Report of a Case of Hypodontia due to Ectodermal Dysplasia: Early Rehabilitation with Overdenture

Maryam Mohajerfar[1] Simindokht Zarrati[2] Yeganeh Memari*[1]

[1]*Dept. of Prosthodontics, School of Dentistry, Shahid Beheshti University of Medical Sciences, Tehran, Iran.*
[2]*Dept. of Prosthodontics, School of Dentistry, Tehran University of Medical Sciences, Tehran, Iran.*

Abstract
Alveolar ridge is underdeveloped in ectodermal dysplasia (ED). The available treatment plans include fixed, removable or implant-supported prostheses, alone or in combination.
A 5 year-old boy was referred for treatment to the Department of Prosthodontics, Tehran University of Medical Sciences with the chief complaint of missing teeth. Prosthodontic treatment was performed to improve mastication, esthetics, phonetics, and psychological support. Altering the alveolar height also provided a more pleasant musculocutaneous profile. Series of overdentures were fabricated in the same conventional manner to compensate for the patient's growth.
Early treatment had a major impact on self-esteem, masticatory function, speech and facial esthetics of our patient. However, long-term success depends on regular recall appointments.

*Corresponding Author:
Memari Y.
E-mail: y_memari@yahoo.com

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Introduction

Ectodermal dysplasia represents a group of inherited conditions in which two or more ectodermally derived anatomic structures fail to develop (1). Ectodermal dysplasia (ED) is defined as a disorder involving structures developing from the ectoderm as well as structures that interact with ectoderm. This condition is usually transmitted as an X-linked recessive trait and includes up to 154 distinctive syndromes and 11 subgroups, labeled from one to four depending on the affected system (hair, teeth, nails or sweat glands). The most common types of ED include hypohidrotic (anhidrotic) and hidrotic forms (2). Sweat glands in hypohidrotic ED decrease significantly or are completely absent; but in hidrotic ED, they are often normal. The prevalence of disorders in ectoderm-derived structures is 1 to 7 in 10,000 births (2, 3). Oral manifestations in ED range from minimal to complex involvement. Dental abnormalities are seen in 80% of the cases (2). If teeth are not present, normal development of alveolar bone would be impaired and it may be partially or totally absent. Teeth, when present, can range in size from normal to small, and in shape from tapered and conical to pointed in incisors. Molars may have a reduced size (3, 4).

In primary dentition, maxillary second molars, maxillary canines, maxillary central incisors and mandibular canines are the teeth most commonly present (5). Lack of tooth buds causes hypoplastic alveolar bone, leading to a reduced vertical dimension of occlusion (VDO) and old-age appearance (4). Clinical case studies have indicated that

jaw growth in ED patients is within the normal range (6, 7). More studies are needed before any definitive conclusions can be drawn (8).

Dental problems are best managed by oral rehabilitation with complete denture, overdentures or fixed appliances depending on the number and location of the remaining teeth (9, 10). Ideally, restorations should be placed before the children start school. The most common treatment modality for dental management of ED involves the use of removable prosthesis. Because anodontia or hypodontia is typical in individuals with this condition, treatment may involve complete dentures, partial dentures or overdentures (11, 12). Long edentulous span does not permit fixed partial dentures either. Telescopic denture by incorporating the best of both fixed and removable prostheses can be an efficient treatment alternative for ED patients with compromised dentition and limited budget (13).

Efficient dental treatment of ED patients requires adequate knowledge about growth and development, behavioral management, prosthetic and restorative technical skills, the ability to motivate patients and parents, and long-term follow up for modification and replacement of prosthesis (14). Early oral rehabilitation is generally recommended from the age of 5 years. If the child is cooperative, oral rehabilitation can be successful and dentures can be fabricated for children as young as 2 or 3 years of age (15). Early treatment is critical for children because it brings about a significant improvement in mastication, esthetics, phonetic function and psychological support. Early prosthetic treatment also alters the alveolar height and provides a better musculocutaneous profile (15, 16).

Common problems are usually related to loss of prosthesis retention or occlusal changes caused by erupting teeth or growth of the jaws (9). As the children with ED grow up, removable prosthesis may need to be relined, rebased or replaced when a decreased VDO and an abnormal mandibular posture are detected due to growth in order to comply with growth changes and maintain masticatory function (15, 16, 17). After completion of growth in older ED patients, osseointegrated implants can be placed to support, stabilize and retain prosthesis (15, 18). Suitable time for implant placement could not be predicted based on chronological age, but it correlates well with the degree of adolescent growth observed on wrist and head radiographs (18).

Osseointegration in growing jawbone can make it difficult to maintain a satisfactory implant position and may compromise esthetics and function. A major disadvantage is that the implant cannot follow jaw growth. As the result, the prostheses would have infraocclusion (18).

This clinical report describes the characteristics and prosthetic treatment of a patient with ED with three years of follow-up.

Case Report

A 5-year-old boy was referred for treatment to the Department of Prosthodontics of Tehran University of Medical Sciences. The patient had been diagnosed with ED (Figure 1). His chief complaint was missing teeth. His parents seemed to be normal and

they denied any history of similar conditions in their respective families. The medical history of the child was unremarkable. The child had scant eyelashes and eyebrows. His skin appeared to be soft, thin, dry and hairless. Examination of nails revealed no abnormality. He had an age-appropriate behavior in dental office. Patient showed reduced vertical dimension of the lower third of face as evidenced by deep folds in the commissures of the mouth and by the patient's thin lips. The patient also showed retruded lips, protruded chin and underdeveloped alveolar ridges, which were small and thin (Figure 1).

Figure 1- Frontal and profile view of the patient shows the loss of nerfic at hight

Oral examination revealed six primary teeth. Central incisors were malformed and decayed both on clinical and radiographic examinations. The patient had a stainless steel crown on his right maxillary molar. Radiographic examination revealed three tooth buds, two in the maxilla and one in the mandible (Figure 2).

Figure 2- Panoramic view of the patient shows the present teeth

When examined, he demonstrated loss of VDO. Extra-oral examination revealed no facial asymmetry, and palpation did not reveal muscle tenderness. The patient had no symptoms of temporomandibular joint dysfunction.

According to the prosthodontic diagnostic index, both maxillary and mandibular arches were class IV, with multiple missing teeth and altered vertical dimension.

Dental treatment:

Composite restorations were performed for maxillary central incisors. Primary impressions were made with irreversible hydrocolloid impression material (Alginoplast, Heraeus Kulzer GmbH, Hanau, Germany) and diagnostic casts were poured and then articulated to evaluate VDO.

Custom acrylic trays were fabricated and border molding was performed with modeling impression compound (Impression compound, Kerr, Orange, CA, USA).

Final impression was made with silicone impression material (Panasil monophase medium, Kettenbach GmbH & Co.KG, Aarbergen, Germany) and final casts were fabricated (Figure 3). Wax occlusal rims were fabricated as well. Jaw relations were based on fullness of the lower face, appearance of the lips, occlusal plane relative to the tongue, freeway space and phonetic tests (19).

For establishment of correct VDO, overdenture was considered as the treatment of choice. A centric relation record was obtained with GC compound bite registration material (GC Corporation, Tokyo, Japan), and face bow recording was

taken. Master casts were mounted and the denture teeth were arranged.

Figure 3- master casts indicative of teeth and locations

After wax try-in approval, overdentures were fabricated. At the delivery appointment, fitness of overdentures was verified using pressure indicator paste. The overdentures were delivered and the patient and his parents were instructed on its proper maintenance.

Follow ups were scheduled at 72 hours, one week, two weeks, one month, three months and then every six months. The patient was satisfied with treatment. Speech was improved and a remarkable improvement in social activities of the patient occurred as well. At the 6-month recall, fitness of overdentures, especially the mandibular overdenture had decreased and the patient reported discomfort as the result of changes in his maxillary and mandibular arches; therefore, reline was performed using Mollosill (DETAX, Ettlingen, Germany). In the recall session at 6 years 4 months, the patient could no longer use the overdentures. It was decided to fabricate new dentures to compensate for the patient's growth. An occlusal window was created on the maxillary denture on both sides for primary second molars (20). For the same reasons, a third set of overdentures was made at 7 years 6 months in the same conventional manner as discussed earlier. Three sets of prostheses fabricated for the patient are shown in (Figure 4).

Figure 4- Three sets of prosthesis fabricated for the patient

Discussion

Provision of early prosthodontic treatment to replace the missing teeth and restore VDO is important in ED patients. Treatment of ED varies and generally depends on the child's age, dental agenesis, degree of malformation of teeth, the growth and development of the stomatognathic system of the patient and patient's cooperation (21).

Removable prosthesis is indicated initially for dental treatment of ED since it can be easily modified during rapid growth periods. Complete dentures are an alternative for patients with complete anodontia. Overdenture is another treatment modality to restore ideal occlusion and preserve the existing dentition. When teeth are present in the mouth, overdentures are the most desirable treatment option and have an added advantage since they preserve the alveolar bone. Crum (22) reported significant reduction in alveolar bone loss in overdenture patients after two years. Preservation of alveolar bone is important in ED patients, because prosthesis support is dependent on alveolar ridges from an early

age. For our patient, a conventional overdenture was the treatment of choice to preserve the remaining dentition and to restore function and esthetics (8, 23). Few existing primary teeth remained as potential overdenture abutments. It is recommended that the denture for the arch with the best prognosis be delivered first, to enhance patient's compliance. Second denture can be delivered two to four months later (8).

The prosthetic treatment was either modified and adjusted or reconstructed to comply with tooth eruption and jaw growth in our patient. In the recall session at 6 years 4 months, a new set of dentures, with one occlusal window for the primary second molars was fabricated because of changes in oral hard and soft tissue structures as recommended by Bonilla et al (23).

An improvement in muscular tone of both masticatory and perioral muscles occurred in our patient after delivery of prosthesis. Early prosthetic treatment improves nutrition and prevents delay in speech development (24). Normal sagittal and vertical position of the mandible is achieved by early treatment; because in ED patients, upward and forward displacement of the chin results in reduction of the lower third height of the face and tendency to class III skeletal relationship (24). Major problems with early treatment are related to periodic adjustments due to child's growth. Preadolescent growth in jaw dimensions and wear of acrylic teeth, leading to under-extension of dentures and posterior open bite, require regular adjustments and construction of new sets of dentures (24, 25). Excellent oral hygiene is crucial for successful treatment of ED patients. Daily topical fluoride should be advised for prophylaxis to prevent caries formation (26).

In patients with ED, under-development of maxillary tuberosity and alveolar ridges make it difficult to obtain adequate resistance and stability of the prosthesis (27). Endosseous implants can also be considered as an alternative treatment. Although age does not appear to affect the ability to establish osseointegration, it clearly impacts treatment planning in growing patients. It is important to place dental implants as early as possible, usually at late adolescence, depending on the state of edentulism (16, 17, 28). Early implant placement in patients as young as 5 to 6 years of age has been reported (29, 30). It is advised to postpone implants in most situations, particularly in partially edentulous patients in growth phase, due to relatively unpredictable consequences and effects of jaw growth and development on implant position and jaw relations. An acceptable time for implant placement could not be predicted by chronological age; but it correlates well with the degree of adolescent growth observed on wrist and head radiographs (17). Prosthesis with mini-implants has been satisfactorily used for prosthetic retention in younger patients (11, 31). However, clinical studies with long-term follow-ups are needed to test the efficacy of mini-implants as an alternative for oral rehabilitation of children with ED.

Conclusion

Regular periodic examinations are essential for compensation of growth changes in patients in growth phase.

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References:

1- Neville BW, Douglas DD, Allen CM, Bouquot JE. Oral and maxillofacial pathology. 3rd ed. 2009:741-3.

2- Shah R, Shah S. Oral rehabilitation of a patient with ectodermal dysplasia: A multidisciplinary approach. J Nat Sci Biol Med. 2014 Jul;5(2):462-6.

3- Glick M. Burket's oral medicine. 12th ed. PMPH-USA. 2015: 672.

4- Hekmatfar S, Jafari K, Meshki R, Badakhsh S. Dental management of ectodermal dysplasia: two clinical case reports. J Den Res Den Clin Den Prospect. 2012; 6(3):108-12.

5- Tarjan I, Gabris K, Roza N. Early prosthetic treatment of patients with ectodermal dysplasia: A clinical report. J Prosthet Dent. 2005 May;93(5):419-24.

6- Sarant BG, Brodie AG, Kubacki WH. Fourteen-year report of facial growth in case of complete anodontia with ectodermal dysplasia. AMA Am J Dis Child. 1953 August; 86(2):162-9.

7- Nomura S, Hasegawa S, Noda T, Ishioka K. Longitudinal study of jaw growth and prosthetic management in a patient with ectodermal dysplasia and anodontia. Int J Paediatr Dent. 1993 Mar;3(1):29-38.

8- Pigno MA, Blackman RB, Cronin RJ, Cvazos E. Prosthodontic management of ectodermal dysplasia: a review of the literature. J Prosthet Dent. 1996 Nov;76(5):541-5.

9- Ramos V, Giebink DL, Fisher JG, Christensen LC. Complete dentures for a child with hypohidrotic ectodermal dysplasia: a clinical report. J Prosthet Dent. 1995 Oct;74(4):329-31.

10- 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 Summer;6(3):108-12.

11- Toomarian L, Ardakani MR, Ramezani J, Adli AR, Tabari ZA. Using implants for prosthodontic rehabilitation of a 4-year-old with ectodermal dysplasia. Gen Dent. 2014 Sep-Oct;62(5):e1-5.

12- Gupta C, Verma M, Gupta R, Gill S. Telescopic overdenture for oral rehabilitation of ectodermal dysplasia patient. Contemp Clin Dent. 2015 Sep;6(Suppl 1):S258-61.

13- 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.

14- Nowak AJ. Dental treatment for patients with ectodermal dysplasias. Birth Defects Orig Artic Ser. 1988;24(2):243-52.

15- Hickey AJ[1], Vergo TJ Jr.. Prosthetic treatments for patients with ectodermal dysplasia. J Prosthet Dent. 2001 Oct;86(4):364-8.

16- Nabadalung DP. Prosthetic rehabilitation of an anhidrotic ectodermal dysplasia patient: a clinical report. J Prosthet Dent. 1999 May;81(5):499-502.

17- Imirzalioglu P, Uckan S, Haydar SG. Surgical and prosthodontics treatment alternatives for children and adolescents with ectodermal dysplasia: a clinical report. J Prosthet Dent. 2002 Dec;88(6):569-72.

18- Bryan SR. The effects of age, jaw site and bone condition or oral implant outcomes. Int J Prosthodont. 1998 Sep-Oct;11(5):470-90.

19- Zarb G, Hobkirk JA, Eckert SE, Jacob RF. Prosthodontic treatment for edentulous patients. 13th ed. St. Louis (MO): CV Mosby 2013: 190-92.

20- Rathee M, Malik P, Dua M, Yadav V. Early functional, esthetic, and psychological rehabilitation of preschool child with nonsyndromic oligodontia and anodontia in mixed dentition stage through conservative systematic approach: A case report with 5-year follow-up. Contemporary Clin Dent. 2016 Apr-Jun;7(2):232–5.

21- Bhargava A, Sharma A, Popli S, Bhargava R. Prosthodontic management of a child with ectodermal dysplasia: A case report. J Indian Prosthodont Soc. 2010 Jun;10(2):137-40.

22- Crum RJ. Rationale for the retention of teeth for overdentures. In Brewer AA, Morrow RM, editors. Overdentures. 2nd ed. ST Louis: CV Mosby, 1980: 3-11.

23- Bonilla ED, Guerra L, Luna O. Overdenture prosthesis for oral rehabilitation of hypohidrotic ectodermal dysplasia: A case report. Quintessence Int. 1997; 28(10): 657-65.

24- Franchi L, Branchi R, Tollaro I. Craniofacial changes following early prosthetic treatment in a case of hypohidrotic ectodermal dysplasia with complete anodontia. ASDC J Dent Child. 1998 Mar-Apr;65(2):116-21.

25- Khazaie R, Berroeta EM, Borrero C, Torbati A, Chee W. Five year foolow up treatment of an Ectodermal Dysplasia patient with maxillary anterior composites and mandibular dentures: A clinical report. J Prosthodont. 2010 Jun;19(4):294-8.

26- Yavuz I, Baskan Z, Ulka R, Dulgergil TC, Dari O, Ece A, Yavuz Y, Dari KO. Ectodermal dysplasia: retrospective study of fifteen cases. Arch Med Res. 2006; 37:403-9.

27- Shaw RM. Prosthetic management of hypohidrotic ectodermal dysplasia with anodontia. Case report. Aust Dent J. 1990 Apr;35(2):113-6.

28- Mello BZ, Silva TC, Rios D, Machado MA, Valarelli FP, Oliveira TM. Mini-implants: alternative for oral rehabilitation of a child with ectodermal dysplasia. Braz Dent J. 2015 Jan-Feb;26(1):75-8.

29- Guckes AD, McCarthy GR, Brajim J. Use of endosseous implants in a 3-year-old child with ectodermal dysplasia: case report and 5-year follow up. Pediatr Dent. 1997 May-Jun;19(4):282-5.

30- Moorrees CF, Gron AM, Lebret LM, Yen PK, Frolich FJ. Growth studies of the dentition: a review. Am J Orthod. 1969 Jun;55(6):600-16.

31- Mello BZ, Silva TC, Rios D, Machado MA, Valarelli FP, Oliveira TM. Mini-implants: Alternative for Oral Rehabilitation of a Child with Ectodermal Dysplasia. Braz Dent J. 2015 Jan-Feb;26(1):75-8.