



## Ectodermal dysplasia with partial anodontia of primary and permanent dentition: A case report.

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### Abstract:

Hereditary ectodermal dysplasia represents a large group of conditions in which two or more ectodermally-derived anatomic structures fail to develop. Patients with ectodermal dysplasia are characterized by hypoplasia or aplasia of structures such as skin, hair, nails, teeth, nerve cells, sweat glands, parts of the eye and ear and other organs. 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 case report presents a child affected by ectodermal dysplasia with partial anodontia. Oral rehabilitation was accomplished with removable acrylic prostheses. Treatment had major impacts on self-esteem, masticatory function, speech and facial esthetic of the patient.

**Keywords:** Ectodermal dysplasia, Oral rehabilitation, Removable prosthesis.

### INTRODUCTION

Ectodermal dysplasia, as first described by Thurman<sup>1</sup>, is a hereditary disorder occurring as a consequence of disturbances in the ectoderm of the developing embryo. It is a congenital, diffuse and non-progressive disease.

Ectodermal dysplasia represents a large and complex group of diseases comprising more than 170 different clinical conditions.<sup>2</sup> The incidence of this condition is 1:100,000, with a mortality rate of 28% in males up to 3 years of age<sup>1</sup>.

From the clinical point of view, two main forms have been distinguished: (1) Hypohidrotic form/Christ-Siemens-Tourian Syndrome that exhibits the classic triad of hypohidrosis, hypotrichosis and hypodontia.<sup>3</sup> Asthma and

eczema are occasionally reported in these patients.<sup>4</sup> It is X-linked recessive inheritance in which males are affected severely, while females show only minor defects. (2) Hidrotic form/Clouston syndrome in which hair and nails are affected. The sweat glands are usually spared. It is inherited as an autosomal dominant trait.<sup>3</sup>

Hypohidrotic ED is the most common phenotype<sup>5</sup>. Dental manifestations include conical or peg shaped teeth, hypodontia or complete anodontia and delayed eruption of permanent teeth.<sup>6</sup> Due to missing teeth and reduced alveolar ridges the patient has characteristic senile facial appearance.

Tanner<sup>7</sup> states that in the case of ectodermal dysplasia such abnormal appearance may affect normal social and psychologic development in

young patients. This case report discusses the features and prosthetic treatment plan (upper and lower partial denture partial dentures) for ectodermal dysplasia which will help in the establishment of quality dental health as well as will provide psychological upliftment to the patient.

### CASE REPORT:

A 5year old boy reported to the department of Paedodontics and Preventive dentistry, Yamuna institute of dental sciences and research, Yamunanagar with absence of teeth as chief complaint. The lack of primary and permanent teeth in the oral cavity resulted in dietary problems. According to familial history, mother of the patient was also afflicted with the same condition but to a lesser extent.

Extraoral examination revealed dry skin, prominent lips, short lower facial third height, sparse fine hair and thin eyebrows. Patient had history of skin dryness and recurrent allergies in skin and eye which are most commonly seen in anhidrotic ectodermal dysplasia.

The intraoral examination revealed two spaced conical shaped maxillary primary central incisors, primary second molars in both the arches, erupting mandibular 1<sup>st</sup> molars, thin alveolar ridge, reduced alveolar bone height and decreased depth of vestibule in lower jaw. Panoramic radiograph also revealed two unerupted permanent maxillary central incisors and erupting permanent 1<sup>st</sup> molars in both arches, therefore, confirming the clinical diagnosis of Ectodermal dysplasia.

Treatment plan included making removable prosthesis for the upper and lower jaws so as to enhance the appearance and impart efficacy in function and speech. Due to limited mouth opening it was very challenging to make preliminary diagnostic impression. In order to avoid gagging and aspiration of impression material, impression was taken on perforated trays using elastomeric impression material. Mandibular impression was taken first so as to avoid gagging and reduce the anxiety of the patient. Teeth's setting was done and was verified during the try-in appointment. After

acrylisation, the dentures were delivered to the patient. Patient was instructed to maintain proper oral hygiene. Follow up after every 6 months was advised for adjustment or replacement of old dentures.



Figure1: (Pre-operative)



Figure: 2: Panoramic view



Figure 3: Maxillary arch



**Figure 4: Mandibular arch**



**Figure 6: Post-operative**



**Figure 5: Removable Prosthesis**

## DISCUSSION:

In oral rehabilitation of the ectodermal dysplasia patient, it is necessary to improve both the sagittal and vertical skeletal relationship during craniofacial growth and development as well as to provide improvements in esthetics, speech, and masticatory efficiency<sup>8</sup>. Multidisciplinary management is needed throughout childhood and prosthetic rehabilitation of patients with ectodermal dysplasia is a routine process.

Dental prostheses improve the tone of the muscles of mastication and may compensate for the reduced vertical dimension thus improving facial profile and expression significantly<sup>9</sup>. In cases of existing teeth, the advantages with regard to retention, stability, function, and the phonetics of the denture should be considered. In addition, existing teeth help protect the proprioception mechanism and prevent the formation of residual alveolar ridges<sup>4</sup>; in addition, mastication and dietary patterns also gets improved.<sup>1</sup>

As the child grows, the denture will have to be modified and replaced, because it is indicated in the longitudinal studies of anodontia that the growth of jaws is independent of existence of teeth. Replacements will be needed at least 3 times between the period of early and late mixed-dentition and permanent-dentition.<sup>10</sup> When the patient is in the last stage, the removable prosthesis may be replaced by a fixed type restoration using Osseo integrated implants. The implant-supported dentures reported are excellent in stability and retention, but the placement of dental implants in young children generates several problems. If given in preadolescent stage, vertical growth may result in submersion of the implants, necessitating prosthetic revision or possible use of longer abutments<sup>11</sup>.

The prognosis of the ectodermal dysplasia is very good and the life span of the patient is usually normal except for case of ectodermal dysplasia with immunodeficiency<sup>12</sup>. This case reports highlight the importance of treatment planning as well as the influence of anodontia

on the diagnosis of ectodermal dysplasia. A multidisciplinary approach must be carried out to establish functional occlusion and aiding to improve the nutrition of patient.

### Conclusion:

Treatment of ectodermal dysplasia in pediatric patients with removable partial or complete denture is an acceptable, easily available and cost-effective method thus improving speech, esthetics, function and psychosocial condition. Hence, we conclude that early identification and intervention to the problems caused due to this disease should be done to help the patients in improve their quality of life and ease their psychological fears. They can also be advised to undergo genetic counseling to prevent the disease from passing on to future generations by educating the family and their descendants.

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