Prosthodontic Management of a Patient with Ectodermal Dysplasia

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ABSTRACT

Ectodermal dysplasia is a rare congenital disease that affects the ectodermal structures. It is characterized by hypotrichosis, hypohidrosis and hypodontia. A 14-year-old boy with ectodermal dysplasia presenting with oligodontia and marked resorption of the maxillary and mandibular alveolar ridges is reported. Prosthetic rehabilitation in the form of a maxillary and mandibular partial denture was made with metal crowns on existing lower teeth to achieve appropriate vertical dimension. Significant improvement in speech, masticatory function and facial esthetics was achieved. Removable prosthodontics can provide an acceptable solution to esthetic, functional and psychological rehabilitation in patients with ectodermal dysplasia.


INTRODUCTION

Ectodermal dysplasias (ED) are a group of heritable conditions characterized by congenital defects of two or more ectodermal structures. They are relatively rare and occur with a frequency varying between 1:10,000 and 1:100,000 live births. Two patterns of ED - the hypohidrotic and hidrotic form have been described. Hypohidrotic ED can either be X-linked recessive (mutations in EDAREDA gene) or autosomal recessive (mutations in the EDARADD gene) with resultant defective formation of ectodysplasin protein. Autosomal dominant (mutations in the DL gene that encodes ectodysplasin receptor) variant has also been described but are rare. Hidrotic ED is autosomal dominant disorder caused by mutations in GJB6, which encodes connexin 30, a component of intercellular gap junctions.

The tissues primarily involved are the skin, hair, nails, sweat glands, and teeth. The most striking feature in the oral cavity is oligodontia. Teeth in the anterior region of maxilla and mandible are conical in shape; and one molar tooth in the second molar region usually exhibits a bud crown form. Oral rehabilitation of the ED patient is recommended to improve both the sagittal and coronal skeletal relationship during craniofacial growth which is necessary for better esthetics, speech and masticatory efficiency.

This case report describes prosthodontic management of oligodontia in a young boy with ectodermal dysplasia.

CASE REPORT

A 14-year-old boy presented with the chief complaints of failure of development of dentition with difficulty in mastication and poor dental esthetics. There was no history of heredo-familial diseases. He exhibited features of ED in the form of saddle nose, prominent forehead, protuberant lips and dry skin (Figure 1A). The nails were abnormal in contour and texture. He had two permanent first molars and a retained deciduous molar in the mandibular arch (Figure 1B) and two permanent first molars in the maxillary arch (Figure 1C), the remaining teeth/tooth buds were congenitally missing and the alveolar ridges were underdeveloped. The palate was well developed but flattened and the tongue appeared to be abnormally large. Radiographic examination revealed well-formed roots of the erupted teeth and the absence of any other teeth or tooth buds in either arch (Figure 1D).

Interocclusal rest space determined by measuring the difference in the distance between the nose tip and the chin tip at rest and in occlusion was found to be 4 – 5 mm; hence, it was decided to increase the vertical dimension of occlusion (VDO). Maxillary and mandibular removable partial dentures were considered to be the treatment of choice. However, to increase the VDO (Figure 1E), individual metal crowns on the teeth present in the lower arch in combination with the removable partial denture were planned.

The two first molars and a retained deciduous molar, in the mandibular arch were prepared to receive all metal crowns. Preliminary impressions were made with irreversible hydrocolloid. Custom trays were fabricated with autopolymerizing acrylic resin and definitive impressions were made. A centric relation record at the increased vertical dimension of occlusion was made and the casts were mounted in a semi-adjustable articulator.
A clinical photograph showing protuberant lips and saddle nose deformity with frontal bossing, (B) Intra-oral view of mandibular and maxillary arches showing oligodontia, (D) Lateral cephalogram showing well-formed roots of the erupted teeth and the absence of any other teeth or tooth buds in either arch (E) Use of metal crowns on mandibular teeth to increase the occlusal vertical dimension, (F) Intra-oral view of the removable partial dentures in maxillary arch, (G) in occlusion and (H) in mandibular arch, (I) clinical photograph showing esthetic outcome.

Artificial teeth were chosen to achieve an age-appropriate appearance and were arranged in wax for trial evaluation. The occlusion and position of the teeth were checked intraorally to evaluate the patient's tolerance to the increased occlusal vertical dimension as well as esthetics, phonetics, and occlusal harmony. The dentures were processed with heat-activated acrylic resin (Lucitone, Dentsply International) using the conventional short curing cycle. After processing, the dentures were remounted and adjusted for laboratory processing error. Partial dentures were inserted (Figures 1F – 1H) and instructions on the regular cleaning and maintenance was explained to the patient and the parents. The patient was instructed to remove the dentures at night and to present the following day and once a week for a period of one month for possible corrections and adjustments. The accommodation to partial dentures by the patient occurred with considerable improvements in esthetics, speech and masticatory function (Figure 1I).

DISCUSSION

Majority of the ED cases follow the autosomal-recessive mode of inheritance, but they can also be autosomal-dominant or X-linked.1 Any structure derived from the ectoderm can be defective in ED. The skin is usually dry, scaly and easily irritated as a result of poorly developed or absent sebaceous glands. Sweat glands can be absent, reduced in number, or non-functioning (hypo-
implants, removable partial dentures are a better treatment option to obtain the desired functional and esthetical benefits. In addition, an understanding of the ED patient's psychosocial status is crucial to any prosthodontic treatment effort. The unesthetic appearance that accompanies ED syndrome often has a negative psychological effect on the patient. Providing expedient prosthodontic treatment to manage orofacial disfigurement may afford the patient some measure of confidence. Therefore, early intervention is advised in these cases.

This patient had oligodontia with absent anterior teeth and the retained deciduous molar was attrited, which resulted in loss of VDO. The VDO was increased by restoring the teeth in the mandibular arch with individual metal crowns. The patient remained comfortable at the increased VDO and early placement of removable partial denture was planned in order to maintain the relationship of the maxilla to the mandible and allow normal growth pattern of the face. Dental implants were not considered due to reduced bone volume and young age. Removable prosthesis was cost effective; it provided rapid and painless result. Follow-up and recall check up at three monthly intervals showed significant improvement in speech, masticatory function and facial esthetics, thereby minimizing the emotional and psychosocial stress on the patient and his family.

REFERENCES