

Hypohidrotic Ectodermal Dysplasia: A Case Report

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Abstract

Ectodermal dysplasia is a hereditary disorder characterized by developmental dystrophies of ectodermal derivatives. It is characterized by triad of signs comprising sparse hair, abnormal or missing teeth and inability to sweat. The case of a six year old child with hypohidrotic ectodermal dysplasia and complete anodontia of both primary and permanent dentition is presented. Owing to the need for treatment at an early age for the anodontia, the prosthodontic management of such a young child can be difficult. Complete dentures were provided to encourage normal psychological development and to improve the function of the stomatognathic system. (*International Dental and Medical Disorders December 2008; 1: 11-14*)

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Introduction

Ectodermal dysplasia is the term used to describe a group of rare, inherited disorders characterized by dysplasia of tissues of ectodermal origin-primarily, nail, teeth and skin, and occasionally, dysplasia of mesodermally derived tissues¹. The condition is thought to occur in approximately 1 of 100,000 live births^{1,2}. Clinically, ectodermal dysplasia may be divided into two broad categories: the Hypohidrotic form (x-linked recessive) and the Hydrotic form (Autosomal inherited).

Hypohidrotic Ectodermal dysplasia, also termed as Christ-Siemens Tourine syndrome^{3,4} is more common and is characterized by a triad of signs comprising sparse hair (hypotrichosis), abnormal or missing teeth (hypodontia or anodontia), and an inability to sweat because of the lack of sweat glands (anhidrosis or hypohidrosis). Hypodontia, in turn, causes reduced alveolar bone growth and lack of development of the alveolar ridges which often appear clinically to be extremely narrow and concave lingually. Teeth, if present, are often

conical in shape, malformed and widely spaced. Other symptoms include, 'saddle' nose, prominent lips, linear wrinkles and hyperpigmentation around the eyes, atopy, mild facial dysmorphic features and increased susceptibility to respiratory infections. The phenotype is seen in its full form only in affected males^{5,6}.

This case report essentially emphasizes the prosthodontic management of appearance and functionality of treatment in the form of a complete denture provided to the patient.

CASE REPORT

A six year old boy reported to the department of Paediatric and Preventive Dentistry, with the complaint of absence of teeth, inability to eat and difficulty in speech. The general medical history and family history was non-contributory.

The child exhibited the classical features of ectodermal dysplasia: anodontia, hypohidrosis, hypotrichosis, prominent forehead, saddle nose, diminished lower facial height, sparse scalp hair, missing eyelashes and eyebrows and protuberant lips.

The intra-oral examination revealed complete absence of primary teeth, as a result of which the edentulous alveolar ridges were deficient in both height and width. The mandibular ridge was of the knife edge variety and the maxillary ridge was depressed posteriorly with a shallow palate. The oral mucosa had a slight dry appearance and the tongue seemed relatively large.

Radiographic examination revealed complete absence of permanent tooth germs and

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underdeveloped maxillary and mandibular alveolar ridges.

A removable upper and lower complete denture was planned for the patient, considering the patient's need for mastication, esthetics, speech and overall psychological development. Although routine procedures for construction of complete dentures were used, case specific modifications were required, as described below.

The maxillary primary impression was recorded using a stock metal tray and polyvinyl siloxane putty impression material. Owing to the typical thin knife edge shape of the mandibular ridge, in place of conventional stock metal tray, primary impression was recorded by supporting the polyvinyl siloxane putty on an 18 gauge orthodontic wire, which had been adapted to conform with the shape of the mandibular arch.

Acrylic custom trays with spacer were fabricated on the primary casts and single step border molding was carried out using putty impression material. Conventional border molding using low fusing compound was avoided for better patient compliance, avoidance of discomfort, irritation and injury to the mucosa. Final impressions were then recorded using light body-injection type polyvinyl siloxane impression material and master casts were obtained.

Temporary record bases and wax rims were fabricated on the master casts. The jaw relation was recorded conventionally, after assessing the phonetics and aesthetics. In order to reduce the forces directed onto the compromised ridges and to consequently reduce the amount of alveolar resorption, the teeth from deciduous tooth mould were reduced in width and height. The cuspal inclines were reduced to a non-anatomic form as it was difficult to obtain an accurate centric relation record. Following the try-in appointment, the waxed up dentures were acrylicized, finished and polished.

After the final insertion, routine post insertion instructions were given to the patient and parents. At recall appointments, pressure spots were identified and relieved. Retention of the maxillary denture was good and the patient gradually adapted to the lower denture. The patient's self esteem improved, as did his socialization skills. Further recalls have been scheduled every three months. Further treatment will include modification of the dentures by relining or replacement of the dentures according to the observed skeletal growth.



Fig.1 Preoperative appearance



Fig. 2 Knife edged ridges

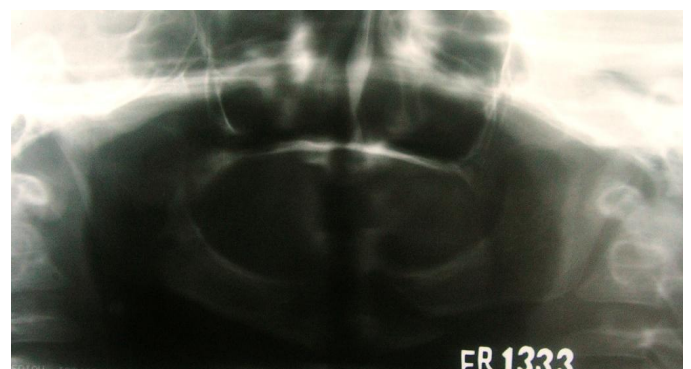


Fig. 3 Radiographic appearance



Fig. 4 Denture insertion

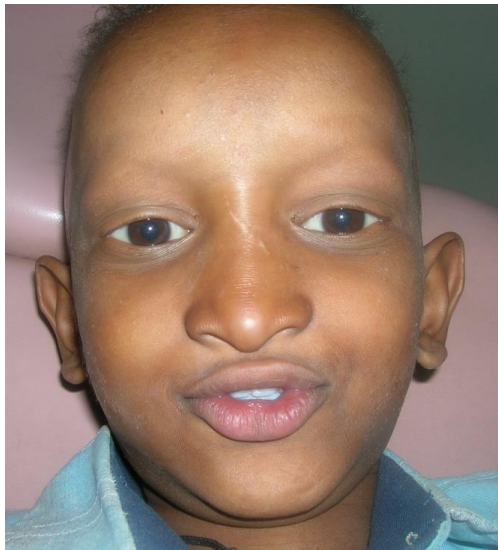


Fig. 5 Post operative appearance

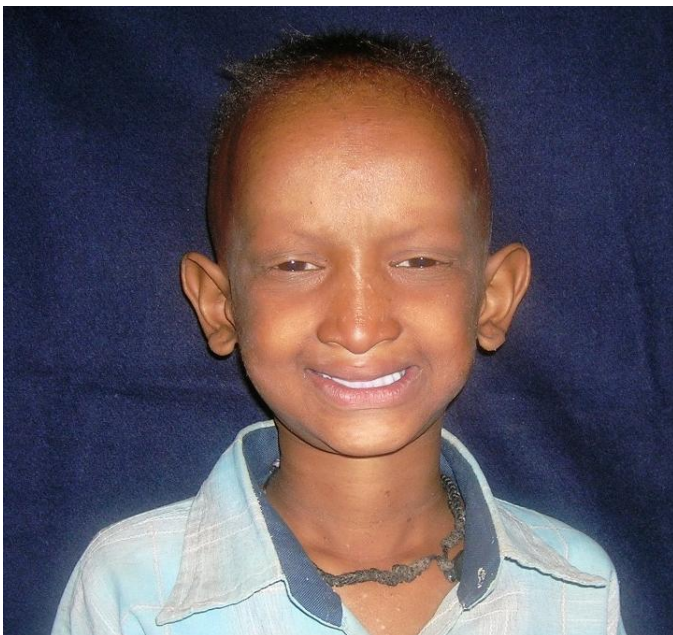


Fig. 6 Aesthetics and confidence restored.

Discussion

The typical decreased vertical facial height of this patient resulted from the congenital absence of teeth leading to underdevelopment of the alveolar ridges. In order to enable a further unrestricted development, the dentures have to be adapted to the changing intra-oral situation due to growth^{7,8,9}.

Historically, prosthetic treatment for ectodermal dysplasia patients involved removable partial dentures, removable partial or complete overlay dentures and fixed partial dentures. The advent of dental implants and implant borne total telescopic dentures has provided an additional treatment modality for restoration of the dentition in this group of patients².

There are aesthetic, functional and psychological reasons that make it important to start oral rehabilitation early in life. However, this is usually a difficult condition to manage prosthodontically, because of the typical oral deficiencies and patient's age. Numerous clinical reports have demonstrated the importance of prosthetic dental treatment in ED patients for physiologic and psychosocial reasons¹⁰.

Serial sets of dentures may be required as a child matures. Shaw describes the need for these to restore vertical dimension and prevent the undesirable protruding lips secondary to overclosure, thereby improving the profile⁴. Ideally, a restored dentition should be in place before the child attends primary school as oral rehabilitation has a major psychological impact on the patient's self esteem and facilitating social acceptance.

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