-2024 **Accepted:** 27-Mar-2024 **Published:** 26-Apr-2024
Bibliographic details: Journal of Orofacial Rehabilitation Vol. 4(1), Apr 2024, pp. 28-32.

Introduction

Ectodermal dysplasia (ED) represents a diverse array of inherited conditions marked by abnormalities impacting multiple structures derived from the ectoderm. With a wide spectrum of presentations, it's estimated that around 1 in 100,000 live births are impacted by ED, encompassing more than 150 recognized variations. These abnormalities can affect a range of ectodermal tissues, including the teeth, skin, hair, nails, and sweat glands.

The primary classifications of ED are hypo hidrotic and hidrotic types, distinguished by the functionality of sweat glands. The X-linked hypo hidrotic form, also known as Christ-Siemens-Touraine syndrome, is distinguished by peculiar features such as hypodontia (partial, or complete absence of teeth), hypo hidrosis (reduced ability to sweat), and hypotrichosis (sparse or absent hair). The main cause of this type of ED is mutations in the X chromosomes ectodysplasin gene (EDA). On the other hand, mutations in genes like GJB6 or connexin-30 cause the hidrotic type, sometimes called Clouston syndrome, which usually does not involve sweat glands and is inherited as an autosomal dominant feature.

A timely and accurate diagnosis of ED is crucial for initiating comprehensive management strategies. This involves a multidisciplinary approach, encompassing dental interventions to address hypodontia or malformed teeth, physical therapies to manage associated physical anomalies, social support for affected individuals and their families, and psychological rehabilitation to address any emotional challenges stemming from the condition. Early intervention is essential not

only for addressing immediate physical needs, but also for promoting overall well-being and improving the long-term prognosis for individuals living with ED.

This study reports 2 cases of hypo hidrotic ED in a same family.

Case Report 1

Accompanied by his father, a 6-year-old boy visited the Department of Prosthodontics, Crown and Bridge at K.D. Dental College and Hospital, Mathura, complaining of missing teeth since birth.

Case Report 2

The elder brother of the first patient, an 8-year-old boy, also suffered from similar symptoms such as frequent fevers, heat intolerance, inability to sweat, and. General and extraoral evaluation evidently showed the standard characteristics of ectodermal dysplasia. An intraoral examination revealed missing teeth in both the maxillary and mandibular arches as well as a comparatively dry mucosa.
Radiographic examination (orthopantomogram) confirmed missing permanent tooth buds.

Based on their history and clinical and radiological findings, the patient was diagnosed with hypo hidrotic ectodermal dysplasia with complete anodontia. A prosthetic evaluation was performed on both patients with the goal of enhancing speech, mastication, and appearance.

Treatment approach:

To improve the patient's psychological and social well-being, full dentures were suggested for both maxillary and mandibular arches. Though Osseo integrated implants were thought to be essential for a permanent occlusion restoration, their implantation was delayed until after the conclusion of jaw growth.

Procedure:

Primary impressions were made using elastomeric impression material, followed by pouring casts with type II dental plaster.
Cold cure acrylic resin was used to fabricate the special trays, providing a consistent 2 mm entire arch wax spacer to ensure an optimal fit.

Low-fusing impression compound was used to seal the border, and zinc oxide eugenol was used to take secondary impressions.
Following the fabrication of master casts, occlusal rims with temporary denture bases were made and jaw relations were obtained. Non-anatomic teeth with a flat, atrophied ridge and impaired neuromuscular control were chosen.

After the final trial, heat-polymerized denture base resin was used to manufacture waxed dentures. Patients then received advice on maintaining dental hygiene and denture care along with their finalised dentures.
Subsequent recall appointments were scheduled for necessary adjustments.

Discussion:

The management of paediatric patients with ectodermal dysplasia (ED) presents numerous challenges, requiring proficiency in a number of areas, including behavioural management, prosthesis fabrication skills, growth and development, and inspiring the patient and parent into using their prosthesis. In presented cases, the initial prosthetic intervention aimed to address the functional and aesthetic concerns of the patients, which are crucial for their social integration and psychological well-being.
To assess the stability and efficacy of the prosthetic intervention, long-term follow-up is crucial, and it may be necessary to make modifications or replacements. Ensuring the

best possible care for children with ED requires collaborative efforts among a multidisciplinary team that includes an oral and maxillofacial surgeon, prosthodontist, orthodontist, and paediatric dentist.

While there isn't a predetermined period for initiating treatment in paediatric ED cases, early intervention is recommended to address functional and aesthetic concerns before the child starts school. However, initiation of the treatment should be made collaboratively, involving the treating dentist, parents, and patient. Given the young age of individuals with ED, behavioural management techniques are essential for ensuring cooperation and compliance during treatment procedures.

Various treatment modalities may be employed in paediatric ED cases, including removable prosthodontics, fixed partial dentures, direct composite restorations, orthodontic therapy, and dental implants. Every treatment plan should be customised to meet the needs of each patient, taking into account things like age, orthopaedic concerns, tooth loss, and abnormalities. Removable prosthodontics, such as complete dentures and overdentures, are commonly used to treat tooth loss resulting from trauma, cavities, or congenital absence of teeth.

To align teeth and fix occlusal abnormalities, orthodontic procedures might be necessary, while dental implants offer a promising solution for restoring missing teeth. However, concerns regarding implant placement in developing alveolar bone must be carefully considered, with implant placement typically deferred until maximum jaw growth is achieved.

A multidisciplinary approach is necessary for the comprehensive therapy of paediatric patients with ED, with careful consideration of growth and development, behavioural management, and Treatment approaches customized to meet the specific needs of each patient. Long-term follow-up and collaborative endeavours are vital in guaranteeing ideal results and enhancing the well-being of ED patients.

Conclusion:

Ectodermal dysplasia (ED) comprises a diverse array of hereditary conditions with familial inclinations. Within this study, two siblings exhibited characteristics indicative of hypo hidrotic ectodermal dysplasia . Dentists wield considerable influence in promptly identifying the disorder, thus facilitating timely interventions crucial for the successful patient management.

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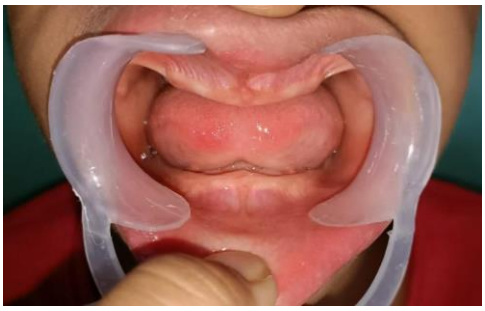
FIGURES

Figure 1



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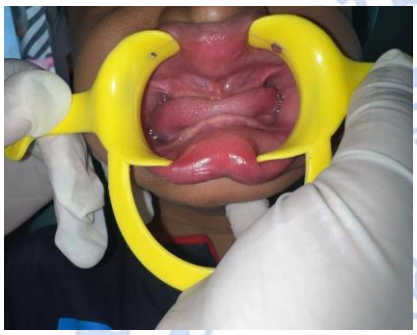

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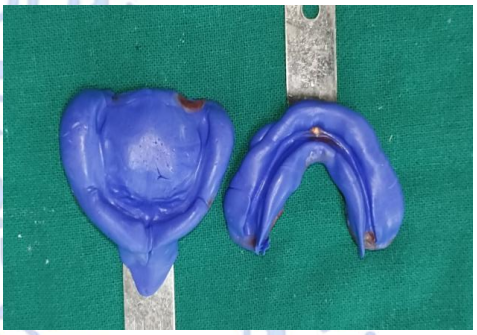

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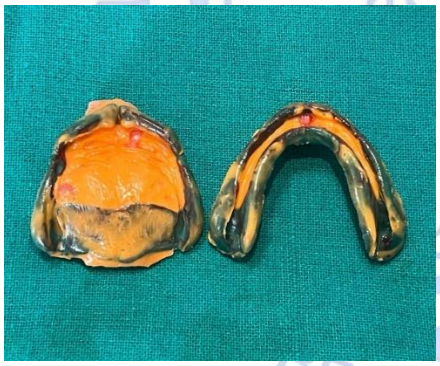

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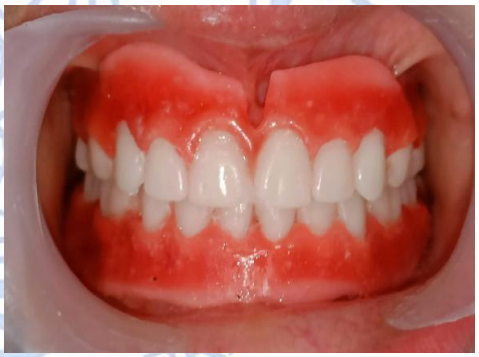

Figure 6

Figure 7

Figure 8