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Case Report

Prosthodontic management of hypohidrotic ectodermal dysplasia: A case report with 4 years follow up

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Abstract

Hypohidrotic ectodermal dysplasia (HED) is a rare genetic disorder characterized by defects in ectodermal structures, including teeth, hair, nails, and sweat glands. This case report describes the prosthodontic management of an 8-year-old male child diagnosed with HED, who presented with missing teeth and difficulty chewing. The patient exhibited characteristic features of HED, such as a low nasal bridge, prominent chin, sparse hair, and dry, scaly skin. Radiographic examination confirmed the absence of retained deciduous teeth and permanent tooth germs. The patient was initially rehabilitated with an interim removable partial denture in the maxillary arch and a complete denture in the mandibular arch. Behavioral modification techniques were employed to help the child cope with the dental treatment. After four years, the patient was re-evaluated and a new set of prostheses was fabricated, consisting of a Cu-Sil denture in the maxillary arch and a conventional complete denture in the mandibular arch. The treatment aimed to improve the patient's masticatory function, speech, and social acceptance. This case highlights the importance of a multidisciplinary approach and long-term follow-up in the management of HED patients, as well as the role of prosthodontic rehabilitation in improving their quality of life.

Keywords: Ectodermal dysplasia, Cu-sil-denture, Early childhood denture, Overdenture

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1. Introduction

Ectodermal dysplasia (ED) is defined by National Foundation for Ectodermal Dysplasia as a large heterogeneous group of inherited disorder that is characterized by primary defect in the development of two or more tissues derived from embryonic ectoderm.[1] Freire-Maia defined ED as any syndrome that exhibits at least two of the following features: Trichodysplasia (abnormal hair), abnormal dentition, onchodysplasia (abnormal nails) and dyshidrosis (abnormal or missing sweat glands).[2]Other orofacial characteristic of this syndrome includes anodontia or hypodontia, hypoplastic conical teeth, hypotrichosis, and protuberant lips. Combinations of features were first reported by Danz in 1792. It was also found its mention in Wedderburn's letter to Charles Darwin in 1838.[3] It was Thurnam who reported hypohidrotic form of ED in 1848. Thadani assigned the X chromosome to have the causative association to ED in 1921. The term hereditary ectodermal dysplasia was coined by Weech in 1929. Felsher suggested the term hypohidrotic in 1944.[4]

Mutations in the ectodysplasin-A and ectodysplasin-A receptor genes are responsible for X-linked and autosomal HED.[5] ED is inherited by autosomal dominant, autosomal-recessive or X-linked genetic transmission. The most frequently reported manifestations of ED are hypohidrotic ectodermal dysplasia (HED) (Christ-Siemens-Touraine syndrome). Hypodontia is known as one of the major factors of ED and is almost always present in affected individuals.[6] These disorders are relatively rare and occur in 1 in 10,000 to 1 in 100,000 births.[7] Dental treatment is often necessary in patients with ectodermal dysplasia and children may need dentures in the early childhood. The remaining teeth should be preserved and modifications should be made in the treatment plan to include them in the intended form of

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prosthesis. Prosthetic options for management of ED can include fixed, removable or implant-supported prosthesis, that may be employed individually or in combination to provide an optimal result. This case report describes management of a 8 year old child diagnosed with hypohidrotic ectodermal dysplasia, who has recently been rehabilitated with Cu Sil denture in maxillary arch and conventional complete denture in mandibular arch.

2. Case Report

A 5 year old male child diagnosed with hypohidrotic ectodermal dysplasia **Figure 1:** Pre-op extra oral) was referred to the dental centre four years ago with the chief complaint of lack of teeth and difficulty in chewing (**Figure 2**). According to parents, upper arch contained few deciduous teeth initially but exfoliated spontaneously leaving only 55 and 65. The lower arch never contained any tooth. The child was fearful and anxious when he reported to the dental setup but he was cooperative. He was moderately built and nourished and his vitals were within normal range. However his parents complained about his intolerability to heat. On physical examination it was found that he had low nasal bridge, pronounced chin, sunken cheeks, prominent and everted cracked lips, circum-oral and periorbital hyper pigmentation, sparse, hypopigmented hairs, sparse eyebrows and eye lashes and his skin was dry and scaly. He also had defective fingernails. Intraoral examination revealed absence of all mandibular teeth, presence of teeth numbers 55 and 65 in the maxilla. 55 was carious and exposed. The panoramic radiograph confirmed the absence of retained deciduous teeth and there were no permanent tooth germ in either the maxilla or mandible (**Figure 3**).

Parents were sensitised and counselled about this condition, its maxillofacial and nutritional consequences and the scope of prosthodontic management according to age. The need for multidisciplinary management was also emphasised. As the child was fearful and aversive towards the dental team, behavioral modification in the form of desensitisation exercises and oral motor exercises like stretching, massage and vibration palpation and passive range of motion exercises were performed to modulate the child.[8]

Subsequently 55 was extracted after consultation with endodontist and pedodontist. A minimally invasive treatment plan was formulated and the child was rehabilitated with an interim removable partial denture in the maxillary arch and complete denture in the lower arch. One denture at a time was inserted with a gap of two weeks in between so as to enable the child to cope with the new prostheses. Well-structured follow up plan was devised and the post insertion instructions were provided to the parents. The child was also referred to a speech therapist for articulation training. The prosthesis was adjusted every month for the first 3 months and then half yearly follow up schedule was followed.

After four years the child was re-evaluated for replacement of the prosthesis as he complained the prosthesis to be very loose. The child was calm, conscious and alert this time and co-operated with the dental team. It was found that his profile has been improved in terms of proportional vertical and horizontal growth. His speech was also found to be well articulated. His parents reported that he was well accepted amongst his peers in school without any sign of discrimination. A fresh set of investigations in the form of OPG and CBCT was advised to evaluate the availability of bone in the maxilla and mandibular arch. It was found that 65 was in perfect health. CBCT data showed that there was inadequacy of bone for implant placement in the maxilla and mandible and conventional implants could not be placed without any adjunctive augmentation surgery. Keeping in mind the age of the patient and future compliance towards dental treatment it was decided to undertake removable prostheses in the form of Cu Sil denture in maxillary arch and conventional complete denture in mandibular arch to rehabilitate him at the age of 9 years.

A preliminary impression was made using stock edentulous tray and putty consistency polyvinyl siloxane impression material (**Figure 4**). The final impression was made with a custom tray fabricated with autopolymerizing acrylic resin and border-moulded with the help of green stick impression compound. Final impression was made subsequently with light body consistency polyvinyl siloxane impression material (**Figure 5**). Master casts were poured using type III dental stone. They were mounted on a Hanau wide vue semi-adjustable articulator with a Hanau spring bow arbitrary facebow instrument, using condylar guidance of 30°, Bennett angle of 15° and standard intercondylar distance (110 mm). Occlusal vertical dimension was established using the physiological rest position associated with phonetic and esthetic. Centric occlusion was established according to dynamic records based on unforced movements of the jaws in the terminal hinge position performed by the patients under the supervision and manual guidance (**Figure 6**). The artificial teeth were arranged in wax occlusion rims and trial was taken. The denture was fabricated in centric occlusion with balanced articulation and anatomically shaped acrylic teeth with a 33 degree cuspal inclination°.

The patient and his parents accepted the arrangement of teeth. The dentures were waxed, processed, finished and polished. A silicone ring was provided around the tooth 65 to maintain the peripheral seal and to aid in comfort to the patient (**Figure 7**). Following denture insertion, the child and parents were counselled about its use, cleaning procedures and the importance of follow up. The patient was monitored for verification of improvement in physical and emotional state, with appropriate adaptation to new denture and subsequent progress in speech and esthetics (**Figure 8**). Future recall was also planned at 6 months to monitor bone growth and for eventual relining or construction of a new denture.

Figure 1: Pre-op extra oral

Figure 2: Pre-op intra oral

Figure 3: OPG

Figure 4: Primary impression and cast

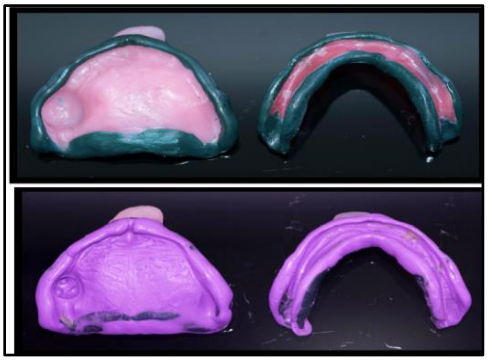

Figure 5: Secondary impression and cast

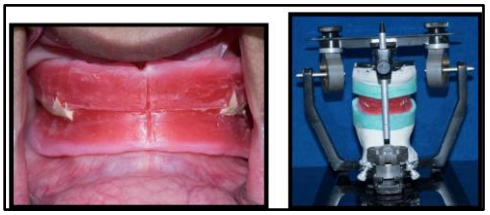

Figure 6: Jaw relation & mounting on semi adjustable articulator

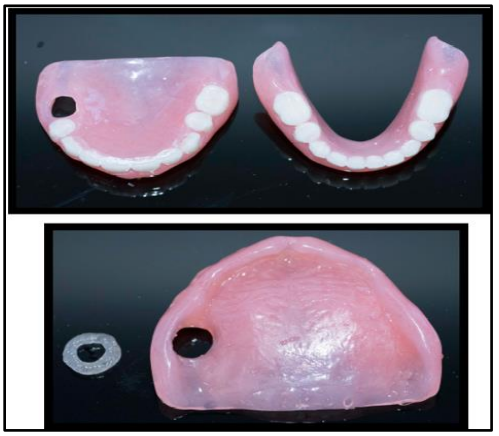

Figure 7: Final dental with ring

Figure 8: Post op extra oral

3. Discussion

The main objectives of prosthodontic rehabilitation in patients with ED are to improve the esthetics, phonetics and masticatory function thereby ensuring the overall psychosocial well-being of the individual.[9] Prostheses also improve the tone of the muscles of mastication and compensate for the reduced vertical dimension. It is crucial for a child to be able to develop physically, emotionally, and socially in the same way as other healthy children. This is especially important for a child's overall well-being and growth. It is imperative to prepare a long-term treatment plan that includes regular reviews to ensure that the patient with ED maintains an adequate level of oral health care. Pedodontist, orthodontist, maxillo - facial surgeon, prostho-dontist as well as speech therapist should be included in the treatment of patients with ED. The objectives of orthodontic treatment during period of growing and development, are improving the sagittal and vertical skeletal relations that could be achieved with modification of the functional appliances simultaneously with eruption of the maxillary and mandibular molars and dental arch expansion. The successful use of any prosthesis depends on the cooperation and communication between members of the multidisciplinary team and the patient.

Nowak[10] stated that "treating the paediatric patient with ED requires the clinician to be knowledgeable in growth and development, behavioral management, techniques in the fabrication of prosthesis, the ability to motivate the patient and parents in the use of the prosthesis, and the long-term follow-up for the modification and/or replacement of the prosthesis."

Till and Marques[11] emphasised the requirement of an initial prosthesis to be delivered before the child begins school so that the child has a normal appearance and time to adapt to the prosthesis. Ultimately, the decision to commence the treatment should be planned by the treating prosthodontist along with the parents and the child.[10]

Prosthodontic rehabilitation for an ED patient consists of various combinations of overdenture, complete or partial removable prostheses, fixed dental prostheses and implant prostheses.[12]Removable prosthesis is the most frequently reported treatment modality for the dental management of ED.[13] In the present case, the decision to rehabilitate the child with removable prosthesis was made initially as the primary management strategy because it enabled easy modification of the prosthesis during rapid growth periods. It also offered the ED patient and his family an easy, affordable and reversible method of oral rehabilitation. Cooperation of the patient as well as the support of the family made the rehabilitation outcome successful. The child became more cooperative towards the dental team in the past 4 years and complied with the follow up schedule efficiently.

The need to change the prosthesis has been attributed to the continual jaw growth. The present rehabilitation strategy of providing a Cusil denture in the maxillary arch stems from the salient advantages of preserving the natural teeth. Natural teeth maintains the alveolar bone, proprioceptive feedback retaining postural positional relationship of jaws, offers improved retention through the silicone ring. Also the retained tooth can be used to develop a future implant site in the strategic maxillary first molar region.

The use of endosteal implants has gained wide acceptance in prosthodontic management of ED. Ekstrand and Thomsson[14] Bergendal et al.[15], Smith et al.[16] and Cronin et al.[17] have reported situations where endosteal implants were successfully used in the prosthodontic management of ED. Several investigations have demonstrated improved physiological and psychological function in individuals with implant-supported dentures, as compared to their condition prior to implant placement or to an edentulous control group wearing complete dentures.[18,19,20,21]Ectodermal dysplasia doesn't have negative effect on survival rate of implants when inserted on adult patients.

UCSF has developed a classification system to simplify the protocol to be followed while considering implant therapy in the young patients afflicted with Ectodermal dysplasia.[22]

1. **Group-1:** Missing single tooth with permanent teeth adjacent to the site- To be restored by transitional prosthesis until growth is complete.
2. **Group-2:** Missing multiple teeth anteriorly or posteriorly with permanent teeth adjacent to the edentulous areas- Advised to wait until growth is complete but implants can be considered in selected patients before the cessation of growth.
3. **Group-3:** Essentially edentulous- Implants are indicated at an early age if the patient has the potential to be compliant.

However, According to Cronin et al.[17] possible consequences of early implant placement include implant submergence, implant exposure due to bone resorption, implant movement because of jaw growth and limitation of jaw growth if the implants are connected by a rigid prosthesis that crosses the midline. When implant placement in young ED patients is being considered, their dental and skeletal maturity, not their chronological age, should be the determining factor. Lekholm[23] concurs with criteria concerning maximum jaw growth, giving age guidelines of 14-15 years of age for girls and a year later for boys. He also recommends that an individual's growth curve be studied before any implant placement procedure is started. According to Mackie and Quayle[24] implant placement in children younger than 16-18 years must be avoided or they will remain in infra-occlusion due to adjacent alveolar bone growth.

The use of conventional removable prostheses before undergoing invasive implant therapy is recommended. It helps to condition the growing child and provides esthetic and functional information for the subsequent implant treatment planning.[25,26] The use of a conventional prosthesis is also important until the patient achieves a more appropriate level of maturity. The literature available for fabrication of overdenture in hypohidrotic ectodermal dysplasia doesn't involve Cu Sil ring as additional support or retention to the denture. It is to be kept in mind that an ED patient requires a life time commitment in terms of dental treatment which is ongoing, progressive and sometimes cumulative in nature. So injudicious and overzealous invasive treatment may predispose a child to become antipathetic towards the dental team and can have serious negative impact on the future compliance and outcome of dental therapy.

4. Conclusion

This case report emphasizes the importance of early diagnosis, interdisciplinary management, and regular follow-ups in the prosthodontic rehabilitation of children with hypohidrotic ectodermal dysplasia. Removable partial and complete dentures can be a valuable tool in restoring oral function, improving social acceptance, and enhancing the quality of life for these patients. Ongoing management is essential to adjust the prostheses as the child grows, ensuring continued success and adaptation to their evolving needs.

5. Source of Funding

None.

6. Conflict of Interest

None.

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