

Prosthodontic Management of Ectodermal Dysplasia: 2 Case Reports

Hajer Zidani*, Hanen Boukhris, Najla Taktak, Narjes Hassen, Abdellatif Boughzela

Farhat Hached Hospital Sousse, Faculty of Dentistry, Tunisia

DOI: [10.36347/sjds.2019.v06i10.003](https://doi.org/10.36347/sjds.2019.v06i10.003)

| Received: 13.10.2019 | Accepted: 22.10.2019 | Published: 30.10.2019

*Corresponding author: Hajer Zidani

Abstract

Case Report

The hereditary condition known as ectodermal dysplasia is characterized by the absence or defect of 2 or more ectodermally derived structures, including teeth, skin, nails, hair and sweat glands. The dental characteristics of this syndrome include anodontia or hypodontia of the primary and/or permanent teeth, hypoplastic conical teeth, and Underdevelopment of the alveolar ridges. The options for a definitive treatment plan may include fixed, removable, or implant-supported prostheses, singly or in combination. This article aims to shed light on the prosthetic rehabilitation of two brothers presenting with hypohidrotic ectodermal dysplasia.

Keywords: Ectodermal dysplasia, hypodontia, removable partial denture, overdenture, fixed prosthesis.

Copyright © 2019: This is an open-access article distributed under the terms of the Creative Commons Attribution license which permits unrestricted use, distribution, and reproduction in any medium for non-commercial use (NonCommercial, or CC-BY-NC) provided the original author and source are credited.

INTRODUCTION

Ectodermal dysplasia (ED) is a rare heterogeneous group of inherited disorders in which two or more structures derived from embryonic ectoderm are affected, including teeth, skin, nails, hair and sweat glands [1].

The disease is broadly classified into hypohidrotic and hidrotic type of ED. The most frequently reported ED syndrome is X-linked hypohidrotic dysplasia (HED), also known as Christ-Siemens-Touraine syndrome, in which sweat glands are either absent or significantly reduced in number [2]. It mainly affects males, whereas females show only minor defects [3]. The hidrotic ED (Clouston syndrome) usually spares the sweat glands, can affect the teeth, hair, nail and is inherited as an autosomal trait [4].

Hypodontia affects around 80% of ED patients and leads to atrophic alveolar ridges and reduced occlusal vertical dimension. Although the prosthodontic treatment is still a big challenge for the dental team, it is of utmost importance in the management of these patients, as it repairs function, aesthetics and psychological issues, and improves their quality of life [5, 6]. A multidisciplinary approach is usually required to provide optimal care for the patients [7]. This article aims to shed light on the prosthetic rehabilitation of two brothers presenting with HED.

CASE REPORT

Two brothers aged 17 (Case 1) and 18 years old (Case 2) presented to the department of prosthodontics, Farhat Hached Teaching Hospital, Sousse complaining of lack of teeth, inability to eat properly and poor aesthetics. The parents denied a consanguineous marriage or having a family member with similar features as their children. The boys exhibited the classical features of HED: hypodontia, hypohidrosis, and hypotrichosis. They were intolerable to hot climate with a history of reduced sweating.

Extraoral examination showed fine sparse hair, slight eyelashes and eyebrows, prominent forehead, saddle nose, depressed nasal bridge and dry and sensitive skin (Fig 1, 2).

Intraoral examination in case 1 revealed hypodontia, with only permanent central incisors and first molars in the upper arch and permanent canines and first molars in the lower one (Fig 3, 4). Retained second primary molars were identified (Fig 5). In case 2, analysis of the dental arches showed the presence of six permanent maxillary teeth (central incisors, canines, first molar and rotated second bicuspid) and the absence of mandibular incisors and first premolars (Fig 16, 17). In both cases, a central diastema was observed. Maxillary central incisors and canines were conical in shape and the patients exhibited aplasia of alveolar bone in the edentulous area.

Based on the history, clinical features, and examination, the brothers were diagnosed as a case of HED.



Fig-1: Frontal view of case 1



Fig-2: Frontal view of case 2

Case 1

The treatment plan included removable partial dentures (RPD) with tooth retained overdenture using 11, 21, 33

And 43 as abutments for attachments and single metallic crowns on molars.

Maxillary and mandibular impressions were taken using alginate and Casts were mounted on articulator for diagnostic study. The diagnostic wax-up and the RPD design were completed. A putty matrix of the completed diagnostic wax-up was made to assess the existing space for the attachment.

After endodontic treatment of the anterior teeth, the abutments were prepared for short copings and the post space preparation was done. Maxillary and mandibular molars were also prepared to receive metal

crowns. Impressions were taken with polyvinyl siloxane (PVS) (Fig 6), jaws relation record was obtained using wax rims oriented to established vertical dimension occlusion.

Prefabricated plastic patterns of matrix were attached to the waxed up copings on abutments using parallelometer and were subsequently casted (Fig 7). Precision attachments with copings and metal crowns were then tried and pick up impressions were made with special trays and polyether impression material (3M ESPE Impregum soft) (Fig 8, 9). Wax patterns were prepared for metal denture bases on refractory cast and casted in cobalt chromium (Fig 10). Frameworks were checked for fit (Fig 11). After recording maxillomandibular relation, try-in was done and dentures were fabricated. The RPD were delivered after cementation of the copings and crowns (Fig 12, 13,14,15).



Fig-3: Pre-operative maxillary view



Fig-4: Pre-operative mandibular view



Fig-5: Pre-treatment panoramic radiograph



Fig-6: Conventional dental impression

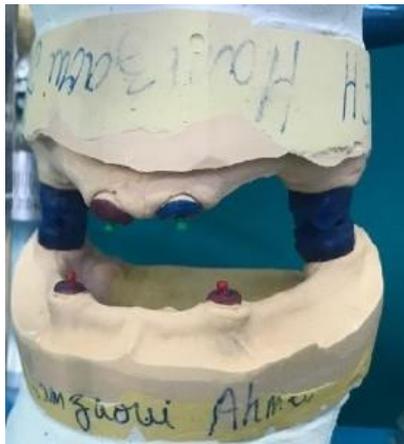


Fig-7: Wax Pattern with Patrices attached

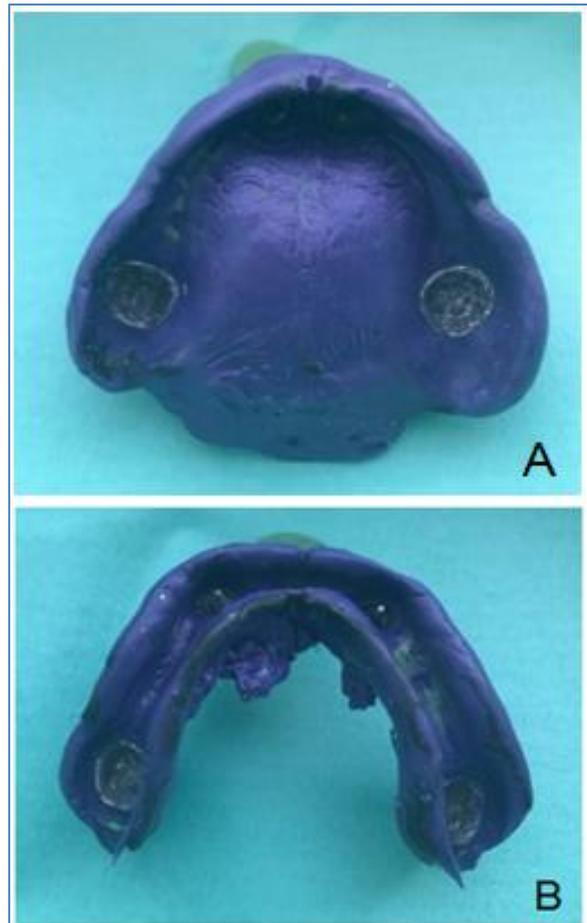


Fig-9: Pick up impression. A: Maxilla, B: Mandible

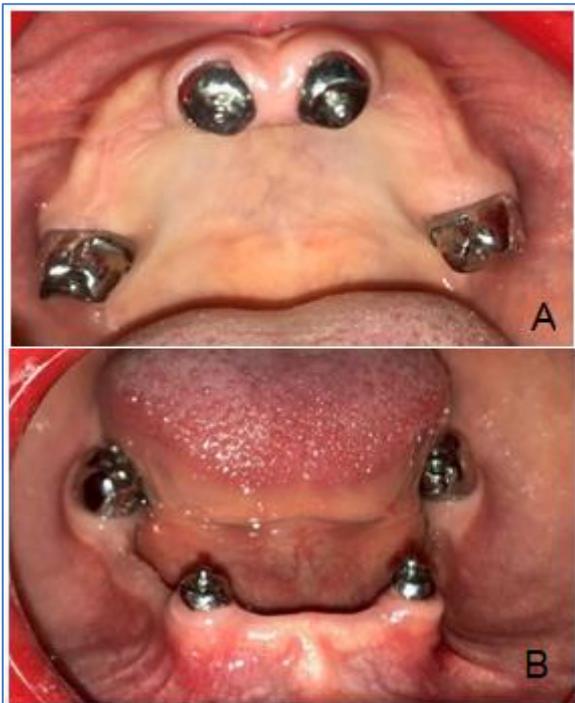


Fig-8: Copings and metallic crowns checked for fit. A: Maxilla, B: Mandible



Fig-10: Fabrication of mandibular framework



Fig-11: Framework trial



Fig-12: Maxillary occlusal view of final prosthesis



Fig-13: Mandibular occlusal view of final prosthesis



Fig-14: Lateral view of final prostheses



Fig-15: Postoperative frontal view

Case 2

In the maxilla, the proposed treatment plan included first, orthodontic treatment to close the diastema between

The maxillary central incisors, a 6-unit fixed Porcelain fused to metal (PFM) bridge from canine to canine, with two pontics in the area of 12 and 22 and a single PFM crown on 26, with a cast partial denture in the posterior edentulous region. In the mandible, we opted for an 11-unit PFM bridge extending from 45 to 36 with seven pontics replacing 35, 34, 32, 31, 41, 42 and 44.

While designing the maxillary partial denture, it turned out that a clasp would have to be placed on the canines. But finally 2 custom precision attachments were preferred for aesthetic reasons without compromising the stability and retention of the denture.

Preliminary maxillary and mandibular impressions were made using alginate and the diagnostic casts were mounted on semi-adjustable articulator. A putty matrix of the completed diagnostic wax-up was made to evaluate the existing space for the extracoronal attachment. After extraction of tooth 15 and diastema closure, teeth were prepared for PFM Bridge and new impressions were made with PVS (Fig 18, 19).

Since the loss of occlusal vertical dimension is a common oral manifestation of ectodermal dysplasia, a correct vertical dimension is paramount to make enough space for the teeth arrangement. The patient's vertical dimension was established and interocclusal records were made.

Wax patterns were prepared for PFM crowns and the patrices were added to the distal aspects of canines using a dental surveyor (Fig 20). The maxillary and mandibular crowns were fabricated and veneered. All of the fixed components were tried and the lower PFM bridge was cemented (Fig 21, 22).

A maxillary pick-up impression was made with polyether impression material (3M ESPE Impregum soft) (Fig 23). The matrices of the attachments were connected to the patrices on the refractory cast. The framework of the RPD was fabricated and tried in the patient's mouth.

Occlusal relations were recorded then a try-in was done. The RPD was delivered after acrylisation and crowns cementation (Fig 24, 25, 26, 27).



Fig-16: Pre-operative mandibular view



Fig-17: Panoramic radiograph



Fig-18: Orthodontic treatment in progress



Fig-19: Frontal view after orthodontic



Fig-20: PFM crowns with patrices attached



Fig-21: PFM crowns checked for fit



Fig-22: Final mandibular prosthesis in situ



Fig-23: Maxillary pick up impression



Fig-24: Fabrication of maxillary prosthesis



Fig-25: Maxillary view of final prosthesis



Fig-26: Lateral view of definitive prostheses



Fig-27: Patient smile with the final prostheses in place

DISCUSSION

For HED patients, partial or complete absence of teeth is one of the most frequently occurring oral symptoms [8]. This condition is a major cause of frustration that impacts the intellectual and psychological maturity of the patient who may suffer from low self-esteem, insecurity, and depression due to his unusual physical appearance and lack of social acceptance [9, 10]. Hence, prosthodontic rehabilitation is fundamental in the management of ED patient, as it provides a functional and esthetic solution that improves the patient's quality of life [11, 5].

The prosthetic treatment should be performed as soon as possible in order to control the vertical dimension and avoid possible resorption and atrophy of the alveolar ridges, which can be severely affected by the total or partial lack of teeth [11].

There is no definitive time to begin dental treatment, but Till and Marques recommend that an initial prosthesis should be provided before school age.

Early prosthetic rehabilitation provides normal appearance, satisfactory masticatory and normal temporomandibular joint functions [2, 8]. According to Li *et al.* it is recommended that prosthodontic rehabilitation for HED patients should be commenced when the patient is 2 to 3 years old [8].

Many treatment options have been described in the literature and may include single crowns, fixed partial dentures, complete dentures, RPD, overdentures, and implant supported denture. These approaches may be used either singly or in combination to provide optimal results [5, 12]. Exclusively fixed prostheses are seldom used as the number of teeth present is usually minimal [13]. However, a fixed solution is closer to the normality and allows the patient to integrate more easily into the society [14].

In the present cases, the patients were skeletally mature and ready for definitive restorative treatment. The final decision was in favour of RPD which is considered to be the most common treatment plan. Nevertheless, depending on the remaining available alveolar bone and the pattern of missing teeth, the implant retained prosthesis have become the treatment of choice with high success rate [15, 16, 9]; Guckes *et al.* evaluated the survival of implants placed in patients affected by ED with mean age of 20.5 years (8-68 years) and showed survival rates of 91% in the mandible after 24 months, while the success in the maxilla was 71% [17, 18]. Due to financial issues of our patients, implant retained prostheses could not be proposed.

Pretreatment extraoral aesthetic evaluation of the patient is very important. The facial appearance of affected individuals is frequently characterised by improper lip support. Therefore, prosthetic rehabilitation with overdentures and RPDs may be the treatment of choice such as in case 1 because the presence of the anterior denture flange can achieve optimal level of aesthetics [5, 10]. Furthermore, the atypical conical anterior teeth whose shape may jeopardise the stability of the RPD could be better used as abutments for an overdenture [15]. The retention can be augmented by various attachments anchored to the available teeth [2]. All RPD with attachments, especially the extracoronal type (case 2), are considered more efficient in providing retention and restoring function and aesthetics [19].

In order to achieve optimal level of aesthetics and function, it was important to complete orthodontic treatment to align the teeth into acceptable position prior to prosthetic rehabilitation (case 2) [2, 17].

The treatment decision should be carried out on an individual basis and based on each patient's needs and also on the economic possibilities [11, 5]. The final

goal is to find a cost-effective treatment, minimise invasive risks and improve the life quality of the patient [8].

In such cases, multidisciplinary team is generally advocated to be the most appropriate approach, involving oral medicine, oral surgery, periodontics, orthodontics and prosthodontics [17, 20].

CONCLUSION

Management of oral manifestations associated with ED could be challenging. It requires a broad knowledge base to handle the special issues associated with the treatment [2]. Several decision factors should be taken into account by the dental team including patient's needs, wishes, the willingness to undergo minor or major treatment and also the economic possibilities [21].

For this reason, a multidisciplinary team approach is usually recommended for optimal dental management of this condition [2].

REFERENCES

- Anuroopa A, Abdulla J, Lovely M. Oral rehabilitation of a young patient with hypohidrotic ectodermal dysplasia: A clinical report. *Contemp Clin Dent*. 2012; 3(Suppl 1):33-6.
- Pigno MA, Blackman RB, Cronin RJ, Cavazos E: Prosthodontic management of ectodermal dysplasia: a review of the literature. *J Prosthet Dent*. 76:541-45,1996.
- Pozo-Molina G, Reyes-Real J, Mendoza-Ramos MI, Villalobos-Molina R, Garrido-Guerrero E. Novel missense mutation in the EDA1 gene identified in a family with hypohidrotic ectodermal dysplasia. *Int J Dermatol*.2015; 54: 790-794.
- Shigli A, Reddy RP, Huger SM, Deshpande D. Hypohidrotic ectodermal dysplasia. A unique approach to esthetic and prosthetic management: A case report. *J Indian Soc Pedod Prev Dent*. 2005; 23:31-4.
- Maroulakos G, Artopoulou II, Angelopoulou MV, Emmanouil D. Removable partial dentures vs overdentures in children with ectodermal dysplasia: two case reports. *Eur Arch Paediatr Dent*. 2016 Jun;17(3):205-10.
- Hickey AJ, Salter M. Prosthodontic and psychological factors in treating patients with congenital and craniofacial defects. *J Prosthet Dent*. 2006; 95:392-396.
- Artopoulou I, Martin JW, Suchko GD. Prosthodontic rehabilitation of a 10-year-old ectodermal dysplasia patient using provisional implants. *Pediatr Dent*. 2009; 31:52-57.
- Li S, Xiao X, Wu R, Zeng B, Yu D and Zhao W. Early Prosthetic Treatment of Children with Hypohidrotic Ectodermal dysplasia: Six Case Reports. *Int J Oral Dent Health*. 2017, 3:039
- Pinto AS, Conceicao Pinto ME, Melo do Val C, Costa Oliveira L, Costa de Aquino C, Vasconcelos DF. Prosthetic Management of a Child with Hypohidrotic Ectodermal Dysplasia: 6-Year Follow-Up. *Case Rep Dent*. 2016; 2164340.
- Meshram GG, Kaur N, Hura KS. A case report of Hypohidrotic Ectodermal Dysplasia: A mini review with latest updates. *J Family Med Prim Care*. 2018,7:264-6.
- Vallejo AP, Monje ELA, Garcia MG, Fernandez MM, Buylla FBMA. Treatment with removable prosthesis in hypohidrotic ectodermal dysplasia. A clinical case. *Med Oral Patol Oral Cir Bucal*. 2008 1;13:E119-E123.
- Pavarina AC, Machado AL, Vergani CE, Gianpaolo ET. Overlay removable partial dentures for a patient with ectodermal dysplasia: a clinical report. *J Prosthet Dent*. 2001;86:574-7.
- Mittal M, Srivastava D, Kumar A, and Sharma P. Dental management of hypohidrotic ectodermal dysplasia: A report of two cases. *Contemp Clin Dent*. 2015 Jul-Sep; 6(3): 414-417 20.
- Chaiban RO, Chaiban W. Ectodermal dysplasia: Dental management and benefits, a case report. *European Journal of Paediatric Dentistry*. 2011;12(4):282-4.
- Hekmatfar S, Jafari K, Meshki R, Badakhsh S: Dental management of ectodermal dysplasia: two clinical case reports. *J Dent Res Dent Clin Dent Prospects*. 2012, 6: 108-112.
- Rashedi B. Prosthodontic treatment with fixed prosthesis for a patient with ectodermal dysplasia: a clinical report. *J Prosthodont*. 2003;12:198-201.
- Grover R, Mehra M. Prosthodontic Management of Children with Ectodermal Dysplasia: Review of Literature. *Dentistry*. 2015; 5: 340.
- Guckes AD, Scurria MS, King TS, McCarthy GR, Brahim JS. Prospective clinical trial of denture implants in person with ectodermal dysplasia. *J Prosthet Dent*. 2002; 88: 21-25.
- Shetty NB, Shetty S, Nagaraj E, Shetty O, and D'souza R. *J Clin Diagn Res*. 2014 Jan; 8(1): 268-270.
- Gupta A, Karki S, Thapa D, Shrestha RM: Multidisciplinary Rehabilitation of Ectodermal Dysplasia: A Case Report. *Orthodontic Journal of Nepal*.2016; 6(2).
- Suja J, George JC, Jose J and Alex KV. Multidisciplinary management of hypohidrotic ectodermal dysplasia – a case report. *Clin Case Rep*. 2015;3(5):280-286.