

## A COMPLETE PROSTHODONTIC SOLUTION FOR PATIENT WITH ECTODERMAL DYSPLASIA: A REVIEW

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### Abstract

The management of the patients with ectodermal dysplasia has always been a challenge for clinicians because of very thin ridge and complete or partial anodontia. Dental management of patient with ectodermal dysplasia is multidisciplinary team approach. Prosthetic Management of patients with hypohidrotic ectodermal dysplasia includes complete dentures, removable partial dentures, fixed partial dentures, and endosseous implants. Removable prosthesis is the common treatment options for the patients with ectodermal dysplasia. Implants and fixed partial dentures cannot be given in a growing child as it can obstruct the craniofacial growth. Hence, Prosthodontic treatment of each patient should be customized according to the patient's age and clinical situation. This article describes various treatment options for the patients with ectodermal dysplasia.

the first time, and hidrotic type shows autosomal dominant pattern of inheritance<sup>2</sup>.

Patient with Ectodermal dysplasia presents with a classic triad of hypohidrosis, hypotrichosis and hypodontia<sup>4</sup>, i.e they have soft, smooth, thin dry skin, Scalp and body hair are sparse and fine textured, diminished or missing sweat glands cause heat intolerance and hyperpyrexia<sup>5</sup>. In patients with ectodermal dysplasia alveolar bone is missing and a very thin ridge is surrounding a rare tooth<sup>6</sup>.

Hypohidrotic type is the most common type of ectodermal dysplasia which affects the patient's aesthetics often, which makes prosthodontic rehabilitation very important. Prosthodontic rehabilitation of ectodermal dysplasia patient will improve patient's function and esthetics and also provide patients with confident poise in society. Prosthodontic treatment of each patient should be customized according to the patient's age, growth and development<sup>7</sup>.

This review article is aimed at prosthodontic rehabilitation of patients with ectodermal dysplasia.

### Introduction

Ectodermal dysplasia(ED) is a hereditary dysplasia of one or more ectodermal structures, which was first described by Thurman<sup>1</sup>. At present there are around 170 types of ectodermal dysplasia, hidrotic ectodermal dysplasia and hypohidrotic ectodermal dysplasia are the two main groups<sup>2</sup>.

Hypohidrotic is also called anhidrotic ectodermal dysplasia or Christ-Siemens-Touraine Syndrome, hidrotic ectodermal dysplasia is also known as Clouston syndrome<sup>1</sup>. Major difference in these two types is sweat gland manifestation, hypohidrotic ectodermal dysplasia (HED) show drastic reduction in the number of sweat gland or it may have defective sweat glands<sup>3</sup>. Hypohidrotic ectodermal dysplasia show x-linked recessive type of inheritance, this pattern of inheritance was described by "Darwin" for

### Search strategy:

PubMed/MEDLINE and google search were conducted using the terms – Ectodermal dysplasia, anodontia, complete denture prosthesis, removable partial denture prosthesis and over-denture prosthesis, fixed partial denture prosthesis and implant retained prosthesis, as well as combinations of these and related terms. Then few articles judged to be relevant were reviewed.

Multidisciplinary approach for management of ectodermal dysplasia:

Patients with ectodermal dysplasia will have to visit the dental clinic at a very young age<sup>8</sup>. According to Till and Marquasz children suffering from ectodermal

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dysplasia should be provided with initial prosthesis before they begin going to school, so that they have adequate time to adapt to the prosthesis<sup>9</sup>. Dental management of patient with ectodermal dysplasia is multidisciplinary team approach with the clinical knowledge of growth, development and behavior management<sup>10, 11</sup>. Pedodontics, prosthodontics, orthodontics and maxillofacial surgery are the dental faculties that play an important role in the treatment of patient with ectodermal dysplasia<sup>11</sup>.

Early treatment of children with ectodermal dysplasia is a combined effort of prosthodontist and pedodontist, as pedodontists are expertised in the psychological management of children<sup>11</sup>. Behavioral management techniques like "tell-show-do" can be used in training young patients to overcome their fear, anxiety and to establish a trustworthy relationship with them. Sedation may be used in Uncooperative young patients for extensive prosthodontic Procedures<sup>10</sup>. Prosthetic Management of patients with HED includes complete dentures, removable partial dentures, fixed partial dentures, and endosseous implants<sup>12</sup>.

Removable prosthesis (complete denture prosthesis, removable partial denture prosthesis and over-denture prosthesis):

Removable prosthesis which includes Complete dentures, partial dentures and over-dentures are the most frequently used treatment modalities for the dental management of children suffering from ectodermal dysplasia in growing age<sup>13</sup>. Patients with ectodermal dysplasia will have complete or partial anodontia, which will lead to hypoplastic alveolar bone, which in turn will, lead to the lack of development of jaw(s) and a reduced occlusal vertical dimension (OVD)<sup>14</sup>.

Conventional removable prosthesis will be the best options for the young patients with ectodermal dysplasia as they require repeated change of prosthesis as the patients grow<sup>10</sup>. But the over-denture is advantageous as it helps in preserving the alveolar ridge for future implant placement and utilizes the natural undercuts in the teeth for retention<sup>10</sup>.

Fabrication of the prosthesis involve making of impression, recording jaw relation and all conventional steps involved in the fabrication of complete dentures<sup>15</sup>. Making of impression, obtaining accurate jaw relation is challenging in a

child patient<sup>15</sup>. The OVD is collapsed in patients with ectodermal dysplasia and the removable prosthesis should restore the OVD to normal. Esthetics, phonetics, freeway space, and swallowing should be analyzed using Conventional methods<sup>15</sup>. Centric relation should be recorded using fast setting materials<sup>15</sup>. Due to the limited advantage and cumbersome procedure, face bow transfer and protrusive records are not necessary in denture fabricated for children; an accurate centric relation record will be sufficient<sup>15</sup>.

Use of lingualized and bilaterally balanced occlusion schemes have been reported in the literature. Bidra et al, recommend a monoplane occlusion due to its simplicity and it will provide the child with freedom of mandibular movement which will allow uninterrupted growth of child<sup>15</sup>.

Patients with ectodermal dysplasia will have under-developed maxillary tuberosities, alveolar ridges and dryness of the oral mucosa, these factors will compromise resistance and stability of the dentures. When fabricating dentures for these patients, denture base must be extended optimally to obtain a wider distribution of occlusal loads<sup>3</sup>. Removable prosthesis requires regular adjustments and should be replaced when a decreased vertical dimension of occlusion and an abnormal mandibular posture are detected due to growth.<sup>3</sup>

Ildiko Tarjan treated 2 young boys suffering from hypohidrotic ectodermal dysplasia. They had partial maxillary and complete mandibular anodontia of the primary dentition. Removable maxillary partial and mandibular complete dentures with individualized occlusion and age-appropriate artificial teeth were employed in the treatment. This resulted in significant improvements in speech, masticatory function, and facial esthetics, contributing to the development of normal dietary habits, and the improved and more rapid social integration of these children<sup>12</sup>.

Candice Zemnick stated that complete dentures and implants could impede normal maxillary and mandibular growth. So complete denture should be given in patients with ectodermal dysplasia only when abutment teeth cannot take up the load or when patient is completely edentulous<sup>16</sup>.

Fixed prosthesis (fixed partial dentures and implant retained prosthesis):

*Conventional fixed prosthodontic treatment* should not be used in children with hypohidrotic ectodermal dysplasia as they have a minimal number of teeth. The fixed partial dentures (FPDs) with rigid connectors should not be used in young, growing patients<sup>17</sup>. Individual crown restorations can be used, but larger pulp sizes and shorter clinical crown heights will be of concern<sup>18</sup>.

*Implant retained prosthesis:* In the last decades oral rehabilitation of partially or totally edentulous patients with dental implants has become common practice<sup>19</sup>. But implants should not be placed in growing children before completion of craniofacial growth as it will lead to several problems in routine practice<sup>20</sup>. This is especially crucial in the maxilla, where implants can be submerged by the downward growth of investing tissues<sup>21</sup>.

At a Scandinavian Consensus Conference in Sonkoping, Sweden, there was a general conformity that implant placement should be postponed until skeletal growth is completed or nearly completed in normal adolescents<sup>22</sup>. Sennerby et al demonstrated, endosseous implants placed in young pig behaved like ankylosed teeth and arrested both eruption and alveolar bone growth and will not adapt to changes secondary to alveolar bone growth leading to unpredictable implant dislocations during growth<sup>20,23</sup>. If, implants are fixed together with fixed prosthesis crossing the mid line, maxillary growth disturbances will occur, as transversal growth of maxilla occurs at mid palatine suture<sup>20</sup>. The anterior mandible was considered as a site for implant placement during active growth, as the lateral growth of the anterior mandible is usually completed by 3 years of age and transversal skeletal or alveolo-dental changes are less remarkable in the mandible<sup>24,13</sup>.

But Cronin et al and Oesterle et al discussed the possible consequences of implant placement in growing individual, which included implant submergence caused by mandibular growth, implant exposure caused by bone resorption associated with mandibular growth, implant movement caused by mandibular growth, and inhibition of mandibular growth, especially, if the implants are connected by a rigid prosthesis that crosses the midline<sup>7</sup>. Impending risks included: loosening of the implants; decementation of the interim crowns; aspiration of the prosthetic parts; plaque

accumulation, if inadequate oral hygiene is performed; and interference with the patient's skeletal growth if a close follow-up schedule is not kept. Repeated refabrication of the prosthesis according to the patient's growth should also be informed.

Mark Pigno et al found that immediate implant placement was beneficial to preserve alveolar bone and those implants which were placed at an age of 15 years in girls and 18 years in boys showed a good prognosis<sup>25</sup>. Alan K.W. Yap et al conducted survey on implant survival and found that Implant survival rates vary between 88.5% and 97.6% in patients with ED<sup>26</sup>. Guckes et al conducted clinical trial evaluated the survival of implants placed in individuals with a form of ectodermal dysplasia and severe hypodontia. They found that Of the 243 implants placed in the anterior mandible, 221 (91%) survived. Of the 21 implants placed in the anterior maxilla, 16 (76%) survived. The results of this study support the continued use of endosteal dental implants in this patient population with appropriate precautions in the maxilla<sup>20</sup>.

#### **Discussion**

Prosthodontic management of a young patient with ectodermal dysplasia will have positive effect, as ability to look and feel like one's peer is essential for psychological development of the child<sup>27</sup>. Multidisciplinary approach in patients with ectodermal dysplasia will help to regain lost function and esthetics<sup>28</sup>. But there are few divisive questions regarding the treatment of patient with ectodermal dysplasia like resorption of the alveolar ridge, if removable prosthesis is given and submergence of implant due to the skeletal bone and alveolar process growth<sup>8</sup>.

According to Hickey Cosmetic and prosthodontic treatment should be started by the age of 4-5 years to help the child look like peer. Prosthodontic treatment can be initiated at the early age of 2-3 years if the child is co-operative<sup>29</sup>. Early prosthodontic treatment enhances Masticatory muscles tonicity, delays alveolar bone resorption associated with the absence of teeth, compensates for the decrease in vertical dimension and prevents angular cheilites<sup>8</sup>. Sagittal and vertical skeletal relationship during craniofacial growth and development are improved by prosthetic rehabilitation of patient with ectodermal

dysplasia<sup>30</sup>. Removable partial dentures or complete dentures, fixed partial dentures, over-dentures and implants are the prosthodontic treatment options for children with ectodermal dysplasia. These approaches used either individually or in combination to provide optimal results<sup>31</sup>.

Considerable improvement is seen in patients with HED with the use of endosseous implants in comparison with conventional prosthetic methods<sup>32</sup>. But according to Sennerby et al. 1993, endosseous implants placed in young pigs act like ankylosed teeth, implants did not participate in growth processes, resulting in multidimensional dislocation and infraocclusion compared with the developing teeth<sup>20</sup>. Yap and Klineberg found that implants placed in ectodermal dysplasia patients below the age of 18 years had a higher failure risk and implants placed in adolescent patients with ectodermal dysplasia did not show significant effect on craniofacial growth<sup>33</sup>. Thus, implant should be considered as treatment option in patients where craniofacial growth is complete.

Fixed prosthodontic treatment should be considered rarely in the treatment of ED. Primarily because many children affected will have a minimal number of teeth and are often quite young when first treated<sup>33</sup>. Fixed partial dentures with rigid connectors should be avoided in young, growing patients<sup>33</sup>. Fixed partial dentures with rigid connector will interfere with jaw growth, especially if the prosthesis crosses the midline<sup>33</sup>. Individual crown restorations can be given, but larger pulp sizes and shorter crown heights may cause concern<sup>33</sup>.

Removable prosthodontics is a common mode of treatment in ED<sup>10</sup>. Complete denture and removable partial denture can provide an acceptable aesthetic & functional result. There may be compromised denture retention & stability due to underdevelopment of the edentulous alveolar ridges<sup>34</sup>. Selection of right impression technique and occlusal scheme which provides broad distribution of occlusal load will help to overcome the problem of retention and stability to considerable extent<sup>8</sup>. Over dentures and removable partial dentures supported by natural teeth are considered as treatment options for preservation of alveolar bone. Prosthesis in the growing age may need a periodic renewal as the child grows and a definitive prosthesis is fabricated<sup>35</sup>.

## Conclusion

Ectodermal dysplasia patients with anodontia should be treated at the earliest, which will help the patient in improvement of the aesthetic, function and phonetic. Treatment options for the patients should be considered based on the clinical situation, number of teeth present, age of patient and the alveolar bone development. According to literature available, implant placement and fixed prosthesis with rigid connector crossing the mid line should be postponed till the craniofacial growth is completed in young patients. Thus removable prosthesis with periodic renewal becomes the treatment of choice in growing patients with ectodermal dysplasia.

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