PROSTHODONTIC REHABILITATION OF ECTODERMAL DYSPLASIA WITH PARTIAL ANODONTIA
Jalaleddin Hamissi, Ebrahim Mosavi Sajad, Hesameddin Hamissi

ABSTRACT
Ectodermal dysplasia usually manifests as X-linked recessive hypohidrotic ectodermal dysplasia (HED) and has a full expression in males, while females show little to no signs of the disorder. This case report discusses the management of a young boy with hypohidrotic ectodermal dysplasia that was treated with removable prostheses.

Keywords: Hypohidrotic ectodermal dysplasia; Partial anodontia; Prosthetic therapy

Introduction
Ectodermal dysplasias (ED) are congenital disorders that are described as large and heterogeneous and were initially described by Thurnam in 1848 and then Darwin in 1875. ED was assigned to the X chromosome in 1921 by Thadani, who later reported that carrier females could manifest signs of the condition. The occurrences are relatively rare (1/100000 births) and it is characterized by the abnormal development of the ectodermal derivatives. The triad of nail dystrophy (onychodysplasia), alopecia or hypotrichosis (scanty, fine light hair on the scalp and eyebrows), and palmoplantar hyperkeratosis is usually accompanied by a lack of sweat glands (hypohidrosis) and a partial or complete absence of primary and/or permanent dentition. Oral health findings include total or partial anodontia affecting both the primary and the permanent dentitions with conical or peg shaped teeth and underdeveloped alveolar ridges resulting in reduced vertical dimension, and a senile facial appearance along with protuberant dry lips. This paper describes the prosthetic rehabilitation of a child with ED presenting severely resorbed alveolar ridges.

Case Report
A male patient with 7 years of age was reported to the outpatient clinic with his parents. The history reveals that missing teeth from infancy except the two deciduous teeth in the upper jaw. He had a history of intermittent fever throughout infancy and childhood. The family history of missing teeth was noncontributory. The physical appearance was of moderately built and well nourished. On examination the skin was extremely dry and he hardly had hypotrichosis. His facial appearance showed periorbital pigmentation, frontal bossing, a depressed nasal bridge, protuberant lips, low-set ears, and scanty scalp hair similar to the description of ED given by various authors. Nails were not very brittle. Intra-oral examination shows dry mucosa.

The mandibular arch was fully edentulous with poorly developed alveolus and the maxillary arch had two standing permanent molars (Figure 1, 2). Since hypohidrosis, hypotrichosis, and hypodontia were evident in the physical examination, the boy was diagnosed with Hypohidrotic Ectodermal Dysplasia (HED). The treatment plan includes removable partial denture in the maxilla and complete denture in the mandible. Using alginate preliminary impressions were made and special trays were fabricated using autopolymerizing acrylic resin. Secondary impressions were made using the Zinc Oxide Eugenol Impression (ZnOE) paste for the mandible arch and light viscosity addition silicone for the maxillary arch (Figure 3, 4). Acrylic dentures were processed through compression molding and the polished dentures were inserted (Figure 5, 6).

The prosthodontic management of children suffering from HED depends on the degree of anodontia/hypodontia. In complete anodontia, the treatment would comprise of complete dentures, either conventional or implant supported ones. In patients with partial anodontia, removable/fixed partial dentures and over dentures may be considered. In the present case, a removable partial denture in the maxillary arch and a conventional complete denture in the mandible arch were selected. Preliminary impressions were made using alginate. Special trays were fabricated using autocopolymerizing acrylic resin after applying spacers to relieve the mid-palatine raphe, incisive papilla, and the standing molars in the maxillary arch and the crest of the ridge in the mandible arches.

Border molding was done using a green stick compound. Secondary impressions were made using the Zinc Oxide Eugenol Impression (ZnOE) paste for the mandible arch and light viscosity addition silicone for the maxillary arch (Figure 3, 4). Record bases were made and jaw relations registered. The selection of teeth for children and adolescents requires more attention as the commercially available moulds do not satisfy esthetic requirements in most cases; obviously because these moulds mimic adult dentitions. Instructions regarding the insertion and removal of dentures was given.

Patient was instructed to be under liquid/semi-solid diet until he was completely at ease while eating with the dentures and emphasize was given on the importance of the existing deciduous maxillary molars and its maintenance. He was recalled 24 hours after denture insertion for his first post insertion checkup. Later, he was kept on monthly recall visits. He reported inconvenience in mastication for more than two weeks even with a semisolid diet. But the esthetic satisfaction was so immense that he wore the dentures and willingly adapted to masticating and speaking with them.
Discussion

The main objective of the prosthodontic rehabilitation in any ED child patient is to improve the sagittal and vertical skeletal relationship during craniofacial growth and development and this improves esthetics, speech, and masticatory efficiency.15 Removable complete or partial dentures are the preferred treatment and implants are largely indicated in cases with complete anodontia. Children with severe midface hypoplasia require early growth modification and implants. In the absence of occlusal stops (or dentures), the anterorotation of the mandible causes an upward and forward displacement of the chin, with a reduction in the height of the lower-third of the face; a tendency to C1 III malocclusion and dentures help to position the chin.15 Frequent followup are indicated in young patients due to the continuing growth and development of jaws which cause adjustments in the prostheses.16 Early rehabilitation of children with ED improve the social and psychological well being of the child. Restoration or prosthesis should offer dentition that match the age of the patient. It helps normalize the function of masticatory and perioral muscles and as a result, the growth pattern of basal bones.3

Conclusion

In conclusion early prosthetic therapy for children with ED provides a unique opportunity for a cooperative effort between the pedodontist, periodontist and the prosthodontist. Prosthodontic treatment may begin at an early age of 3-4 years as a condition for growth and development of orofacial structures.

Authors Affiliations

1. Jalaleddin Hamissi DMD MDS(Perio) PG Dip Perio(UK), Associate Professor, Department of Periodontics and Preventive Dentistry, Faculty of Dentistry, Qazvin University of Medical Sciences, Qazvin, Iran, 2. Ebrahim Mosavi Sajad DMD MDS, Assistant Professor, Department of Prosthetic Dentistry, Faculty of Dentistry, Qazvin University of Medical sciences, Qazvin, Iran, 3. Hesameddin Hamissi, Dental Student, Faculty of Dentistry, Qazvin University of Medical Sciences, Qazvin, Iran.

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Address for Correspondence
Jalaleddin Hamissi DMD, MDS (Perio), PG Dip Perio(UK), Associate Professor, Department of Periodontics and Preventive Dentistry, Faculty of Dentistry, Qazvin University of Medical Sciences, Qazvin, Iran.
E-mail: jhamissi@qums.ac.ir

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