

## **Dental Treatment Modality for Ectodermal Dysplasia: A Case Report**

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### **Abstract**

Ectodermal dysplasias are a diverse group of genetic disorders that involve defects of the hair, nails, teeth, skin and glands. Other parts of the body, such as the eyes or throat, may be affected as well. The combination of physical features a person has and the way in which it is inherited determines if it is an ectodermal dysplasia. Ectodermal dysplasias can occur in any race but are much more prevalent in Caucasians than any other group and especially in fair Caucasians. Such an ectodermal dysplasia case is represented here along with its dental management.

**Keywords:** Ectodermal Dysplasia, oral rehabilitation, dental implant, genetic disorder.

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### **Introduction**

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<sup>1</sup>. Ectodermal dysplasia might be inherited in any form of several genetic patterns including autosomal-dominant, autosomal-recessive, and X-linked modes.<sup>2</sup> Although more than 170 different subtypes of ectodermal dysplasia have been identified, these disorders are considered to be relatively rare with an estimated incidence of 1 case per 100,000.<sup>3</sup> The most common form of ED is X-linked anhidrotic ectodermal dysplasia (AED),

also known as the Christ-Siemens-Touraine Syndrome, involving a mutation in the AED gene<sup>4</sup>

ED is characterized by the triad of signs comprising sparse hair (atrachosis or hypotrichosis), abnormal or missing teeth (anodontia or hypodontia) and inability to sweat due to lack of sweat glands (anhidrosis or hypohidrosis). Most patients with EDA have a normal life expectancy and normal intelligence. However, the lack of sweat glands may lead to hyperthermia<sup>5</sup>.

Clinically, the forehead appears square, with frontal bossing, and there is a prominent supra-orbital ridge. The nose has a depressed nasal bridge and is called a saddle nose. The midface is depressed and hypoplastic, giving it a “dished-in” appearance. The cheekbones are high and broad, although they appear flat and depressed as well. The chin may be pointed and the lips everted and protuberant<sup>6</sup>. Missing teeth or the delay in teething often starts to worry the parents and leads to the diagnosis of ED in the second year of life<sup>7</sup>. The enamel layer is thin and the cervical area of the tooth is constricted. Enamel is rarely hypoplastic<sup>8</sup>.

In general, the skin of affected children is lightly pigmented and appears thin and almost transparent; surface blood vessels are easily visible. Other manifestations include fine sparse hair, reduced density of eyebrow and eyelash hair. When hair is present, it may be fragile, dry, and generally with unruly appearance as a result of poorly developed or absent sebaceous glands. Fingernails and toenails may also show faulty development and be small, thick or thin, brittle, discolored, cracked, and/or ridged<sup>9</sup>.

In this article we present the case of ectodermal dysplasia and its oral rehabilitation with fixed prosthesis in maxilla and implant supported denture in mandible.

### **Case 1**

A 19 years old male patient reported to the Department of Periodontics, CSMSS Dental College and Hospital, Aurangabad with a chief complaint of missing lower teeth and inability to masticate the food. The patient showed no family history. The skin of the patient was dry. The patient had history of lack of sweating and diagnosed as ED in childhood. Parents were taking care since then. Patient also had palmer planter hyperkeratosis (dry scaly patches on the skin of the palms and the soles). The mandible was small, underdeveloped and atrophic. Intraoral examination showed presence of permanent maxillary central incisors and first molars of both sides and an impacted first premolar of right side with deciduous canine and first molars which were firm enough. In mandible only deciduous right lateral incisor was present which was grade III mobile. OPG of the patient showed absence of all mandibular permanent teeth and remaining maxillary teeth (Fig 1). Mandible was atrophic and mandibular canal was approaching towards crest.

Extraction of the mandibular right lateral incisor was done because of grade III mobility and implant supported complete lower denture was placed (Fig 2, 4). Implants were placed in the canine region (Fig 2). In the maxilla, fixed prosthesis was given with the support of remaining teeth which were strong enough to be an abutment (Fig 3).

### **Discussion**

The course of the treatment for ectodermal dysplasia is to restore the function and the aesthetics of the teeth, normalize the vertical dimension and support the facial soft tissues. The denture must be periodically modified as alveolar growth; erupting teeth and rotational jaw growth change both the alveolar, occlusal and basal dimensions. In children, breakage and even loss of removable prostheses is quite common. So in young age we prefer for crown and bridges<sup>10</sup>.

Implant-supported denture is also suggested as the ideal reconstruction modality for adolescents over 12 years. When implant therapy is indicated, the main problem is insufficient bone; if bone atrophy progresses in these already alveolar deficient patients, implant placement may not be possible without bone grafting<sup>11</sup>.

However For some authors, it is even recommended that implant treatment is complete before the age of puberty to ensure aesthetics, as well as optimal functional and psychosocial development<sup>12,13</sup>.

The ideal timing of implant placement in children is a matter of debate. For the young patient with severe oligodontia or anodontia, such as individuals with ED, the oral rehabilitation has the impact of improving the patient's masticatory efficiency, quality of life, self-confidence, and social acceptability. Results of the B.R. Chrcanovic study review showed that dental implants placed in children with ED have relatively low failure rates (5.3%-7.2%, depending on the age group) after reasonable mean follow-up times (52.8-70 months, depending on the age group). Implants placed in adult ED patients presented a slightly lower failure rate (46/1030; 4.5%) than children<sup>14</sup>.

Cronin *et al* observed that if implants are placed during active growth, they may be displaced or malpositioned by continued growth and may require removal and replacement. Implants placed after age 15 for girls and age 18 for boys have the most predictable prognosis. Implants placed before these ages may not be permanent and may have to be re-implanted<sup>15</sup>.

The study by Balshi *et al* illustrates the use of implants to rehabilitate a 20-year-old ED patient who initially presented with only 2 permanent and 6 primary teeth in the maxilla. Along with conventional endosseous implants, 2 specially designed zygomatic implants were

utilized to avoid the need for bone grafting in the patient's severely resorbed maxilla<sup>16</sup>.

The study by Nurhan Güler *et al* presents a 15-year-old female with hypohidrotic ectodermal dysplasia who had surgical removal of bilaterally impacted teeth in the coronoid process and was rehabilitated with a dental implant-retained fixed prosthesis in the mandible and over-denture in the maxilla<sup>17</sup>.

The most suitable site of insertion seems to be the anterior mandible; insertions in the maxilla should be avoided or at least should not cross the midline. In order to determine the optimal individual time point of implant insertion, the status of skeletal growth, the degree of hypodontia and extension of related psychosocial stress should be taken into account in addition the status of the existing dentition and dental compliance of the pediatric patient<sup>18</sup>.

However, removable partial or complete dentures require regular adjustments and should be replaced when a decreased vertical dimension of occlusion and an abnormal mandibular posture are detected due to growth.<sup>19</sup>

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.<sup>20</sup>

### Conclusion

Ectodermal dysplasia is very rare condition and management of clinical manifestations presents a unique challenge for dentist. Treatment of such patients with removable partial or complete denture or implant supported denture improves function, speech, esthetics and psychosocial condition. However, their long-term success depends on regular oral hygiene and follow up maintenance.

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Legend Figures



Fig. 1: OPG



Fig. 4: Implant supported mandibular denture



Fig. 2: mandibular ridge with implant



Fig. 3: Fixed prosthesis with maxillary teeth