Biology and Medicine

Jain et al., Biol Med (Aligarh) 2016, 8:6
DOI: 10.4172/0974-8369.1000335

Case Report Open Access

Full Mouth Rehabilitation of an Ectodermal Dysplasia Patient with Hypodontia and Reduced Vertical Dimension Using Metal Ceramic Restorations: A Case Report

Ashish. R. Jain[1,*] and T. Janani[2]

[1]*Research Scholar & Reader, Department of Prosthodontics, Saveetha Dental College and Hospital, Saveetha University, Chennai, India*

[2]*CRRI, Department of Prosthodontics, Tagore Dental College and Hospitals, Chennai, India*

***Corresponding author:** Dr. Ashish R. Jain (MDS, MD.ACU.VARMA), Research Scholar & Reader, Department of Prosthodontics, Saveetha Dental College and Hospital, Ponamalle High Road, Chennai 600127, India. Ph: +91-9884233423; Email: dr.ashishjain_r@yahoo.com

Received: Jun 21, 2016; **Accepted:** Aug 16, 2016; **Published:** Aug 31, 2016

Abstract

Oral rehabilitation of the hypohidrotic ectodermal dysplasia (HED) patient is recommended to improve both the sagittal and vertical skeletal relationship as well as to provide improvements in esthetics, speech, and masticatory efficiency. It has also proven to boost the self-esteem of the patient. This clinical report describes the full mouth rehabilitation of a HED patient who was clinically monitored for a period of 3 months to evaluate her adaptability to the new increased vertical dimension at occlusion achieved with the help of an overlay removable partial denture (ORPD) for a period of 3 weeks, and provisional restoration for an additional period of 3 months to determine esthetic and functional outcome with provisional restoration, which was followed by the placement of final definitive prosthesis, designed based on shortened dental arch (SDA) scheme, which eliminates the need for invasive implant surgeries. The occlusal cant was corrected by a conservative approach using the macroesthetic elements of smile design, which helps to establish the occlusal plane in a simple and reliable technique.

Keywords: Full mouth rehabilitation; Hypohidrotic ectodermal dysplasia (HED); Vertical dimension at occlusion (VDO); Shortened dental arch (SDA); Macroesthetic elements

Introduction

Ectodermal dysplasia (ED) is a group of hereditary disorders involving an absence or deficiency of tissues and structures derived from the embryonic ectoderm. ED has been divided into 34 subgroups and over 100 variations have been identified. The disease may also be inherited by autosomal dominant, autosomal recessive, or X-linked genetic transmission. Clinically, ectodermal dysplasia may be divided into two broad categories—the hypohidrotic form (X-linked recessive) and the hydrotic form (Autosomal inherited). Hypohidrotic ectodermal dysplasia, also termed as Christ–Siemens–Touraine syndrome is more common and is characterized by a triad of signs comprising sparse hair (hypotrichosis), abnormal or missing teeth (hypodontia or anodontia), and an inability to sweat because of the lack of sweat glands (anhidrosis or hypohidrosis). The most common oral characteristic is hypodontia or anodontia in many cases, reflecting the complete suppression of dental ectoderm. A few teeth may be present but with retarded eruption. Because of the lack of teeth and resultant loss of vertical dimension, the lips are protuberant, the vermilion border is indistinct, and the alveolar process does not develop in the absence of teeth and, hence, is missing. This is often accompanied by characteristic facial features including a low nasal bridge, small nose with hypoplastic alae nasi, full forehead, prominent supraorbital ridges, prominent lips, and sparse hair [1-6].

Case Presentation

A 23-year-old female reported with the chief complaint that she could not eat anything because her teeth were worn too much, were hypersensitive and had an unesthetic appearance. On examination, the patient presented with multiple missing teeth (hypodontia), and on interrogation she revealed reduced sweating response (hypohidrosis). Since hypodontia and hypohidrosis are the characteristic features of hypohidrotic ectodermal dysplasia (HED), the patient was suspected to be affected. Extraoral examination revealed remarkable facial features such as depressed nasal bridge, frontal bossing, retruded midface, decreased lower anterior facial height, flat mandibular plane, prominent chin, and a resultant concave facial profile (Figure 1). Deviation of the mandible towards the left was also noted. There was insufficient incisor show on smiling. Intraoral findings included hypodontia, mandibular overclosure with excessive vertical overlap, and xerostomia. The patient had anterior cross-bite and scissor-bite posteriorly on the right side (Figure 2). Orthopantomogram (OPG) revealed generalized spacing, retained deciduous teeth (71,81,85), abnormal crown formation, multiple missing teeth (14,17,21,22,26,27,31,34,37,41,44,46,47) (Figure 3). Cephalometric appraisal revealed mandibular prognathism due to overclosure and forward rotation. The hypodivergent facial pattern with restricted vertical development of the lower anterior face height, coupled with the forward rotation and prognathic mandible, compounded the skeletal Class III pattern. Both upper and lower lips were protruded, and the chin was prominent. A decrease in vertical dimension at occlusion (VDO) was observed. These cephalometric and anthropometric findings add on to the confirmation that the patient is affected by HED. Multiple treatment options were given to the patient, such as removable of partial dentures, full mouth rehabilitation with metal ceramic restoration, and orthodontic correction of spacing and posterior scissors-bite, followed by implant-retained replacement of missing teeth. The patient did not want a removable partial denture as she had already used and found it to be esthetically unappealing and functionally unstable, neither willing for orthodontic treatment nor implant placement as she was anxious about painful invasive and long-term procedures. Therefore the patient ultimately opted full mouth rehabilitation with metal ceramic restoration.

Figure 1: Extraoral examination showing depressed nasal bridge, decreased lower anterior facial height, prominent chin, slight deviation of the mandible towards the left, protuberant lips, frontal bossing, and retruded mid face

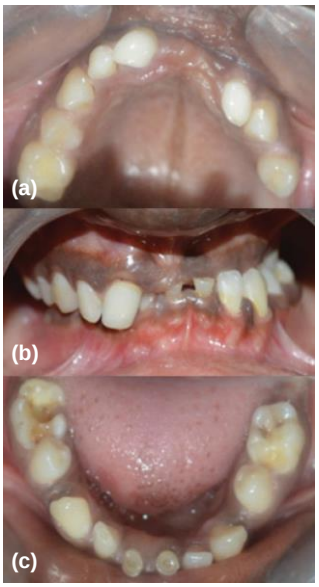

Figure 2: Initial oral examination. a: Frontal view. b: Maxillary occlusal view. c: Mandibular occlusal view

Treatment Course

Retained deciduous teeth 71, 81 and 85 were extracted. Alginate impressions (Tulip Alginate Impression Material; Cavex Holland BV, Haarlem, The Netherlands) of the maxillary and mandibular arches were made, and the diagnostic cast was poured using dental stone

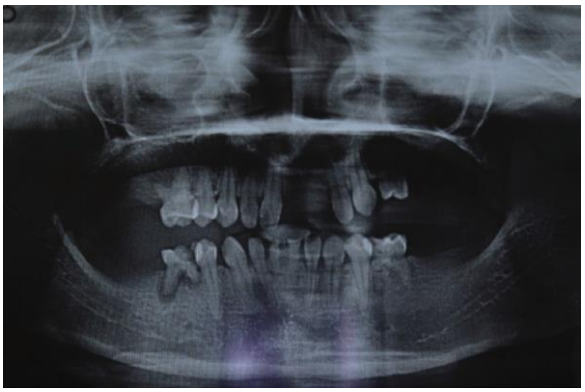

Figure 3: Preoperative OPG

Figure 4: Maxillary overlay removable partial denture to increase VDO

(Ultrarock; Kalabhai Karson Pvt. Ltd., Mumbai, India). The patient's casts were mounted on a semi-adjustable articulator (Hanau™ Modular Articulator; Whip Mix Corp., Louisville, USA) using a face-bow record, and an interocclusal record was made with the aid of a Lucia jig and polyvinyl siloxane occlusal registration material (EXABITE II; GC Corp., Tokyo, Japan). Vertical dimension at rest (VDR) was found to be 65 mm by using facial measurements after swallowing and relaxing and was verified using phonetics. VDO was found to be 58 mm using Niswonger's and Thomson's technique. Freeway space was 7 mm. The existing VDO was increased by 3 mm using the incisal guidance pin of the articulator to a new VDO of 61 mm. An ORPD was designed for the maxilla replacing 21, 22 and 26 (Figure 4). The appliance was designed in such a way that it replaces the missing teeth as well as increases the VDO to 61 mm. The adaptation of the patient to the increased VDO was evaluated for a period of 3 weeks using the ORPD. Centric relation was recorded using Lucia jig and wax-rim; diagnostic wax-up was performed (Figure 5). When wax-up was done according to the patient's occlusal plane, canting was observed since she had underdeveloped alveolar ridge on the left side (Figure 6). A midline shift towards the right side was also noted. Therefore establishing an esthetically appealing occlusal plane was required. The modified occlusal plane was established using the macroesthetic elements of smile design. The position of the six maxillary anterior teeth, their axial inclination, and incisal edge position were designed according to the macroesthetic principles. In

Figure 5: Luted final provisional restorations; lip lines during smile and at rest

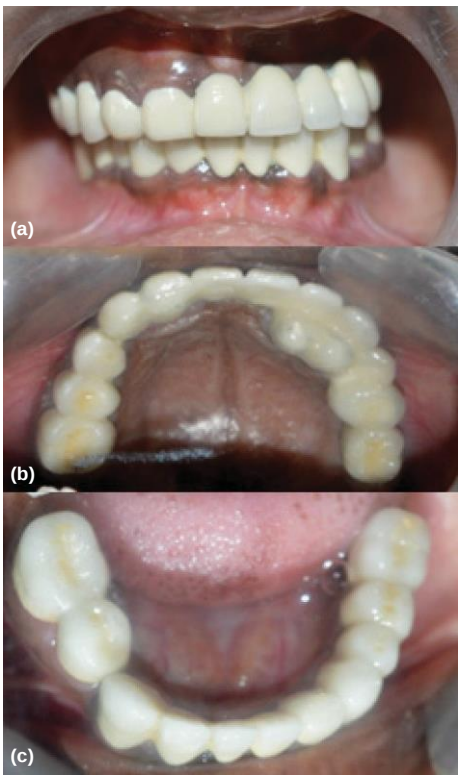

Figure 6: Definitive restorations were luted. a: frontal view, b: maxillary occlusal view, c: mandibular occlusal view

this case, the midline deviation was greatly reduced and it was made parallel to the facial midline of the patient. The proportions of the maxillary six anterior teeth were designed according to the principles of golden proportion in dentistry. The simultaneous arch technique was employed for rehabilitation. All existing teeth were prepared to accept full veneer metal ceramic restorations with equigingival chamfer margins. The maxillary anterior teeth were prepared in accordance with the modified wax-up. The interim prostheses were fabricated from autopolymerizing acrylic resin (ALIKE™; GC America, ALSIP, USA) on a cast using the indirect technique that was duplicated from the cast with the modified wax-up. The provisional restorations were cemented with temporary cement (FREEGENOL TEMPORARY PACK; GC Corp., Tokyo, Japan), and the patient's adaptation was monitored (Figure 6). Interim restorations were observed for 3 months, and used as a guide for definitive oral rehabilitation. After evaluation of 3 months, bite registration was performed using occlusal registration material (StoneBite; Dreve Dentamid GmbH, Unna, Germany) by first removing the provisionals on the right side of the arch while maintaining the provisionals on the left side of the arch and vice-versa. Definitive impressions were made with polyvinyl siloxane impression material (Extrude; Kerr Corp., Romulus, Germany). The definitive prosthesis was designed based on SDA concept. Each quadrant consisted of only one molar. First and third quadrant consisted of two premolars each and fourth quadrant consisted of one premolar. In the second quadrant, the lateral incisor and canine were replaced with premolars, in order to camouflage the anterior cross-bite on the left side. However, the facial surfaces of these teeth were designed as lateral incisor and canine only. This is done so that the bucco-palatal width of these teeth will be increased so that it can be brought into a normal occlusion with the opposing mandibular teeth. After 1 week, metal trial was done, and porcelain fused to metal restorations were fabricated utilizing

Figure 7: Postoperative OPG

Figure 8: Preoperative, during provisionalization showing cant, postoperative view of the patient

the duplicated provisional restoration casts, and cemented with resin-modified glass-ionomer cement (FujiCEM; GC America, Alsip, USA) (Figure 7). Oral hygiene instructions were given, and regular reviews were scheduled. Three reviews were completed in 6 months. At the sixth month an OPG was obtained (Figure 8). Improvement in the facial esthetics of the patient after treatment at various intervals was recorded. One-year follow-up of the patient was done during which the patient did not have any complaint regarding the prostheses.

Discussion

Dental defects associated with ED can cause severe esthetic and functional problems. The treatment objective of this case was to create a more favorable starting point for the prosthodontic phase of rehabilitation by improving the sagittal and vertical skeletal relationships and facial esthetics. This was accomplished by restoring the vertical dimension at occlusion with the help of ORPD. It was used as a diagnostic aid to judge adaptation to altered VDO. The waiting period to judge adaptability is between 3 and 5 weeks for the partial denture, and 2-6 months for the provisional prosthesis [7,8]. In this case, the patient was carefully monitored for 3 weeks to evaluate the adaptation to the removable partial denture. Also, the patient's adaptation to the provisional restoration was monitored for 3 months. The increase of VDO was measured by facial measurements and also by patient's factors like interocclusal rest space and speech. The patient exhibited canting of the occlusal plane since she had underdeveloped alveolar ridge on the left side which was confirmed with the help of lip lines at rest and during smile and by diagnostic wax-up following the patient's occlusal plane. The patient also had a midline shift towards the right. Principles of macroesthetics were used in this case to determine the position of the maxillary anterior teeth. Macroesthetics attempts to identify and analyze the relationships and ratios between anterior teeth and surrounding tissue landmarks. The starting point of any esthetic treatment plan must be the facial midline. Whenever possible, the midline between the maxillary central incisors should be coincidental with the facial midline. In cases in which this is not possible, the midline between the central incisors should be parallel to the facial midline [9-11]. In this case, though the dental and facial midline could not be made to coincide, the deviation was reduced and the midlines were made parallel to each other. The long axis of the anterior teeth in an esthetic smile follows a progression as the teeth move away from the midline. In this case they were made to tip medially, that is, the long axis of the tooth tips toward the midline. When the maxillary anterior teeth tip medially, the overall esthetic impact is one of a harmonious relationship with the framing of the lower lip [12,13]. In this case, the incisal plane was made to be consonant with the curvature of the lower lip during a smile. In this case, the standardized esthetic golden proportion was used to determine the width of the six maxillary anterior teeth [14]. Since the patient had most of the posterior teeth missing and was not willing for implant surgery, the SDA concept was opted. A shortened dental arch (SDA) is defined as having an intact anterior region but a reduced number of occluding pairs of posterior teeth [15]. The outcome of SDA therapy was found to be acceptable in approximately 82% of patients in terms of the oral function, comfort, and well-being [15]. Full mouth rehabilitation in this patient using the SDA concept showed long-term occlusal stability and improved masticatory function.

Therefore, a multidisciplinary team approach is necessary for optimal dental management of the HED patients. The treatment not only improved the masticatory function but also enhanced esthetics, boosted the self-esteem, and helped the patient to develop socially.

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