



ECTODERMAL DYSPLASIA –AN UNUSUAL PROSTHETIC PROBLEM

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Article Info <i>Received 18/09/2015</i> <i>Revised 29/09/2015</i> <i>Accepted 13/10/2015</i>	ABSTRACT Hypohidrotic Ectodermal dysplasia is a hereditary disorder of two or more ectodermal structures. It may include hypodontia, hypotrichosis and hypohidrosis. Patient with this disease often need complex prosthetic treatment. The option for a definitive treatment plan may include removable, fixed or implant supported prosthesis, single or in combination. A case of a 6 year old child with Hypohidrotic Ectodermal dysplasia with complete anodontia is presented .Clinical management consisted of fabricating complete denture to help in psychological development and to restore the vertical dimension, esthetic and functioning of the stomatognathic system.
Key words: Ectodermal dysplasia, Anodontia, Prosthetic treatment.	

INTRODUCTION

Ectodermal dysplasia (ED), as first described by Thurman, is a hereditary disorder with abnormal development of tissue and structures of ectodermal organ [1]. Over 170 different forms of ED have been described [2,3]. Two clinically distinguished forms are hydrotic form (Clouston's syndrome) and a hypohidrotic form (Christ – Siemens-Touraine Syndrome).⁴ The later form is most common type seems to be an X-linked recessive trait, with an incidence of this syndrome is estimated to be 1 to 7 per 100,000 live births [4]. Hypodontia, hypotrichosis and hypohidrosis which form a triad are the characteristic feature of the hypohidrotic form [5].

The oral manifestation of this disorder creates treatment challenges for the dentist [6]. The most frequent prosthetic treatment for the dental management of ectodermal dysplasia is removable prosthesis. The need for partial or complete dentures is critical during the preschool years and continues into adulthood. Hypodontia leads to an atrophy of the alveolar bone so it becomes more challenging than usual to construct dentures and restore the form and function [7]. In such cases, multidisciplinary team is generally advocated to be the most appropriate

approach [8]. The present case report describes the prosthetic rehabilitation of a young boy with hypohidrotic ectodermal dysplasia associated with complete anodontia.

CASE REPORT

A 6 year old boy was presented to the department of Pediatric dentistry because of lack of teeth, speech problems and mastication difficulties, which resulted in a very restricted diet and esthetic concerns. The child himself seemed anxious to improve his appearance, and his attitude enhanced his compliance with the treatment. The patient had been diagnosed with anhidrotic ectodermal dysplasia when he was 3-year old. The patient had typical characteristics such as protuberant lips, a saddle nose, fine sparse hair, and scanty eye lashes and eyebrows (Fig 1). In addition the patient showing aged profile with increased nasolabial fold and intolerance to heat. The patient father and mother were normal and not consanguineous. Her elder sister was also normal and none of the parent's relatives was known to have a similar condition.

Intraoral examination revealed the edentulous condition (Fig 2). The alveolar ridges were rather atrophic.



The color of alveolar mucosa and other oral mucosa was normal. The flow of saliva was observed in the floor of oral cavity. Panoramic radiographs revealed complete absence of teeth or tooth buds and aplasia of the alveolar processes of the maxilla and the mandible (Fig 3).

Removable complete maxillary and mandibular complete denture was provided to the child .Removable prosthodontic treatment for children followed the same basic principle as would have been followed for an adult patient. Preliminary impressions were made with irreversible hydrocolloid, and then custom trays were prepared for functional impression. Functional impression were made using zinc oxide eugenol impression paste .On the master cast ,acrylic bases with wax rims were made and jaw relation were registered and then they were mounted on an articulator. Teeth selection require more

attention. Primary tooth forms, are smaller in size does not fulfill the adequate vertical dimension because the patient is in the transition period from mixed to permanent dentition, so permanent tooth forms were selected. Trial dentures were checked for retention, freeway space, occlusion; esthetics (appearance of lips, fullness of lower face) phonetics .Acrylic dentures were processed (Fig 4). The child was given post insertion instruction and was advised to maintain a soft diet for the first few days, and to remove the dentures at night to promote healing of the oral tissue. The patient was recalled every 3 month. After 12 months the dentures still fit well with an excellent occlusion and the parents reported a significant improvement in terms of speech and mastication (Fig 5). Further treatment will include modification or replacement of the dentures according to the observed skeletal growth.

Fig 1. Frontal view of the patient



Fig 2. Intraoral view of the patient



Fig 3. Panoramic radiograph of the patient

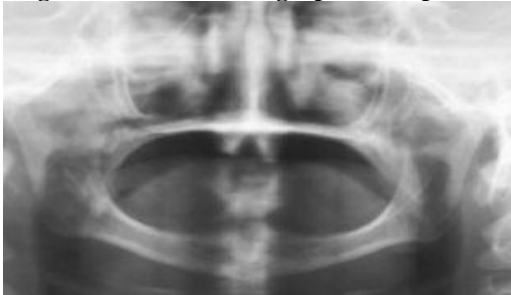


Fig 4. Upper and lower prosthesis



Fig 5. Frontal view of the patient after treatment



DISCUSSION

Prosthetic treatment modes using RPDs or complete dentures and dental implants are the primary treatment alternatives for the clinical management of E D young patients with severe hypodontia or anodontia. It is

often characterized by underdeveloped residual ridges and loss of vertical dimension of occlusion [9]. 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 dento- facial region is taking place and definite rehabilitation following completion of jaw growth [10, 11].

There is no definitive time to begin treatment, but Till and Marques [12] recommend that an initial prosthesis could be delivered when the child starts school, so that the child could have a better appearance and have time to adapt to the prosthesis. This early restoration of facial appearance is essential for normal psychological development. In each situation, the parents should be aware of the possibility that the young patient may refuse the treatment procedure initially and may not wear the dentures.

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. They require regular adjustment and should be replaced when a decreased vertical dimension of occlusion and an abnormal mandibular posture are detected due to growth. Without a dentures, the antero-rotation 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 and a tendency to class III malocclusion. The presence of dentures allows a backward-downward rotation of the mandible, with consequent normal positioning of the chin in space [13,14]. Retention and stability of the prosthesis are also difficult to obtain. In HED patients, dryness of the oral mucosa and the underdeveloped maxillary tuberosities and alveolar ridges are problematic factors for resistance and stability of dentures. When planning dentures in these patients, care should be taken to obtain a wide distribution of occlusal load fully extending the denture base [15].

When full growth is achieved, treatment planning may include implant-retained prosthesis. Early implant placement in patients aged up to 5 to 6 years have been

reported in the literature [16]. However it must be kept in mind that early implant placement in a growing child may cause cosmetic problems, as the implants act like ankylosed teeth [17]. According to I.P.Sweeney [18] the mean age of patients receiving maxillary implant was 18 years and mandibular implant was 17 years.

In this case as the child was 6 years old when first reported to the department, implant could not be considered as the treatment of choice. It was decided to fabricate a complete denture. The patient and parent should be instructed in the home care of prosthodontic appliances. They need to be informed of the need for periodic recall and refabrication of the dentures as growth occurs. The treatment not only improved the patient functional and esthetic status, but also improved the psychological health and social life.

CONCLUSION

This clinical report describes the prosthodontic management of a child with anhidrotic ectodermal dysplasia. The use of acrylic removable prostheses is an interesting and practical alternative that provides a relatively quick, easy, acceptable and economical solution in these patients. New alternatives for rehabilitation such as the use of implants, must be carefully considered, taking into account the cost factor, presence of underdeveloped, thin alveolar bones and age.

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CONFLICT OF INTEREST

No conflict of interest.

STATEMENT OF HUMAN AND ANIMAL RIGHTS

All procedures performed in human participants were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. This article does not contain any studies with animals performed by any of the authors.

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