



Removable prosthetic treatment of an ectodermal dysplasia patient: A case report

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ABSTRACT

Ectodermal dysplasia is an inherited disorder with partial or complete anodontia. Early dental treatments are mandatory for improving child mastication, communication, and esthetic. There are various treatment approaches such as implant-retained prosthesis, fixed and removable denture which should be selected according to remained teeth, oral and facial parameters. This clinical study describes the prosthetic treatment of a child with ectodermal dysplasia with combination of complete overdenture and partial removable denture.

Keywords: Ectodermal dysplasia; Hypodontia; Children; Removable prosthodontic denture; Overdenture.

Introduction

Ectodermal dysplasia (ED) is an inherited disorder that affects the structures of the fetal ectoderm, such as skin, nail, hair, glands, and teeth [1]. Clinical manifestation in literature shows 91% has trichodysplasia (abnormal hair), 80% has tooth agenesis, 75% has onychodysplasia (abnormal nails), and 42% has dyshidrosis (abnormal sweat glands) [2]. Two common forms of ED are the Hidrotic (Clouston's syndrome) and Hypohidrotic (Christ-Siemens-Touraine syndrome). Inheritance patterns showed for ED are Sex-linked, autosomal dominant,

and autosomal recessive [3]. X-linked form is the most common form of the disease with 95% abundance. Oral demonstrations include partial or complete anodontia, abnormal shape of the teeth, enamel hypoplasia, reduced asymmetric alveolar ridge height, maxillary retrusion, and high palatal arch [4]. Dental characteristics in ED are peg-shaped or conical front teeth, which those teeth cannot be distinguished from incisors.

Young cases with hypohidrotic ED have problems with mastication, communication, esthetic, and psychology that cause a decrease in quality of life [5], namely ectodysplasin A1 (EDA-A1). Many prosthodontic treatments depending on the age and growth progress of the stomatognathic system and treatment approaches include single crown, fixed partial denture (FPD), complete denture (CD), removable partial denture (RPD), overdenture (OD), and implant-retained prosthesis. Financial restrictions and other preferences may limit patients from choosing the most favorable treatment [6]. A removable prosthesis is the most common treatment for young patients with ED [7]. RPD can significantly improve function and esthetics and also allow adjustments or remade at intervals for permanent tooth eruptions [8]. Implant-supported denture as the ideal reconstruction modality can be used for adolescents over 12 years. The main problem in implant restoration treatment is insufficient bone; if bone atrophy progresses in these already alveolar deficient patients, implant placement may not be possible without bone grafting. Early prosthetic treatment generally recommends from the age of 5 years. Concerning child cooperation, dentures can also fabricate as early as 3 to 4 years of age. It seems necessary to follow up and periodic recall due to developmental changes in the oral cavity in young patients [9]. This clinical report describes the features and prosthetic treatment of a patient suffering from ectodermal dysplasia.

Case Study

A 9-year-old male child diagnosed with hidrotic ectodermal dysplasia referred to the Prosthodontics Department of Tehran University of Medical Sciences. He was a student in a family with a good socio-economic situation. The principal chief complaint of the patient was the generalized irritation of soft tissue, some pain spots under existing removable denture, long-lasting inflammation, and wound in the corner of lips and inconvenience. The medical history of the patient was clear, with no taking medication. The diagnosis of ectodermal dysplasia confirmed due to his features as following: soft and hairless skin, lack of sweat, sparse eyelash and eyebrow, frontal bossing, saddle nose, low set ears, and hypodontia [3]. Previous dental treatments were reported as following:

The first primary maxillary incisors underwent root canal therapy and composite operative treatment, and they shed at seven years old, and the primary maxillary second molar on the left side was treated by pulpotomy and stainless steel crown. At five years old, for the

first time, he underwent dental prosthetic treatment. That time he had just six teeth: two primary maxillary central incisors, two primary maxillary second molars, and two primary mandibular second molars. He received his first dentures at five, which were acrylic overdentures that cover the primary teeth. After 16 months (at six years and four months), due to alveolar growth, lack of retention, and patient discomfort, the second removable dentures were fabricated. The second prosthesis had an opening on top of the upper and lower molars. After a while, primary maxillary incisors shed, and the denture became loose. The third overdentures fabricated 14 months later (at seven years and six months). The third prosthesis was complete overdentures with full coverage of remaining teeth.

Extraoral examination revealed normal lymph nodes, temporomandibular joints, and masticatory muscles. The lips were competent with angular cheilitis due to vertical dimension (VD) reduction. The lip support was insufficient. In the intraoral examination, he had two primary maxillary second molars, two primary mandibular second molars, one permanent right first molar and one erupted maxillary permanent central incisor at left, which was a conical shape and without rotation, and the right maxillary permanent central incisor was impacted. The maxillary and the mandibular arches had no abnormal spacing; also, the alveolar ridges were underdeveloped, sharp, and thin. Panoramic radiography confirmed clinical data [3]. According to the prosthodontic diagnostic index, maxillary and mandibular arches were class IV because of restoring the vertical dimension and replacing multiple tooth loss. The proposed treatment plan was to fabricate removable acrylic prosthesis. Decision about the partial prosthesis or overdenture could be made after VD analysis and occlusal plan determination [6].

Primary impressions made with irreversible hydrocolloid (Alginoplast, Heraeus Kulzer GmbH, Hanau, Germany) and diagnostic casts prepared with type III dental stone [3]. Custom maxillary and mandibular acrylic trays were fabricated with uniform space and three tissue stops for stability. Some holes were prepared in all aspects for impression material retention. Trays were border molded with modeling impression compound (Impression compound, Kerr, Orange, CA, USA). The final impression of the maxillary and mandibular arches were made with light silicone impression material (Spidex, Coltene AG, Altstätten, Switzerland) and final casts were poured, also wax occlusal rims were fabricated. Wax rims were adjusted based on

providing sufficient lip support, the height of maxillary occlusal plane was determined according esthetic consideration and the mandibular occlusal plane was mapped out into a line crossing from lower lip corner to two third of the retromolar pad for supplying the maximum stability. Vertical dimension was determined according to facial proportions, patient phonetic, and providing 2mm free-way space in rest position. Centric relation was recorded with GC compound bite registration material (GC Corporation, Tokyo, Japan), Master casts then were mounted on a average value articulator. The determined occlusal plane indicated the fabrication of partial acrylic denture for the mandible and complete overdenture for the maxilla. The teeth were set up according to occlusal plan. In the posterior part of the maxilla due to presence of primary second molars and lack of enough vertical space for teeth set up over them, the first maxillary molar teeth were waxed up. After teeth try in and patient confirmation, the dentures were processed with heat cure acrylic by compression mold method in long curing cycle. At the delivery appointment, the dentures fitness were checked, the occlusion was adjusted and hygiene and maintenance points were instructed to patient and his parents. Patient was followed up after 1 day, 1 week and 2 weeks. He was satisfied with new set of dentures and his appearance, phonetic and self-confidence were improved. The difference between dimension of new and previous dentures were shown to patient parents and they became aware of need for periodic recalls for denture adjustment due to eruption of permanent teeth and alveolar growth.

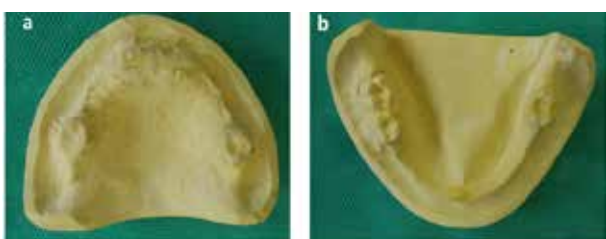


Fig 1. Primary dental casts a: upper jaw, b: lower jaw.



Fig 2. Border molded special trays a: upper jaw, b: lower jaw.

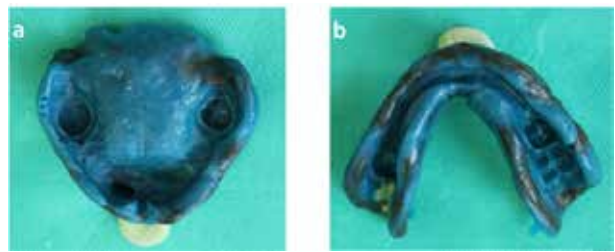


Fig 3. Final impression making a: upper jaw, b: lower jaw.

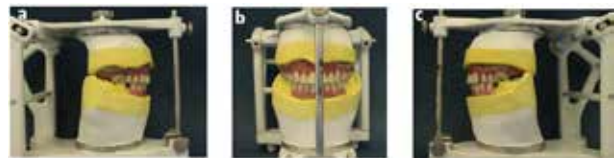


Fig 4. Teeth set up a: right lateral view, b: frontal view, c: left lateral view.

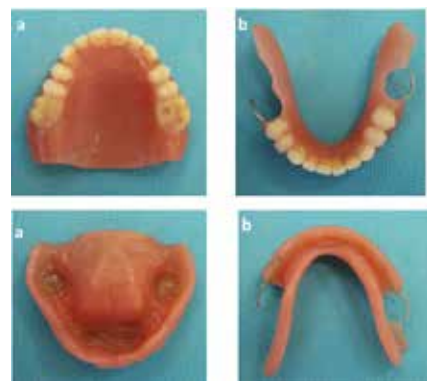


Fig 5. Removable dentures ready for delivery a: upper complete overdenture, b: lower partial denture.



Fig 6. The difference between dimension of new and previous dentures a: external surfaces, b: intaglio surfaces.

Discussion

An early prosthodontic intervention helps children to adapt to the prosthesis and promote normal appearance, mastication, and swallowing [10]. ED treatment is different and generally depends on the child's age, the amount of malformation of teeth, the growth and development of the stomatognathic system, and the patient's collaboration [11,12]. Removable prosthodontics is the most common modality used for den-

tal treatment of ectodermal dysplasia because of many advantages such as the prevention of alveolar ridge resorption [7,13]. The importance of alveolar ridge preservation in ED patients is obvious and the ridge should supply the denture support from early childhood. Complete dentures are an option for patients with complete anodontia. Partial removable denture is considered for patient with reasonable numbers of abutment teeth with adequate lip support and stable occlusion in terms of centric relation and vertical dimension [7]. The partial removable prosthesis is usually fabricated with acryl and wire retentive clasps and it is avoided to use metal framework in order not to limit the teeth movement and alveolar growth. The other modality is overdentures, which provide ideal occlusion and preserve existing dentition [14]. It is proper for partially dentulous patients with few numbers of malform and malposed abutments, unstable occlusion and need of restoring the CR and VD.

In this case, the patients came with defective prostheses that revealed the need of fabrication of new sets of removable denture to compensate the jaw growth, VD and CR. After diagnostic work up and occlusal plan determination, the treatment of choice was upper jaw overdenture and lower jaw partial denture. As obtaining adequate resistance and stability of the prosthesis in ED patients is difficult due to under-development of maxillary tuberosity and alveolar ridges, remaining teeth act as the partial prosthesis and overdenture abutments provide retention, support and stability [15]. Improvement in masticatory muscles in our patient had occurred immediately after prosthesis delivery. Prosthetic treatment resulted in better nourishment and speech. Sagittal and vertical position of the mandible became normal. The patient was satisfied and the point to preserve this comfort regular visit is recommended. Schnalb et al. reported the interval of 3-6 months for long-term follow-up [16]. The aim of periodic follow up is to check alveolar changes, occlusion adjustment, need for relieving around teeth and give the required growth space. ED patients have to keep good oral hygiene. Topical fluoride in daily dose is advised as a prophylaxis [17]. which include dental ageneses, and describe the oral habilitation. Hypohidrotic ectodermal dysplasia patients had a clinical examination and underwent radiographic and Steiner's analyses and a respiratory capability test before assessment and treatment. Fifteen patients (eight males and seven females, aged 5-45 years new removable prosthesis should be made every 1.5-2.5 years [16]. The use of digital denture is reported in management of ED [1].

Improvements in digital dentistry have simplified the design and manufacture of dental prostheses [18,19]. In the last five years, computer-engineered complete denture fabrication has incorporated significant advancements in materials and technology [20,21,22]. The advantages of digital dentures are the virtual block-out, trial dentures the patient can take home, better material properties offered by monolithic bases, and information archived for future use which is recommended for ED patients with need of making new sets of prosthesis every 2 years 1.

Treatment progression can utilize endosseous implants as an alternative treatment [14]. Age affects treatment planning in growing patients but does not seem to affect the establishing ability of osseointegration [13]. In most situations, it is desirable to postpone endosseous implants, especially in partially edentulous patients in the growth phase, due to unpredictable effects of jaw growth and development on endosseous implant site and jaw relations but early implant placement reported in edentulous patients as young as 5 to 6 years of age [9,23]. Agreeable time for implant placement could be associated with the status of adolescent growth observed on the wrist and head radiographs [10]. Additionally, there are mini-implants prostheses that are suitably used for prosthetic retention in pre-adolescent patients, but they needed long-term follow-ups to test the efficacy in oral rehabilitation of children with ED [24,16].

Conclusion

Treatment of the pediatric patients with ED is a challenging task in terms of behavior management, restorative and prosthodontic techniques. Treatment plan should be determined in each case based on the remaining teeth, oral and facialsituation. Regular follow-up is recommended.

Conflict of Interest

There is no conflict of interest to declare.

References

- [1] Punj A, Kattadiyil MT. Management of ectodermal dysplasia with tooth-supported computer-engineered complete overdentures: A clinical report. *J Prosthet Dent.* 2019; 121(2):195-199. doi:10.1016/j.prosdent. 2018.03.011.
- [2] Gupta C, Verma M, Gupta R, Gill S. Telescopic overdenture for oral rehabilitation of ectoder-

- mal dysplasia patient. *Contemp Clin Dent.* 2015; 6:S258-S261. doi:10.4103/0976-237X.166821.
- [3] Soares L, Vilanova R, Sánchez-ayala A, Ribeiro GR, Campos CH, Farias-neto A. Case Report Ectodermal Dysplasia. *Case Rep Dent.* 2015; 2015:1-4doi.org/10.1155/2015/714963.
- [4] Alnuaimi R, Mansoor M. Prosthetic rehabilitation with fixed prosthesis of a 5-year-old child with Hypohidrotic Ectodermal Dysplasia and Oligodontia: A case report. *J Med Case Rep.* 2019; 13(1):1-6. doi:10.1186/s13256-019-2268-4.
- [5] Bashyam MD, Chaudhary AK, Reddy EC, et al. A founder ectodysplasin A receptor (EDAR) mutation results in a high frequency of the autosomal recessive form of hypohidrotic ectodermal dysplasia in India. *Br J Dermatol.* 2012; 166(4):819-829. doi:10.1111/j.1365-2133.2011.10707x.
- [6] Pavarina AC, Machado AL, Vergani CE, Giampaolo ET. Overlay removable partial dentures for a patient with ectodermal dysplasia: A clinical report. *J Prosthet Dent.* 2001; 86(6):574-577. doi:10.1067/mpr.2001.119981.
- [7] Maroulakos G, Artopoulou II, Angelopoulou M V., Emmanouil D. Removable partial dentures vs overdentures in children with ectodermal dysplasia: two case reports. *Eur Arch Paediatr Dent.* 2016; 17(3):205-210. doi:10.1007/s40368-015-0212-x.
- [8] Snawder KD. Considerations in dental treatment of children with ectodermal dysplasia. *J Am Dent Assoc.* 1976; 93(6):1177-1179. doi:10.14219/jada.archive.1976.0286.
- [9] NaBadalung DP. Prosthodontic rehabilitation of an anhidrotic ectodermal dysplasia patient: a clinical report. *J Prosthet Dent.* 1999;81(5):499-502. doi:10.1016/S0022-3913(99)70200-3.
- [10] Jain N, Naitam D, Wadkar A, Nemane A, Katoch S, Dewangan A. Prosthodontic Rehabilitation of Hereditary Ectodermal Dysplasia in an 11-Year-Old Patient with Flexible Denture: A Case Report. *Case Rep Dent.* 2012; 2012:1-5. doi:10.1155/2012/489769.
- [11] Mohajerfar M, Zarrati S, Memari Y. Report of a Case of Hypodontia Due To Ectodermal Dysplasia: Early Rehabilitation With Overdenture. *J Dent Sch Shahid Beheshti Univ Med Sci.* 2016; 34(4):274-281.https://www.sid.ir/en/VEWSSID/J_pdf/82520160408.pdf.
- [12] Khazaie R, Berroeta EM, Borrero C, Torbati A, Chee W. Five-year follow-up treatment of an ectodermal dysplasia patient with maxillary anterior composites and mandibular denture: A clinical report. *J Prosthodont.* 2010; 19(4):294-298. doi:10.1111/j.1532-849X.2010.00579x.
- [13] Bhargava A, Sharma A, Popli S, Bhargava R. Prosthodontic management of a child with ectodermal dysplasia: A case report. *J Indian Prosthodont Soc.* 2010; 10(2):137-140. doi:10.1007/s13191-010-0026-9
- [14] Pigno MA, Blackman RB, Cronin RJ, Cavazos E. Prosthodontic management of ectodermal dysplasia: A review of the literature. *J Prosthet Dent.* 1996; 76(5):541-545. doi:10.1016/S0022-3913(96)90015-3.
- [15] Shaw RM. Prosthetic management of hypohidrotic ectodermal dysplasia with anodontia. Case report. *Aust Dent J.* 1990; 35(2):113-116. doi:10.1111/j.1834-7819.1990.tb05873x.
- [16] Toomarian L, Ardakani MRT, Ramezani J, Adli AR, Tabari ZA. Using implants for prosthodontic rehabilitation of a 4-year-old with ectodermal dysplasia. *Gen Dent.* 2014; 62(5):e1-e5.https://www.researchgate.net/profile/Zahra_Tabari/publication/265343015_Using_implants_for_prosthodontic_rehabilitation_of_a_4-year-old_with_ectodermal_dysplasia/links/555c3bf908aec5ac2232ae50/Using-implants-for-prosthodontic-rehabilitation-of-a-4-year-old-with-ectodermal-dysplasia.pdf.
- [17] Yavuz I, Baskan Z, Ulku R, et al. Ectodermal dysplasia: Retrospective study of fifteen cases. *Arch Med Res.* 2006; 37(3):403-409. doi:10.1016/j.arcmed.2005.06.016.
- [18] AlHelal A, Goodacre BJ, Kattadiyil MT, Swamiclass R. Errors associated with digital preview of computer-engineered complete dentures and guidelines for reducing them: A technique article. *J Prosthet Dent.* 2018; 119(1):17-25. doi:10.1016/j.prosdent. 2017.02.023.
- [19] Goodacre BJ, Goodacre CJ, Baba NZ, Kattadiyil MT. Comparison of denture base adaptation between CAD-CAM and conventional fabrication techniques. *J Prosthet Dent.* 2016; 116(2):249-256. doi:10.1016/j.prosdent. 2016.02.017.
- [20] Kattadiyil MT, AlHelal A, Goodacre BJ. Clinical

- complications and quality assessments with computer-engineered complete dentures: A systematic review. *J Prosthet Dent.* 2017; 117(6):721-728. doi:10.1016/j.prosdent. 2016.12.006.
- [21] Kattadiyil MT, AlHelal A. An update on computer-engineered complete dentures: A systematic review on clinical outcomes. *J Prosthet Dent.* 2017; 117(4):478-485. doi:10.1016/j.prosdent.2016.08.017.
- [22] Saponaro PC, Yilmaz B, Heshmati RH, McGlumphy EA. Clinical performance of CAD-CAM-fabricated complete dentures: A cross-sectional study. *J Prosthet Dent.* 2016; 116(3):431-435. doi:10.1016/j.prosdent. 2016.03.017.
- [23] Imirzalioglu P, Uckan S, Haydar SG. Surgical and prosthodontic treatment alternatives for children and adolescents with ectodermal dysplasia: A clinical report. *J Prosthet Dent.* 2002; 88(6):569-572. doi:10.1067/mpr. 2002.130146.
- [24] Mello BZF, Silva TC, Rios D, Machado MAAM, Valarelli FP, Oliveira TM. Mini-implants: Alternative for oral rehabilitation of a child with ectodermal dysplasia. *Braz Dent J.* 2015; 26(1):75-78. doi:10.1590/0103-6440201300111.

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