

# Ectodermal Dysplasia: Prosthodontic Considerations

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## ABSTRACT

Ectodermal dysplasia is a congenital hereditary disorder associated with abnormal development of ectodermally-derived organs including teeth, nails, hair and sweat glands during embryonic growth. The most common oral finding being the Hypodontia of the primary as well as the permanent dentition. Therefore, affected patients need prosthetic treatments during their developmental years. Treatment options for such challenging situations includes fixed, removable, or implant prosthodontics, either singly or in combination. Dental function and esthetics in these patients are extremely challenging due mainly to impact on the patients' self-esteem.

**KEYWORDS:** Ectodermal Dysplasia, Oral Rehabilitation, Removable Prosthesis

## INTRODUCTION

This syndrome was first reported in 1792 by Danz and Wedderburn found in inbred Indian group in 1838.

According to NFED which is The National Foundation for Ectodermal Dysplasia Ectodermal Dysplasia is defined as a genetic disorder in which there are congenital birth defects of 2 or more Ectodermal structures. These structures may include skin, hair, nails, teeth, nerve cells, sweat glands, parts of the eye and ear / parts of other organs.<sup>1</sup>

According to NFED there are 20 common types of the disorders and the severity differs, even among the people affected with the same type of ED.

## TYPES

They can be mainly classified into 3 categories:

1. Hidrotic (autosomal dominant)
2. Hypohidrotic (anhidrotic, X-linked recessive)
3. Others : Rothmund Thomson syndrome, Chondroectodermal dysplasia, Coffin-Siris syndrome.<sup>2</sup>

## INHERITANCE PATTERN

- Usually transmitted as an X-linked recessive trait
- Gene carried by female & manifested by male

**Can occur in association with other anomalies like:**

- Cleft palate, Cleft lip,
- Palatal paralysis,
- Dwarfism,
- Congenital hypothyroidism,
- Mental retardation,
- Diabetes insipidus,
- Agglobulinemia
- Allergy disease

- Hearing loss
- Ocular manifestations

## CLINICAL FEATURES

- Absence or incomplete or delayed development of one or more appendages derived from epidermal tissue or oral ectodermal origin during embryogenesis
- Abnormal hair – scanty, fine, light hair on scalp & eyebrows (hypotrichosis)
- Abnormal or missing teeth (hypodontia, oligodontia or anodontia)
- Abnormal nails (onchodysplasia)
- Abnormal or missing sweat glands (dyshidrosis)
- Other associated features include pronounced supraorbital ridge & frontal bossing, depressed nasal bridge, protuberant everted lip due to decreased facial vertical dimension.<sup>3</sup>

Oral findings often are significant which may include anodontia, Hypodontia, malformed and widely spaced peg - like teeth, protuberant lips, loss of occlusal vertical dimension, and underdeveloped alveolar ridge.<sup>4, 5</sup> With little or no dental support, a hypoplastic maxilla and mandible result in bite collapse and narrowing of the alveolar ridges.

## PROSTHODONTIC TREATMENT MODALITIES

Several prosthodontic treatment procedures like, Complete Denture, Removable Partial Denture, Overdentures, Fixed partial dentures, and Implants can be used individually or in combination. The ideal treatment is by a multidisciplinary approach. Though there is no definitive time to begin treatment, but for the child to

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have a normal appearance and to get adapted to the prosthesis, it is advised to deliver the initial prosthesis before the child begins school. And there is a need for regular adjustments or replacements in growing children<sup>6</sup>.

## TREATMENT CONSIDERATIONS

### Treatment include:

1. Fixed prosthetic option.
2. Removable prosthetic option.
3. Implant prosthetic option.

**Fixed Prosthetic Option:** It is seldom used because of the presence of only minimal number of teeth. Patients are quite young & FPD's with rigid connectors should be avoided in young, actively growing patients as rigid FPD's interferes with jaw growth, especially if prosthesis crosses the midline. Hence, Individual crown restorations can be done, but larger pulp sizes and shorter crown heights have to be considered for either doing root canal treatment or the reduction of teeth. In early teens, orthodontic treatment is indicated as consolidation of spaces may better prepare mouth for FPD or implants in future, they fall within this parameters.

**Removable Prosthetic Option:** This is the most frequently used treatment modality as it is safe and provides time to treating phases besides being cost effective ( Fig 1)<sup>7</sup>. Due to under developed edentulous ridges, there will be a compromise with complete denture retention & stability. If the ridge is insufficient for complete denture support, vestibuloplasty & ridge augmentation should be considered. Periodic recalls will be necessary for young patients for modification of prosthesis or replacement. As the child matures removable prosthesis is relined, rebased or remade to accommodate growth changes & maintain function like speech, chewing & swallowing.



Fig 1 (a) Pre-operative view (b) Post-operative view

Flexible dentures are the latest treatment of choice as it is very easy to remove and insert (Fig 2)<sup>8</sup>. In these kind of dentures there is no need for tooth or tissue preparation



Fig 2 a) Preoperative intraoral view (b) Post insertion (flexible denture in maxillary and complete denture in mandibular arch)

for gaining retention. Retention is achieved by the denture base material by using RetentoGrip tissue bearing technique for retention. Flexible dentures are the good alternative to the conventional cast partial dentures because they are light weight, have good retentive and fracture properties and also esthetically accepted by the patient.

**Implant Prosthetic Option:** 90% success rate at second stage surgery in ED patients 13 to 69 years (Guckes et al. in National Institute of Health) is seen. There is an improvement in physiologic & psychosocial function of adult patients. Implant treatment also has a beneficial impact on the preservation of alveolar bone.

Root form implants are most widely used in prosthodontic management (Fig 3)<sup>9</sup>. In treatment planning for implant dentistry in these patients, extra care must be taken to determine whether the available bone is adequate to receive the implants. Also, the vertical dimension of bone to support the implants need to be adequate. A cone beam tomography will be ideal to determine the quantity and quality of bone. Diminished bone volume may limit the success of implants, especially in the maxilla.<sup>10</sup>

While planning for implant treatment in young ED patients, chronological age of the patient should be the determining factor than their dental and skeletal maturity.

There is a concern regarding placement of implants in a growing jaw. Guckes et al. stated that implant placement should be postponed until age 13. A 1989 Consensus Conference on Implantology concluded that the maximum jaw growth occurs by the age of 15 years, and hence the implants placement should be done after maximum jaw growth has occurred. An individual's growth curve should be studied before any implant placement procedure is commenced.<sup>10</sup>

Cronin et al. and Oesterle et al stated that the possible consequences of early implant placement can include implant submergence and implant movement both of which occur due to jaw growth, implant exposure due to bone resorption associated with jaw growth, and there will be limitation of jaw growth if the implants are connected by a rigid prosthesis that crosses the midline.<sup>11</sup>



Fig 3 a) Patients' occlusion before treatment (b) Definitive cement-retained restorations in place (c) Preoperative panoramic radiograph (d) Postoperative panoramic radiograph of osseointegrated implants.

**Overdentures:** Overdentures are amenable to long-term maintenance and allow progressive changes to be made to the prosthesis. Implant-supported overdentures are also the best treatment option. (Fig 4)<sup>12</sup>

**There are several advantages of Overdentures:**

- Improved support & stability
- Superior restoration of vital oral functions of speech, swallowing chewing with improved comfort
- Increased sense of security
- Preservation of alveolar bone
- Preservation of tooth structure & psychological support for patients

Oral and prosthesis hygiene are important to prevent caries. Fluoride should be used once per day. Early treatment allows a child to develop normal forms of speech, chewing & swallowing, normal facial support & improved TMJ function.

In early teens, orthodontic treatment is indicated as consolidation of spaces and prepare mouth better for FPD or implants in future.



Fig 4 a)Preoperative showing complete anodontia, (b) implants placed in mandible, (c) with maxillary complete denture and mandibular over denture

## CONCLUSION

A multidisciplinary team approach is needed for the dental management of the condition. As the treatment begins at the very young age, any prosthesis made must be monitored for regular adjustments or for a replacement prosthesis made necessary by growth and development. A 6- to 12-month recall is advised till the complete skeletal growth. Treatment can involve fixed, removable, or implant prosthesis on either jaw. Young children with anodontia have difficulties in eating and speaking. These patients have the psychological effect that their appearance is different than others. The use of dental

implants in the rehabilitation of ED patients can provide excellent support for dental rehabilitation, both functionally and esthetically once growth is completed.

The clinician should have a thorough knowledge about growth and development and the need for adjustments or replacement of the prosthesis as dictated by growth and development. The clinician should also have behavioral management skills and the ability to motivate the patient in the use of the prosthesis. Regular long-term follow-up for the modification and/or replacement of the prosthesis is crucial.

The desire to be like others who have teeth can motivate even the young child. The dentist has a significant role to play in the overall development and well-being of a child with HED.

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