

Oral rehabilitation of a hypohidrotic ectodermal dysplasia patient: An unusual prosthetic problem

Clinical Report

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ABSTRACT

The aim of this clinical report is to describe the prosthodontic management of a young boy aged 7 years affected by ectodermal dysplasia (ED). Dental treatment can vary depending on the severity of the disease (tooth size, morphology, and the amount of available alveolar bone). New technologies, such as adhesive dentistry, overdenture, and complete denture represent some of the options in the management of the rehabilitation of the patient affected by ED. The conical shaped central incisor was modified using composite resin followed by complete denture. Prosthodontic and restorative treatment was provided for the psychological and social comfort of the young patient.

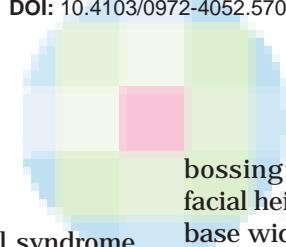
KEY WORDS: Ectodermal dysplasia, prosthetic problem

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INTRODUCTION

Ectodermal dysplasia (ED) is a congenital syndrome characterized chiefly by abnormalities of tissue of ectodermal origin, namely skin, nails, hair, and teeth.^[1] There are more than 150 different variants of ED.^[2] ED is usually described as being hypohidrotic or hidrotic depending upon the degree of sweat gland function. Hypohidrotic ectodermal dysplasia (HED) frequently exhibits the most severe dental anomalies; thus clinical management is of interest to the prosthodontist. HED is characterized by hypohidrosis, hypotrichosis, and hypodontia and occurs with a frequency of 1/100000 births.^[3] Exact data are not available concerning the prevalence of ectodermal dysplasia although it is comparatively rare. Finn in 1944 reported the cases of 81 patients. Nyegaard studied 80 reports; 3 of these patients had complete anodontia and the rest had varying numbers of primary and permanent teeth.

The classical facial features of HED include frontal



bossing; depressed nasal bridge reduced vertical facial height and facial depth, small palatal and cranial base width, small malar processes, and small high set orbits.^[6] The most striking oral feature of HED is the absence of most deciduous and permanent teeth. The average number of missing permanent teeth is reported as 23.7.^[7] The maxillary central incisor incisors, maxillary first molars, mandibular first molars, and maxillary canines are the teeth most often present.^[8,9] The maxillary central incisors and the maxillary and mandibular canine are usually conical in shape. The alveolar process fails to develop at edentulous sites^[10,11] and may be poorly developed even at dentate sites.^[12] The deficient alveolar growth and decreased vertical dimension of occlusion results in a small lower vertical face height.^[13] These features give the child a distinctly aged facial appearance similar to an edentulous patient. Diminished salivary secretion^[10], dry oral mucosa, and hoarse voice quality have also been reported.^[14] The psychological and functional effects of HED can have a tremendous negative impact on a young individual.

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Figure 1: Extra-Oral view



Figure 2: Intra-Oral view showing conical shaped tooth



Figure 3: Orthopantomogram



Figure 4: Conical shaped tooth contours with composite resin



Figure 5: Poly vinyl siloxane impression material used



Figure 6: Poly vinyl siloxane impression, boxing was done



Figure 7: Cast prepared with improved die stone



Figure 8: Complete denture in patient's mouth

Dental management of ectodermal dysplasia patients traditionally focuses on provision of a series of complete or removable dentures partial (RPD) during the growing years.^[15] Early and extensive dental treatment is needed throughout childhood because of the absence of most of the deciduous and permanent dentition. A multidisciplinary team approach to treatment is recommended.^[16] In addition, osseointegrated implants are an alternative treatment in older persons with anhidrotic ectodermal dysplasia.^[17,18]

A 7-year-old boy [Figure 1] with HED was referred due to missing of all teeth except upper right deciduous central incisor. On examination, the patient presented with the classical triad of hypohidrosis, hypotrichosis, and hypodontia mandibular over closure. The erupted central incisor was conical in shape [Figure 2]. An important feature was decreased height of the lower 3rd of the face. The orthopantomogram X-ray [Figure 3] revealed the presence of a permanent tooth bud beneath the erupted deciduous right central incisor. No tooth was present in mandibular arch neither was any presence of underlying tooth bud.

Treatment options considered for this child included extraction of upper right deciduous central incisor followed by construction of complete denture. However, due to deficient alveolar growth in edentulous area and even near the dentate area the option of extraction was not considered in order to preserve whatever alveolar bone was present. Hence the conical shaped deciduous central incisor was contoured with the help of composite resin [Figure 4] to attain the correct morphology of deciduous central incisor. The contouring of conical shaped deciduous central incisor also enhanced the retention of upper denture due to favorable guiding plane achieved. Maxillary and mandibular dentures were fabricated in conventional manner. A polyvinyl siloxane (PVS) [Figure 5]^[19,20] impression material was used for this young patient with dry mucosa, because PVS is highly biocompatible, clean, and pleasant for the patient. It is highly elastic and the setting time is less which help in easy removable from thin undercut area and causes less discomfort for the patient.

Anterior deciduous resin teeth were used for esthetic reasons. Anatomic 33° resin posterior teeth were used to develop bilateral balanced occlusion. Maxillary and mandibular trial denture was tried in centric relation position and Vertical dimension of occlusion (VDO) were verified, and protrusive record was obtained. Prior to the pouring of the PVS impression, boxing was done [Figure 6]. The cast was formed in improved die stone

for greater strength and abrasion resistance [Figure 7].

After the approval of the tooth arrangement by the patient and his parents, the waxed dentures were processed in a heat-polymerized denture base resin (Lucitone 199. Dentsply International Inc, York, Pa.) The complete denture [Figure 8] was delivered and the patient and his parents were instructed on the proper maintenance of oral hygiene and application of a fluoride gel on the deciduous tooth two times a day. The patient put on a 72 hours follow up schedule for any adjustments. The patient was then scheduled for 1 week, 1 month, and 3 months follow up. Since that time, the patient had been observed without major complication since 2 years.

DISCUSSION

After 2 years, with the exception of several minor adjustments, there have been no problems with the complete denture. No caries has developed in the tooth present in the oral cavity. The maxillary right deciduous central incisor may be exfoliated eventually and the permanent central incisor will be allowed to erupt into position. At that stage, further treatment is to be considered. Moreover, the erupting tooth remained as potential overdenture abutment and hopefully to stimulate the growth of alveolar bone. Complete denture prosthesis can provide acceptable results for esthetics, psychological support, and function. But underdevelopment of the alveolar ridges and xerostomia in the patient of hypohidrotic ectodermal dysplasia makes denture retention and stability difficult to achieve. Because of erupting tooth and growing jaws of the young patient, prosthodontic treatment was modified and adjusted or reconstructed as necessary. Endosseous implants can also be considered an alternative treatment. However, the consideration of osseointegrated implants was postponed because of the age and potential growth of this young patient.

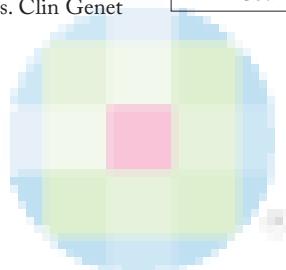
SUMMARY

This clinical report describes the characteristics and prosthodontic restoration of a young male patient with hypohidrotic ectodermal dysplasia. With proper care and prosthodontic treatment the patient can enjoy a relatively normal life. It is important that the patient and his parents fully understand the dental problems related to his physiologic, psychologic, and social conditions. The need for continued dental treatment is necessary.

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