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Prosthetic Treatment of Hypohidrotic Ectodermal Dysplasia With Partial Anodontia: A Case Report

Dr.Anju Bansal Mds[1] Dr.Sovendu Jha Mds[2] Dr.Chitrita Gupta Mukherjee Mds, Mfdsrcs (Eng)[3] Dr.Madhushree Mukhopadhyay Bds[4]

[1] *Reader, Department Of Pedodontics & Preventive Dentistry Buddha Institute Of Dental Sciences & Hospital, Patna Bihar, India*
[2]*Senior Lecturer, Department Of Orthodontics & Dentofacial Orthopedics Buddha Institute Of Dental Sciences & Hospital, Patna Bihar, India*
[3]*Professor & Head, Department Of Pedodontics & Preventive Dentistry Buddha Institute Of Dental Sciences & Hospital, Patna Bihar, India*
[4]*Post-Graduate Student, Department Of Pedodontics & Preventive Dentistry Buddha Institute Of Dental Sciences & Hospital, Patna Bihar, India*

Abstract: *The hereditary condition known as ectodermal dysplasia is characterized by the absence or defect of 2 or more ectodermally derived structures. A case of 8 year old child with hypohidrotic ectodermal dysplasia with partial anodontia is presented here. Common dental, oral, and physical conditions were taken into consideration. Clinical management consisted of removable complete denture to improve psychological development and to promote better functioning of the stomatognathic system*
Keywords: *child, partial anodontia, hypohidrotic ectodermal dysplasia, prosthetic treatment*

I. Introduction

The term ectodermal dysplasia indicates a heterogeneous group of hereditary diseases involving the epidermis and its appendages. Freire-Maia-Pinheiro have described 154 patterns of ectodermal dysplasia, divided them into 11 subgroups, and then classified them according to the involved structures (the hair, the teeth, some or all the sweat glands). **[1]** When at least 2 types of abnormal ectodermal features occur (for example, malformed teeth and extremely sparse hair), the person is identified as being affected by an ED syndrome.**[2]**
ED syndrome affects both males and females of all races and ethnic groups. Prevalence is estimated to be 7 cases in 10.000 births. The mortality rate is 30% in infancy or early childhood because of intermittent hyperpyrexia**[3]**.

Two different forms are clinically distinguished: an autosomal inherited form (Clouston syndrome) and a hypohidrotic form (Christ-Siemens syndrome).**[4]** Hypohidrotic ectodermal dysplasia (HED) is usually an X - linked recessive genetic trait, in which case the disorder is fully expressed in men only.**[5]** However, women who carry a single copy of the disease gene, may exhibit some of the symptoms and findings associated with the disorder. The etiology of this disease is unknown, nevertheless genetic studies showed ectodermal dysplasia is due to gene 'EDA'(Ectodermal Dysplasias Anhydrotic). Patients with hypohidrotic ED generally have skin which is usually dry, scaly and easily irritated as a result of poorly developed or absent oil glands. Sweat glands can be absent, few in number, or non-functioning, which may result in a high body temperature. Scalp hair may be absent, sparse, very fine pigmented, or abnormal in texture. Eyebrows, eye lashes, and other body hair may also be sparse or absent.**[2]**

Orofacial characteristics include anodontia or hypodontia, hypoplastic conical teeth, underdevelopment of the alveolar ridges, frontal bossing, a depressed nasal bridge, protuberant lips, and hypotrichosis. Patients may present with a marked mandibular protrusion on closure and deep vertical overlap, giving old age appearence.**[5]** Depending on the severity of the condition, various prosthodontic treatments are available to improve appearance, mastication, and speech. Prosthetic treatment is of great value to these patients, from the functional standpoint as well as for psychological and psychosocial reasons.

The present report describes the prosthetic management of a young child manifesting HED with partial anodontia and shows the importance of prosthetic therapy in short term welfare of the patient.

II. Case report

An 8-year old boy presented with a complaint of missing teeth to the Department of Pedodontics and Preventive Dentistry, Sardar Patel Postgraduate Institute of Dental & Medical Sciences, Lucknow. He was accompanied by his mother and father, who gave a history of the boy's missing teeth since infancy, except for two teeth in the upper jaw, which erupted when the boy was 3-years old. They also mentioned that he was

intolerant to heat and takes frequent dips in summer to keep cool. There was no family history of missing teeth except his sibling, a 3 years old brother. The boys were moderately built and poorly nourished. Their skin was dry and the body hair was scanty. During the extraoral examination a facial physiognomy typical of HED were observed, exhibiting sparse scalp hair, missing eyelashes and eyebrows, prominent forehead and ears, saddle nose, and protuberant and everted lips. The skin was soft, thin, and dry with linear wrinkles and a hyperpigmentation around the eyes and the mouth. The nails were normal. A diminished lower facial height contributed to a senile facial expression (Figures 1 and 2).

Intraoral examination revealed a relatively dry mucosa. Mandibular arch was completely edentulous with poorly developed alveolus and the maxillary arch had two standing deciduous central incisors (mobile) in elder brother (Figure 3). Radiographic investigations (OPG and lateral cephalogram) showed malformed incisors impacted in the maxillary arch (Figure 4). Since hypohidrosis, hypotrichosis, and hypodontia were very evident on physical examination, the boys were diagnosed with Hypohidrotic Ectodermal Dysplasia (HED) with partial anodontia.

To improve the child's appearance, mastication, and speech, he was provided with removable complete maxillary and mandibular dentures. Considering the age and uncooperation the treatment plan was postponed for the younger child. In this case, since deciduous teeth were mobile, they were planned for extraction followed by complete removable denture.

Initial impression was made with addition-cured silicone for fabrication of diagnostic casts. Then custom trays were prepared and functional impression was made (Figure 5). On the master casts, acrylic bases with wax rims were made to establish maxilla-mandibular relations, and then they were mounted on an articulator. (Figure 6) The maxilla-mandibular recordings were carefully verified at the try-in stage. (Figure 7) After the final insertion routine, hygiene instructions for the dentures were given to the patient and his parents. Initially, the patient had some difficulty in keeping the mandibular denture in his mouth. To improve and facilitate its adaptation, he was advised to use a denture adhesive paste for the first few days. A week later, he was completely at ease with his new denture. At recall appointments some adjustments were made to eliminate a little interference.

The child began to feel the effect of his appearance, and his parents reported a significant improvement in speech and mastication. Further recalls have taken place every 3 months. The dentures were advised to be replaced after 12 months or after eruption of permanent central incisors.

III. Discussion

Patients with ED present a characteristically thin and underdeveloped residual ridge, covered by thin mucosa. These features, along with the decreased quantity of saliva, are the main problems encountered in treating such cases. [6] There is no definitive time to begin treatment, but Till and Marques [7] recommend that an initial prosthesis could be delivered when the child starts school, so that the child could have a better appearance and have time to adapt to the prosthesis.[8] Premature loss of several primary teeth may also result in speech abnormalities. At the beginning of therapy the patient was quiet and withdrawn. He became communicative and his speech skills improved after the insertion of the complete maxillary and mandibular dentures.

Development of a good psychologic self-image was achieved through the esthetic improvement and transformed the patient into a socially acceptable individual. Periodic recalls of young patients with ED are also important, because prosthesis modification or replacement will be needed as a result of continuing growth and development.[9, 10]

IV. Conclusion

Prosthetic management of children with complete anodontia associated with hypohidrotic ectodermal dysplasia is important because: it provides good esthetics, phonetics, and masticatory comfort and helps the patient develop a positive self-image.

New alternatives for rehabilitation for children with hypohidrotic ectodermal dysplasia, such as the use of implants, must be carefully considered, taking into account the presence of underdeveloped, thin alveolar bones and age.

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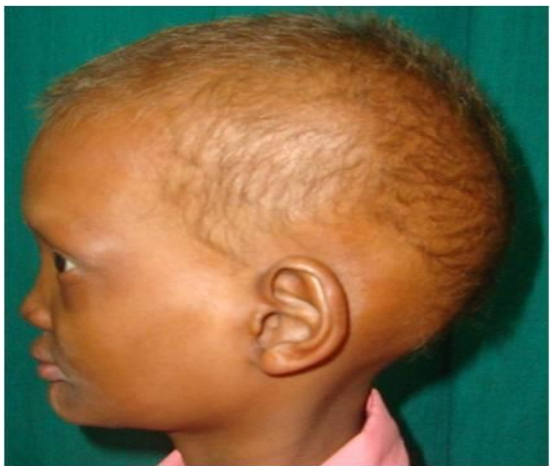
Figure Legends

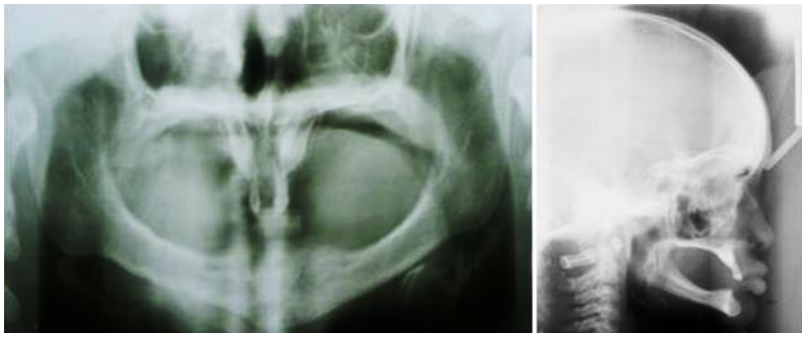
1. Figure 1: Pre-operative frontal profile

2. Figure 2: Pre-operative lateral profile



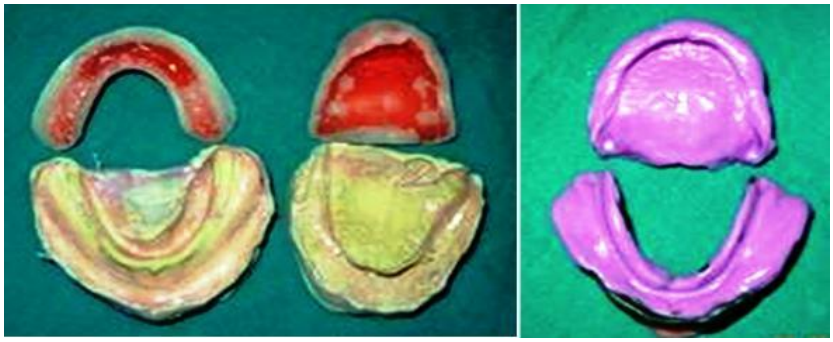
3. Figure 3: Intra-oral photograph

4. Figure 4: Extra-oral radiograph



5. Figure 5: Fabrication of special tray and secondary impression

6.



7. Figure 6: Recording of maxilla-mandibular relationship

8.

9. Figure 7: Try-in & final dentures

10. Figure 8: Post-operative photographs