“Dental Care and Complete Denture Therapy for Ectodermal Dysplasia Patients: A Comprehensive Approach”

Rupal Shah¹, Sanjay Lagdive¹, Ekta Chheda¹, and Bansri Tank¹

¹Government Dental College and Hospital Ahmedabad

August 27, 2023

"Dental Care and Complete Denture Therapy for Ectodermal Dysplasia Patients: A Comprehensive Approach"

Abstract

Ectodermal dysplasia is a genetic disorder that affects various aspects of a person’s development. It is characterized by a triad of symptoms: hypodontia (lack of some teeth), hypotrichosis (sparse hair), and anhidrosis (inability to sweat properly). Individuals with this condition often face challenges related to their oral health, appearance, and ability to regulate body temperature.

Managing ectodermal dysplasia through prosthodontics poses challenges due to inherent craniofacial dysmorphology, and a diverse range of dentofacial irregularities, compounded by the fact that affected individuals are often evaluated for treatment at a young age. Early dental intervention is crucial for this population’s physiological and psychosocial reasons.

The oral expression of ectodermal dysplasia (ED) can encompass conditions such as anodontia or hypodontia, sometimes in conjunction with cleft lip and palate. The absence of teeth contributes to the diminished height of the alveolar ridges, causing a decrease in the vertical dimension of the lower face. This can lead to the vanishing of the vermilion border and the prominence of the lips, ultimately giving the affected individual’s countenance a likeness to that of an older person.

This article provides an overview of dental treatment choices for individuals with ectodermal dysplasia and presents an in-depth case study illustrating the tailored prosthodontic approach involving complete denture therapy, specifically adapted for a younger patient who reported issues with chewing and an unesthetic jaw appearance.

Keywords

ectodermal dysplasia, anodontia, oligodontia, overdenture, complete denture

Introduction

According to Freire-Maia’s classification, the category of Ectodermal dysplasia encompasses syndromes that demonstrate a minimum of two of the subsequent characteristics: abnormal hair (trichodysplasia), irregular dentition, unusual nails (onchodysplasia), and abnormal or absent sweat glands (dyshidrosis) [1]. Thurman’s initial report of an ED patient dates back to 1848, although Weech officially coined the term in 1929 [2,3].

Ectodermal dysplasia can be inherited through three main patterns: X-linked recessive, autosomal recessive, and autosomal dominant. Ectodermal dysplasia (ED) is categorized into two main forms: Hidrotic (Clouston’s syndrome) and Hypohidrotic (Christ-Siemens-Touraine syndrome) [4]. The key distinction between these two forms lies in their sweat gland manifestations. Hidrotic ectodermal dysplasia is characterized by autosomal dominant inheritance and normal sweat glands, whereas Hypohidrotic ectodermal dysplasia, with
X-linked recessive inheritance, presents with absent sweat glands. Among these, Hypohidrotic ectodermal dysplasia (HED) is the most prevalent, predominantly affecting males and displaying a higher severity [4].

The characteristic facial features of ED, often not identified until infancy, encompass frontal bossing, hollowed cheeks, saddle nose, thick outward-turned lips, and periorbital skin with wrinkles and hyperpigmentation. Dental implications involve conical or peg-shaped teeth, partial or complete absence of teeth (hypodontia or anodontia), and delayed eruption of permanent teeth [5]. The average count of missing permanent teeth averages 23.7, with the most frequent presence observed in maxillary central incisors, maxillary first molars, mandibular first molars, and maxillary canines. These teeth tend to exhibit a conical shape, particularly the maxillary central incisors and maxillary/mandibular canines. The alveolar process may not develop in areas devoid of teeth, and even in dentate regions, its development can be suboptimal [6].

Traditional dental management for ED patients revolves around providing a sequence of complete or removable partial dentures during the developmental stages [6]. Definitive rehabilitation is typically pursued after the cessation of jaw growth [7]. The patient’s age, dysplasia pattern, existing tooth count and morphology, and alveolar ridge structure play pivotal roles in determining the appropriate dental interventions.

Case Report

A 14-year-old male patient, accompanied by his grandfather, reported a chief complaint of inability to chew food and an unaesthetic appearance. None of his family members had Ectodermal dysplasia.

On examination, the patient displayed the characteristic traits of ED, including hypodontia, anhydrosis, hypotrichosis, a prominent forehead, scarce eyebrow, darkly pigmented skin around the periorbital area, reduced lower anterior facial height, a flat mandibular plane, saddle nose, thick upper lip, everted lower lip, prominent chin, and resulting concave facial profile (Figure 1).

An intraoral assessment revealed the presence of three teeth in the maxilla: deciduous right maxillary first and second molar and deciduous left maxillary second molar. He had a completely edentulous mandibular atrophic ridge and macroglossia. Furthermore, he displayed a loss of vertical dimension, absence of alveolar processes, and irregular development of alveolar ridges (Figures 2a and 2b). OPG indicated alveolar bone aplasia and the absence of other tooth buds (Figure 2c).
Figure 1: Pre-operative extra-oral view.
Given the patient’s age and the limited amount of available alveolar bone, the treatment approach for the younger child included crafting an interim overdenture for the maxillary arch without any modification to existing teeth as the child was uncooperative. A complete denture was considered for the mandibular arch. This therapeutic strategy aimed to enhance the psychological and social well-being of the patient.

Although osseointegrated implants were contemplated as a crucial aspect of definitive occlusion restoration, their placement was postponed until after the completion of jaw growth.

A preliminary impression was taken with irreversible hydrocolloid (DPI Imprint Alginate). The casts were poured with type II dental plaster. A special tray was prepared from cold cure acrylic resin, featuring a uniform 2 mm full arch wax spacer that covered the natural teeth and ensured a proper fit. Peripheral border sealing was achieved by low-fusing impression compound (DPI Pinnacle Tracing Stick), and secondary impressions were taken using light body condensation silicone (Coltene Speedex). Subsequently, master casts were created, and occlusal rims with temporary denture bases were fabricated. Jaw relations were established by manually guiding the mandible into a centric position. The patient was provided with a reduced vertical dimension due to the instability of the mandibular denture resulting from macroglossia. The jaw record was mounted on a three-point articulator.

Non-anatomic teeth were selected and provided to the patient due to compromised neuromuscular control and a flat, atrophied ridge. Following the final trial, the waxed dentures were processed using heat-polymerized denture base resin. The finalized complete dentures were then provided to the patient (Figure 3 and 4), accompanied by instructions on oral hygiene and denture care. Subsequent recall appointments were scheduled for necessary adjustments.
Figure 3: Intra-oral front view post-denture insertion
Within 6 months, the dentures, particularly the mandibular one, began to fit poorly. The patient could no longer wear the lower denture while eating due to discomfort. Both jaws had increased in size. To address this, a new complete denture was crafted for the lower arch, and an upper maxillary overdenture was retained. The fabrication process followed the conventional steps.

**Discussion**

Providing treatment to a pediatric patient with ED necessitates the clinician's proficiency in various areas such as growth and development, behavioral management, prosthesis fabrication techniques, modifying existing teeth using composite resins, motivating both the patient and parent in prosthesis use, and ensuring
long-term follow-up for potential prosthesis adjustments or replacements. Collaborative efforts involving a multidisciplinary team consisting of a pediatric dentist, prosthodontist, orthodontist, and oral and maxillofacial surgeon have been proposed in certain reports to ensure optimal care for young individuals with ED.

While there is no definitive timeframe to initiate treatment, Till and Marquesz [8] suggest delivering an initial prosthesis before the child starts school, allowing for a normal appearance and adaptation time. Ultimately, the decision to commence treatment should be a joint decision involving the treating dentist, parents, and patient. Considering that individuals with ED are typically young during treatment evaluation, the treating dentist should possess skills in the behavioral management of pediatric patients. Nussbaum and CarreP [9] suggest sedation for managing challenging cases requiring extensive prosthodontic work. In contrast, Nowak [10] opposes sedation, asserting that success hinges on patient comprehension and compliance. Instead of sedation, Nowak advocates a “tell-show-do” approach to familiarize patients with impending dental procedures. This conditioning approach has been effectively utilized by other authors as well.

Based on factors like teeth abnormality, loss, orthopedic issues, and patient age in ED cases, various treatments have been applied individually or combined, including complete dentures (CD), removable partial dentures, fixed partial dentures (conventional or resin), implant-supported prostheses, autotransplantation, direct restorations, orthodontic therapy, and orthopedic/orthognathic interventions [11].

Removable prosthodontics is a frequently employed treatment approach for individuals with ED, particularly those experiencing anodontia or hypodontia. The absence of congenital teeth, coupled with tooth loss due to caries or trauma, often necessitates the use of removable prosthetics for young ED patients. Treatment options may encompass complete dentures, removable partial dentures, overdentures, or a combination thereof. Complete dentures offer satisfactory functional and aesthetic rehabilitation, although their retention on severely hypoplastic ridges can be challenging. In such cases, vestibuloplasty and ridge augmentation can enhance both hard and soft tissue support. Overdentures have the added benefit of preserving alveolar bone, as confirmed by studies. Overdentures utilize existing natural undercuts and incorporate precision attachments to enhance stability and retention. Alveolar bone preservation is crucial for individuals with ED, who rely on the ridges for prosthesis support early on [1].

In individuals with ED, who typically have a limited number of teeth, caution is advised when considering fixed partial dentures (FPDs) with rigid connectors, particularly in young patients still undergoing active growth. The rigidity of FPDs could potentially disrupt jaw development, especially if spanning the midline. An illustrative case by Hogeboom highlighted the impact of jaw growth on a detachable fixed prosthesis, which separated at the midline due to transverse growth [1].

While individual crown restorations, factors like larger pulp sizes and shorter crown heights may warrant consideration. Despite these considerations, crowns are frequently used in treating young ED patients. The emergence of direct composite restorations offers a preferred method for restoring natural morphology to hypoplastic teeth commonly seen in ED cases. The incorporation of crowns and direct composite restorations becomes necessary to achieve proper tooth contours for RPD abutments [1].

Orthodontic interventions may be necessary to align teeth. Functional appliances could be advised for correction of overjet, overbite, infra-occlusion, and tilted abutments. Occlusal plane therapy can be initiated when there is a requirement for posterior tooth eruption before commencing prosthodontic treatment. [12].

Dental implants are now being utilized more frequently in managing ED patients. However, concerns have been raised about placing osseointegrated implants in developing alveolar bone. Implants in young ED patients might become submerged due to ongoing alveolar process growth, potentially leading to issues like infra-occlusion and frequent adjustments. The risk of peri-implantitis and inadequate crown/implant ratios could also arise. Implant placement might require bone grafting in cases of severe bone atrophy. As a result, dental implants are typically contraindicated for children under 6 years old [13,14]. A consensus conference on implantology in 1989 suggested that implant placement should be deferred until maximum jaw growth is achieved, typically around 15 years of age. For girls, implant placement after age 15, and for boys after age
18, yielded the most predictable outcomes [1].

This study highlights the comprehensive approach to managing a child with anhidrotic ectodermal dysplasia, with a focus on immediate esthetic and functional restoration while considering the ongoing growth of the jaw. Enhancing the patient’s facial aesthetics and masticatory function played a crucial role in facilitating their social integration with peers, averting potential psychological consequences, and preserving their self-esteem. The ultimate goal is to implement an implant-supported prosthesis with bone augmentation when necessary, representing the first phase of a longitudinal study for this particular case.

Conclusion

Patients with Ectodermal dysplasia are difficult to manage. They require regular follow-up every six months and it is very important to keep them motivated to lift their self-esteem. Encouraging and educating them about dental care is of utmost importance so that they report back for regular check-ups and accept complex dental procedures.

Acknowledgment

The authors would like to thank the study participants for their participation and kind cooperation throughout the study.

Conflict of interest

The authors declare that they have no conflict of interest.

Source of funding

Not applicable.

References

2. Thurnam J. Two cases in which the skin, hair and teeth were very imperfectly developed. Med Chir Trans. 1848;31:71-82.