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Case report:

Prosthodontic rehabilitation of a hypohydrotic ectodermal dysplasia patient with extracoronal attachments: A case report

Mohammed Ziauddeen Mustafa BDS, MDS

Corresponding Author: moh.z@mu.edu.sa, drzia01@gmail.com; Contact: +966-557921606; +91-9789940310

Mohammed Ziauddeen Mustafa

Assistant Professor in Department of Prosthodontics, College of Dentistry, Majmaah University, Al- Majmaah, 11952, Saudi Arabia.
ORCID No. 0000-0003-1090-1200

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Abstract

Prosthodontic rehabilitation in a patient with hereditary ectodermal dysplasia (ED) is often challenging due to compromised intraoral supporting structures. The recent advances in dental materials and improved techniques provide an array of options in restoring both the aesthetics and function of the stomatognathic system, especially in complicated scenarios. The literature search provided diversified modalities of treatment approaches for individuals affected with ED. This article describes the use of an extra coronal attachment in the prosthodontic management of a hypohydrotic ectodermal dysplasia patient presenting with severe oligodontia.

Keywords: Ectodermal dysplasia; oligodontia; dental prosthesis; extracoronal attachments

Introduction

Ectodermal dysplasia (ED) connotes a heterogeneous group of rare, inherited disorders mainly characterized by dysplasia of ectodermal tissues and occasionally of mesodermal tissues of the developing embryo.[1-3] It is thought to occur in approximately 1 in a million live births, with a mortality rate of 28% in males.[4,5] There are two major types of ED depending on the number and functionality of the sweat glands, namely, hypohydrotic and hydrotic.[5] The most common form (80%) of ED is hypohidrotic ectodermal dysplasia (HED), and is often inherited as an X-linked disorder, characterized by hypohidrosis (hypoplasia of sweat glands), hypotrichosis (sparseness of scalp and body hair), and hypodontia.[4,6,7]

The prosthodontic management of ectodermal dysplasia for children affected with hypodontia, oligodontia, and anodontia have been reported in the literature.[8-10] In hypodontic patients, there is no consensus whether removable partial dentures should be fabricated as early as two to three years of age or whether orthodontic treatment and implant-supported overdentures are better treatment options at an early age.[9,10] In patients above ten years of age, a consensually feasible treatment modality includes an effective orthodontic treatment combined with removable partial dentures or implant-supported fixed prostheses.[15] For patients older than 18 years, fixed implant-supported prosthesis is recommended.[15] Prosthodontic rehabilitation in patients with anodontia, includes dentures, dental implants, and overdentures. However, a multidisciplinary treatment protocol is essential in the effective management of ED individuals.[26,28,33,39]

This article describes the application of castable semi-precision extra coronal attachment in the prosthodontic management of a hypohydrotic ectodermal dysplasia patient with severe oligodontia.

Case Report:

A 16-year-old male patient reported to the outpatient department of prosthodontics complaining of difficulty in mastication and unaesthetic appearance due to missing teeth. The patient had an average build, and his gait was normal. He had generalized dryness of skin, with no family history of relatives presenting with a similar condition, and his birth was from a no consanguineous marriage.

Extraoral examination revealed sparseness of hair in the scalp area and also on both eyebrows. There was hyper-pigmentation around the eyes, pronounced supraorbital ridge, and depressed nasal bridge. He had hoarseness in his voice. The patient's face form was tapering, and his lateral facial profile was straight. The lips were dry, scaly, and protuberant. [Figure 2a-2d] Temporomandibular joint examination revealed no abnormalities, and the mandibular movements were coordinated with no deviations observed on mouth opening.

Intraoral examination revealed severe oligodontia with only two permanent teeth present in the maxillary arch and a completely edentulous mandibular arch. The teeth present were morphologically resembling permanent canines with a favorable crown/root ratio and were located in the right and left maxillary central incisor region. [Figure 1, 2e-2g] The residual alveolar ridges were severely resorbed, and the mucosa overlying them was delicate and friable. The maxillary labial frenal attachment was prominent and highly placed, encroaching in between the two teeth. The patient's saliva was reduced in quantity.

The risks and benefits of all the treatment options were explained to the patient, and the patient opted for an attachment-supported removable maxillary prosthesis and conventional removable complete denture mandibular prosthesis. Primary impressions of maxillary and mandibular arches were made using alginate impression material, and diagnostic casts were fabricated.

A tentative jaw relation was recorded using wax occlusal rims on a temporary denture base. The record was then mounted onto an articulator, which showed a Class-1 ridge relation and the presence of adequate interocclusal distance for over denture attachment. [Figure 3a] A soft tissue laser was used to perform maxillary labial frenectomy.

Tooth preparation was completed using diamond burs on a high-speed air-rotor handpiece to receive porcelain fused to metal crowns. [Figure 3b] Low fusing green stick compound was used to perform the border molding procedure on both maxillary and mandibular arches [Figure 3d, 3e] using a custom tray fabricated on the diagnostic casts.

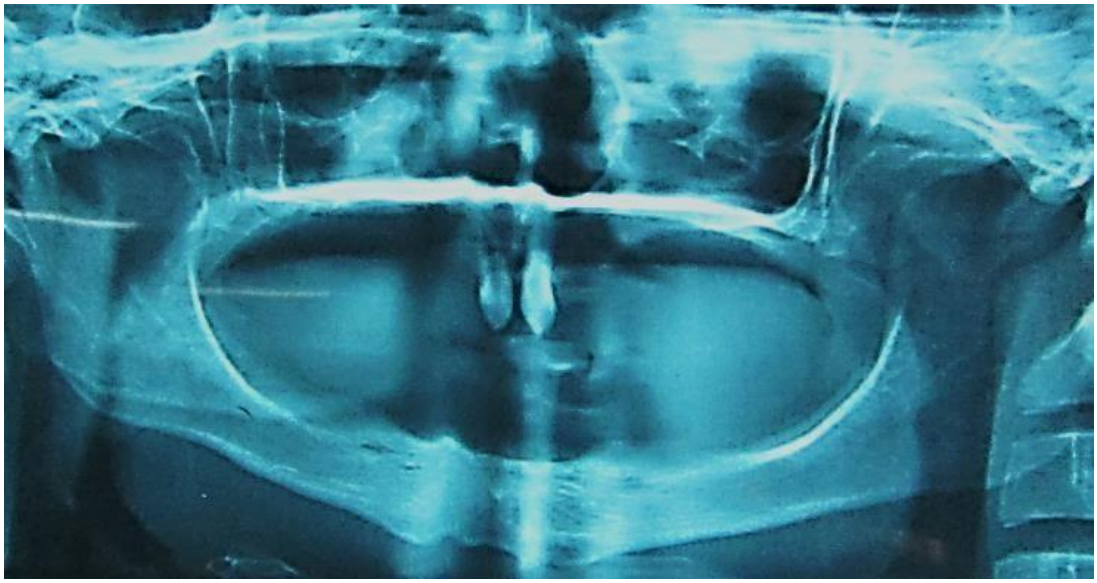

Figure 1 – Orthopantomogram of the patient showing presence of maxillary two central incisors.

Figure 2. (A) Frontal view, (b) Lateral view, (c) scarce eyebrows, (d) dryness of the skin, (e) Labial view of teeth present, (f)Palatal view of teeth present along maxillary alveolar ridge, and (g) mandibular alveolar ridge.

The final impression of the arches was then made with light-bodied vinyl polysiloxane impression material. The master casts were then made using type III dental stone. Two extra coronal attachments [Rhein83 OT-CAP, Rhein83 attachment systems, Italy] were carefully positioned on the distal surface of the wax pattern using a dental surveyor. [Figure 3f]

The wax pattern was then cast into a metal framework and subsequently layered with porcelain. The splinted final crowns and the cast attachments on the distal surface were then cemented onto the prepared teeth using a type 1 glass ionomer luting cement [GC Corporation, Tokyo, Japan]. [Figure 3c] Maxillary and mandibular acrylic complete dentures were fabricated using routine, conventional techniques.

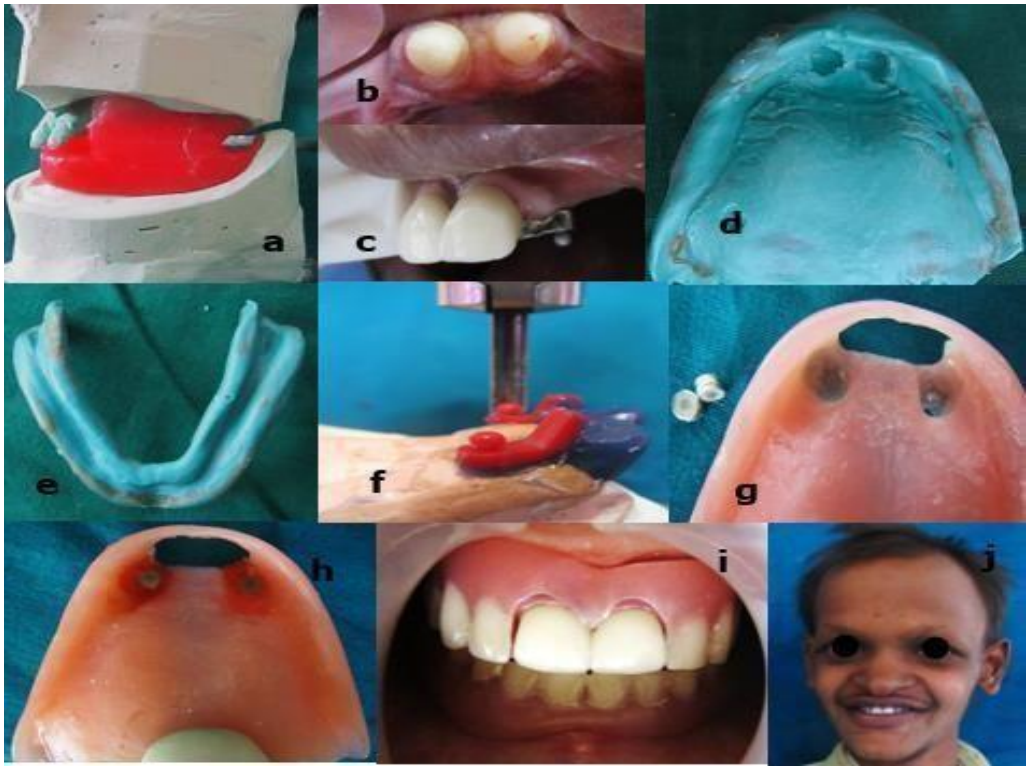

Figure 3 (a) Tentative jaw relation record, (b) Teeth preparation to receive PFM crowns, (c) Final cementation of splinted PFM crowns with extra coronal attachment, (d) (e) Maxillary and mandibular arch border molding and final impression respectively, with light-body addition silicone impression material, (f) Rhein83 OT-CAP male attachment with wax pattern, (g) Maxillary denture prepared for chair-side pick-up along with female attachments, (h) Chair-side pick-up done with pattern resin, (i) Final prosthesis delivery, and (j) Satisfied patient.

The female attachment's chair-side pick-up [Figure 3g, 3h] was carried out during the final insertion appointment using a pattern resin material [GC Corporation, Tokyo, Japan]. The prosthesis was retrieved from the patient's mouth, and any excess resin material was removed, the denture was polished again, and occlusion was verified before final delivery of the prosthesis. [Figure 3i, 3j] The patient was recalled for minor adjustments required for the prosthesis on a subsequent day. Recall checkups were completed after one week, one month, and three months after delivering the prosthesis to the patient.

Discussion:

Early intervention with dental prostheses is necessary in patients affected by ED to improve the quality of life in such individuals.[34-37] The clinical presentation of decreased lower facial height (poor vertical growth of maxilla and mandible) and Class-3 skeletal relation (less maxillary growth than mandible in the sagittal plane) makes prosthetic treatment planning more difficult.[40-43] In children presenting with oligo- or anodontia, one must consider craniofacial growth since a favorable maxillo-mandibular relation can improve aesthetic appearance and facilitate permanent rehabilitation as the individual grows. [16,27,48,50,51]

A systematic review [15] has proposed prosthodontic treatment recommendations by categorizing the patients depending on the age group. Early prosthodontic treatment is reported in most children at less than five years of age, affected with severe oligodontia or anodontia.[32,34,47,48] Fixed teeth supported prosthesis (resin-bonded bridges) were frequently used in patients with more than six teeth present.[15] Clasp retained removable partial dentures mainly represented are a standard restoration in children with oligodontia, whereas overdentures were considered an alternative option.[44-46]

Dental implant treatment is generally contraindicated in growing children [9,11,12], but mini dental implants can provide certain advantages such as reduced costs, less traumatic surgical placement, lesser implant diameter, and immediate loading with prosthetic restoration.[13,17] They should be considered as feasible provisional options until the completion of growth.[47,49] In children with ED, it is also reported that lesser bone volume and extremely hard bone have

been shown to result in a higher implant failure rate.[14] Schnabl et al. [15] suggested that concerning craniofacial development and chewing ability, a fixed implant-supported prosthesis in the mandibular arch for a younger age group does not seem to be a recommended treatment option. In adults aged >17 years, placing eight implants in the edentulous maxilla and six to eight implants in the edentulous mandible was a more standard treatment approach. The desire for better aesthetics and comfort directly reflects on the motivation to undergo extensive surgery in this age group. [29-31]

A removable dental prosthesis remains a more feasible alternative compared to fixed teeth-supported or implant-supported prostheses because the removable prosthesis is less invasive, more affordable, and it is easier to maintain oral hygiene.[38] Swelem et al.,[21] reported on patient satisfaction, and quality of life assessment among removable dental prosthesis wearers suggested that comfort, general satisfaction, and masticatory efficiency were significantly higher after conventional treatment and continued to improve significantly after the use of attachments. The use of such attachments increases prosthesis retention and stability in an aesthetically more pleasing way and may improve the patient's acceptance.[22-24] Persic et al. [25] reported that incorporation of attachments resulted in a more significant impact on mastication, speech, appearance, and psychological comfort. They also suggested that attachments provide an excellent cost-effective treatment option for partially edentulous individuals from a quality of life and patient satisfaction perspective. The drawbacks of the treatment option described in this report include periodic replacements of the rubber retentive caps in the metal housing, less stability of removable mandibular prosthesis, and refabrication or relining of prostheses with changes in the alveolar ridges.

Conclusion:

Prosthetic rehabilitation of patients with rare diseases such as ectodermal dysplasia requires an interdisciplinary treatment approach. The objectives of treatment are dictated by the age and demands of the patient. An effective treatment plan, continued guidance, and follow-up of such patients goes a long way in improving patient motivation and, thereby, increased compliance for treatment, resulting in improved quality of life in such individuals.

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