

# Prosthetic rehabilitation for a patient with hypohidrotic ectodermal dysplasia: a clinical case

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## Abstract

Hypohidrotic ectodermal dysplasia (HED) is a hereditary syndrome, characterized by a classic triad of hypotrichosis, hypodontia and hypohidrosis. The case of an 8-year-old girl with HED presenting oligodontia and marked resorption of maxillary and mandibular alveolar ridges is reported. A specially designed aluminum wire tray was used for taking impressions of both dental arches. Prosthetic rehabilitation included a removable maxillary overdenture and a mandibular conventional acrylic resin denture made tissue-compatible by means of a soft denture liner attached to denture base. The proposed treatment plan provided improved esthetics, function, and quality of life.

**Keywords:** ectodermal dysplasia, oligodontia, soft liner, mucosa supported dentures.

## Introduction

Ectodermal dysplasia (ED) is a group of hereditary diseases with abnormal development of two or more structures of ectodermal origin<sup>1</sup>. These disorders are relatively rare with an incidence of 1 in 10,000 to 1 in 100,000 live births<sup>2</sup>. ED can present numerous clinical alterations and up to 154 different types and 11 subgroups have been observed depending on the involvement of the hair, teeth, nails or sweat glands<sup>3</sup>. ED can be hypohidrotic or hidrotic depending on the sweat gland function<sup>2</sup>.

Hypohidrotic ectodermal dysplasia (HED) is characterized by dry and scaly skin due to absence of sweat and sebaceous glands and the affected child will develop intolerance to heat due to absence or reduced sweat glands causing elevated body temperature<sup>1</sup>. Skeletal manifestations in individuals affected by HED include decreased lower facial height and depth, small cranial base width and calvarial height, prominent forehead and depressed nasal bridge<sup>4</sup>.

Oral findings include total or partial anodontia affecting both the primary and the permanent dentitions. The teeth are usually conical or peg shaped. Alveolar ridges are underdeveloped resulting in reduced vertical dimension, thereby giving a senile facial appearance along with protuberant dry lips<sup>1,4-5</sup>. This paper describes the prosthetic rehabilitation of a child with HED presenting severely resorbed alveolar ridges by using specially designed aluminum trays and tissue-supported dentures.

## Case Report

An 8 year-old girl was referred to the Department of Pediatric Dentistry accompanied by her mother with the complaint of difficulty in eating due to

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several missing teeth since infancy. She is the elder of the two siblings with a younger brother, and was born out of consanguineous marriage. Past medical history revealed that the girl had several frequent episodes of fever throughout childhood along with frequent ear and nose infections. The pedigree of the patient could not be researched due to insufficient data from the parents.

Detailed dialogue history with mother revealed that the girl was intolerant to heat and takes frequent dips in summer to keep cool, and that she had difficulty in speech and social interaction. The girl appeared poorly nourished. Extraoral features included frontal bossing, depressed nasal bridge, sunken cheeks, protuberant lips, periorbital pigmentation, dry skin, scanty scalp hair and decreased lower facial height, which suggested typical facial physiognomy of hypohidrotic ectodermal dysplasia (Figure 1).



Fig. 1: Frontal and lateral initial views of the patients face.

Intraoral examination revealed a relatively dry mucosa, bone atrophy of the alveolar ridges, on both mandible and the maxilla. Mandibular arch was completely edentulous and the maxillary arch presented two widely spaced conoid deciduous incisors (Figure 2).

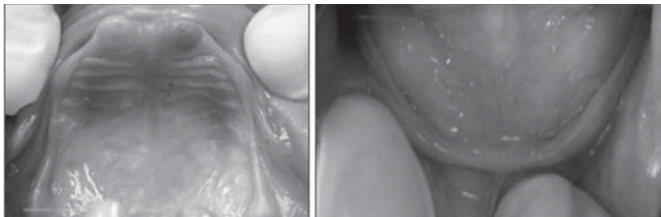


Fig. 2: Maxillary arch with two wide spaced conoid deciduous incisors and completely edentulous mandibular arch.

Radiographic investigations included digital panoramic radiograph, which displayed numerous agenesia of primary and permanent teeth, conical shaped maxillary deciduous incisors and unerupted malformed permanent central incisors. In the mandible, all of the anterior and posterior permanent tooth germs were missing (Figure 3). The patient had a total of 26 congenitally missing permanent teeth excluding third molars.

A clinical diagnosis of HED with oligodontia was made and the treatment plan included fabrication of a removable mucosa supported over denture and a conventional acrylic resin denture in the maxillary and mandibular arches respectively.

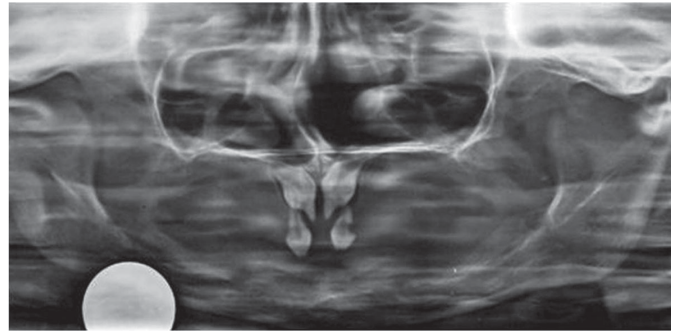


Fig. 3: Digital panoramic radiograph showing multiple agenesia of permanent teeth and malformed unerupted conical maxillary incisors

Impression trays were specially designed and fabricated using an aluminum wire stabilized with green wax (Figure 4) and impressions were made using putty and light viscosity additional silicone, simultaneous mix technique, for both maxillary and the mandibular arches (Figure 5). Master casts were obtained and temporary denture bases were prepared. Jaw relations were recorded and acrylic artificial teeth were modified by trimming down their size to accommodate the jaw size and were then arranged in class I molar relation.

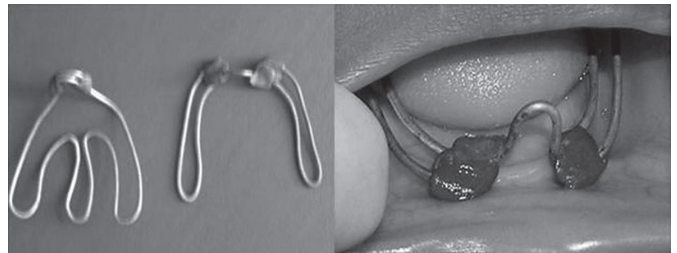


Fig. 4: Maxillary and mandibular custom-made trays made of aluminum wire and tray check for mandibular arch.

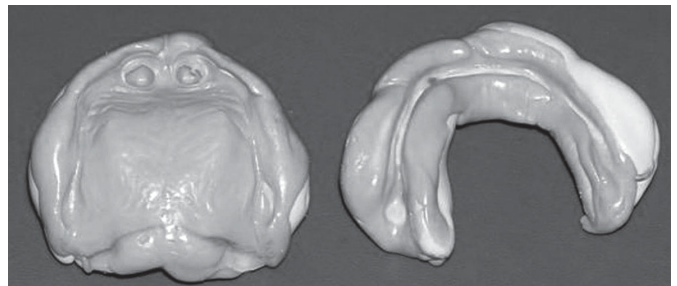


Fig. 5: Master impressions with aluminum trays.

After wax try-in acrylic dentures were fabricated lined with a heat cure soft liner (Molloplast B, Detax GmbH & Co, KG Ettingen, Germany). Insertion and removal of the dentures was taught to the child and post-insertion instructions were given regarding mastication, hygiene and maintenance. Patient was found to be extremely satisfied with her esthetics. She was instructed to speak and read daily for sometime in order to improve phonation. Diet was restricted to semi-solid/liquid, till she was comfortable eating with dentures. During the follow-up visit at the end of 2 months she presented with marked improvement in articulation of speech by means of controlled mandibular movements.

The existing two conoid maxillary anterior teeth were preserved and served as retention for the prosthesis. After a 6- and 12-month follow-up visit, the patient's mother gave a positive feedback regarding peer-group interaction and significant improvement in her phonetics (Figures 6 and 7).

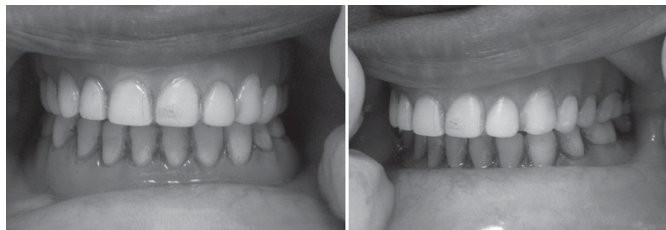


Fig. 6: Frontal and lateral intraoral views of the patient with the dentures at the 12-month follow-up visit.



Fig. 7: Frontal and lateral final extra oral views of the patient's face at the 12-month follow-up visit.

## Discussion

Hypohidrotic type, originally known as HED because of notable reduction of sweat gland function, is clinically characterized by triad of hypotrichosis, hypohidrosis and either hypodontia or anodontia<sup>6-7</sup>. The gene responsible for x-linked recessive trait has been mapped to Xq12-q13.1<sup>8</sup>. Recent findings reported that mutations in the ectodysplasin-A (EDA) gene are responsible for X-linked HED<sup>9</sup> and isolated hypodontia<sup>10</sup>. Van der Hout et al.<sup>11</sup> identified mutations in the ED1 gene in 24 (57%) of 42 unrelated European probands with HED. Genetic counseling of affected patients or parents may be necessary so that they can be informed of the possible recurrent risks and the eventual dental problems that their offspring may encounter.

Majority of these children present multiple ocular, dental, infectious, and dermatologic problems and require multidisciplinary treatment<sup>7</sup>. Various rehabilitation possibilities in patients with ED may include removable partial/complete prostheses, fixed, and /or implant-supported prostheses<sup>12</sup> or a combination of these options.

Treatment should be initiated as soon as possible so that further resorption or atrophy of the alveolar ridges due to partial or complete absence of teeth can be prevented and vertical dimension controlled. Additionally, cosmetic and psychological issues become more important for the patient later in the childhood and adolescence. Therefore early prosthodontic treatment of these patients has been strongly

recommended<sup>13</sup>. The objective of treatment in the present case was not only to reestablish the masticatory function, but also to improve the emotional and social aspect of the child so that the child can lead a normal life-style with positive self esteem.

The removal of the conoid teeth for subsequent placement of a prosthetic appliance would further reduce the alveolar ridge of the patient<sup>6</sup>. Therefore the teeth were not extracted. In the present case, the conical teeth served as an aid in retention of the maxillary prosthesis. The child adapted herself well to the prosthesis with great personal satisfaction.

Severe resorption of the alveolar ridges in ectodermal dysplasia may contribute to instability and discomfort of the conventional acrylic resin dentures. These dentures may frequently irritate residual alveolar ridge tissues, which often are atrophic and minimally resistant to stress<sup>14</sup>. Use of custom-made aluminum wire trays will minimize unbalanced and excessive pressures on an already compromised alveolar ridges, and thereby reduce degenerative changes in bony and soft tissue architecture. As the aluminum wire used in the present case has a thin border, it does not impinge upon muscle attachments, which could produce dislodging forces.

Conventional acrylic resin dentures with a soft liner have proven to be advantageous in patients who has severely atrophic alveolar ridge. The soft liner accommodates ridge irregularities and changes such as excessive resorption, minimal keratinized ridge epithelium and decreased resistance to irritation due to nutritional and physiologic problems<sup>15</sup>. In the present case, the entire acrylic resin base was lined with a soft liner to prevent abuse to the soft tissues and accommodate the jaw growth. These prostheses need to be changed periodically according to the growth and the bone development.

The choice for the treatment approach was based on the child's physical condition and the need of esthetics. The masticatory and phonetic functions were markedly improved, which would otherwise have had a negative effect on the psychosocial development of the patient. Franchi et al.<sup>16</sup> in a serial cephalometric study of patients with ED found that the growth of both maxilla and mandible has been favored by the use of conventional prosthesis.

The use of tissue-supported (soft liner) maxillary overdenture and conventional acrylic resin denture in the mandibular arch in the present case can be considered as a good practical alternative that provided a relatively quick, easy, acceptable and economical solution to the functional and esthetic oral rehabilitation. Moreover, it helps stimulating the alveolar ridges for later treatment with implant-retained dentures, as a more stable alternative.

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