

Prosthetic Management of Patient with Ectodermal Dysplasia: A Case Report

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Abstract

The etiology of congenital absence of teeth is believed to be involved in hereditary or developmental anomalies. It can be isolated or as part of a syndrome. There are a number of options available to restore space created by missing teeth. Dental treatment can vary depending on the severity of the disease and usually requires a multidisciplinary approach. The aim of this report is to describe the management patient with oligodontia including few permanent teeth. A 28-year-old male patient with oligodontia affecting permanent teeth received conservative care that met his as well as his parent's expectations. The existing teeth were endodontically treated and ball attachments were fabricated and restored with overdenture prosthesis on maxillary and mandibular arch. Metal framework was incorporated in the denture to eliminate the risk of fracture. The prosthetic treatment provided psychosocial comfort for the young patient. The loss of teeth in young patients can cause aesthetic, functional, and psychological problems particularly if the teeth of the anterior region are involved.

Key words: Ectodermal dysplasia, Oligodontia, Overdenture.

INTRODUCTION

Congenital absence of teeth is a heritable phenomenon probably most often transferred to each generation by an autosomal dominant pattern with incomplete penetrance and variable expressivity.¹ Individual presents with complete or partial absence of primary or permanent tooth. Oligodontia represents the condition where there is congenital absence of more than six teeth in primary or permanent or in both the dentition. It often presents as part of a syndrome and isolated representations are rare. The genes associated with syndromic oligodontia are MSX1 and PAX9. Ectodermal dysplasia (ED) is one of the conditions commonly associated with partial or complete absence of tooth.

During the third gestational week the embryo which is in bilaminar form turns into three layered oval discs, termed as ectoderm, mesoderm and endoderm. Each cell in these three layers are further distinctly developed into different organs. Dental enamel is originating from the ectoderm. Ectodermal dysplasia consists of a group of conditions related to the abnormal development of at least 2 or 4 classical ectodermal tissues. This disorder often affects the hair, teeth, nails and sweat glands. Dry skin is more prevalent in ectodermal dysplasia than in a healthy person. The oral presentation of ectodermal dysplasia includes severe tooth agenesis and peg shaped tooth. Along with the abnormal tooth size and shape there is complete or partial absence of tooth called oligodontia. Since salivary gland is derived from ectoderm, its hypofunction is commonly seen in ectodermal dysplasia.²

Prosthetic rehabilitation of patients with oligodontia is based on the extent/severity of the condition. An interdisciplinary approach is usually required. Congenitally deformed teeth that are already present can be included in prosthetic planning. Treatment options may include orthodontic therapy, implants, adhesive techniques, and removable prostheses. Treatment of such individuals must be done with constant support and maintenance of proper maxillofacial relationship.

CASE REPORT

A 28year old male patient reported to the Department of Prosthodontics, Annoor Dental College and Hospital, Kerala, with chief complaints of poor aesthetics and difficulty in eating. Patient was aware about his diagnosis of ectodermal dysplasia and is under treatment by the general physician. He is the first-born son of a consanguineous marriage between second degree relatives. The obstetric history revealed caesarean section being done as labour failed to progress. He was full term baby with a birth weight of 2.1kg. During childhood his various developmental milestones were delayed according to his mother, and his younger brother too was diagnosed with ED at an early age. The mother also noticed complaints of heat intolerance in both her children from an early age. Patient gave a history of full complement of primary teeth which were irregular in shape. Few teeth were exfoliated and the successor teeth were conical in shape. Paternal history of mesiodens was reported.

On general examination, the patient was moderately built, adequately nourished showed, characteristic facial abnormalities including a prominent forehead, a sunken nasal bridge (so-called "saddle nose") and everted lips. His hair and nails were normal but the skin was dry and scaly, appearing prematurely aged. On extra-oral examination, the patient had a straight profile and competent lips (Figure:1). Clinical examination revealed the presence of teeth as shown in (Figure2a and b). Deep folds in the commissures of the mouth were seen indicating loss of vertical dimension of occlusion (VDO) (Figure 2c). Niswonger's method was used to verify vertical dimension of occlusion. The teeth appeared abnormal in size and shape, rotated, and without any anatomical features. Some teeth in the maxillary and mandibular arch were periodontally weak. Patient is having fractured overlay denture over the existing teeth which was fabricated 14 years back.



Figure 1: Pre-operative profile photographs

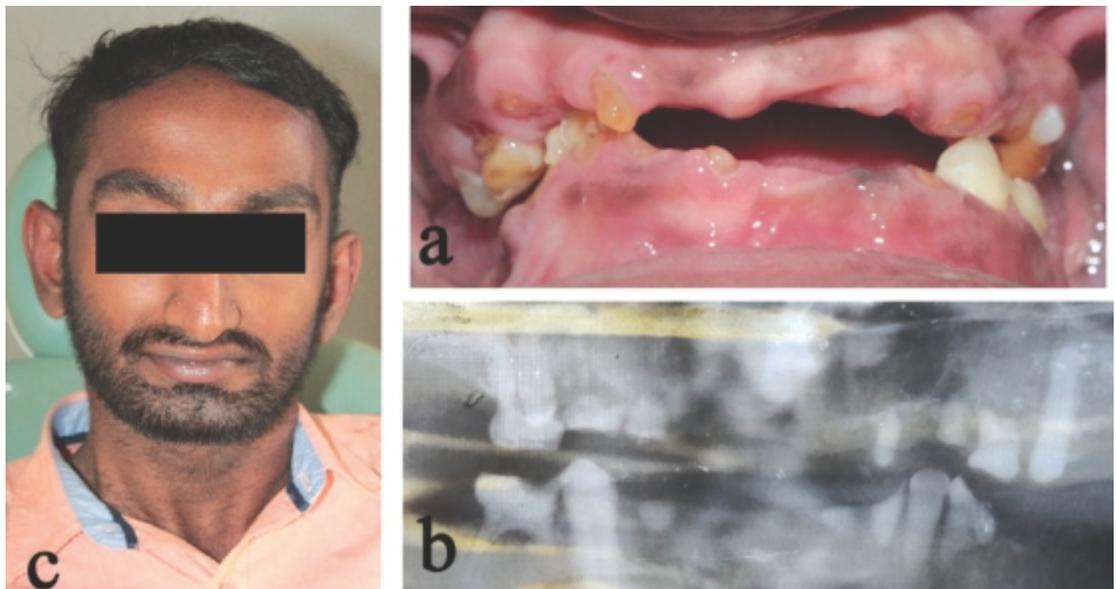


Figure 2:a) Intraoral photograph showing teeth present. b) Pre-operative OPG. c) Pre operative photograph

CASE MANAGEMENT

After the clinical and radiographic evaluation and considering the age of the individual, preservation of the tooth and bone was a priority. Since the patient's finances were limited, it was decided to rehabilitate the maxillary and mandibular arch with tooth supported over denture. It was decided to rehabilitate maxillary and mandibular arch with tooth supported over dentures. A multi-disciplinary approach was planned in association with the specialities of Oral and Maxillofacial Surgery

and Endodontics. It was decided to extract teeth with lack of periodontal support and unfavourable crown to root ratio. In the maxillary arch teeth in the first molar region of the first and second quadrant were preserved and the periodontally weak teeth in the 12,13,14,23,25 regions were extracted. On the mandibular arch the existing teeth were in the region of 32,33,34,35,43,44,46 from which teeth with favourable crown root ratio and tooth anatomy were selected as abutments.

Prosthetic management of ectodermal dysplasia

The selected abutment teeth were single rooted and conical in shape with sufficient periodontal support. In maxillary arch, teeth in respect to 16 and 26 and in mandibular arch 34 and 44 regions were taken as abutments. Intentional root canal treatment was advised for the teeth. Post space preparations were done. Post space impressions were made with direct technique using elastomeric impression material. Custom made stud attachments on dome copings with small root extensions 3-5mm in length were cemented. (Figure 3).

Primary impression for the maxillary and mandibular arch was made with irreversible hydrocolloid impression material. The impressions were poured and custom trays were fabricated with cold-cured acrylic resin. Border moulding was done for both the arches with low fusing compound. Final impression for the maxillary and mandibular arch were

made with light body elastomeric impression material. Master casts were prepared by pouring the impressions in Type IV gypsum. Copings on the master cast were covered with modelling wax and record base fabricated after applying separating media. Placement of wax over abutments prevents the fracture of the cast during removal of the temporary record base at the time of dewaxing. Occlusal rims were fabricated; maxillo mandibular relations recorded and transferred onto the semi-adjustable articulator with the help of face-bow. Teeth setting was done, evaluated in the patient's mouth for phonetics and aesthetics (Figure 3). Vertical dimension was verified; centric and eccentric contacts checked. Patient's approval was taken, and the processing of the final denture was done in heat-cured acrylic resin. Since size of the ridge was very narrow and the metal housing for the over denture would weaken the denture base, a metal

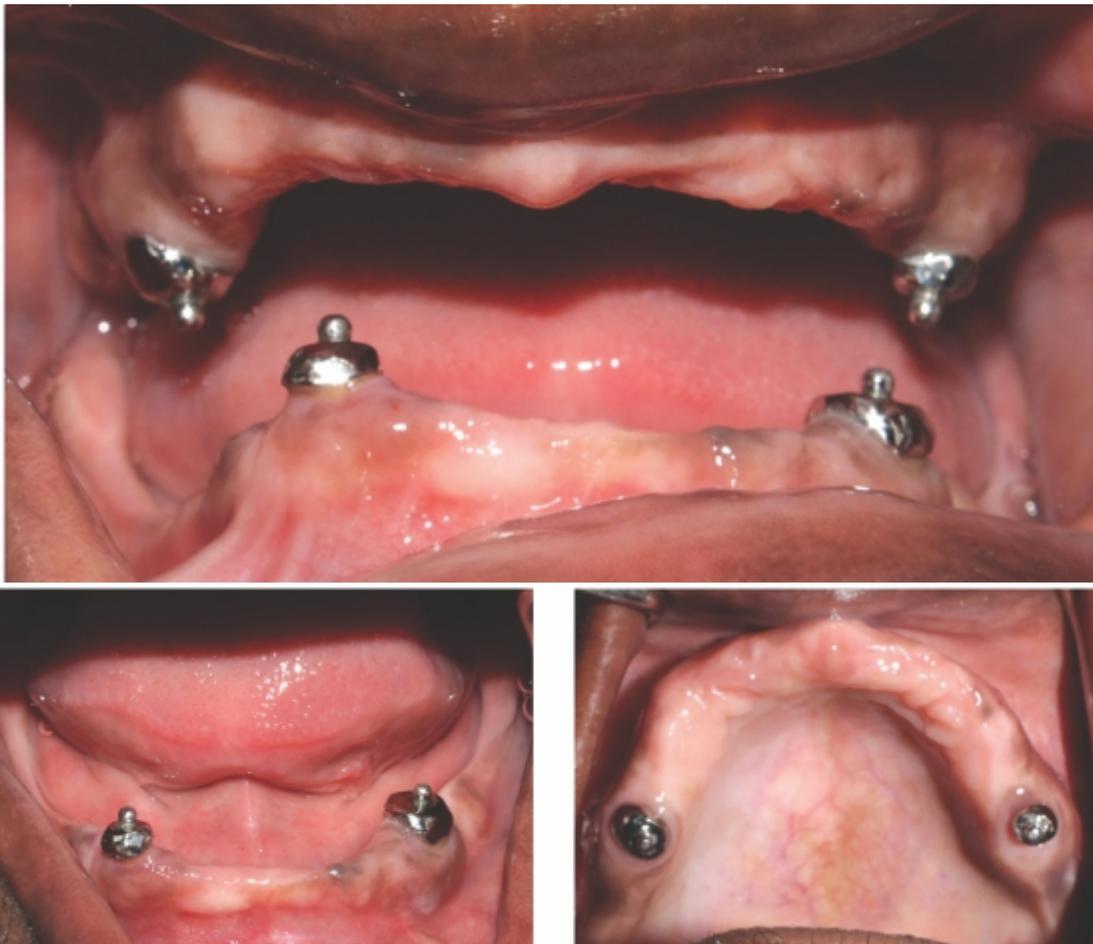


Figure 3: Custom made stud attachments over the abutment tooth.



Figure 3: Fabrication of complete denture

framework was incorporated. Processed denture was polished and finished. Denture stability, retention and support were evaluated and occlusal refinement were done at the time of insertion. Patient was recalled regularly, where denture, abutment, and supporting structures were evaluated, and oral hygiene instructions reinforced. Patient expressed his satisfaction with denture retention, function and aesthetics (Figure 4).

DISCUSSION

Any malformation or defect during the development of ectoderm can lead to a group of phenotypically distinctive entities collectively termed as the ectodermal dysplasia syndromes. These heterogeneous group of disorders with a constellation of findings mainly involve the skin, teeth and appendageal structures.³ Depending on the number and functioning of the sweat glands ED can be classified into X-linked anhidrotic or hypohidrotic, where sweat glands are either absent or significantly reduced in number and autosomal dominant anhidrotic where sweat glands are normal.⁴

Hypohidrotic ectodermal dysplasia (HED) is an uncommon X-linked condition. It is characterized by sparse hair, an inability to sweat due to the lack of sweat glands and anodontia or oligodontia with conical teeth. The two first exons of the gene responsible for this disorder, *EDA1*, were identified by Kere et al. The remaining seven exons have been later published.⁵

Since the patient has hyposecretion or absence of sweat gland, skin appears smooth, dry and thin. Facial features are more distinguishable in the patient. There was hyperkeratosis of the palms of the hands and soles. There is absence of lanugo hair, beard is normal, axillary and pubic hair is generally sparse. The hair over the scalp is mostly fine, stiff and short. Patient has ectodermal dysplasia with primary defect in skin, teeth and nails.

Individual with ectodermal dysplasia and associated oligodontia often requires an extensive and complicated treatment plan to derive a functional and conservative restoration. In such children primary objective is to retain the teeth to preserve the supporting alveolus.

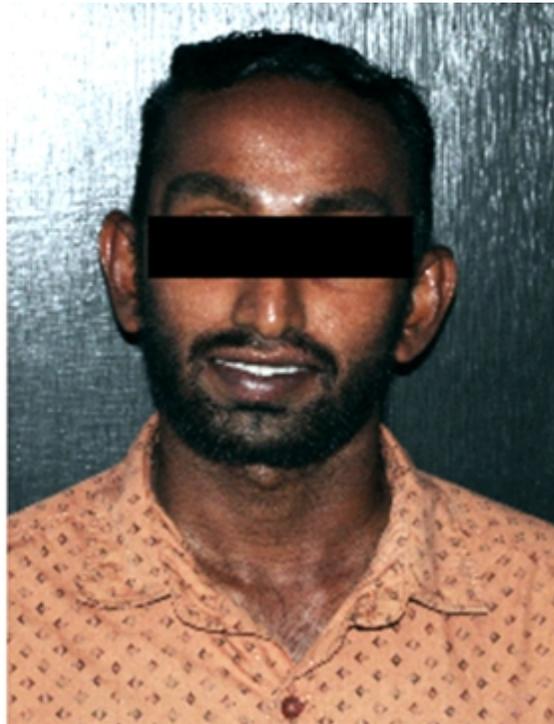


Figure 4: Pre and Post operative photographs

Such an early intervention is critical to create a functional, aesthetic and psychological stability.⁶ Removable complete denture prosthesis is a more practical option for an oligodontia patient. Tooth supported over dentures (overlay dentures) and implant supported dentures are also common options. Agarwal et al were of the opinion that it is preferable to treat children with oligodontia using overlay dentures, a reversible, relatively inexpensive and conservative method.⁷

In ED there is more prevalence of hyposalivary secretion compared to isolated oligodontia. So, the retention for a conventional complete denture is difficult to achieve in these patients. Overlay prosthesis, partially supported by natural tooth, tooth root or dental implants, is the effective treatment for patients with less number of teeth.⁸ The patient had a few remaining teeth with favourable crown root ratio which could be used as abutment for a tooth supported over denture and this lead to his current treatment option. Various attachment systems have

been used in over dentures and no single attachment system is perfect in every case, it varies depending on the interarch space and position of the abutment. The alveolar process is under developed and reduced in size so in corporation of a metal casing for the stud attachment might weaken the denture base. Hence, the final denture is fabricated with metal framework in the denturebase. Ozcelik et al reported that the incorporation of metal to the denture base may prevent or reduce the chance for over denture fracture not only by the reinforcement of the denture base with a metal framework but also the inclusion of the attachment metal housing in the framework design to prevent fractures that could occur at the sites close to the tooth abutments.⁹ Edentulous persons often prefer alternative treatment to the conventional complete denture including tooth or implant supported over dentures, the main advantages being decreased resorption of the residual ridges; psychological benefits for the patient and maintenance of masticatory efficiency.¹⁰

CONCLUSION

Ectodermal dysplasia is an uncommon hereditary defect with different craniofacial malformations. In an early stage of development, the oral problem including oligodontia should be evaluated to provide an ideal treatment. Prosthetic treatments at developmental stages can improve the functional, aesthetic and psychological development of the individual to a greater extent. Most often, treatment for oligodontia is the removable prosthesis which is considered to be most inexpensive, reversible and conservative treatment. The development of osseointegration of dental implants provides new and better possibilities. Since implant options requires a stable and structured bone this option is not recommended in children before skeletal maturity. Treatment options for oligodontia is often complicated and extensive. For a successful, healthy and functional outcome cooperation and effective communication between the dental team and the patient is of paramount importance.

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