Various prosthetic treatment options for ectodermal dysplasia patients – A review

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ABSTRACT

The ectodermal dysplasia (ED) is a group of symptoms, which affects mostly all parts of the body. Despite some of the syndromes having different genetic causes, the symptoms are sometimes very similar. Diagnosis is mainly by clinical observation often with the help of family medical history so that it can be verified whether transmission is autosomal dominant or recessive. Worldwide, around 7000 people have been diagnosed with an ED condition. Some ED conditions are only present in single-family units and derive from very recent mutations. ED can occur in almost all races but is much more common in Caucasians than any other race. In this article, the causes, symptoms, and various forms of treatment are reviewed from various research articles and are summarized below.

KEY WORDS: Ectodermal dysplasia, Hidrotic, Hypodontia, Hypohidrotic, Oligodontia

INTRODUCTION

Ectodermal dysplasia (ED) is not only a single disorder but also a group of syndromes all deriving from abnormalities of the ectodermal structures, especially affecting hairs, nails, sweat glands, mammary glands, oral soft tissue and tooth, skins, lens or retina of the eye, parts of the inner ear, the development of fingers and toes, nerves, and other parts of the body. More than 150 syndromes have been identified. Hidrotic and hypohidrotic EDs are the two common types of ED observed. The primary distinctions between the hidrotic and the more common hypohidrotic forms of ED are based on the defects found in the sweat glands, with the hypohidrotic form being more severe.

Diagnosis

The diagnosis of patients with ED is based fundamentally on the clinical history in dental clinics. Patients with ED usually show relatively reduced number (hypodontia or oligodontia) or complete absence of teeth structures (anodontia) when compared to normal patients. These are of great interest when diagnosing ED. A patient may give a history of dry mouth. Oligodontia is most frequently associated with dominant type hereditary factors, polygenic factors, or X-linked. In some cases, teeth morphology will also be affected such as the presence of peg laterals and conical-shaped canines, which also serves as a factor for diagnosing ED. The clinical diagnosis is followed by radiographic diagnosis. For radiographic diagnosis, patients are advised to take orthopantomogram (OPG), i.e., panoramic view. The radiograph will show multiple missing and impacted teeth structures with malformed dental arches.

Treatment Planning

The treatment planning for patients with ED should be multifactorial as the symptoms are multifactorial. It is

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critical to discuss with each patient the expectations of treatment outcomes, and the patient must have a realistic understanding of treatment length, process, and effort necessary to achieve treatment goals. The patient must take an active role in treatment decisions and should be an involved participant. If the patient would benefit from and be receptive to psychotherapy, then the appropriate referral should be made. [11] Medical and dental interventions improve the appearance and function of a patient with congenital and craniofacial defects with implant-supported fixed and removable prostheses, overdentures, and traditional fixed and removable prosthesis which can provide more normal facial contours, an improved smile line, improved arch relationships, and improved function for teens and young adults with facial defects. [11] Implant-supported prostheses can enhance stability, retention, function, and bone preservation. [11]

For treatment planning, certain investigations are vital to conclude the sequence of treatment. The vital investigations include radiographic examinations such as OPG, cone-beam computed tomography to narrow down the skeletal conditions of the patient and clinical examinations of bone density (using Vernier caliper), Mucosal thickness and conditions, Salivary flow etc., helps in drawing out the correct sequence of treatment suited for patient.

Patients with ED will have severe skeletal deformities such as cleft lip and palate and malformed maxilla and mandible, who might need surgical intervention such as orthognathic surgery. However, in this article, we are trying to emphasize only the prosthodontic treatment options available for ED patients.

Various Prosthodontic Options for ED Cases

Temporary partial dentures

Temporary partial dentures are used as a temporary means of replacement of missing teeth in ED patients. Child patients are mostly provided with temporary dentures as a means of the temporary mode of treatment until they are suited to receive implant-supported or other fixed partial dentures.

Dental implants and implant-supported fixed dentures

Dental implants are the most advanced mode of replacing missing teeth nowadays. Guler et al. presented a case report of treating a 15-year-old female ED patient. Intraoral examination showed the presence of four primary teeth in the maxilla, and in mandible, only first molars and second premolars are present with three deciduous teeth structures. On OPG, retrusive dental arches were recorded with impacted molars near the coronoid process. The sequence of treatment was started with removal impacted and retained primary teeth followed by placement of fixed denture with the support of 4 dental implants in the mandible, and full metal crowns were placed followed by overdenture in the maxilla and the prognosis was good with satisfactory functional and esthetic results. [12]

The use of mini dental implants in children with ED seems to be very optimal in some cases where normal implants cannot be incorporated. Two cases were presented by Sfeir et al., in 2014. A 6-year-old boy suffering from anhidrotic ED, on clinical examination, presented with only tooth 11 with a conical peg-like crown. Initially, a removable prosthesis was performed to meet the esthetic and functional requirements. At the age of 8, the upper prosthesis becomes unstable, and clinical examination shows the beginning of an eruption of 21. At the age of 11, it was decided to insert two mini implants following discussions with the patients and his parents. A new prosthesis with support from the mini implants was fabricated, which solved the problem of instability of the previous prosthesis. [13]

A second case was a 10-year-old boy, suffering from a mild form of ED, presented with agenesis of all permanent teeth except teeth 34, 37, and 47. Initially, removable denture was given as a temporary solution like in the previous case. At the age of 12, two mini implants were inserted in the mandible to replace the lateral incisors. At the age of 14, four mini implants were inserted in the alveolar bone to replace the upper four incisors. At the age of 16, 4 years after the insertion of the lower implants, and 2 years after the insertion of the upper implants, the esthetic and functional results were very satisfactory. [13]

However, in patients with severe ED, placing an implant to replace the missing teeth is complicated due to diminished alveolar ridges lacking adequate thickness of bone for an implant placement. Hence, in these some cases, the need of bone grafting is essential to increase the thickness of the bone. Such a case was presented by Bayat et al., in 2011. An 18-year-old male genetically diagnosed hypohidrotic ED came to our clinic for implant rehabilitation of his edentulous maxilla and mandible. Severe hypodontia (only one existing permanent tooth in the maxilla), dry mucosa, loss of vertical dimension, underdeveloped alveolar ridges, and class III jaw relation were detected in the oral cavity. Clinical and radiographic examination showed that only right maxillary second molar was present in the oral cavity. Both the mandibular canines and the left mandibular first premolar were impacted. Severe maxillary and mandibular hypoplasia and mandibular prognathia were present, in addition to a loss of the vertical dimension. A Le Fort I maxillary osteotomy was performed with simultaneous sinus floor augmentation. It was followed by extraction of all impacted teeth in the oral cavity. Radiographic evaluation revealed that the bone quality was poor on the alveolar ridges; therefore, ridge augmentation with onlay graft was done to construct adequate bone.
volume allowing proper implant placement. After 6 months, dental implant insertion was performed.[15]

Implant survival rates vary between 88.5% and 97.6% in patients with ED and between 90% and 100% in patients with tooth agenesis. Implants placed in adolescent ED patients do not have a significant effect on craniofacial growth, while implants placed in ED patients younger than 18 years have a higher risk of failure.[14]

**Overdentures**

In some cases of ED, dentition may be present with severe deformities. In those cases, overdentures are the choice of treatment which are less invasive and expensive when compared to implants. A case report of 3-year-old boy was referred to the Department of Pediatric Dentistry, Shiraz University of Medical Sciences, with the chief complaint of lack of teeth. The intraoral examination revealed complete edentulism in the lower jaw, four peg-shaped anterior teeth in the upper jaw, thin alveolar ridge, reduced vertical bone height, and loss of vestibular depth in the lower jaw. The radiographic findings revealed two unerupted molar teeth in the upper and two unerupted incisors in the lower jaw. Treatment planning included making complete overdentures for the upper and lower jaws to improve appearance, function, and speech. The lower anterior teeth were left unmodified due to short crowns. Instructions were given to maintain proper oral hygiene during the delivery of the prosthesis. Continuous follow-ups every 6 months were planned for adjustment or replacement of old denture.[16]

**CONCLUSION**

Management of clinical manifestations associated with ED presents a unique challenge for prosthodontists. It seems that the treatment of patients with severe hypodontia due to ED will be different according to the unique anatomic and dental status. This article highlights the multidisciplinary treatment planning concepts necessary for successful rehabilitation of these patients. This article suggests that various prosthetic options are available to treat patients with ED to ensure good esthetic appearance and functional stability. However, more researches should be done to provide more state of the art treatment for patients with ED.

**REFERENCES**


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