MANAGEMENT OF HYPOHYDROTIC ECTODERMAL DYSPLASIA WITH REMOVABLE PROSTHESIS IN A PEDIATRIC PATIENT

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ABSTRACT: Rare genetic disorders known as ectodermal dysplasias are characterized by abnormal development of specific ectodermal-derived tissues and structures. Dental features associated with these disorders include congenitally missing teeth (hypodontia or anodontia), malformed (conical or peg-shaped) anterior teeth, ankylosed primary teeth, delayed eruption of permanent teeth, macrodontia or microdontia, and enamel hypoplasia. Affected children require a multidisciplinary approach. Early in childhood, it is advisable to use removable prostheses to enhance speech, mastication, aesthetics, and self-esteem, as well as to prevent bone resorption. Implants are generally recommended only after growth is complete In the case presented, a six-year-old child with hypohidrotic ectodermal dysplasia and partial anodontia was rehabilitated with a partial denture in the maxillary arch and a complete denture in the mandibular arch.

Keywords: Ectodermal dysplasia, prosthetic rehabilitation, removable prosthesis.

INTRODUCTION

Ectodermal dysplasia encompasses a diverse group of inherited disorders that impact the development of epidermal appendages.[1]This condition arises from disruptions in the ectoderm during embryonic development, leading to abnormalities in tissues and appendages derived from the ectoderm.[2, 3] The first documented case of ectodermal dysplasia was reported by Thurnam in 1948. Current estimates indicate that the prevalence of this syndrome is approximately 7 in 10,000 individuals, with a mortality rate of 28% among males by age 3.[1, 4]

The EDA (ectodysplasin A) gene, located on the X chromosome at Xq12–q13.1, plays a crucial role in the development of ectodermal appendages, including keratinocytes, hair follicles, and sweat glands. Mutations in this gene are responsible for the majority of ectodermal dysplasia cases.[5]

It has different modes of inheritance including autosomal dominant, autosomal recessive, and X-linked recessive traits. The x-linked trait also called the 'Christ-Siemens-Touraine syndrome', is the most common [3] and is characterized by a triad of clinical features that includes absent or diminished sweat glands(anhidrosis or hypohidrosis), hypotrichosis, and hypodontia.[6] The second common form, is the hidrotic type also called 'Clouston's syndrome'which is more severe and involves nail dystrophy, hypotrichosis, and palmo-plantar keratosis as its main features.[7]

These syndromic patients can be easily distinguished by their orofacial features including frontal prominence, prominent supraorbital ridges, sunken cheeks, wrinkles and periorbital hyperpigmentation, saddle nose, everted lips, large low-set ears, brittle nails, reduced lower facial height. Due to these features, their faces generally have an elderly appearance. [3, 6]

Dental features may include- congenitally missing teeth(hypodontia or anodontia), malformed (conical or peg-shaped) anterior teeth, ankylosed primary teeth, delayed eruption of permanent teeth, macrodontia or microdontia, and enamel hypoplasia. Generally,these children have normal IQ levels and intelligence[6,7].Based on the literature, early treatment is indicated in these patients, to improve esthetics, speech, and eating habits and to promote emotional and social well-being in them for improved acceptance by family and peers.[2, 5]

The following case report describes the prosthetic rehabilitation of a young patient.

CASE REPORT

A six-year-old male patient presented to the Department of Pedodontics and Preventive Dentistry at Dr. R Ahmed Dental College and Hospital with a chief complaint of missing teeth. The patient was accompanied by his parents. The mother reported a history of missing teeth since infancy, with only the eruption of maxillary molars occurring when the child was around 2 years old. She also mentioned that she had missing lower anterior teeth since birth.

The patient's extraoral examination revealed features such as frontal prominence, a saddle nose, sparse eyebrows and eyelashes, reduced lower facial height, and dry skin over the body (Fig. 1). Intraoral examination showed the presence of teeth 55 and 65, along with a peg-shaped lateral incisor on the left side of the upper arch. The mucosa appeared relatively dry, and there was complete anodontia in the mandibular arch (Fig. 2). Radiographic examination (orthopantomogram) indicated the formation of only the crowns of teeth 16 and 26, with no tooth buds evident in the mandibular arch (Fig. 3). The clinical features suggest a diagnosis of hypohidrotic ectodermal dysplasia, characterized by the classic triad of hypohidrosis, hypodontia, and hypotrichosis.

PROSTHODONTIC MANAGEMENT

It was decided to rehabilitate the patient with a complete removable denture for the lower arch and a partial removable denture for the upper arch. The treatment plan, including the need for frequent follow-ups due to the patient's considerable remaining growth, was explained to the parents. After obtaining their consent, the treatment commenced.

To familiarize the patient with the instruments and materials and reduce his anxiety, preliminary impressions were taken using the smallest stock tray with alginate hydrocolloid impression material. These impressions were poured with dental plaster to create primary casts. Custom trays were then prepared, with relief provided in the thin, knifelike ridge of the mandible using a wax spacer. Border molding was performed with greenstick compound to minimize trauma to the patient (Fig. 4). Final impressions for both arches were made with light-body addition silicone material, and a master cast was created (Fig. 5). Record bases with occlusal rims were fabricated, and jaw relations were recorded (Fig. 6).

The master casts were mounted in a simple hinge articulator using the interocclusal record. Acrylic teeth of the appropriate shade were selected and trimmed to dimensions suitable for primary dentition to ensure optimal occlusion and aesthetics.

Bilateral balanced occlusion was used while arranging the teeth on waxed rims. A trial with these waxed occlusal rims was conducted. After the trial dentures were approved by the child's parents, the dentures were finalized and processed with heat-cured acrylic resins. The dentures were then polished and inserted into the patient's mouth, with the conical tooth reshaped as a lateral incisor for improved aesthetics (Fig. 7). Post-insertion instructions regarding oral hygiene and denture maintenance were provided, and the child was asked to return after one week for further adjustments. Improvements in facial profile and oral functions were noted during subsequent recall appointments.

DISCUSSION

Various treatment strategies have been explored for managing oligodontia and anodontia. Typically, conventional removable complete or partial dentures are recommended for these patients. Early placement of such prosthetic dentures can significantly improve functions like chewing, swallowing, speech, and overall appearance. According to Khazaie and colleagues, prosthetic treatment can be initiated as early as preschool age, allowing young children to adapt to their use. Most experts agree that effective prosthetic intervention can commence around ages 2 or 3 in children who are cooperative.8, 9, 10, 15, 16, 17

The process of creating a denture is akin to that used for adult patients; it involves taking initial and final impressions, registering the occlusion, try-in, and inserting the completed denture. Nonetheless, certain adaptations to the widely recognized methods are typically necessary for children and call for extra care [8].

A child's prosthesis must be made after a thorough diagnostic procedure and a thorough review of their medical and dental histories. To create a prosthesis that is not only functional but also aesthetically pleasing and age-appropriate, all these criteria should be taken into account during the construction process [11]. It is also challenging to achieve retention and stability of the prosthesis during a child's growth period, mostly because of the absence of sufficient undercut zones, generally amorphous tooth structure, and insufficient bone support [16, 17]. In this case, care was given to fabricate a denture with a wide distribution of occlusal force, fully extending the denture base. Additionally, particular attention was given to the impression technique; as in complete dentures, support should encompass the vestibular sulcus in its entirety for a retentive base construction with a sealed border, rather than just the denture base region [15]. For primary impressions, irreversible hydrocolloid material with a higher viscosity is recommended by most authors, which was used for the primary impression for this case. This is likely due to its faster setting time, which helps prevent aspiration of the impression material and improves patient comfort[14,15]. Making a proper custom tray requires accurate impressions with appropriate anatomic extensions, which contributes to a better final impression[16].

Sustaining a high degree of cooperation from these patients is essential and reliant upon the application of specific management and motivational strategies[18]. Regular periodic recalls of young patients with complete dentures are typically required until more permanent implant-assisted prostheses can be delivered, as when a decreased vertical dimension of occlusion and an abnormal

mandibular posture are detected due to growth, dentures should be adjusted, relined, rebased, or replaced.

Depending on the patient's needs and the severity of the disease, other prosthodontic treatment options include fixed prostheses, implant-supported prostheses, and flexible dentures.

The use of fixed prosthodontic treatment alone to treat ED is uncommon due to the large pulp cavity and the presence of a few teeth in these patients. As a result of its stiff connectors, a fixed prosthesis that crosses the midline interferes with a young patient's growth ability in maxilla[19].

The prosthesis can be stabilized, retained, and supported with dental implants. The age of the patients determines the success of the implant therapy. Growing children are not treated with implants, especially in the maxilla[20]. Implants are advised for the anterior part of the mandible in children older than preschool age (7 years or older), per the National Foundation for ED recommendations. Implants placed after the ages of 15 for girls and 18 for boys offered the most predictable outcome, according to Cronin et al[21].If oral implants are implanted before skeletal growth and dental maturity, there are several outcomes, as Multidimensional limitations on the growth of the craniofacial skeleton, unpredictable implant dislocations because they do not contribute to the process of maxillary growth drift and displacement, implant exposure due to bone resorption, maxillary growth limitation if implants are connected by a rigid prosthesis that crosses the midline, infraocclusal position of inserted implants compared with adjacent natural dentition, and trauma to adjacent teeth buds resulting in abnormalities in dental development [22,23]

Hence, Removable prostheses made with acrylic resin (complete dentures, partial dentures, or over-dentures) are the most frequently reported treatment modality for the dental management of ED in childhood; these are cost-effective and can be easily readapted and modified (relaying) during periods of rapid growth. Early use of oral prostheses on patients requires a long-term commitment from the patient, the clinician, and the patient's parents. Pediatric dentists are a vital component of the multidisciplinary team that cares for these children, helping them to develop emotionally and clinically.

CONCLUSION

Ectodermal Dysplasia should be treated as early as possible to maintain the social and emotional well-being of the child. Removable dentures in such patients serve as a non-invasive, cost-effective, viable, and efficient treatment and help the patient to live a normal life.

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FIGURES WITH LEGENDS

Figure 1: Orofacial characteristics of the patient showing frontal bone prominence, sparse eyebrows

Figure 2: Intraoral picture showing the partially edentulous upper arch and completely edentulous lower arch.

Figure 3: Orthopantomogram showing two deciduous molars in upper arch, one peg-shaped lateral incisor in the upper left quadrant, permanent maxillary 1st molar tooth buds. Lower arch revealed no tooth buds.

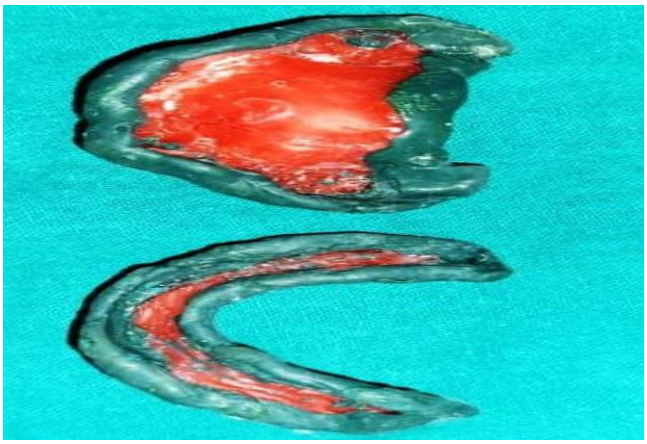

Figure 4: Border moulding

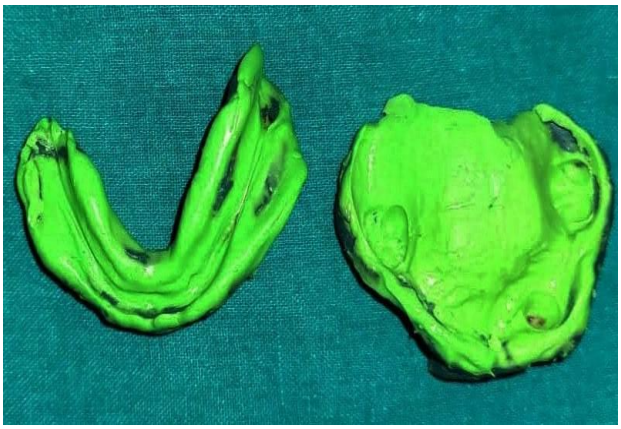

Fig 5: Final impression taken with light body silicone material

Fig 6: Occlusal rims made on acrylic baseplate.

Fig 7: After insertion of denture and reshaping the peg lateral.

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