# PROSTHODONTIC MANAGEMENT OF ECTODERMAL DYSPLASIA PATIENT WITH DENTAL IMPLANTS: A CASE REPORT

**Varun Arya[1], Bhupender Yadav [2], Dayashankar Rao J K [3] Akshay Bhargava[4], Anil Sheorain[5], Manoti Sehgal[6]**

1. Senior lecturer, Dept. of Oral surgery, Faculty of Dental Sciences, SGT University, Gurgaon
2. Reader, Department of Prosthodontics, Faculty of Dental Sciences, SGT University, Gurgaon.
3. Dept. of Oral surgery, Faculty of Dental Sciences, SGT University, Gurgaon
4. Director PG studies, ITS Dental College Greater Noida.
5. Dept. of Oral surgery, Faculty of Dental Sciences, SGT University, Gurgaon
6. Dept. of Prosthodontics, Faculty of Dental Sciences, SGT University, Gurgaon

## ABSTRACT

Ectodermal dysplasia syndrome is a large heterogenous group of inherited disorder in which two or more ectodermally derived anatomic structures fails to develop. These include skin, hair, nails, teeth, sweat glands, nerve (neural) cells and constituent parts of the ear and eye. A common oral finding in these patients is oligodontia, which results in underdeveloped alveolar process with highly resorbed ridges. This unfavorable ridge anatomy can hamper the prosthetic rehabilitation with conventional procedures in these cases. Thus the purpose of this report is to illustrate the oral rehabilitation of a 24 year old male patient affected by ED and treated with implant placement in mandible and flexible dentures in maxilla. Fixed implant-supported cement retained prostheses in the mandible were delivered to the patient, which improved his self-esteem and quality of life.

**Keywords:** Ectodermal Dysplasia, Dental Implants, Cement retained Prosthesis

## INTRODUCTION:

Ectodermal dysplasia syndrome is a large heterogenous group of inherited disorder in which two or more ectodermally derived anatomic structures fails to develop. These structures primarily are skin, hair, nails, teeth, and eccrine glands. It was first identified by Charles Darwin in 1860s. The disorders are congenital, diffuse and non progressive with more than 150 different subtypes. The most common within the group is hypohidrotic ectodermal dysplasia[1]. It has an X linked inheritance pattern and shows male predominance.

Any structure derived from the ectoderm can be defective in ED. Affected individual typically displays heat intolerance because of reduced number of sweat glands. Other signs of this disorder include fine, sparse blonde hair, including a reduced density of eyebrows and eyelash hair[2]. The skin of

Corresponding Author: Dr. Varun Arya Email: drvarunarya@yahoo.com

affected children is lightly pigmented and appears thin and almost transparent; surface blood vessels are easily visible. Pigmentation is heaviest around the eyes (usually wrinkled) and on the elbows, palms, and soles, with the latter 2 areas hyperkeratotic in nature[2]. The skin is usually dry, scaly, and easily irritated as a result of poorly developed or absent oil (sebaceous) glands[2]. Mid face hypoplasia is frequently observed often resulting in protuberant lips. Salivary glands also show less activity and patient suffers from xerostomia. The nails may also appear dystrophic and brittle. Dental abnormalities have been associated with 80% of cases and may include anodontia or hypodontia. The crown shapes are characteristically abnormal and the incisor crown usually appears tapered, conical or pointed and the molar crowns are reduced in diameter[3]. Even when complete anodontia exists growth of jaws is not impaired. Hypodontia is associated with lack of development of the alveolar ridge and results in less volume of bone for support of conventional prostheses and also reduced vertical dimension resulting in protuberant lips[3]. In addition the palatal arch is frequently high and a cleft plate may be present. It is not uncommon for the face of an affected child to take on the appearance characteristic of old age.

Conventional prosthodontic treatment for ED has consisted of various combinations of overdentures, complete or removable partial denture, or fixed patial dentures. Numerous case reports have discussed the use of such traditional approaches for ED patients[4]. However these conventional prosthesis are associated with poor retention and stability due to resorbed, thin alveolar ridges and conical teeth. Poor retention and stability may cause progressive resorption of alveolar bone and lead to subsequent prosthetic problems.

In the recent years endosseous implants have become recognized alternative for ED patients[5-7]. At the time of second stage surgery, rates of clinical immobility for implants placed in ED patients have been shown to be comparable to those for non ED edentulous patients. For both adults and children the stability offered by implant supported prosthesis results in better function and esthetics and may avoid the drawbacks of normal dentures.

Orofacial rehabilitation of ED patients can be benefited from the modern treatment concepts. These may include the use of dental implants, bone grafting and advanced prosthodontic procedures. The purpose of this paper is to present a case report of implant placement in a patient 24

years of age, subsequent prosthodontic treatment and 2-year follow-up.

**CASE REPORT:**

A 24 year old male patient who had been diagnosed with hypohidrotic ectodermal dysplasia at the age 4 reported to department of prosthodontics for replacement of missing teeth. The patient was otherwise in good general health and there was no significant medical history. Extraoral examination revealed sparse hair, depressed nose, small and retrusive malar and maxillary region, lip thickening and prominent chin (fig.1). The patient had severe intolerance to heat, which may be attributed to under developed sweat glands.

Clinical and radiographic examination revealed significant underdevelopment of alveolar bone and severe hypodontia in maxilla, with only 5 teeth present in the arch (12, 13, 14, 22 & 26). The anterior teeth present were malformed and conical in shape with abrasion present in cervical part of the coronal portion. Bone support around the teeth was good with a crown root ratio of 2/3. There was deposition of plaque, presence of bleeding on probing and gingival inflammation in relation to the teeth present in the maxillary arch. Patient was suffering with problem of severe halitosis which may be attributed to decreased secretion of saliva. There was severe resorption of bone in mandible with inferior alveolar canal located at the superior border of the mandible (atwood order VI). There was presence of mucosal thickening and flabby tissues in relation to mandibular anterior region. As the mandible was completely edentulous, it resulted in loss of vertical dimension and forward closure of the lower jaw. Masticatory efficiency of the patient was severely hampered due to the absence of teeth. The patient had not received complete dental care because of financial constraints. Complete rehabilitation of dentition was planned and the patient was informed of the treatment necessary to restore the dentition.

The treatment plan focused on the devans principle i.e “perpetual preservation of what remain is more important than the meticulous replacement of what is missing”. Flexible denture in relation to maxilla was planned as it was not advisable to alter the existing teeth present in the arch which were already compromised in terms of shape, presence of cervical abrasion and poor gingival and periodontal health. Also flexible denture can engage the soft and hard tissue undercuts better than the conventional partial denture or cast partial dentures without putting undue forces on the remaining teeth. The mandibular arch was

deficient in both bone height and width with inferior alveolar canal lying at the superficial border in the posterior region. In the anterior interforamina region bone height was around 10-12 mm and bone width was in a range of 3.4- 3.6 mm. Since placement of implants were not possible in the posterior region without complex procedures such as nerve repositioning and iliac bone graft, 4 two piece narrow diameter implants (3 mm x 10 mm) were planned to be placed in the interforamina region. 8 unit Cement retained prosthesis was planned as a final prosthesis over the implants. Cement retained prosthesis was preferred over screw retained because stresses on implant in cement retained is less as compared to screw retained[8]. Also in this case there was severe resorption of bone and in order to restore the lost vertical dimension the crown to implant ratio would have to be increased which would put increased stresses on the implant fixtures, so cement retained restoration was preferred over screw retained prosthesis. To restore the missing posterior teeth flexible denture was planned, cast partial denture was avoided since placement of rest and clasp on the cement retained prosthesis would have put extra stresses which would either have resulted in dislodgement of the prosthesis or failure of the implants. The posterior flexible denture in the mandible was planned as an interim prosthesis for the time being. The final prosthesis would be implant retained prosthesis for posterior teeth after nerve repositioning and iliac bone grafting. The family decided to go with the treatment plan decided.

Primary impressions of upper and lower arch were made with impression compound and alginate respectively and poured with dental stone. Custom trays were fabricated and final impression were made using zinc oxide eugenol in mandible, dual impression using ZOE and alginate was made in maxilla. Denture base, occlusal rims were fabricated over the master cast. Facebow transfer was done and and cast were mounted on semiadjustable articulator using interocclusal records(fig 2). Try in was done and final denture was cured in heat cure denture base resin and insertion was done(fig 3). This complete denture will serve as a provisional restoration during the osseointegration period of implants. The denture also provided the prototype positioning of the teeth and clarified the appropriate lip and cheek support needed, along with the required vertical dimension.

The denture was duplicated in clear heat cure denture base resin, GP points were placed at the proposed implant site and CBCT was done to know the exact bone width, height and location of mental

foramen. The duplicate denture will also act as a stent during placement of implants for the positioning of the pilot drill (fig 4).

Four dental implants (3 x 10 mm) were placed in anterior mandible between the mental foramina(fig 5). Cover screws were placed and after thorough irrigation the site were closed with sutures. To minimize swelling and post surgical discomfort patient was prescribed standard course of antibiotics and pain killer for the first 48 hours after implant surgery. The patient was also advised to use antimicrobial mouthwash. Ten days later, the patient was recalled for suture removal, sutures were removed and the prefabricated lower complete and upper partial dentures were relined with soft liner and seated in patient's mouth. The patient was recalled after 3 months for implant second stage surgery, healing abutments were placed and alginate impression was made for the fabrication of special tray. Patient was recalled after one week for final impression. At the time of final impression, impression copings were placed and splinted using arch wires and pattern resin. Special tray was adjusted and open tray impression was made using polyether impression material. Master cast was obtained, denture base and occlusal rims were fabricated and jaw relations were done. Mounting was done on a semiadjustable articulator using interocclusal records. Abutments were placed on the cast and a one piece nickel chromium casting was fabricated and metal try in was done(fig 6, fig 7). The final metal ceramic prosthesis was fabricated using previously determined arch form and vertical dimension (fig 8). Flexible denture was fabricated for the maxillary arch which filled the labial and buccal spaces which were void as a result of underdevelopment of the maxilla(fig 9) .

The patient was educated about the proper methods to achieve acceptable oral hygiene after implants through models and videos. He was also instructed to continue oral hygiene visits every 4-6 months during post operative years. Patient was recalled for follow-up 1 year after the loading of implants. On clinical examination the peri-implant soft tissues were found to be healthy with no signs of bleeding on probing or pathologic probing depths. The panoramic radiograph confirmed the clinical findings (fig 10). The marginal bone levels around the implants were stable with no bone loss at the mesial and distal aspects of each implant or any peri-implant radiolucency.

**DISCUSSION:**

The treatment of patients with ectodermal dysplasia will vary according to the unique anatomic, dental, and patient realted

factors. The different treatment options to restore missing dentition for patients with ED include tissue borne complete or removable partial denture, tooth supported fixed partial dentures and implant supported prosthesis. Removable dentures can be used to restore complete anodontia as well as less severe case and are cost efficient for the patient. However removable dentures are dependent on anatomic factors such as an adequate ridges or healthy adjacent teeth. Second viable option is fixed partial denture which can be used to restore short edentulous spans, however it requires abutment teeth which are free from caries, periodontally sound and have good crown ratio. Frequently fixed partial dentures cannot be used to restore the long edentulous span often encountered in ED patients. Furthermore ED patient presents with dental dysmorphic features that limit the structural reliability of the existing malformed dentition in adequately supporting fixed partial dentures. The third treatment option available is dental implants. This treatement modality has enjoyed a high success rate and can be used to replace one or several teeth or provide retention for prosthesis in partial or completely edentulous arches with the unique advantage of conserving adjacent dentition and provide additional support for masticatory function. Various authors have described a positive outcome with dental implants in the oral rehabilitation of ectodermal dysplasia patients [9,10,11]. The hypodontia associated with ED makes these patients particularly deserving candidates for dental implant reconstruction; however, the lack of bone volume in young patients, owing to failure of development of the alveolar ridges, is a major challenge in providing implant treatment.

In partially edentulous patients who have ED, multiple implant placement is not possible because the bone height and width is not sufficient for implant insertion without advanced surgical approaches. Application of removable dentures may be the only restorative option in these patients. However, the insufficient bone support, typically amorphous tooth structure, and lack of sufficient undercut zones may affect the retention and stability of the prosthesis. In the clinical situation described in the case report the maxillary arch was partially edentulous with only 5 teeth present, also there was severe bone defects in the anterior and posterior maxillary region. Fixed treatment with implants was not possible without complex bone grafting procedure in the anterior region and sinus augmentation in the posterior region, so flexible denture was planned for the maxillary region as it

restored the lost lip and cheek support and also provided good retention and support by engaging the available soft tissue undercuts.

In the present case report mandibular arch was completely edentulous and sufficient bone was available only at the mid-symphysial area, where implants were used to provide support for the prosthesis. Guckes *et al*. [12] achieved 24-month survival rates of 91% in the mandible of patients with ectodermal dysplasia, whereas the success in the anterior maxilla was 71%. Statistically, they found that implants placed in the anterior maxilla were 2·8-fold more likely to fail than those placed in the anterior mandible; however, they caution over-interpretation of their results owing to the smaller number of maxillary implants placed. The same authors [13] had previously described the successful use of osseointegrated implants in the anterior mandible of ED patients. They reported 203 of 243 cylindrical threaded implants placed primarily in the anterior mandible of 52 patients aged between 7 and 68 years to have integrated and remain in function after 3 years of follow up. Their success rates were variable with age. The preadolescent group (aged 7–11) showed an 87% success rate, and adolescents (12–17) a 90% success rate, whereas the adult group showed a 97% success rate

An understanding of the ED patient's psychosocial status is crucial to any prosthodontic treatment effort. The unesthetic appearance that accompanies ED syndrome often has a negative psychological effect on the patient. Poor self-image, peer pressure, and school/job related discrimination have been directly related to psychological scarring experienced by ED patients. Providing expedient prosthodontic treatment to manage orofacial disfigurement may afford the patient some measure of confidence. Early prosthodontic intervention can be accomplished with a removable or implant supported prosthesis, which can affect a rapid and painless result and, at the same time, minimize the onset of emotional and psychosocial problems for the patient and his/her family.

**CONCLUSION:**

The present case report underscores the importance of considering implant assisted dental rehabilitation combining implant retained and conventional treatment options as a practical solution that can provide an acceptable functional and esthetic solution within a reasonable treatment time when dental malformations dictates the reconstructive procedures.

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**FIGURES:**

Fig 1: extra oral preoperative view

Fig 2: Facebow transfer

Fig 3: insertion of provisional prostheses

Fig 4: diagnostic stent with gutta percha

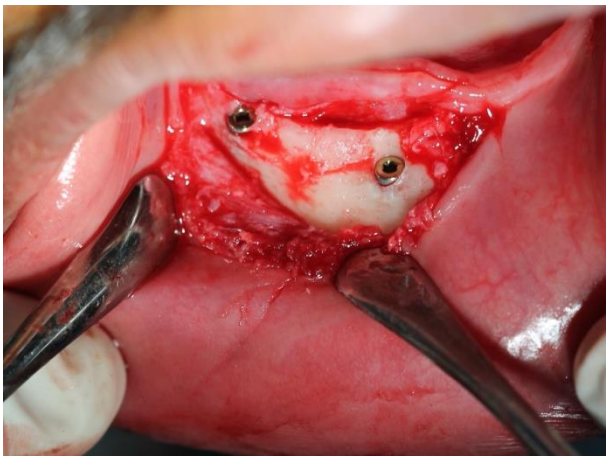

Fig 5: surgical placement of dental implants

Fig 6: abutment in place

Fig 7: metal try in done

Fig 9: metal ceramic cement retained prosthesis with flexible dentures

Fig 8: metal ceramic cement retained prosthesis

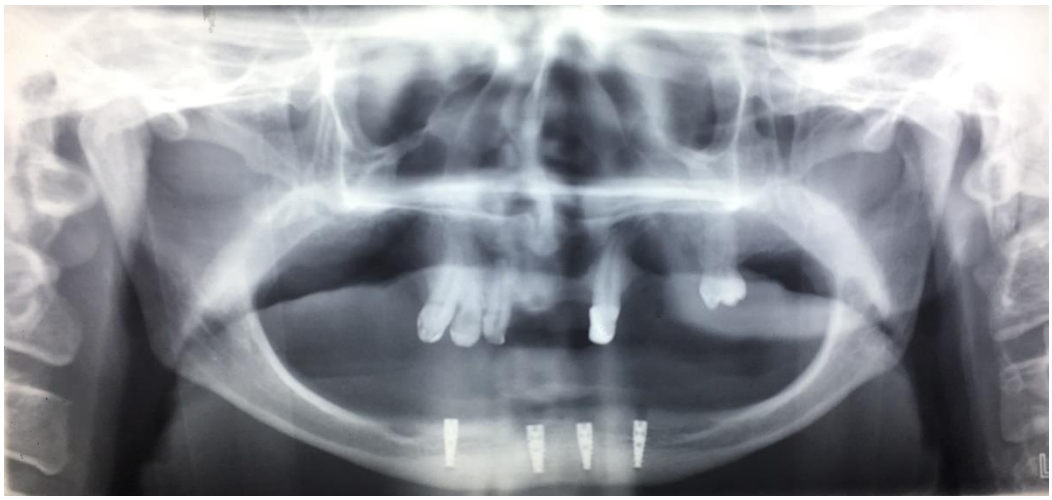

Fig 10: post-operative OPG after 1 year follow up