Case Report

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Prosthodontic Rehabilitation of Anhydrotic Ectodermal Dysplasia

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ABSTRACT

Background: Ectodermal dysplasia syndrome represents a rare group of congenital disorders that exhibits at least two of the following features: trichodysplasia, onchodysplasia, abnormal dentition and/or dyshidrosis. This condition manifests mainly in two major forms: (1) Anhidrotic or hypohidrotic (Christ-Siemens-Touraine syndrome) and (2) hidrotic (Clouston syndrome). The former is more severe form and is associated with severe dental defects. Children suffering from hypohidrotic ectodermal dysplasia may benefit utmost from implant supported prosthesis in terms of their quality of life, social behavior and psychological profile. Hence, dental implants should be considered as a superior modality of management in such patients. This case report advocated the habilitation of a 10 years old child with Christ-Siemens-Touraine syndrome by means of mandibular implant supported overdenture and maxillary conventional complete denture.

Keywords: Anodontia, ectodermal dysplasia, overdenture, dental implant.

INTRODUCTION

The prosthodontic habilitation of children with anodontia is usually a complicated endeavor involving special challenges to the prosthodontic fraternity. Active search for definitive diagnosis should be accomplished in patients with oligodontia or anodontia since this condition, at times, may be related to rare disorders involving organs with ectodermal origin [1]. Ectodermal dysplasia is one such rare disorder with over 170 different subtypes and wide range of clinical features [2] demanding utmost multidisciplinary medical and dental care. The incidence of this disorder is 1 per 1,00,000 live births [3]. The syndrome represents a complex group of genodermatoses that primarily affects organs with ectodermal origin such as skin, nails, hairs, sweat glands and teeth [4,5,6]. Ectodermal dysplasia syndrome usually manifests in two forms: (1) Hypohidrotic/anhidrotic (Christ-Siemens-Touraine syndrome), and (2) Hidrotic (Clouston syndrome) [7,8,9]. The former usually inherits as X-linked recessive trait with severe oral manifestations while latter is an autosomal dominant disorder with no specific dental defect [4]. Christ-Siemens-Touraine syndrome is characterized by hypohidrosis, hypotrichosis, onychodysplasia and anomalous dentition. The condition manifests as frontal bossing, saddle nose, thick everted lips, anodontia or oligodontia, conical or hypoplastic teeth with delayed eruption of deciduous and permanent teeth. However, the clue for the diagnosis is often the unexplained fever and dry scaly skin in neonates. The intelligence level of the patients is usually normal. Ectodermal dysplasia syndrome could be attributed to mutations in genes encoding cell to cell signals, adhesion molecules or those regulating transcription [2]. The patients with total anodontia can be afforded better quality of life by way of dental implants. This case reports highlights the habilitation of a 10 year old child with Christ-Siemens-Touraine syndrome and anodontia with an *implant supported mandibular overdenture*

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using ball and socket O-ring attachments and a conventional maxillary complete denture.

CASE REPORT

A 10 year old male child was referred from a primary health center for habilitation of his dentition. The child was accompanied by his parents who gave history of child's missing teeth since birth. The parents thought that the permanent teeth would come in a matter of time. The child's awareness and insecurity among peers for the absence of teeth motivated the parents to see a dentist. The parents complained of reduced sweating and lack of tolerance to heat in child. Discussion with the parents revealed that he was getting good grades in school implying good mental health. There was no family history of this condition. He was second kid of his family with one elder sister. The child was very curious to get his teeth for the purpose of his competence and security among peers in addition to those for esthetics, mastication and speech.

Extraoral examination (Fig. 1) revealed sparse scalp hairs, frontal bossing, protuberant lips and hyperpigmented and dry scaly perioral skin; the typical features of Ectodermal Dysplasia Syndrome. The lower facial height was reduced contributing to senile facial expression. Intraoral examination (Fig. 2a) revealed completely edentulous maxillary and mandibular arches. The alveolar ridges, especially mandibular, were found to be thin and atrophic. An Orthopantomogram (Fig. 2b) was taken to confirm the absence of tooth buds in jaw bones. A Cone Beam Computed Tomogram mandible was requested as an aid in treatment planning for endosseous implants. A multidisciplinary team arrived at the diagnosis of Hypohidrotic Ectodermal Dysplasia or Christ-Siemens-Touraine syndrome with total anodontia after correlating the history, clinical and radiographic findings.

The treatment plan was then formulated to habilitate the child and was discussed with parents and child who agreed for the treatment. An informed consent was taken. The habilitation was planned with mandibular two implant supported overdenture (Overdenture Option 1) with ball and socket attachments and maxillary conventional complete denture. The endosseous implants (3.8mm × 11.5 mm, MIS system) were placed in A and E column [10] of anterior mandible with flap technique. The silk sutures (3-0 Mersilk, Ethicon) were placed and patient was recalled after one week. During this appointment, sutures were removed and thorough irrigation over implant site was done. The appointments were then rescheduled for fabrication of conventional maxillary and mandibular complete dentures. Preliminary impressions were made with putty bodied condensation silicone (Speedex, Coltene Whaledent, Switzerland) in anterior sectional trays as routine stock trays could not be inserted in patient's mouth.

The border molding was done in custom tray (with full arch spacer) with single step technique using putty consistency elastomer. The low fusing green stick compound was avoided for the reason of intolerance to hot. The final impressions (Fig. 3) were then made in light bodied silicone impression material. Special care was taken to pour the impressions immediately so as to prevent dimensional changes in set impression. This was followed by recording of maxillomandibular relationship. Face bow transfer was done using earpiece facebow and casts were mounted in Hanau Wide Vue articulator. Teeth selection was then done. It was decided to use a mold with permanent tooth forms omitting the premolars in arrangement of teeth because of reduced basal seat area available for denture. A smaller size of teeth was selected considering patient's facial form and age. Try-in was done and patient's and parent's consent for esthetics was taken.

The trial dentures were evaluated for retention, phonetics, esthetics and occlusion. The dentures were fabricated with fiber-reinforced heat cure acrylic resin (Leucitone, Dentsply, USA) using injection molding technique. After finishing and polishing of the denture, they were inserted in patient's mouth and occlusion was verified. Post-insertion instructions were given to the child and his parents. The child responded happily with the mere insertion of dentures in his mouth. The recall appointments were scheduled after 24 hours followed by one appointment every week till 4 months of implant placement. IOPA X-rays were taken to depict the changes in bone around implants. Second stage surgery was performed and

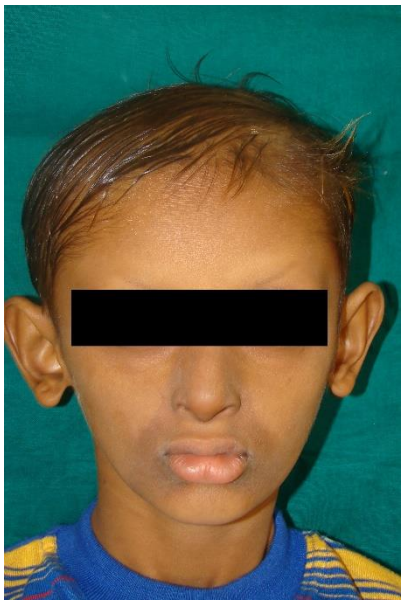

Fig 1: Pre-habilitation frontal view.

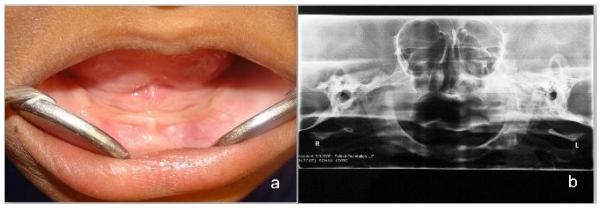

Fig 2a & b: Intraoral examination and Orthopantomogram.

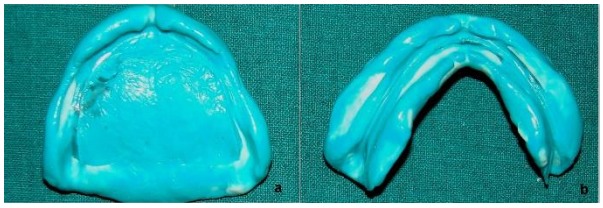

Fig 3a & b: Maxillary final impression and Mandibular final impression.

ball attachments with longer transmucosal height were screwed in place (Fig. 4a). Corresponding areas were drilled in mandibular complete denture to accommodate those attachments. The area around implants was lined in denture with a silicone based soft liner (GC Reline, GC America Inc.). After 2 weeks of progressive bone loading, the soft liner was removed and female attachments (Fig. 4b) were inserted in corresponding positions in mandibular denture using autopolymerizing resin with direct technique. The tissues were prevented from burning sensation of monomer by placing a small piece of rubber dam sheet with a small hole

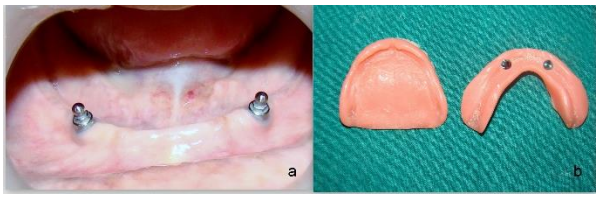

Fig 4a & b: Implants with ball attachments and Denture with female attachments.

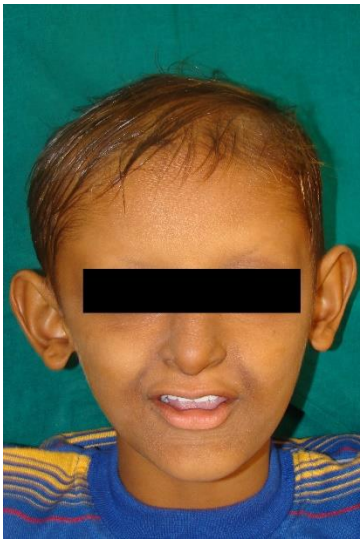

Fig 5: Post-habilitation frontal view.

onto the ball attachments. The rubber dam sheets were then removed. Every effort was made to allow the passive fit of ball and socket O-ring attachments to prevent undue stresses on implants. Further follow up appointments were scheduled after 24 hours, 1 week and every 6 months. The parents reported a real improvement in quality of his life in terms of interaction with society. Such follow ups are being done every 6 months. There is an obvious improvement in facial and dental esthetics (Fig. 5), phonetics, mastication and above all, the psychological development. The dentures were found to be acceptable even after five years.

DISCUSSION

Every patient is an individual and requires a particular treatment. A generalized treatment plan cannot be speculated for all patients [11]. The psychological development of a person is, to an extent, dependent on the teeth and face that increase his/her sense of well-being. Any dentofacial deformity usually manifests psychologically as a sense of inferiority, especially in adolescent period which may lead to inadequacy,

incompetence and a painful emotional depressive state [12]. Fiske J et al., after interviewing edentulous patients, concluded that absence of teeth deprived such persons from self-confidence leading to feeling of shame or bereavement [13]. Smile as an emotion signifies pleasure. Any aberrations in smile can be a source of anxiety for obvious reasons. Thus, patients with hereditary ectodermal dysplasia require special counselling and treatment sessions by a team of multidisciplinary medical and dental experts [1,14,15]. Prosthodontist should have acute perception of actual dental problems of patient seeking treatment [1]. The patient's own assessment of his problems and his reaction to them are equally important.

The ectodermal dysplasia manifests in full form in males. Female patients show the partial expression of abnormal gene owing to its X-linked recessive mode of inheritance. This has been explained by Lyon hypothesis with half of the female patient's X-chromosomes expressing normal gene and the other half expressing the mutated gene [2]. The female carriers of ectodermal dysplasia do not require any dental treatment but they could know that they might give birth to a child with typical ectodermal dysplasia syndrome with full expression [1]. Thus, genetic counselling for the parents and patients should be considered as a golden rule in the management of hereditary ectodermal dysplasia [1].

The diagnosis of ectodermal dysplasia is usually based on manifestations resulting from dysfunction of two or more organs with ectodermal origin such as teeth, skin, nails, hairs, salivary glands and sweat glands. The diagnosis is often not arrived at in spite of extensive prosthodontic rehabilitation [1]. Being a rare disorder with different clinical presentations, diagnosis often is overlooked by pediatricians, dermatologists, ENT specialists and dentist. Lack of function of sweat glands can be one of the first alarming feature which can be diagnosed early in infancy. Hyperthermia resulting from sweat gland dysfunction proves to be dramatic and severe. The early diagnosis is helpful because it may avoid the possibilities of hyperthermia in infancy [1]. Moreover, the growth-adapted measures can be used in treatment planning process. The salivary glands, being ectodermal in origin, may be hypoplastic or absent. Thus, the patient experiences varying degree of xerostomia. Nordgarden H et al. [16] recommended that salivary flow rate testing should be done in patients with ectodermal dysplasia syndrome.

The goal of modern dentistry is to return patients to oral health in a predictable fashion. Management of dental problems can be done by bonded restorations, fixed and removable partial dentures, complete dentures or overdentures, depending on the age and number and location of remaining dentition. The most common and reliable treatment modality for growing children is removable dentures: partial or complete as they do not interfere with growth and can be fabricated again, if required [6,17]. Fixed prostheses are contraindicated during periods of growth as they interfere with jaw growth owing to rigid connectors and their frequent dislodgement [6]. In cases with anodontia, the only options are conventional complete denture and implant-supported complete denture. Conventional complete denture often lack retention and stability because of anatomical abnormalities which justifies their use only as a temporary prosthesis to provide esthetics and function during implant treatment [18]. Hence, dental implants have been suggested to ensure esthetics, function and psychological rehabilitation of affected patients. Various authors have opined for and against the use of dental implants in growing children [5,7,8,19,20]. National Foundation for ectodermal dysplasia recommends that implants can be placed in anterior mandible in children after the age of 7 years [21]. A case report describing the placement of endosseous implant in 3 year old child has been presented in earlier literature with 5 year follow up after implant placement [19]. Implant prostheses often offer a more predictable treatment course than traditional restorations. The anterior mandible has been suggested as ideal site for implant placement as the lateral growth of mandible is usually completed by the age of three years. However, the growth of maxilla is more dramatic than mandibular growth which may result in unpredictable dislocation of implant after skeletal maturation [5]. Furthermore, it is recommended not to splint the implants by rigid bar as it may interfere with growth of jaws [22]. It is prudent to state that favorable results can be achieved by placing ball

and socket attachments over endosseous implants without any rigid splinting. Besides other routine benefits afforded, there is improvement in the quality of life of children in terms of psychological development and security [10]. Considering the above protocols, it was decided to fabricate mandibular implant supported overdenture [14] and maxillary conventional complete denture at this stage. Implant supported overdentures should be considered a standard treatment in these patients owing to reduced basal seat area and lack of salivation which further deteriorate the prognosis of conventional complete denture, especially in mandible.

Ideal sites for placement of endosseous implants for mandibular overdenture are B and D column of bone. Placement of implants in these regions reduces stress on implants [10]. However, the reduced width of bone precluded implant placement in these columns of mandible. The objective of this mode of treatment is the preservation of bone. In this case, the bone was found to be satisfactory at approximately A and E columns following Cone beam computed tomography (CBCT). So it was decided to place implants in A and E locations. Furthermore, because of reduced arch size, the rotation of denture was expected to be minimal with lesser destructive forces on implants. Prosthesis movement should be kept as much as possible to reduce leverage on implants.

SUMMARY AND CONCLUSION

Dental implants offer a predictable treatment outcome for patients with Ectodermal Dysplasia Syndrome with total anodontia. They serve as a connecting link between insecure edentulous state and a state of esthetics, function and psychological maturation of such children. Every effort should be made to habilitate the patient with dental implants after thorough diagnosis and treatment planning for available bone and other factors concerned with implantology. It is essential to monitor the growing patients of ectodermal dysplasia with regular recall visits to monitor the results and growth changes. The growth of jaws and wear of prosthesis may necessitate remaking of prosthesis at regular intervals.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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