



## ECTODERMAL DYSPLASIA-A CASE REPORT

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**ABSTRACT** Ectodermal dysplasia is a hereditary disorder associated with abnormal development of embryonic ectodermally-derived organs including teeth, nails, hair and sweat glands. Hypodontia of the primary and permanent dentition is the most common oral finding. Therefore, affected patients need dental prosthetic treatments during their developmental years. This report A 13 year-old girl was presented to the dental clinic due to missing of teeth. She was suffered to difficulty in chewing and esthetics presents children affected by ectodermal dysplasia with anodontia. Oral rehabilitation was accomplished with removable acrylic prostheses.

**KEYWORDS :****INTRODUCTION**

Ectodermal dysplasia are inherited conditions characterized by defects of the hair, teeth, nails, and sweat glands. The modes of inheritance are X-linked recessive, autosomal recessive, and autosomal dominant. The most common mode is X-linked recessive; therefore, more males are affected. "The ED syndromes are a remarkably diverse group of human disorders in which other parts of the body may be affected. 186 types of ectodermal dysplasia have been identified using the Freire-Maria classification. The current estimate of the incidence of ED is 7 in 10,000 newborns."(1) .The defects associated with ED are not life threatening when managed appropriately; however, all can significantly affect the quality of life. The most prevalent form of ectodermal dysplasia is hypohidrotic ectodermal dysplasia (HED), also known as Christ-Siemens-Touraine syndrome (CST). Eruption of the teeth may be delayed, or only a few teeth may erupt in those affected by HED.(3) the National Organization For Rare Disorders, and Geneskin.(1,2,4) Due to missing, abnormally shaped, and malpositioned teeth, a wide scope of dental procedures, including all aspects of restorative dentistry are required for dental rehabilitation of patients affected by ED. A team of dentists with training in pediatric dentistry, orthodontics, prosthodontics, and oral and maxillofacial surgery are often involved in establishing and carrying out a treatment plan(5-8 ).The goals of dental treatment for individuals affected by the ED syndromes are to provide an age-appropriate dentition that optimizes chewing function (and thus nutrition), oral/facial development, speech, swallowing, and esthetics. Comprehensive treatment also aims to enhance physical, emotional and psychosocial development for affected individuals. Since the dental manifestations of the ED syndromes persist throughout life, dentists must anticipate working closely with children, adolescents, and adult(9, 11) .



Extraoral aspects of patients with ectodermal dysplasia(Figure -1(a))



Facial profile showing prominent supraorbital ridges, absent eyebrow, depressed nasal bridge, broad columella, high hair line and dry skin(figure-1(b))



An ectodermal dysplasia patient with abnormal hand



An ectodermal dysplasia patient with abnormal foot

**Case Report**

A 13-year-old girl was referred to the Department of prosthodontics dentistry, representing lack of teeth as the chief complaint. According to familial history, a brother of the patient also suffered from oligodontia and heat intolerance. Extraoral examination revealed dry skin, hypoplastic midface, prominent lips, and sparse fine hair. These findings matched typical features of anhidrotic ectodermal dysplasia (Figure 1(a,b)). The intraoral examination revealed complete edentulism in the lower jaw, two teeth present in molar right and left upper jaw, thin alveolar ridge, reduced vertical bone height. The radiographic findings also confirmed the clinical diagnosis. Panoramic view revealed two erupted molar teeth in the upper jaw and no teeth present lower jaw (Figure(2)).Treatment plan included making complete overdentures for the upper and complete denture on lower jaws to improve appearance, function and speech. Due to small arch size, preliminary impression was made by alginate impression material and custom tray made and border moulding with green stick compound and final impression elastomeric impression material (light body putty) material. Jaw relationship was recorded using temporary base and wax rim (figure-4). The arranged teeth were verified in the mouth during the try-in appointment (figure-5). After laboratory processing, the upper overdentures and lower denture were delivered to the patient (Figure-6 ). In the delivery appointment, instructions were given to maintain proper oral hygiene. Continuous follow-ups every six months were planned for adjustment or replacement of old denture.



Ectodermal dysplasia is characterized by the absence and/or malformation of teeth (from hypodontia to anodontia (figure-2))



Panoramic radiography showing the upper permanent molar right and left side and edentulous of lower jaw

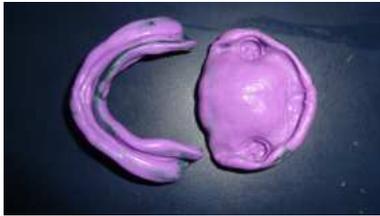


Figure-(3)



Upper and lower bite plate to record jaw relation (figure-4)



Wax up upper and lower prosthesis (figure-5)



After laboratory processing, the upper overdentures and lower denture was delivered to the patient (figure-6)



Patient happy with prosthesis

## DISCUSSION

Oral rehabilitation of patients with ectodermal dysplasia is necessary to improve sagittal and vertical skeletal relationships during craniofacial growth and development as well as esthetics, speech, and masticatory efficiency. A persons affected with ectodermal dysplasia includes case reports of patients treated at various ages with removable, fixed, and implant-supported prostheses.(12-15) Follow-up periods vary. In this case dental treatment planning include age, psychosocial environment, teeth present, oral hygiene, occlusal vertical dimension, bone volume, jaw growth and development. Parents or guardians of young persons with ED often desire to have treatment of children begin as soon as possible. The most common

treatment plan is removable prosthesis. Because Implant-supported denture is given when sufficient bone present. Positive effects include more self-confidence, facial esthetics, speech and masticatory function improvement(13). However, removable when a decreased vertical dimension of occlusion and an abnormal mandibular posture are detected due to growth(14). Retention and stability for the prostheses are also difficult to obtain. In patients with ectodermal dysplasia, dryness of the oral mucosa and the under-developed maxillary tuberosities and alveolar ridges are problematic factors for resistance and stability of the dentures (15 ).When fabricating dentures for these patients, care should be taken to obtain a wider distribution of occlusal loads by extending the denture base as much as possible(16) .In this situation, special attention must be paid to the impression making technique. The occlusion of removable partial denture should be in harmony with the patient's occlusion.. This treatment modality improves the patient's quality of life and it can be regarded as an acceptable treatment modality for functional and esthetic rehabilitation.

## CONCLUSION

A ectodermal dysplasia patient is challenging for prosthodontists . Treatment of ectodermal dysplasia edentulous patients with removable partial or complete denture is a most acceptable to improve sagittal and vertical skeletal relationships during craniofacial growth and development as well as esthetics, speech, and masticatory efficiency available and cost effective modality and psychosocial condition. However, its long-term success depends on regular recall appointments and proper maintenance of oral hygiene .

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