Dental management of hypohidrotic ectodermal dysplasia: A report of two cases

Meenu Mittal, Dhirendra Srivastava[1], Ashok Kumar, Poonam Sharma[2]

Abstract

Ectodermal dysplasia (ED) represents a group of inherited conditions characterized by anomalies in two or more structures of ectodermal origin, which can be presented as problems related to hair, nail, teeth, sweat glands, and sebaceous glands. Based on clinical findings, there are two major types of this disorder: (1) Hypohidrotic/anhidrotic and (2) hidrotic ED. The anhidrotic/hypohidrotic ED (HED) is the more severe form and is associated with more dental defects. This article presents with prosthetic rehabilitation including removable partial and complete denture and implant supported overdenture of two male children of a family presenting with HED.

Keywords: Dental management, hypohidrotic ectodermal dysplasia, implant overdenture

Introduction

Ectodermal dysplasia (ED) represents a group of inherited conditions characterized by anomalies in two or more structures of ectodermal origin, which can be presented as problems related to hair, nail, teeth, sweat glands, and sebaceous glands. EDs are a large group of syndromes, and nearly 200 different conditions have been described under this term.[1]

Based on clinical findings, there are two major types of this disorder: (1) Hypohidrotic/anhidrotic ED in which sweat glands are either absent or significantly reduced in number; the condition shows mainly X-linked recessive inheritance severely affecting males, while females show only minor defects and (2) hidrotic ED in which sweat glands are normal; the condition is inherited as autosomal dominant trait. The dentition and hair are affected similarly in both types, but the hereditary patterns nail and sweat gland manifestations tend to differ.[1]

The anhidrotic/hypohidrotic ED (HED) is the more severe form and is associated with more dental defects.[2] This article presents with the prosthetic rehabilitation of two male children of a family presenting with HED.

Departments of Pediatric Dentistry, and [1]Oral Surgery, [2]Orthodontics, ESIC Dental College, New Delhi, India

Correspondence: Dr. Meenu Mittal,
A 29, Ground Floor, Hauz Khas, New Delhi - 110 016, India.
E-mail: meenu20feb@gmail.com

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Case Reports

Two brothers aged 5 years (Case 1) and 9 years (Case 2) reported to the Department of Pediatric and Preventive Dentistry, ESIC Dental College, New Delhi with the complaint of lack of teeth and inability to eat properly. The general medical and family histories were noncontributory. Neither their sister nor their parents had any missing teeth.

The boys exhibited the classical features of HED-severe ologodontia/anodontia, hypohidrosis, and hypotrichosis. They were intolerable to hot climate with a history of reduced sweating.

Extraoral examination [Figure 1] shows the presence of thinning of scalp hair, frontal bossing, scarce eyebrows and eyelashes, prominent supraorbital ridges, saddle nose, depressed nasal bridge, large ears, protuberant lips with reduced facial height. Mid-face was depressed, and the profile was concave. Perioral, periorbital, and perinasal pigmentation was present. The skin was dry with no nail dystrophy. Hyperkeratosis in soles and palms was seen.

Intraoral examination in Case 1 [Figure 2a], revealed the presence of conical right lateral incisor and both second primary molars in maxillary arch and absence of all the teeth in mandibular arch. In Case 2 [Figure 3a] complete anodontia of both the arches was seen.

Panoramic radiographs of both the children [Figures 4 and 5a] show no other evidence of tooth formation. Clinical diagnosis of HED was made.

Case 1

It was decided to fabricate removable maxillary partial denture and mandibular complete denture. For partial denture irreversible hydrocolloid (alginate) impression was taken. For complete dentures initial impression was taken with alginate on which custom tray was prepared and border

Figure 1: Extraoral views. (a) Case 2, (b) Cases 1 and 2 together, (c) Case 1

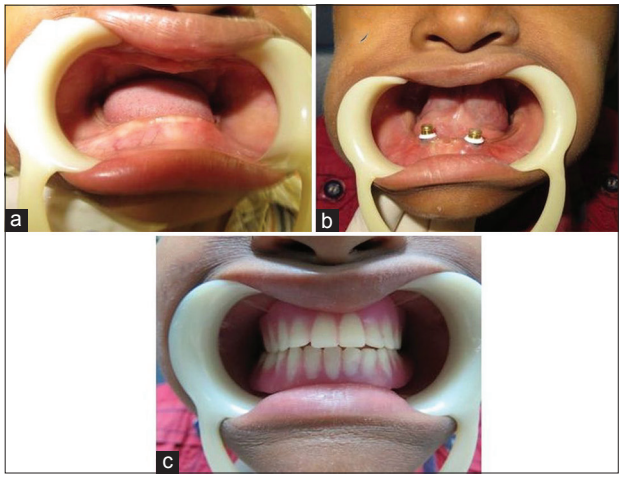

Figure 3: Intraoral views of Case 2. (a) Preoperative showing complete anodontia, (b) implants placed in mandible, (c) with maxillary complete denture and mandibular over denture

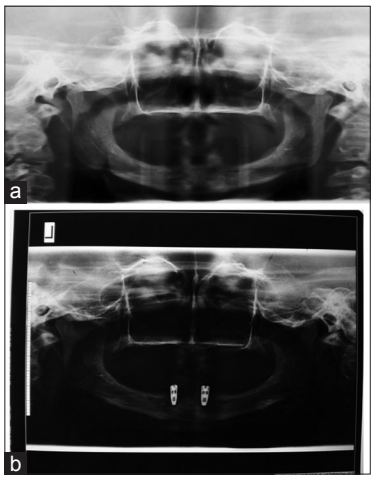

Figure 5: Panoramic radiographs of Case 2. (a) Preoperative, (b) with implants

molding done using heavy body polyvinyl siloxane (Aquasil, Dentsply) material. The final impression was taken with the light body of the rubber base material. Maxillo-mandibular relations were recorded, and teeth arranged as per balanced occlusion. The thermal curing acrylic resin was used for fabrication of dentures.

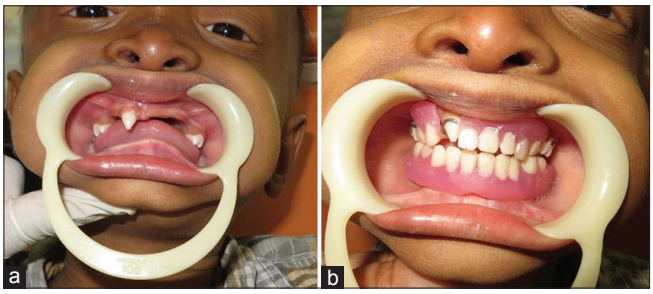

Figure 2: Intraoral view of Case 1. (a) Preoperative, (b) with removable partial dentures

Figure 4: Panoramic radiograph of Case 1

Case 2

Removable maxillary complete denture and mandibular implant supported over denture were planned. Cone beam computerized tomography of mandible was taken [Figure 6]. Two implants (Nobel Biocare Replace, Nobel Biocare Company, Sweden) of size 4.3 mm × 10 mm were placed in the mandibular anterior region [Figures 5b and 3b]. Tissue level implants were placed to prevent abutment surgery on the relatively thicker gingiva. Denture fabrication was done 2 months after the placement of implants. Resilient attachment system (locator abutment 4.3 mm wide with collar size 1 mm) was used for the prosthesis, which allowed some movement of the prosthesis during the function. A technique similar to Case 1 (mandibular denture) was used for fabrication of dentures.

During delivery of the prostheses [Figures 2b and 3c], parents were instructed about the home care of the prosthesis and explained about the need for periodic recall for re-evaluation and remaking of dentures as growth occurred.

Discussion

ED is characterized by the absence or defects of two or more ectodermally derived structures. Most striking dental manifestation is anodontia or hypodontia. Of the two major clinical types of ED, hypohidrotic anhidrotic (HED) type, and hidrotic more prevalent is HED. HED is caused by a mutation

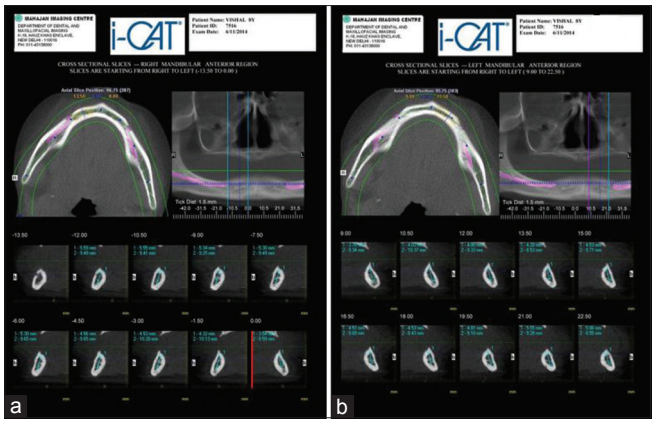

Figure 6: Cone beam computerized tomography right (a) and left, (b) mandible of Case 2

in ectodysplasin A gene located at Xq12-q13.1.[1] HED can be X-linked or autosomal hypohidrotic with males affected more frequently than females.[3] The X-linked recessive has a full expression in males. Female carriers though more than affected men, their clinical identification is difficult, as they have little or no signs of the condition. The classical triad of hypotrichosis, hypodontia/anodontia, hypohidrosis, and almost all other features of HED were found in our patients. Both patients were males and their sister had no symptoms of ED. Hence, clinical diagnosis of HED with X-linked recessive pattern of inheritance was made.

It is imperative to provide oral rehabilitation of ED children to improve both sagittal and vertical skeletal relationship during craniofacial growth and development; to improve esthetics, emotional well-being, stomatognathic efficiency, and temporomandibular joint functions.[3,4] A multidisciplinary approach is required for optimal esthetics and functioning of the stomatognathic system. It is important to commence optimal dental treatment as soon as possible to control the vertical dimension and to avoid possible resorption and atrophy of the alveolar ridge which can be severely affected by total or partial lack of teeth.[3,5]

As such there is no definitive time for starting dental treatment, it is suggested that initial prosthesis should be provided before school age of the patient.[2] Early prosthetic treatment is generally recommended from the age of 5 years though dentures can also be fabricated as early as 3–4 years of age of the child.[6]

Oral rehabilitation treatment plan for ED may include fixed, removable or implant supported prosthesis, used individually or in combination to provide an optimal result.[2] Exclusively fixed prostheses are seldom used as the number of teeth present are minimal and also jaw growth may be interfered in young patients due to rigid connectors if the prosthesis crosses the midline.[2,7] The most common treatment plan is removable prostheses (complete dentures, partial dentures or over dentures), because of the need to modify intraoral prosthesis during the rapid growth period.[2,8] Furthermore, removable rehabilitation methods are easy, affordable, and reversible.[8]

As per the guidelines of National Foundation for ED, removable or nonrigid fixed prostheses including complete dentures or over dentures may be considered for children aged 0–6 years with hypodontia.[7] Thus, for younger sibling (Case 1), who was aged 5 years, removable partial denture for maxillary arch and complete denture for the mandibular arch were constructed. It was planned to follow it with an implant retained mandibular over denture in subsequent years.

For the elder child - Case 2 (aged 9 years), removable maxillary complete denture and implant retained mandibular overdenture with implants in the mandibular anterior region were constructed. Many authors have similarly used mandibular implant retained over dentures.[9,10] There was a general agreement at a Scandinavian consensus conference in Sonkoping, Sweden that implant placement should be postponed until skeletal growth is completed or nearly completed in normal adolescents. However, earlier intervention could be indicated, especially in the mandible, in individuals with oligodontia or anodontia.[11] For children 7–12 years of age, implants can be used for missing mandibular teeth and are recommended for anterior portion of the mandibular arch.[7,11] Prior to the cessation of growth, implant placement in the symphyseal region of the anterior mandible may be performed with caution.[12] In our patient, two implants were placed in the anterior mandible. Implants placed in anterior mandible move with the mandible as growth occurs in the condyles and rami.[13] Provisional implants placement can be an option as these do not osseointegrate and thus do not interfere with growing bones, but additional surgery trauma, cost, and time are the limitations.[14] Neither these implants nor interim implants are recommended for the posterior portion of the mandibular arch or elsewhere in the maxillary arch because substantial growth is still expected.[7,11] Transverse growth of maxilla occurs mostly at the midpalatal suture, hence fixed implants prosthesis crossing the midpalatal suture will result in transversal growth restriction of the maxilla.[11] Thus, implants were not placed in the maxilla in our patient. In case of severe maxillary bone atrophy, transzygomatic implantation technique can be considered once growth in the maxilla is complete.[15] Many cases are reported in literature placing implants in children as young as 3, 4, 5 years;[13,16,17] but in addition to psychological effects, particularly in young children, implant surgery is accompanied by higher risk of failure compared to that of more conservative prosthetic treatment.[6] Hence, the use of implants in young children should be considered carefully.[6]

Even young children can cooperate for the denture making process although complete denture fabrication requires multiple patient appointments and good cooperation. The desire to be like others who have teeth can be a motivator

for cooperation in even the young child. Regular adjustments are required in removable partial, or complete dentures and dentures are to be replaced when the vertical dimension of occlusion is decreased or abnormal mandibular posture is detected due to growth.[6] Retention and stability for the prosthesis are also difficult to obtain.

Conclusions

Management of oral manifestations associated with ED is challenging and satisfying and require multidisciplinary involvement. Implant supported over denture can be a functional and esthetic alternative in young children. Periodic recall for prosthetic modification and adjustment is necessary due to continuing growth and development. The interim prosthesis should be replaced by more definitive prosthesis once skeletal growth is complete.

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