Oral findings in patients with ectodermal dysplasia (ED) include complete or partial hypodontia, anodontia, loss of vertical dimension of occlusion, protuberant lips, malformed and widely spaced conical-shaped teeth and underdeveloped alveolar ridges. These patients present a substantial challenge in dental treatment. This case report presents oral rehabilitation of a 22-year-old male patient diagnosed with ED using an implant-tooth supported telescopic partial denture at mandible and a tooth supported telescopic partial denture at maxilla. Implants in the mandible were placed at the sites of the right and left lateral incisor teeth. Following implant placement, the remaining buccal bone dehiscence was filled with deproteinized bovine bone graft and covered with resorbable membrane. To manage the vestibular insufficiency and to increase the keratinized mucosa in maxilla, bilateral acellular dermal matrix allograft was used on the right and left buccal aspects. Treatment described here improved the patient's functional and esthetic status while significantly restoring his oral health and self-esteem.
Full-Mouth Rehabilitation of a Patient with Ectodermal Dysplasia Patient with Dental Implants

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Oral findings in patients with ectodermal dysplasia (ED) include complete or partial hypodontia, anodontia, loss of vertical dimension of occlusion, protuberant lips, malformed and widely spaced conical-shaped teeth and underdeveloped alveolar ridges. These patients present a substantial challenge in dental treatment. This case report presents oral rehabilitation of a 22-year-old male patient diagnosed with ED using an implant-tooth supported telescopic partial denture at mandible and a tooth supported telescopic partial denture at maxilla. Implants in the mandible were placed at the sites of the right and left lateral incisor teeth. Following implant placement, the remaining buccal bone dehiscence was filled with deproteinized bovine bone graft and covered with resorbable membrane. To manage the vestibular insufficiency and to increase the keratinized mucosa in maxilla, bilateral acellular dermal matrix allograft was used on the right and left buccal aspects. Treatment described here improved the patient’s functional and esthetic status while significantly restoring his oral health and self-esteem.
Ectodermal dysplasia (ED) describes a large and complex group of disorders defined by the abnormal development of two or more structures derived from the embryonic ectodermal layer; i.e., skin, hair, nails, nerve cells, sweat glands, and parts of the eye and ear (1). The incidence of the disease is with a frequency varying between 7 in 10,000 and 7 in 100,000 live births (1,2). ED is classified as 2 major types: hypohidrotic, in which the sweat glands are absent or significantly decreased; and hidrotic, in which the sweat glands are normal. Hypohidrotic ED (HED) is the more common and severe form associated with dental defects (3). HED is inherited as an X-linked autosomal recessive disorder, whereas other forms of ED are inherited as autosomal dominant disorders (4).

In addition to the complete absence of sweat glands, clinical features of the HED include; abnormal or missing teeth, absent or very thin hair and abnormal nails. HED may also present with increased pigmentation around the eyes and mouth, saddle nose and fine-line wrinkles around the eyes, and heat intolerance. Children with the disease may have difficulty controlling fever. Mild illnesses can produce extremely high fevers due to lack of sweating and appropriate temperature control. At an adult age, the affected individual is usually unable to tolerate a warm environment and needs special measures to keep a normal body temperature (5, 6). Clinical findings suggest that there may be an inflammatory process associated with the complications in ED. Oral characteristics include complete or partial hypodontia, anodontia, impacted teeth, loss of vertical dimension of occlusion, protuberant lips, malformed and widely spaced conical-shaped teeth and underdeveloped alveolar ridges (6, 7, 8, 9). Absence of teeth as a result of hypodontia alters transverse bone growth, sagittal maxillary and mandibular growth while vertical growth can be affected by reduced alveolar height (3). Thus, ED patients often need a multidisciplinary approach in dental treatment planning to restore function, aesthetics and comfort. Conventional prosthodontic treatment including complete dentures, overdentures, or a combination of fixed and removable partial dentures present considerable problems in ED patients (10). The irregular distribution and the conical-shaped teeth often limit the integration of
bridges and crowns in patients with ED (11). Hypodontic mandibles of children with ED exhibit underdeveloped alveolar ridges (12) making very difficult to gain adequate retention and support for conventional prosthesis (13). Finally, progressive resorption of basal bone when the edentulous ridge is loaded at an early age may lead to prosthetic difficulties in later years (14). Therefore, there is a need for alternative therapeutic approaches beyond conventional dental techniques for restoring the function, esthetics, and phonation in patients with ED. Dental implants have been suggested as an important alternative for ED patients to support, stabilize and retain the prosthesis (15, 14, 3, 12, 16, 9, 17, 18). The present report describes a severe case of HED and full-mouth restorations using osseointegrated dental implants.

CASE DESCRIPTION

A 22-year-old male patient with HED was referred to Department of Periodontology, Faculty of Dentistry, University of Istanbul with esthetic complaints and not being able to eat properly. Extraoral examination revealed fine sparse thin hair, scant eyelashes and eyebrows, frontal bossing, a depressed nasal bridge, protuberent lips, a prominent chin and a resultant concave facial profile (Figure 1A). Patient reported that he sought frequent medical care because of high fever and hydrated lesions on his skin due to a lack of sweat glands and was diagnosed with HED. He also reported absence of primary and secondary teeth and treatment with maxillary and mandibular over denture prosthesis fabricated when he was 7 years old. Patient had never adapted to the removable dentures.

Intraoral examination revealed all permanent teeth were missing except for the maxillary central incisors and mandibular right canine and the vertical dimension of occlusion was increased because of the severe alveolar resorption of jaws (Figure 1B). The patient had hypoplasia of salivary glands and dry mucosa. Panoramic radiography (Cranex 3+ Ceph, Sorodex, Finland) and dental volumetric tomography (E-WOO Tecnology CL, Republic of Korea) were used to measure the bone density and to observe anatomic restrictions (Figure 1C). After a comprehensive clinical and radiographical
examination and detailed discussion of treatment options with the patient and his family; the treatment was planned as fabrication of a mandibular and maxillary implant-tooth supported telescopic partial dentures. The patient declined implant prosthetic treatment for the maxilla. Therefore, maxilla was rehabilitated with tooth-supported telescopic partial denture while the mandible was restored with implant and teeth supported prosthesis as planned. **Alveoloplasty** was necessary due to ‘knife-edge’ morphology of the edentulous mandibular ridge prior to implant placement during the same surgery. Implants (Tapered Screw Vent, Zimmer Dental, San Diego, USA) were placed in the mandible at the sites of right (3.7x13 mm) and left (3.3x13 mm) lateral incisor teeth using a torque of 50 Newton. Following implant placement, the remaining buccal bone dehiscence was filled with deproteinized bovine bone graft (Bio-Oss: Geistlich Pharma AG, Wolhusen, Switzerland) and covered by a resorbable collagen membrane (Bio-Gide: Geistlich Pharma AG, Wolhusen, Switzerland). Patient was prescribed with amoxicillin+clavulanic acid (1gr; 2x1) for 1 week and chlorhexidine gluconate (% 0.2; 2x1) for 2 weeks. After a submerged healing period of 6 months, implants were uncovered and healing abutment connections were placed. At the same time, free gingival graft was placed to increase the attached gingival around the implants as and autogenous graft. Graft healed uneventfully (Figure 2A). In maxilla, a frenectomy was performed between the upper santral incisors. In order to increase the keratinized mucosa and vestibular depth for preparation to the placement of partial dentures, bilateral soft tissue grafting was planned. **Acellular dermal matrix allograft** (AlloDerm: Biohorizons, LifeCell Co., Branchburg, USA) was used to simultaneous grafting of the right and left buccal aspects of the maxilla (Figures 2B and 2C) since the intraoral sites were not going to be sufficient for this procedure. A large wound was not favored at the donor sites. After complete healing of the soft tissues in edentulous sites and around teeth and implants for 8 weeks, prosthodontic procedures were started. An impression of the abutments was made with polyvinylsiloxane impression material (Affinis Putty Soft: Coltène/Whaledent GmbH + Co. KG Langenau/Germany) using an open-tray technique. Primary and secondary copings of telescopic crowns were prepared with gold alloy due to it is compatibility with gingival tissues and high precision casting and retentive
properties (Figure 3A). A daily maintenance care was instructed with an inter-dental brush (Curaprox Soft Implant Brush CPS508, CURADEN International, Istanbul, Turkey), superfloss (Curaprox Bridge & Implant Floss DF844, CURADEN International, Istanbul, Turkey) and a single toothbrush (Curaprox Single CS 1009, CURADEN International, Istanbul, Turkey).

Professional maintenance program was instituted with periodontal and peri-implant re-call visits every 3 months following delivery of definitive restorations (Figure 3B). Periodontal and peri-implant conditions were stable over 12- and 24-month follow-up appointments with no signs of attachment or bone loss associated with implants and the remaining teeth. Radiographic control was done with panoramic radiographs first and second year after the definitive prosthesis (Figures 3C and 3D). In order to evaluate the qualitative as well as quantitative changes in the alveolar bone, DVT were repeated (Figures 4 A and 4B). No signs of resorption were observed.

**DISCUSSION**

In this paper, we report full-mouth restoration of a patient with severe ED. The treatment included a combination of dental implants and remaining natural teeth and telescopic partial dentures. The case was followed for 2 years. Patient has been highly satisfied with the outcomes at the functional, esthetic, and phonation levels with restored social acceptance and self-esteem. This case supports the notion that using a multidisciplinary approach, severe cases of ED can successfully be restored and treated in the dental practice.

The most common complaint of childhood and adolescence in patients with ED is the concern about the dental anomalies and facial appearance (19). The treatment of choice is often a removable partial denture or complete denture since these can be easily modified during periods of rapid growth (20, 21). Early implant placement and fixed prosthesis were suggested as a good treatment option in ED cases with anodontia or hypodontia and with poor co-operative child (22) while this has not been supported with
strong evidence. Likewise, a recent review has recommended the insertion of implants in pediatric ED patients who suffer from syndromal hypodontia suggesting that the most suitable site of insertion to be the anterior mandible while insertions in the maxilla should be avoided or at least should not cross the midline (9). Nevertheless, clinical evidence in implant treatment of young children with ED is still limited. Implant surgery in ED patients presents special challenges, mainly because of the small dimensions of the jaws and the dense cortical bone. Thus, such treatments should always include dental tomographic imaging and the use of small-diameter implants postponing the treatment until jaw size is sufficient (23). In adult ED patients, dental implants have increasingly become the treatment of choice, because growth has stabilized by adulthood and the implants can be used to support, retain, and stabilize the prosthesis (3, 12, 18, 24, 25).

Implants placed in ED patients younger than 18 years have a higher risk of failure but implant survival rates vary between 88.5% and 97.6% in adolescent ED patients (26). Garagiola et al. suggest that dental implants inserted successfully in ectodermal dysplasia patients in association with GBR and bone grafts (27). In this report, we have used a similar approach to facilitate the placement of implants in a prosthetically guided position. The survival data for telescopic systems with both types of abutments used, ie., teeth and implants have been comparable to prostheses supported solely by implants (28). Therefore, we have combined the dental implants with retaining teeth and telescopic partial removable dentures. The strategy used in combination with soft tissue grafting to increase the vestibular depth and keratinized gingival width have been stable for 2 years and patient satisfaction was achieved.

Dental treatment of ED has a marked financial effect on patients and their families and varies depending on the type and duration of treatment. Murdock et al. reported in their study that the costs ranged from $2038 to $3298 for those who received prosthodontic treatment only and from $12,632 to $41,146 for those who received a combination of prosthodontic, orthodontic, and implant treatment (29). This is a limiting factor and has also been the case in our patient. Advanced augmentation techniques and surgeries (sinus lifting, autogenous bone blocks from the iliac crest, distraction osteogenesis, etc.) could not be performed due to financial limitations. Still, we were able
to restore the dentition with the support of 2 dental implants and build a functional and esthetically acceptable prosthetic restoration. The main goal was to replace missing teeth and bone, establish the normal vertical dimension and provide support for the facial soft tissues (29). While the follow-up time was limited to 2 years, this was critical to detect any implant failures and alveolar bone resorption. During this period, no implants were lost with no observable bone resorption.

In summary, case report presented in this article demonstrate that the oral rehabilitation of an adult HED patient with a reduced number of teeth and underdevelopment of alveolar bone is possible. This treatment highly enhanced the patient’s functional and esthetic status, significantly increasing his social confidence and self-esteem.
REFERENCES


FIGURE LEGENDS:

**Figure 1:** Preoperative findings. Panel A. Preoperative extra-oral view. Panel B. Preoperative intra-oral view. Panel C. Dental volumetric tomography demonstrating the knife edge morphology of the edentulous ridge.

**Figure 2:** Placement of implants and soft tissue grafting. Panel A. Autogenous free gingival graft placed around implants in mandible. Panel B. Insufficient attached gingiva was grafted at left maxilla using acellular dermal matrix allograft. The recipient site was prepared for graft placement. Keratinized gingiva and vestibular depth were increased. Panel C. Insufficient attached gingiva was grafted at right maxilla using acellular dermal matrix allograft. The recipient site was prepared for graft placement. Keratinized gingiva and vestibular depth were increased.

**Figure 3:** Prosthetic treatment. Panel A. Telescopic crowns were prepared using gold alloy. Panel B. Final prosthetic restoration. Panel C. Panoramic radiograph 1 year after restoration. Panel D. Panoramic radiograph 2 years after restoration.

**Figure 4.** Qualitative and quantitative assessment of alveolar bone in mandible using dental volumetric tomography after 2 years. Panel A. Right mandible. Panel B. Left mandible
Figure 2

A

B

C
Figure 4
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