

# Prosthodontic Management of a Child with Ectodermal Dysplasia: A Case Report

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**Abstract** Ectodermal dysplasia is a rare congenital disease that affects several ectodermal structures. Children with ectodermal dysplasia may have various manifestation of the disease that differ in severity and may or may not involve teeth, skin, hair, nails, sweat glands and sebaceous glands. The most common form of the ectodermal dysplasia syndrome is hypohidrotic ectodermal dysplasia and is usually inherited as an X-linked recessive trait. Female carriers may have a variable degree of clinical manifestations. This case report discusses the management of a 5-year-old girl with ectodermal dysplasia. Clinical management consisted of fabricating upper and lower dentures to help in psychosocial development and to restore the vertical dimension, esthetics and functioning of the stomatognathic system.

**Keywords** Ectodermal dysplasia · Christ Siemens syndrome · Hypohidrosis · Hypotrichosis · Hypodontia

## Introduction

Ectodermal dysplasia is a hereditary disorder associated with dysplasia of tissues of ectodermal origin primarily nail, teeth, hair and skin and occasionally dysplasia of mesoderm derived tissues. As defined by Freire-Maia the nosologic group of Ectodermal dysplasia is any syndrome that exhibits at least two of the following features, that is, abnormal hair (trichodysplasia), abnormal dentition, abnormal nails (onchodysplasia) and abnormal or missing sweat glands (dyshidrosis) [1]. More than 150 different variants of Ectodermal dysplasia have been described [2].

Hidrotic and hypohidrotic are the two forms of Ectodermal dysplasia. In both types teeth and hair are similarly affected but manifestations in nails and sweat glands and the hereditary pattern tend to differ [3]. The X linked hypohidrotic form or Christ Siemens syndrome is characterized by clinical triad of hypohidrosis, hypotrichosis and hypodontia. Hidrotic form is inherited as an autosomal dominant trait and affects teeth, hair and nails but usually spares the sweat glands. Prosthodontic rehabilitation is of great importance to the patient with Ectodermal dysplasia for functional, physiologic and psychosocial reasons [1].

## Case Report

A 5-year-old girl child reported to dental clinic, with reference from her pediatrician for having a problem in mixing with friends and moving around socially (with suggestive signs of clinical depression), also difficulty in eating food due to absence of teeth. The parents wanted a prosthesis which could help the child in eating and improve her social acceptance. The child was diagnosed of Anhidrotic Ectodermal Dysplasia at the age of two. Family

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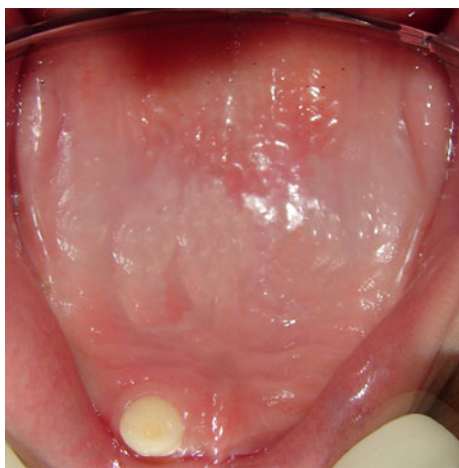
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**Fig. 1** Photograph showing the frontal view of the face with periorbital pigmentation and scarce eyebrows

history of Ectodermal dysplasia was negative. Child was moderately built with hypotrichosis, scarce eyebrows and scarce eyelashes, dry anhidrotic skin, depressed nasal bridge, thick lips, dark pigmented skin around periorbital area and nose, facial height was reduced (Fig. 1). The child was very shy and uncooperative. The parents reported that the child doesn't sweat and her lips and tongue remain dry in all climates. There was however no effect on the nails. Intraoral examination revealed presence of partially erupted two peg shaped teeth in the region of 53 and 83 (Figs. 2, 3, 4). The edentulous ridge was atrophic with decreased height. The palate was shallow, oral mucosa was normal and dry due to less saliva, the tongue was relatively



**Fig. 2** Image of atrophied maxillary arch with 53



**Fig. 3** Image of thin and atrophied mandibular arch with 83



**Fig. 4** Intra-oral photographs showing maxillary and mandibular arches

large. OPG showed presence of an impacted tooth in relation to 12 region and erupted 53, 83 (Fig. 5).

### Prosthodontic Management

A preliminary impression was made using irreversible hydro An interim overdenture (maxillary and mandibular) was decided to be made initially without any modification of the existing two teeth in order to gain the child's



**Fig. 5** Digital ortho pantomograph showing maxillary and mandibular ridges

confidence as it was difficult to get the child to cooperate for the clinical procedures—colloid (Zelgan 2002, Dentsply) impression material. A special tray with uniform 2 mm full arch wax spacer covering the natural teeth was prepared. Peripheral border seal was established and secondary impressions were made with non Eugenol Zinc Oxide impression paste. Master casts were made and occlusal rims with temporary denture base were fabricated. Jaw relations were done by manually guiding mandible into centric. Three teeth sets (small size, acrylic teeth of Heraeus–Kulzer) were used. Lower teeth set of size large set was trimmed to shape like maxillary anteriors. The lower anteriors were trimmed similarly from small size lower arch teeth set. Care was taken to shape the trimmed teeth as per the age, sex and size of the patient. Trial was done and dentures were fabricated using heat polymerizing P.M.M.A. (Trevalon-Hi powder and liquid, Dentsply).

It was not possible to give long clinical sittings, as the child was not very cooperative all the time. The impression technique and jaw relation were decided to be improved subsequent to the fabrication of the denture by relining with soft permanent reliner (rather than continue with clinical sittings and lose the cooperation of the child). Therefore to improve the fit of the dentures tissue surface, the maxillary and mandibular dentures were relined with tissue conditioner material (Soft-Liner, GC Corporation). The patient was recalled after 2 days and the dentures were relined with heat cure P.M.M.A (Trevalon powder and liquid, Dentsply). It was noted that the child adjusted to the denture very well. There was improved speech and a marked improvement in the social activities of the patient with the provisional denture. The child was recalled after every 3 months.

After 6 months the dentures especially the mandibular denture was found to be becoming ill fitting. In the 9th month the patient was unable to wear the lower denture and had stopped wearing the denture during eating. The clinical evaluation showed that the lower tooth (83) had erupted and therefore the lower denture was not being able to seat on the ridge. There was also an increase in the size of both the jaws.

It was decided to fabricate a new removable partial denture with clasp in relation to 83 for the lower arch, and a maxillary overdenture (as made previously) for the upper arch (Fig. 6). The upper and lower prosthesis were fabricated in same conventional manner as discussed earlier, except medium phase. Polyether impression material was used to make the final impressions.

## Discussion

The treatment for a patient with ectodermal dysplasia varies and generally depends on child's age, dental



**Fig. 6** Intra-oral photographs showing maxillary and mandibular dentures in occlusion

agenesis, degree of malformation of teeth, the growth and development of the stomatognathic system of the patient and patient's motivation. According to Nowak [1], treating the pediatric patient with ectodermal dysplasia requires the clinician to be knowledgeable in growth and development, behavioral management, techniques in the fabrication of prosthesis, the ability to motivate the patient and parent in the use of the prosthesis, and the long term follow-up for the modification and/or replacement of the prosthesis.

Prosthodontic treatment for children with ectodermal dysplasia includes removable partial denture or complete denture, over denture and implants. These approaches may be used either individually or in combination to provide optimal results.

Complete denture prosthesis given to patient alters the alveolar height, provides a better musculocutaneous profile and brings about a significant improvement in mastication, esthetics, phonetic function and psychological support. The usual treatment for ectodermal dysplasia focuses on a series of complete or partial denture during the years when growth of the dentofacial region is taking place and definite rehabilitation following completion of jaw growth.

Early prosthetic treatment is generally recommended from the age of 5 years and dentures can be fabricated as early as 3–4 years of age for cooperative children [4]. Till and Marques [1] recommended that an initial prosthesis should be delivered before the child begins school so that the child has a normal appearance and time to adapt to the prosthesis. This early restoration of facial appearance is essential for normal psychological development.

The problems associated with early placement of complete denture are mainly associated with periodic adjustment due to growth changes and difficulties in achieving good retention and stability [5]. Difficulties in achieving adequate resistance to lateral and anteroposterior displacement of the denture in hypohidrotic ectodermal dysplasia patient are due to dryness of oral mucosa and underdevelopment of maxillary tuberosities and alveolar ridges [6].



**Fig. 7** Photographs showing esthetics of the denture

When teeth are present in the mouth, overdentures are the most desirable treatment option [1, 3]. Overdenture has an added advantage, that they preserve the alveolar bone. As a result of continuing growth and development, periodic prosthesis modification or replacement is needed.

Fixed Prosthodontic treatment is seldom used because of decreased number of abutment, and moreover the patient is too young. Use of FPD's with rigid connectors should be avoided as it may interfere with jaw growth. In young patients, individual crown restoration and direct composite restorations have been used in combination with removable partial denture [1].

For adult patients with ectodermal dysplasia, dental implants are the treatment of choice because growth has stabilized and implants can be used to support, retain and stabilize the prosthesis [1]. For using implants in young patients, the timing of treatment is of utmost importance to avoid possible complications that may result from jaw growth.

In this case as the child was 5 years old when first reported to the clinic, implants could not be considered as the treatment of choice. It was decided to fabricate an overlay denture. After 6 months it was observed that eruption of 83 and the increase in the jaw size lead to the loss of retention in mandibular denture, while only there

was increase in jaw size in maxilla, and marginal loss in retention and stability of the upper denture, the child had no complaint in the maxillary denture. The treatment not only improved the patient's functional and esthetic status (Fig. 7), but also improved the psychological health and social life—so important for the complete development of the child.

## Conclusion

The study discusses the management of a child with Anhidrotic Ectodermal Dysplasia with only two teeth present and atrophic edentulous ridges. The basis philosophy is to restore the esthetic and function of the child at present time, till the complete growth of the jaw has occurred. The final treatment will be implant supported prosthesis with bone augmentation as and when necessary. This is the first part of the longitudinal studies which are to be undertaken for this case.

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