Prosthetic Treatment of Pediatric Patients with Ectodermal Dysplasia: Two Case Reports

Ektodermal Displazili Çocuk Hastaların Protetik Tedavisi: İki Olgu Raporu

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ABSTRACT

Ectodermal dysplasia (ED) is a hereditary disorder characterized by anomalies in the ectodermal structures. The most common dental anomalies are oligodontia and/or anodontia of the primary and permanent dentition. Therefore, pediatric patients with ED may be need prosthetic rehabilitations. Several prosthodontics treatment options, such as complete, removable partial, overdenture, fixed and implant-supported dentures are avaible to rehabilate patients with ED. The aim of this clinical case report is to present the clinical characteristics and prosthetic treatment of two cases with ED.

Key Words: Dental prosthesis, Ectodermal dysplasia, Pediatric dentistry

ÖZ

Ektodermal displazi (ED), ektodermal yapılardaki anomalilerle karakterize kalıtsal bir hastalıktır. En yaygın görülen diş anomalileri, süt ve daimi dişlerin oligodontisi ve/veya anodontisidir. Bu nedenle ED'li çocuk hastalarda protetik rehabilitasyona ihtiyaç duyulabilir. ED'li hastaları rehabilite etmek için tam, hareketli parsiyel, overdenture, sabit ve implant destekli protezler gibi çeşitli protetik tedavi seçenekleri mevcuttur. Bu olgu raporunun amacı, ED'li iki olgunun klinik özelliklerini ve protetik tedavisini sunmaktır.

Anahtar Kelimeler: Dental protez, Ektodermal displazi, Çocuk diş hekimliği

INTRODUCTION

Ectodermal Dysplasia (ED) comprises clinical and inherited heterogenous group of disorders affecting at least two or more ectodermal-derived tissues, such as nails, hair, sweat glands, and teeth with or without the involvement of the other organs (1). ED can be inherited in various genetic patterns: e.g. autosomal-recessive, autosomal-dominant or X-linked modes, and it affects males more commonly and severely compared with females (2). The estimated incidence of ED is 1 in 100.000 births and considered to be relatively rare. Nearly 200 clinically different pathologic conditions have been identified as ED (3).

ED has two major types considering the number and function of the sweat glands:

1. Hypohidrotic (HED)/anhidrotic ED is characterized by sweat glands that are either missing or severly decreased in number (Christ-Siemens-Touraine syndrome),
2. Hydrotic ED is characterized by normal sweat glands and autosomal-dominant inheritance (Clouston's syndrome) (3,4).

While the dentition and hair are similarly affected in both types, inheritance patterns, nail and sweat gland manifestations tend to differ (5). General clinical findings in patients with ED include dry-lightly pigmented skin, alopecia or hypotrichosis (sparse, shiny hair or eyelashes), nail dystrophy, absence of sweat glands, and palmar-plantar hyperkeratosis (2,5). Complete or partial anodontia of the primary and permanent dentition and tooth malformations are the most common dental findings.

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Furthermore, incisors and canines are typically conical in shape, whereas, second molars, if present, are predominantly affected by taurodontizm (6). Due to multiple missing teeth, the alveolar ridge of both maxilla and manbidula are reduced, which leads to a decrease in the vertical dimension of occlusion (7). As a result, affected children often exhibit an old-age appearance (8). Other orofacial characteristics of ED include a prominent supraorbital ridge and frontal bossing, midfacial hypoplasia, a depressed nasal bridge, disappeared vermillion border and protuberant lips (9,10). Additionally, xerostomia can occur when the major salivary glands are affected in ED patients (10).

Early oral and prosthetic rehabilitation of ED patients is necessary to restore function and esthetics, as well as speech, psychosocial development and well-being (10). A multidisciplinary team involving pediatric dentist, orthodontist, prosthodontist, oral-maxillofacial surgeon, and speech therapist is essential to achieve successful results for the treatment of patients with ED (6,7,11). This case report aimed to present characteristic dental findings and prosthetic treatment of two pediatric patients with ED.

CASE REPORTS

Case 1:

An 11-year-old male patient was referred to Pamukkale University, Faculty of Dentistry, Department of Pediatric Dentistry with a complaint of inability to masticate with previous prosthesis, which be applied two years earlier. He had been diagnosed with HED when he was four years old, and displayed characteristic features of ED including brittle and fine blonde hair, narrow eyeblow-eyelashes, depressed nasal bridge, prominent forehead, periorbital pigmentation, and protuberant lips (Figure 1). An intra-oral examination showed mandibular anodontia, low alveolar ridges, and presence of hypoplastic conic-shaped maxillary central incisors (Figure 2A). Panoramic radiography also verified mandibular anodontia and the absence of maxillary teeth except of two permanent maxillary central incisors (Figure 2B).

Figure 1: Extra-oral view of case 1.

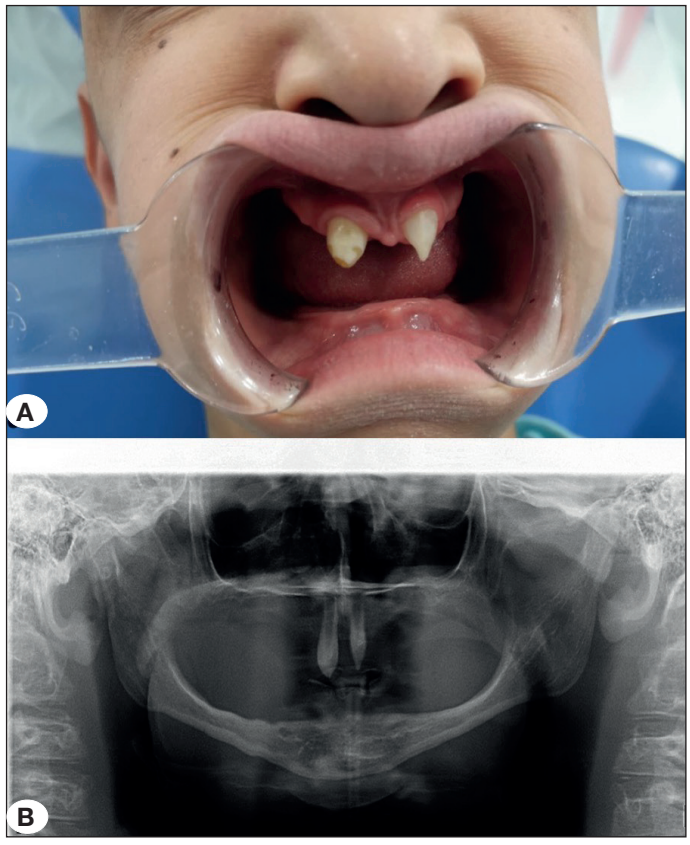

Figure 2: A) Intra-oral view of maxilla and mandibula (conic-shaped maxillary central incisors and mandibular anodontia). **B)** Panoramic radiography of case 1.

Figure 3: Prosthetic treatment of case 1 with a maxillary overdenture and a mandibular complete denture.

Dental treatment plan was discussed with patient and his parents, and informed consent was obtained. To improve function, esthetics, and speech, a tooth-supported maxillary overdenture and a mandibular complete denture were considered the preferred treatment options. After preparing both conic-shaped

permanent maxillary central incisors, preliminary impressions were made using irreversible hydrocolloid (Cavex Tulip, Cavex Holland BV, Haarlem, Holland), and then custom trays were fabricated for functional impression. Occlusal relationship was recored using wax occlusal rims, and the models were mounted on a semi-adjustable articulator. Prosthetic permanent denture teeth (Eray, Eray Inc., Türkiye) were set to provide proper lip support and ensure bilateral balance occlusion. After assessing retention, occlusion and esthetics of trial dentures, dentures were fabricated heat-activated acrylic resin (Imicryl®, Konya, Türkiye). The dentures were inserted and required adjustments were made (Figure 3). Instructions about to maintain a soft diet for the first few days, oral hygiene procedures, and wearing dentures throughout the day except for brushing and sleeping were given to parents. During follow-up visits the patient and his parents reported improvements in mastication, appearance, speech, and social behaviour after prosthetic rehabilitation. Follow-up visits at 3-month intervals were scheduled for adjustments of the dentures, to evaluate growth and development, as well as oral hygiene.

Case 2:

An 11-year-old female patient was referred to Pamukkale University, Faculty of Dentistry, Department of Pediatric Dentistry with the same complaint of case 1. It was revealed through parental history that the patient was intolerant to heat, and her mother and uncle also presented oligodontia. Extra-oral examination revealed typical features of ED including dry anhidrotic skin, diffusely sparse hair, narrow eyebrow-eyelashes, depressed nasal bridge, prominent forehead, periorbital and perioral pigmentation, protuberant lips, and old-age appearance (Figure 4). An intra-oral and radiographic examination showed

Figure 4: Extra-oral view of case 2.

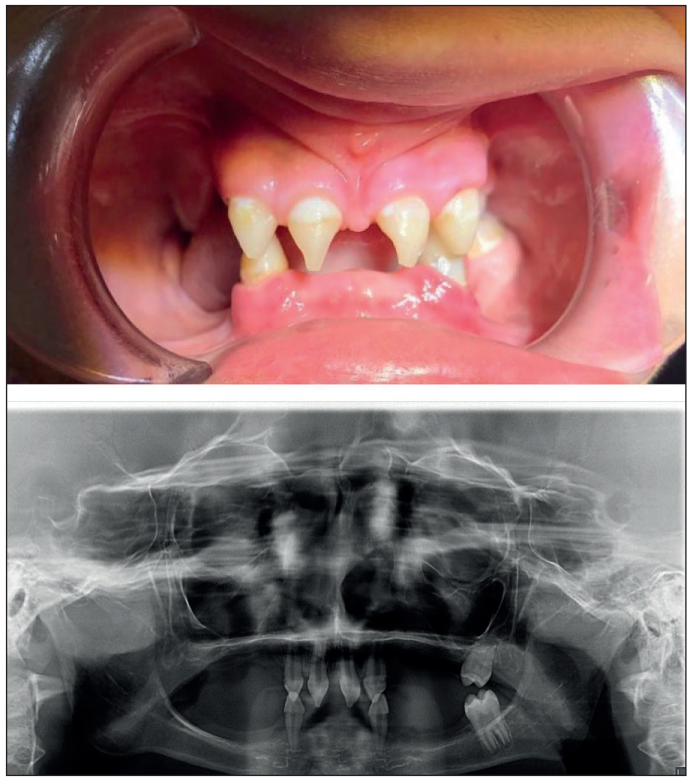

Figure 5: Clinical intra-oral frontal view of case 2. Panoramic radiograph revealing absence of 24 permanent teeth.

Figure 6: The maxillary and mandibular removable partial dentures of case 2.

that only four conic-shaped permanent maxillary anterior teeth, both permanent mandibular canines, left permanent maxillary and mandibular first molars had erupted (Figure 5). The patient also presented low alveoler ridges, loss of vertical dimension, and reduced sulcus depth in the posterior region of the maxilla and mandibula. Caries lesions were detected in both permanent mandibular canines, and her oral hygiene was poor.

Treatment plan included maintaining of appropriate oral hygiene, restoration of carious permanent mandibular canines and conic-shaped permanent maxillary anterior teeth,

and fabricated maxillary and mandibular removable partial dentures. The dental treatment plan was explained to the patient and her parents, and informed consent was obtained. After carious permanent mandibular canines were restored with composite resin (Clearfil MajestyTM Anterior; Kuraray Medical Inc., Okuyama Japan), and conic-shaped permanent maxillary incisors reconstructed with strip crowns (TDV Dental Ltda. Brussels, Belgium), preliminary impressions were made using irreversible hydrocolloid (Cavex Tulip, Cavex Holland BV, Haarlem, Holland). Following fabricated of custom trays, border-molding was established and final impression were made using polyvinylsiloxane impression material. Occlusal record was obtained with the same procedures as in the first case. Prosthetic permanent denture teeth (Eray, Eray Inc., Türkiye) were arranged to ensure age-appropriate appearance, and bilateral balance occlusion was given. Four wrought wire clasps were positioned on the permanent maxillary lateral incisors, permanent maxillary first molar, and permanent mandibular first molar (Figure 6). Removable maxillary and mandibular partial dentures were fabricated as previously described, then inserted and required adjustment were made. Recall appointments were scheduled as described for case 1. Despite the patient and her parents reported improvements in mastication, appearance, speech, and social behaviour during follow-up visits, the patient was unable to maintain proper oral hygiene. The patient was instructed to maintain proper oral hygiene, and further follow-up visits were scheduled every 3-months to evaluate growth and development, and oral hygiene.

DISCUSSION

Patients with ED present a significant challenge with regard to mastication ability, speech problems, esthetic concerns, and potential impacts on social and psychological development (10). Therefore, prosthetic treatment is essential for ED patients to provide improvements in function, speech, and esthetics, as well as increased self-esteem and psychological development. Several prosthetic treatment options including removable (complete/partial or overdentures), fixed partial, and implant-supported dentures are avaible for patients with ED. Treatment choice depends on patient's age, clinical findings, developmental stage, patient's individualised needs, patient's motivation, and also parents' social status (11). It is generally recommended that the first dental prosthesis should be delivered prior to school-age, though there is no certain time period to start dental treatment (9). In general, early prosthetic treatment is recommended from the age of 5, but depending on the cooperation of the patient, dentures can also be made at the age of 3 to 4 (8). As a result, it contributes to normalize the function of the masticatory and perioral muscles, resulting in proper growth of the basal bones, and psychologically improves the child's self-image (12).

Fixed dentures are rarely used in ED patients because of minimal number of teeth, and furthermore, if these dentures cross the dental midline, the rigid connectors may interfere with normal jaw growth in actively growing patients (9). Implant-supported dentures are recommended as a treatment option for adolescents over 12 years (7,8). In a growing child, early placement of implants may lead to cosmetically unfavorable results due to the implants acting like ankylosed teeth. The vertical growth of the jaws may cause implant over-structures to not contact with the opposite teeth, which may lead to prosthetic infraocclusion (7). In 2013, consensus meeting focusing on the rehabilitation of patients with ED was held, and expert teams decided that the earliest age for implant therapy should be 7-8 years old for the anterior mandible, while older ages should be considered for the maxilla (13). When implant therapy is considered as a treatment option, the primary challenge is lack of sufficient bone. The alveolar ridges and basal bone may be insufficient, especially in the maxilla. Therefore, in ED patients who suffering from alveolar deficiency, if bone atrophy progresses severe extent, implant placement may not be achievable without bone grafting (8). Consequently, when implant therapy is considered as a treatment option, it requires extra consideration to determine if there is sufficient bone level to placement the implants, and whether there is sufficient vertical bone dimension to support to implants (14).

Removable dentures (complete/partial or overdentures) are the most common preferred options for the prosthetic rehabilitation of the pediatric ED patients due to their ease of modification during the rapid growth period (3,9). However, retention and stability of dentures can be compromised due to insufficient bone support, dryness of oral mucosa, and the irregular tooth shape (8,13). Conversely, overdentures offer more retentive options when teeth are present to support them, and have several advantages, such as preserving alveolar bone, providing greater support and stability for dentures, improving proprioception and neuromuscular feedback, and increased comfort when compared to complete dentures (10,15).

In both cases, implant therapy was not considered as a treatment option because of ongoing growth and development as well as alveolar deficiency. In case 1, the treatment consisted of a maxillary overdenture and a mandibular complete denture. Due to the conical shape of the maxillary central incisors and their positioning, they were used as abutments for the overdenture. These dentures can preserve sensorial input of periodontal receptors, preserve alveolar bone structure, provide increased retention, stabilization, mastication performance, and also have psychological benefits for the children (11). In case 2, maxillary and mandibular removable partial dentures with wire clasps were fabricated to provide increased retention. Following prosthetic treatment of both patients, notable improvements in esthetics, speech, mastication performance were achieved. However, in case 2, the patient failed to maintain appropriate

oral hygiene at her 1-month follow-up. Oral hygiene instructions were reinforced, and follow-up appointments were scheduled to evaluate oral hygiene, and assess the need for any prosthetic modification/renewal of dentures at 3-month intervals due to ongoing development.

CONCLUSIONS

The management of ED patients is challenging and requires multidisciplinary approach involving pediatric dentist, orthodontist, prosthodontist, maxillofacial surgeon, and speech therapist. When deciding on the choice of treatment, the patient's age, intraoral condition, and patient's/parent's demands should be considered. Prosthetic rehabilitation of pediatric ED patients with removable partial/overdenture or complete denture is a suitable and cost-efficient option that can improve esthetics, speech, and mastication performance. Periodic follow-up is essential to assess the need for any modification or renewal of dentures due to ongoing development. Fixed or implant-supported dentures may be considered as a treatment option after skeletal growth is complete.

Consent

A written informed consent was obtained from all patients and their parents for publication of clinical data and images.

REFERENCES

1. Gholman RR, Kassar WM, El Meligy OA. Dental Rehabilitation of a Child with Ectodermal Dysplasia: A Case Report. Int J Clin Pediatr Dent 2019;12:362-5.
2. Kilic S, Altintas SH, Yilmaz Altintas N, Ozkaynak O, Bayram M, Kusgoz A, et al. Six-Year Survival of a Mini Dental Implant-Retained Overdenture in a Child with Ectodermal Dysplasia. J Prosthodont 2017;26:70-4.
3. Mittal M, Srivastava D, Kumar A, Sharma P. Dental management of hypohidrotic ectodermal dysplasia: A report of two cases. Contemp Clin Dent 2015;6:414-7.
4. Vasconcelos Carvalho M, Romero Souto de Sousa J, Paiva Correa de Melo F, Fonseca Faro T, Nunes Santos AC, Carvalho S, et al. Hypohidrotic and hydrotic ectodermal dysplasia: A report of two cases. Dermatol Online J 2013;19:18985.
5. Tarjan I, Gabris K, Rozsa N. Early prosthetic treatment of patients with ectodermal dysplasia: a clinical report. J Prosthet Dent 2005;93:419-24.
6. Montanari M, Callea M, Battelli F, Piana G. Oral rehabilitation of children with ectodermal dysplasia. BMJ Case Rep 2012;2012:bcr0120125652.
7. Bani M, Tezkirecioglu AM, Akal N, Tuzuner T. Ectodermal dysplasia with anodontia: a report of two cases. Eur J Dent 2010;4:215-22.
8. 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;6:108-12.
9. Ladda R, Gangadhar S, Kasat V, Bhandari A. Prosthodontic management of hypohidrotic ectodermal dysplasia with anodontia: a case report in pediatric patient and review of literature. Ann Med Health Sci Res 2013;3:277-81.
10. Perry JE, Perry S, Soto DM, Cline JA, Mugayar LR. Early Prosthetic Treatment of Young Children with Ectodermal Dysplasia: Two Case Reports. J Dent Oral Biol 2017;2:1052.
11. Bajraktarova Valjakova E, Misevska C, Korunoska Stevkovska V, Gigovski N, Sotirovska Ivkovska A, Bajraktarova B, et al. Prosthodontic management of hypohidrotic ectodermal dysplasia: a case report. South Eur J Orthod Dentofac Res 2015;2:20-6.
12. Kaul S, Reddy R. Prosthetic rehabilitation of an adolescent with hypohidrotic ectodermal dysplasia with partial anodontia: Case report. J Indian Soc Pedod Prev Dent 2008;26:177-81.
13. Ou-Yang LW, Li TY, Tsai AI. Early prosthodontic intervention on two three-year-old twin girls with ectodermal dysplasia. Eur J Paediatr Dent 2019;20:139-42.
14. Celli D, Manente A, Grippaudo C, Cordaro M. Interceptive treatment in ectodermal dysplasia using an innovative orthodontic/prosthetic modular appliance. A case report with 10-year follow-up. Eur J Paediatr Dent 2018;19:307-12.
15. Cardoso JS, Faria Carvalho D, Carvalho Silva C, Moura Teles A, Leal F, Lopes Cardoso I. Hypohidrotic Ectodermal Dysplasia and Its Manifestations in the Oral Cavity. J Dent & Oral Disord 2021;7:1161.