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Case Report

Prosthodontic rehabilitation of a pediatric patient with hypohidrotic ectodermal dysplasia: a case report

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ABSTRACT

The aim of this case report was to prosthodontically rehabilitate and boost the psychological development of the patient. Ectodermal dysplasia is a hereditary condition presented with abnormal development of tissues of ectodermal origin such as hair, skin, nails, and teeth. This condition generally presented with following signs and symptoms; dental abnormalities, onychodysplasia, hypohidrosis, and hypotrichosis. Ectodermal dysplasia usually has an X-linked recessive trait, with most common being of hypohidrotic ectodermal dysplasia also known as Christ-Siemens-Touraine syndrome. This case report describes the prosthetic rehabilitation with conventional dentures in maxillary arch and tooth supported overdenture in mandibular arch of a young boy with hypohidrodic ectodermal dysplasia. Prosthetic treatment is very helpful to these patients from functional, psychological, and psychosocial points of view. Need for continued dental treatment is necessary.

Keywords: Hypohidrotic ectodermal dysplasia, Christ-Siemens-Touraine syndrome, Prosthetic rehabilitation, Multidisciplinary management, Pediatric patient

INTRODUCTION

Ectodermal dysplasias are congenital disorders of the epidermis and its appendages. It was first described by Thurnam in 1948.[1,2] Ectodermal dysplasias are diverse group of disorders with more than 170 clinically distinct symptoms. It is a very uncommon disorder.[1,3]

Patients with ectodermal dysplasia generally presented with aberrant development of tissues and appendages of ectodermal origin.[4-6] This condition is mainly of two types. The first is Christ-Siemens-Touraine syndrome, whereas the second is Clouston's syndrome.[2,5,6] Christ-Siemens-Touraine syndrome is the most common kind of ectodermal dysplasia. It's an X-linked recessive disorder. In this type, sweat glands may be missing and if present they are very less in number. Clinically it is categorized into two types: hypohidrotic and anhidrotic ectodermal dysplasia. In Clouston's syndromes, it is inherited as an autosomal dominant disorder and the sweat glands are normal. Both types of condition affect the teeth and hair in the same way, but the inheritance patterns and sweat gland symptoms tend to be different.[6]

Orofacial characteristics of ectodermal dysplasias may be manifested as anodontia or hypodontia. This condition may manifest itself as reduce in the development of the alveolar ridge; as a result, the lower face height is reduced. Existing teeth may be malformed or may have conical morphology, the oral mucosa may be dry, indistinct vermilion border, protuberant lips, frontal bossing, depressed nasal bridge, and hypotrichosis. Affected children's faces generally have an elderly appearance. This condition may affect primary as well as permanent dentition.[6]

Early intervention is needed for oral and Prosthetic rehabilitation in ectodermal patients to restore function

and aesthetics. Furthermore, it also facilitates the normal emotional and psychological profile as well as improves the self-confidence of the patient. A multidisciplinary team approach is recommended for the treatment.[7,8]

This clinical case report describes the prosthetic rehabilitation of a young patient with ectodermal dysplasia.

CASE REPORT

A seven-year-old male patient was referred to the unit of prosthodontics, faculty of dental sciences, institute of medical science, Banaras Hindu university due to missing teeth, mastication and speech problems. The patient was accompanied by his mother. She gave the history of missing teeth since infancy except for one tooth present in the lower jaw which was erupted when the patient was 3 years old. She also gave the history of recurrent episodes of fever throughout infancy and also mentioned about his heat intolerance and preference for the cold environment. The kid was the only patient in the family, and no other members of the family had ectodermal dysplasia.

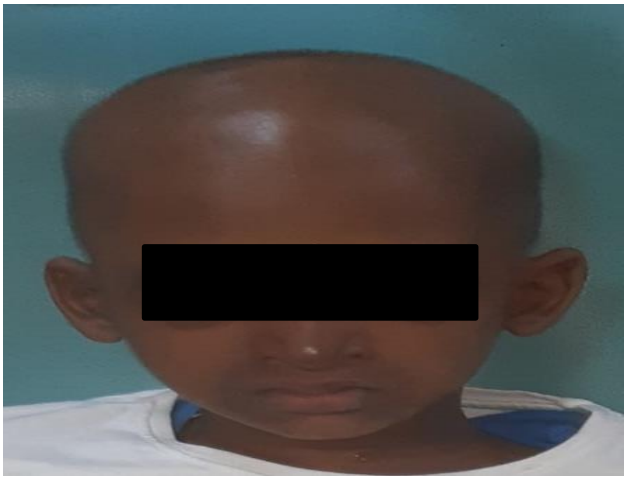

Figure 1: Extra-oral view.

The patient presented with moderate built and was poorly nourished. The patient had dry skin and body hairs were sparse and scanty. On extraoral examination, the patient displayed typical features of ectodermal dysplasia including; frontal bossing, periorbital pigmentation, depressed nasal bridge, protuberant lips, low-set ears, sparse scalp hair, absent eyebrows, and sparse eyelashes and reduced lower facial height which contributed to a senile appearance.

Intraoral examination revealed a relatively dry mucosa. The maxillary arch was completely edentulous and only one canine was present in the Mandibular arch. Alveolus ridges were poorly developed in both the arches (Figure 2). The underdeveloped maxillary and mandibular alveolar processes were further verified by radiographic investigation (OPG). There were four conical tooth buds present, 2 in the maxillary arch and 2 in the mandibular arch. There was also one horizontal impacted canine and one fully erupted canine in the mandibular arch (Figure 3). Patient was diagnosed with hypohidrotic ectodermal dysplasia (HED) because the patient was presented with, hypohidrosis, hypotrichosis, and hypodontia which are the classic features of ectodermal dysplasia.

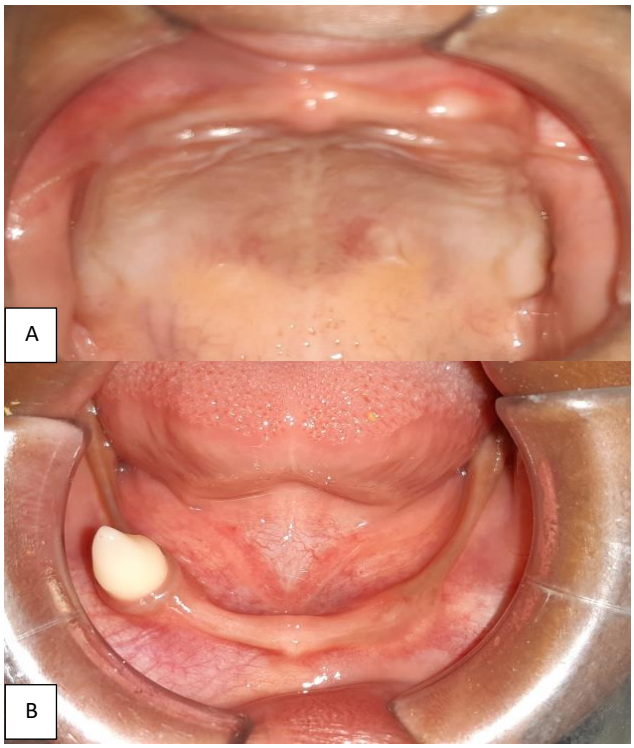

Figure 2 (A and B): Intra-oral view.

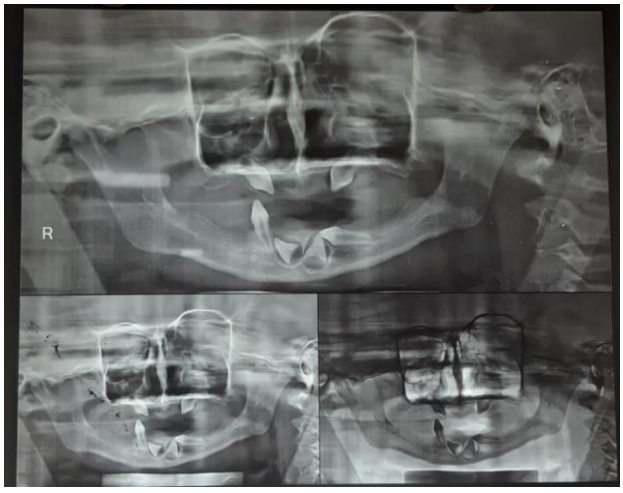

Figure 3: Orthopantomogram.

Prosthodontics management

In this case, a pediatric dentist was consulted regarding the patient's tooth buds and tissue conditions. No treatment was needed in this case. We discussed the treatment plan with the patient and his mother and

informed consent was obtained. The patient was rehabilitated with removable complete denture in the maxillary arch and tooth-supported overdenture in the mandibular arch.

Canine in the right mandibular arch had open apex. Apexification of the teeth was done with biodentine and later on abutment preparation was done. Metal coping was fabricated and was cemented to the abutment (Figure 4). Standard procedures were followed for the fabrication of complete dentures. Preliminary impressions were made with silicone impression (putty consistency) material for this patient with dry mucosa as the material is biocompatible, hydrophobic and requires a dry field. Special trays were fabricated for both the arches. Final impressions were made with light viscosity addition silicone for both the arches (Figure 5). Record bases with wax rims were fabricated on the master cast and jaw relation was registered (Figure 6).

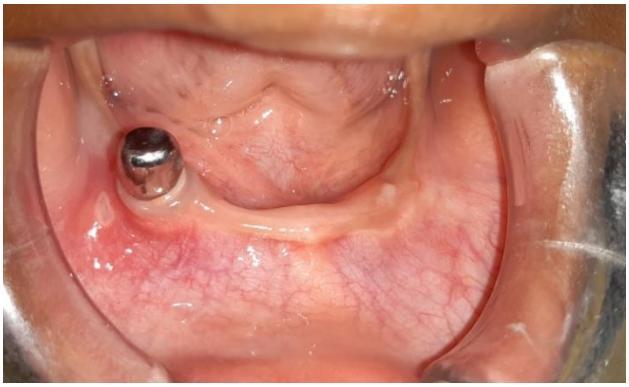

Figure 4: Abutment preparation was done and metal coping was cemented.

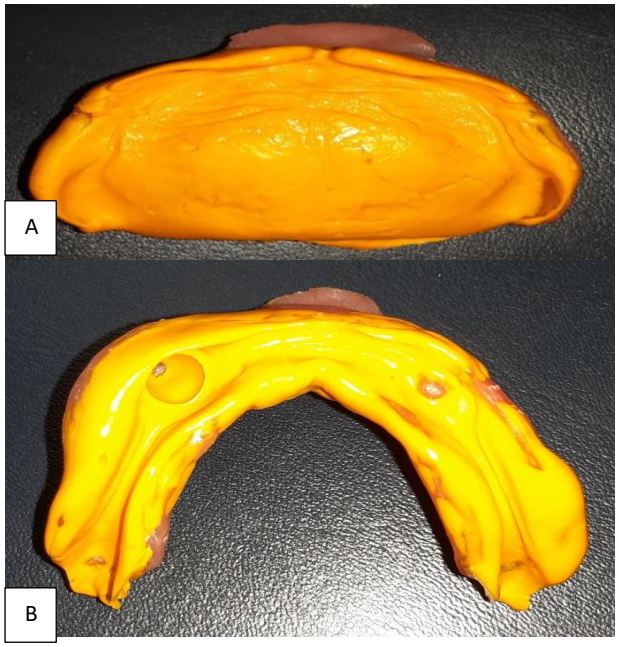

Figure 5 (A and B): Final impression with light consistency addition silicone.

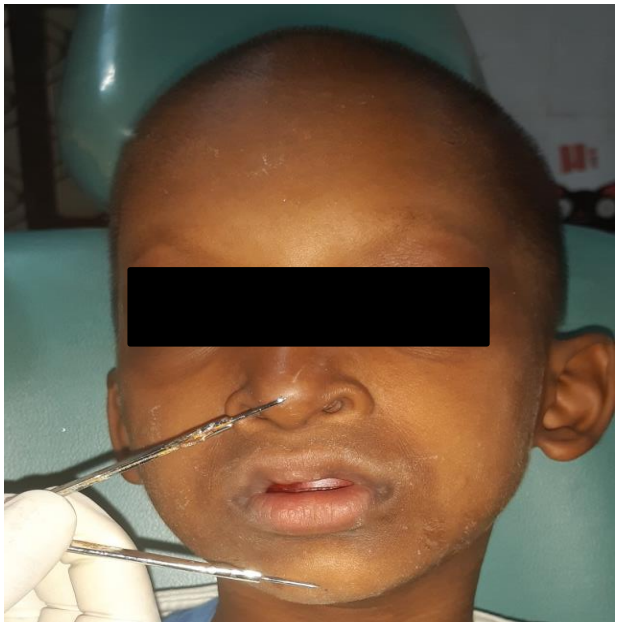

Figure 6: Maxillo-mandibular relation was registered.

Permanent tooth forms were selected instead of primary tooth forms, to provide better occlusion. Bilateral balanced occlusion was established during teeth arrangement. Trial dentures were tried intraorally and approval was taken from the patient and his mother (Figure 7). The waxed dentures were processed in a heat-polymerized denture base resin. The processed dentures were finished and polished and were inserted in the mouth and adjustments were made as needed (Figure 8). Post-installation instructions were given to the patient and his parents. Post denture insertion follow up was scheduled after 24 hours and 1 week for any adjustments. The patient was scheduled for continuous follow-up visits every three months to accommodate growth and development. Recall appointments showed good retention, and parents reported significant improvement in speech and mastication.

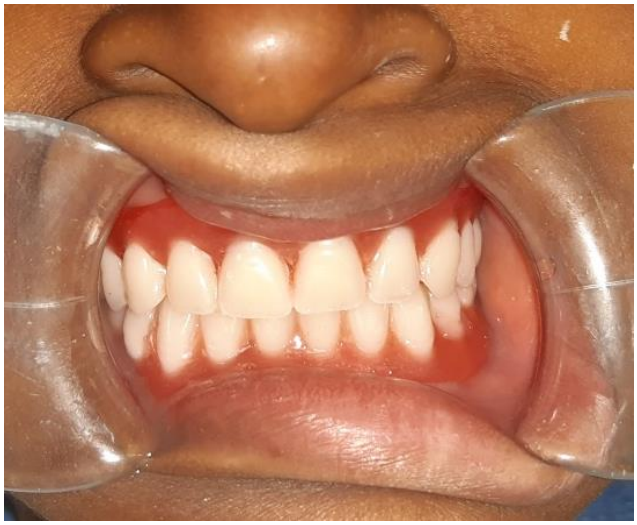

Figure 7: Denture try-in.

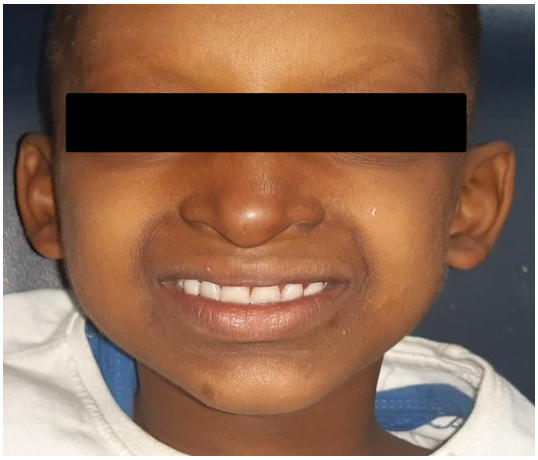

Figure 8: Final denture.

DISCUSSION

Treatment of ectodermal dysplasia in children requires a collaborative effort.[7,8] There is no definitive time to start dental treatment in ectodermal dysplasia patients, however, Pigno et al, suggested to give the first prosthesis before the school age (3-4 years) of patient[6], this helps to normalize the function of muscles of mastication and perioral region and consequently normalize the growth pattern of basal bones and providing a psychological boost to the child[9]. The prosthodontic rehabilitation of ED patients should be on an individual basis, considering the growth and developmental characteristics of each patient. Many therapeutic approaches have been reported, including single crowns, removable partial dentures (FPDs), complete dentures (CDs), removable partial dentures (RPDs), overdentures (ODs), and implant-supported dentures.[6,10-13] These can be used individually or in combination to achieve the best results. The most frequent prosthetic treatment option for ectodermal dysplasia is removable prosthesis as it can be easily changed during periods of rapid bone growth.[6,14] In this clinical report, the erupted teeth remained as potential overdenture abutments and conventional complete dentures in the maxillary arch and tooth-supported overdenture in the mandibular arch were planned. Due to continued growth and development as well as insufficient alveolar bone support, implant therapy was not addressed in this case. Although dentures are not good alternatives to healthy dentition, but they provided the much needed daily dietary nourishment.

CONCLUSION

The characteristics and prosthodontic rehabilitation of a young male patient with HED are described in this clinical report. Prosthetic treatment is very helpful to these patients from functional, psychological, and psychosocial points of view. Need for continued dental treatment is necessary.

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