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Case Report

A simple novel appliance for prosthodontic rehabilitation of a child with hypohidrotic ectodermal dysplasia – A case report

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Abstract

A particular condition known as Ectodermal Dysplasia (ED) is defined by the congenital abnormality of one or more ectodermal components. Symptoms include abnormalities in the skin, nails and sweat glands. Many dental abnormalities, such as hypodontia, anodontia, impacted teeth, and peg-shaped or conical front teeth, inadequate alveolar ridge development are among the oral features. These diseases affect 1 in 10,000 to 1 in 100,000 newborns, which is a relatively low incidence. Clinically, ED can be categorised as Christ Siemens-Touraine syndrome, which is anhidrotic or hypohidrotic X-linked type that is inherited, characterised by the typical triad of hypodontia, hypohidrosis, and hypotrichosis, as well as by facial dysmorphism and other one is Hidrotic (Syndrome de Louis).Hidrotic kind of ED, which is inherited as an autosomal characteristic, can damage the teeth, hair, and nails but often spares the sweat glands. In ED patients, prosthetic dental therapy aims to retain alveolar ridges, promote early perioral musculature development, improve speech, and restore normal face features. This promotes typical emotional and psychological growth. In present case report, the aetiology, clinical symptoms, therapeutic possibilities of this hereditary illness and prosthetic management of oligontia are described and discussed.

Keywords: Anhidrotic, Ectodermal Dysplasia, Hidrotic, Hypodontia.

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1. Introduction

Ectodermal Dysplasia (ED) is an hereditary disorder that is diffuse, congenital, and non-progressive. It is a primary ectoderm-derived tissue developmental abnormality that affects two or more tissues. The principal tissues effected include those of the skin, hair, nails, exocrine glands, and teeth. The two most frequent forms of Ectodermal Dysplasia are Hidrotic and Hypohidrotic dysplasia.[1] Numerous abnormalities, including as hypohidrosis, aberrant dentition, onychodysplasia, and hypotrichosis, can be seen in Ectodermal Dysplasia.[2] Common facial characteristics include a saddle nose, sunken cheeks, hyperpigmented skin around the eyes, low-set ears, and frontal bossing. A few dental features include delayed eruption, hypodontia or anodontia, and conical or peg-shaped teeth. Exocrine sweat glands may be missing in Hypohydrotic Ectodermal Dysplasia or they may be few and poorly developed.[3]

2. Case Report

A 10-year-old male patient with many missing front teeth in reference to the lower arches and poor facial appearance presented to the Department of Paediatric and Preventive Dentistry at KLE VK Institute of Dental Sciences, Belagavi, Karnataka with his mother. There was no history of birth complication during his delivery. His mother gave history of fine hair growth, dryness of mouth and skin, decreased sweating and no tears observed. Additionally, his mother detailed a history of numerous fever attacks during his childhood and indicated that her son was sensitive to heat. According to the boy's family background, he is the third of four children; none of his siblings have any medical conditions. The boy's average weight was around 13.8 kg,

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and he was of average build. The facial features exhibited frontal bossing, a noticeable supraorbital ridge, sunken cheeks, a thick lower lip, thin hair with scanty eyebrows and low-set, overfolding ears on extraoral inspection.(**Figure 1**a) The most remarkable facial features were decreased lower anterior facial height, flat mandibular plane, prominent chin and a resultant concave facial profile.(**Figure 1**b) Intraoral examination revealed presence of sixteen teeth in upper and lower arches– in maxilla: 4 permanent incisors, two first permanent molars and two first primary molars(retained erupting 14 and 24 seen), two deciduous canine and two second primary molars (11, 12, 21, 22, 53, 63, 54, 64, 55, 65, 16, 26), in mandible one permanent lateral incisor, one first premolar and two permanent first molars (36, 46, 32, 85).Maxillary central incisors were widely spaced. The patient exhibited a deep overbite and thin atrophic knife-edge alveolar ridges with loss of vestibular height, especially in the mandibular arch. **(Figure 2**)

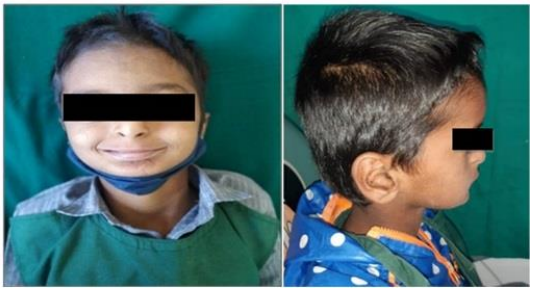

Figure 1: a & **b:** Frontal and Lateral view of the patient

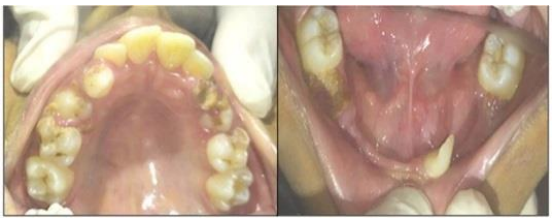

Figure 2: Intra-oral view of the patient showing partial anodontia, conical teeth & poorly developed mandibular ridge

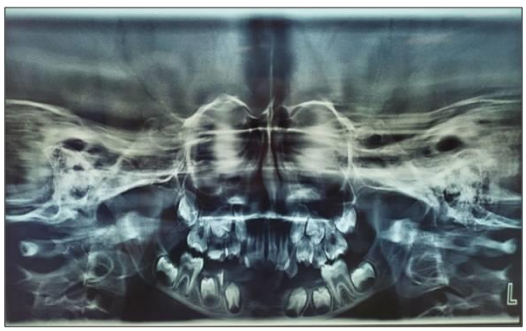

Figure 3: Panoramic radiograph showing congenital absence of lower permanent tooth buds

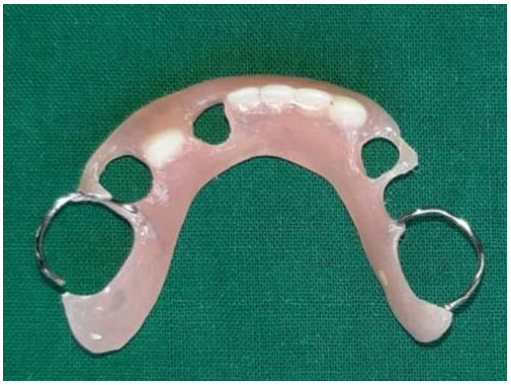

Figure 4: Processed modified acrylic partial denture with windows

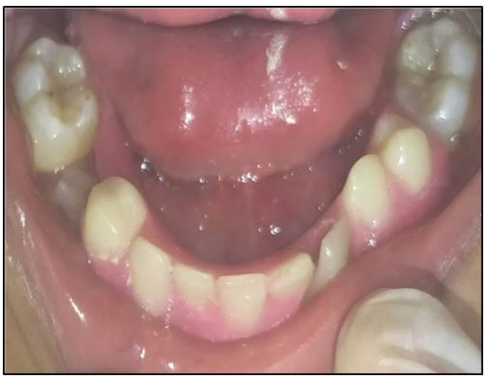

Figure 5: Intraoral view of partial denture in the mouth

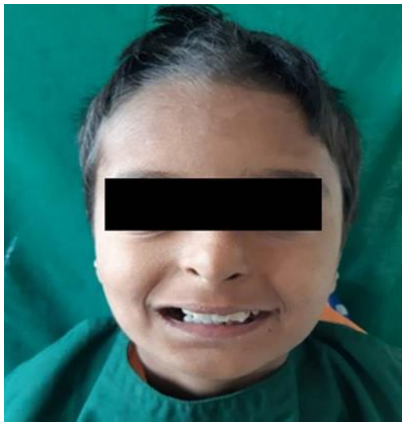

Figure 6: Happy smiling patient

The patient's oral mucosa was slightly dry, and his tongue was enlarged. He had an apparent reduced ability to produce saliva, and the saliva was viscous in nature. Some food debris and plaque accumulation were present, which was attributed to poor oral hygiene exacerbated by reduced cleansing ability of saliva. After routine investigations, the panoramic radiograph confirmed the complete absence of tooth buds in relation 31,41,42,43 and 34 and the extreme underdevelopment of the maxillary and mandibular arches.(**Figure 3**) Patient displayed the classical triad of oligodontia, hypotrichosis and hypohidrosis confirming the diagnosis of Hypohidrotic Ectodermal Dysplasia. The goals of treatment for the dental condition included bone

preservation, the development of normal chewing, speaking, and swallowing mechanisms, establishment of normal facial features and smile, the development of a normal emotional and psychologic profile, and the fitting of a functional prosthesis with adequate retention, stability, and support.

2.1. Oral rehabilitation

The child was scheduled for oral rehabilitation with a removable partial denture in the lower arch. This improves maxillomandibular relationship, phonetics, masticatory efficiency and positive self-image which is prerequisite for normal psychological development of the child. Ectodermal Dysplasia is challenging to handle prosthodontically due to the young age of the patients and atrophic ridge. During his first visit, educated both mother and son about various treatment modalities which is suitable for the patient.After introducing the child to the various instruments, carried out oral prophylaxis and restoration of 16,55 and 65 was done. Further visits extraction of the rootstumps irt 54, 64, 63, 85 and 53 was done. Later, the primary impressions of upper and lower arches were made with alginate. Sharp and thin ridges were covered with a wax spacer. Acrylic custom trays were prepared and border moulded using green stick compound. Final impressions were made using Zinc Oxide Eugenol impression paste.To produce greater muscle tone and lip support, occlusal rims were made and jaw relations were noted with enhanced vertical dimension at occlusion. Acrylic teeth were selected as per the shade, trimmed to the approximate dimensions of the permanent teeth and teeth arrangement was done with bilateral balanced occlusion. Windows were created in the region for erupting 35, 44, 45 and erupted 32 and also to enhance the retention. Try-in was done for retention, aesthetics' and phonetics. Heat cure acrylic resin was used in the processing of removable partial denture.(**Figure 4**) Removable partial denture that had been prepared were finished, polished, and inserted.(**Figure 5**) Areas of discomfort were relieved. Instructions for denture usage, speech and masticatory functions, cleanliness, and maintenance were provided after the procedure. Parents were given instructions on how to care for the denture at home and replace them as growth occurs. Patient was asked to come for the follow up visits.

3. Discussion

A comprehensive approach and understanding of behaviour management are necessary for a child with Ectodermal Dysplasia to be rehabilitated.[4] The degree of anodontia affects how children with Hereditary Ectodermal Dysplasia are treated with prosthetics. Depending on the amount of teeth present, removable/fixed partial dentures and overdentures may be explored for individuals with partial anodontia. Early dental rehabilitation prevents the loss of alveolar bone due to tooth loss and enhances the tone of the perioral muscles and exhibits a lower facial height decrease along with an upward and forward displacement of the chin.[5] A detachable prosthesis option is osseointegrated implants, either an implant or an overdenture secured by one. Implants can become embedded only if they are placed close to adjacent teeth. Because implants will eventually serve as a supported full denture for that patient, improving his quality of life greatly, they should not be positioned next to natural teeth in patients with hypodontia or oligodontia who are still developing.[6] Severely underdeveloped mandible is not a suitable foundation for traditional detachable prosthesis rehabilitation. Another feature that directs the treatment strategy towards an implant supported prosthesis is xerostomia. Even if implants are inserted as early as child turns to 7 year old, but there are no findings in the literature indicating that displacement of implants inserted in the anterior edentulous jaw is a concern. Although a slight lingual rotation is anticipated, it won't jeopardise the rehabilitation of a prosthetic device in the future.[3] Choosing angulated abutments might assist solve this issue. All other oral sites are thought to be locations where implants might be moved to an unrestorable extent due to growth trends. Implants shouldn't be positioned close to natural teeth in hypodontia and oligodontia situations since the patient will eventually develop ankylosed teeth and the implants will be buried in the bone. The patient who is the subject of this case had oligodontia, as there is no alveolar bone and no bone apposition is taking place on the crests of their ridges, patient is not at danger of developing ankylosed teeth. Reduced bone development is another symptom of Ectodermal Dysplasia.[4] The rehabilitation of a prosthetic device would be further hampered by increased resorption of the supporting hard tissues. Alveolar bone could be preserved with the use of implants. However, it should be possible to preserve alveolar bone without sacrificing development. In contrast to a maxillary fixed prosthesis, which must be divided in the midline, the mandibular implant-supported full denture does not need a split design at the midline. However, in people with anodontia, the anterior breadth of the mandible stabilises early; as a result, the jaw's development pattern won't be interfered with.[7] Additionally, the technical aspects of implant placement may be difficult for young patients since they often have a small maximum oral aperture. Additionally, because young patients typically have a limited maximal oral aperture, the technical elements of implant placement may be challenging.[2] The shortest height for a pilot drill would be roughly 30 mm, making this element hard. Additionally, unless significant bone grafting procedures are employed, the quantity and quality of bone only support implant placement in the anterior mandible. In addition to the hazards associated with implant surgery, the implant-supported full denture design also adds a higher level of upkeep.[6] A competitive advantage for the developing patient is provided by the superior biomechanical features of this therapy, which prevent the use of a detachable design that promotes adult behaviours and pictures. It is crucial for the young patient to feel as normal as possible, and the permanent bridge is the only way to do this. In some circumstances, more adjustable implant-retained overdenture designs may be

necessary.[5] The choice of therapy must be made with the full participation of the parents, who must also agree to bear the added time and expense of the treatment. Overdentures with implants offer the benefit of stability and retention, but in young children, dental implants implanted during the development years may get buried. Because there is a chance that the implants will migrate as a result of jaw growth, this procedure should wait until the patient is 13 years old.[8] Early dentures in the preschool years aid with the child's better adaption. The denture will need to be relined and replaced as the kid develops, in primary, mixed, and permanent dentition, until the growth is finished. Due to the lack of teeth and the risk of causing jaw development interference if fixed prosthesis cross the midline, fixed prostheses are seldom used entirely in the treatment of ED.[7] Mandibular removable partial dentures were developed in this case taking into account the patient's age, clinical and radiological results, and financial situation. The patient was obviously shy at his initial visit and was not taking part in any conversations. We made an effort to include him in queries on his broader interests, such as school, games, or friends. It was noted that the patient was an introvert who was experiencing psychological stress as a result of his outward look and speech impairment. Dentures aid in maintaining a satisfying daily diet even if they are poor substitutes for a healthy dentition. Additionally, the lack of occlusal stops (or dentures) results in a decrease in the height of the bottom part of the face and a propensity for Class III malocclusion because of the mandible's anterorotation.[9] Dentures aid with appropriate chin posture in addition to raising the lower face height. Young people frequently require periodic recalls because of their continued growth and development, which necessitates alterations to their prostheses.[10] Patients' personality as a whole significantly changed as a result of the partial denture. At the subsequent recalls, he was unquestionably happier and more engaging. He was also completely at ease wearing a denture and reacted favourably to the oral hygiene instructions. Patient and his mother were delighted with the advancements in speech, function and appearance.(**Figure 6**)

4. Conclusion

Early prosthetic intervention in Ectodermal Dysplasia instances enhances the child's physical and psychological development. In an active quest for a diagnosis, seek for signs and symptoms from ectodermal organs other than the teeth in individuals with dental agenesis, notably oligodontia. Young children with Ectodermal Dysplasia who have partial or complete anodontia experience aesthetic problems in addition to communication and feeding problems. To promote healthy psychological and physiological growth and to enhance the performance of the stomatognathic system, early prosthetic therapy should be given. It is advised to start the patient's rehabilitation with dentures when they are still young and then switch to fixed prosthesis or implants when their growth ceases.

5. Source of Funding

None.

6. Conflict of Interest

None.

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