Case Reports

JRAD Journal of Research and Advancement in Dentistry

J Res Adv Dent 2016;5:1:199-204.

Prosthodontic Rehabilitation of Patient with Ectodermal Dysplasia: Two Case Reports

Bhushan R Bangar[1*] Suresh S Kamble[2] Ajit S Jankar[3] Mohammed Asim[4]

[1]Professor, Department of Prosthodontics, MIDSR Dental College and Hospital, Latur, Maharashtra, India.
[2]Professor and Principal, Department of Prosthodontics, MIDSR Dental College and Hospital, Latur, Maharashtra, India.
[3]Professor and Head, Department of Prosthodontics, MIDSR Dental College and Hospital, Latur, Maharashtra, India.
[4]Post Graduate Student, Department of Prosthodontics, MIDSR Dental College and Hospital, Latur, Maharashtra, India.

ABSTRACT

Background: Ectodermal dysplasia is a heterogeneous group of disorders characterized by developmental dystrophies of ectodermal structures. It is characterized by the triad of signs comprising hypotrichosis, hypohidrosis and hypodontia.

Prosthetic Management of patients with hypohidrotic ectodermal dysplasia includes complete dentures, removable partial dentures, fixed partial dentures and implants. Removable prosthesis is the common treatment option for these patients. Because the disease presents in younger age, implants and fixed partial dentures cannot be planned in a growing child as it may lead to obstruction of the growth of stomatognathic system. This article describes about characteristic and prosthodontic treatment of two patients with hypohidrotic ectodermal dysplasia.

Keywords: Ectodermal dysplasia, Hypodontia, Prosthodontic rehabilitation.

INTRODUCTION

Ectodermal dysplasia is a heterogeneous group of disorders characterized by developmental dystrophies of ectodermal structures [1]. It represents a large and complex nosological group of congenital diseases that was first described by Thurman [2]. The condition is reported to occur in 1 in 1,00,000 live births. The triad of nail dystrophy (onchodysplasia), alopecia or hypotrichosis (scanty, fine light hair on the scalp and eyebrows), and palmoplantar hyperkeratosis is usually accompanied by a lack of sweat glands (hypohidrosis) and a partial or complete absence of the primary and/ or permanent dentition [3].

Oral manifestations of ectodermal dysplasia include hypodontia (or anodontia) of the primary or secondary teeth, malformed or peg like teeth, and underdeveloped alveolar ridges. Oligodontia is commonly found in patients with ectodermal dysplasia, who present with an average of 22 missing deciduous or permanent teeth, leading to decreased alveolar bone volume, presence of large edentulous spans, and poorly developed ridges. So, because of the presence of multiple anatomic and dental abnormalities, the prosthodontic treatment for ectodermal dysplasia patients is challenging and unique, necessitating a multidisciplinary approach to restoring function and esthetics [4].

About 160 clinically and genetically distinct hereditery ectodermal dysplasias have been catalogued[1]. The syndrome involves overlapping features, thereby complicating a definitive classification. Lamartine in 2003 has described various well defined ectodermal dysplasia as

Received: July. 19, 2015: **Accepted:** Nov. 15, 2015
***Correspondence Dr. Bhushan R. Bangar.**
Department of Prosthodontics, MIDSR Dental College and Hospital, Latur, Maharashtra, India.
Email: brbangar@yahoo.com

pISSN 2278-0076
eISSN 2321-9270

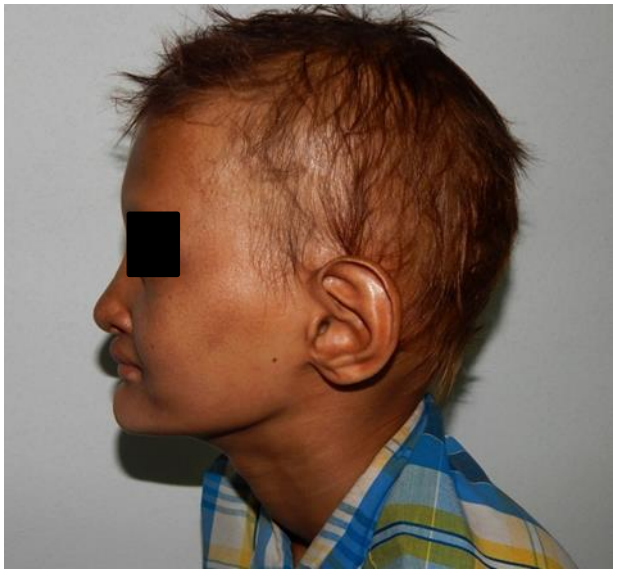

Fig 1: CASE 1 showing Lateral profile view.

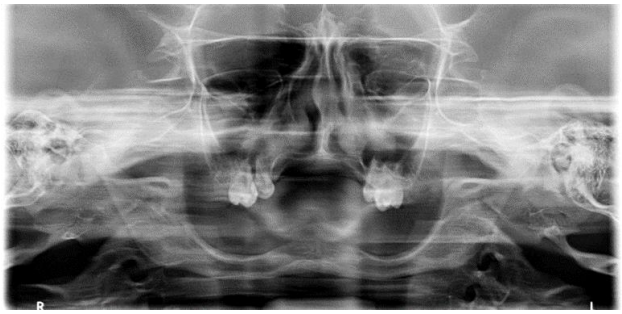

Fig 2: Orthopantomograph of patient showing absence of tooth buds.

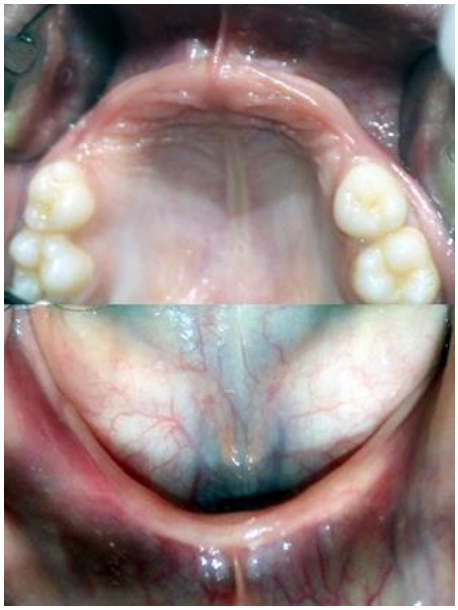

Fig 3: Intraoral photograph showing partially edentulous maxillary and completely edentulous mandibular arch.

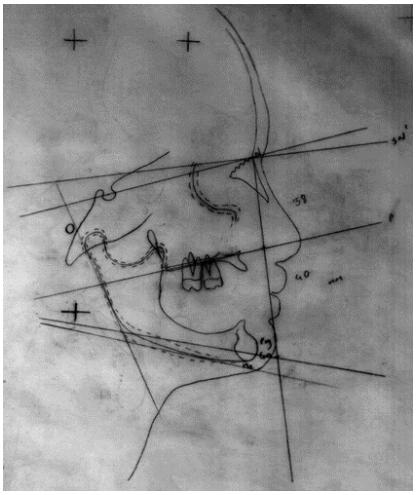

Fig 4: Photograph of lateral cephalogram tracing before treatment.

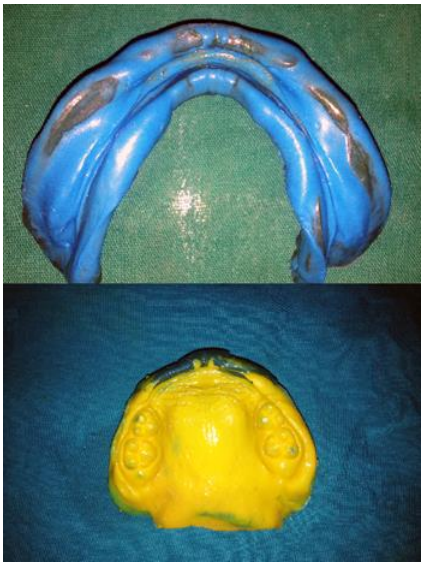

Fig 5: Photograph showing maxillary and mandibular final impression.

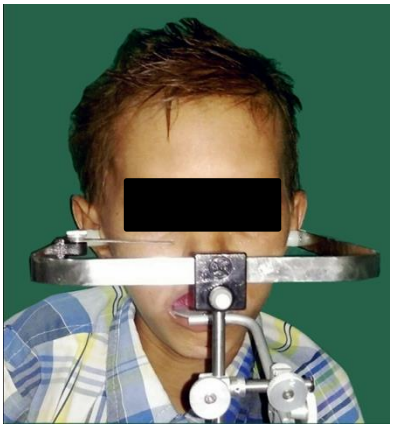

Fig 6: Photograph showing facebow records of patient.

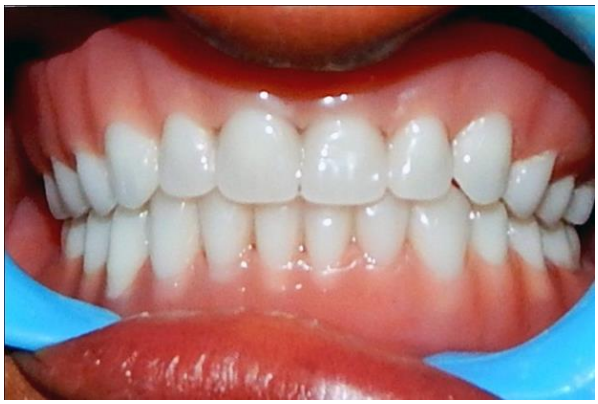

Fig 7: Intraoral photograph showing complete denture insertion.

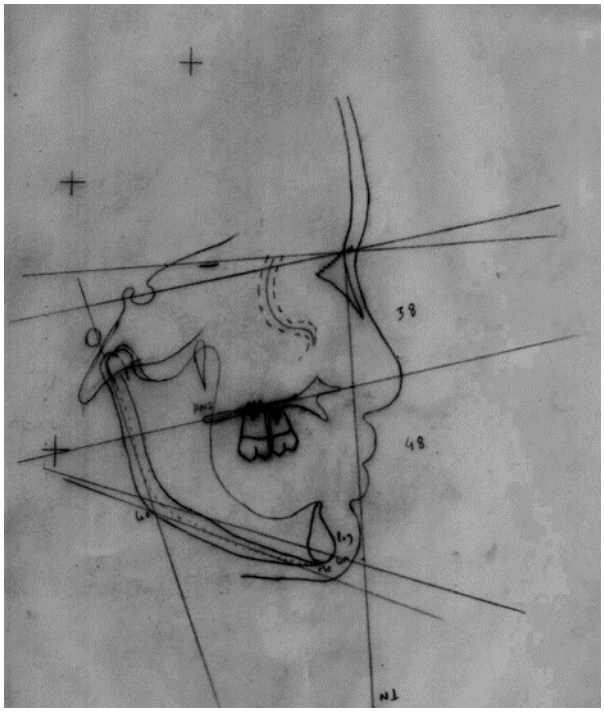

Fig 8: Lateral cephalogram tracing post treatment.

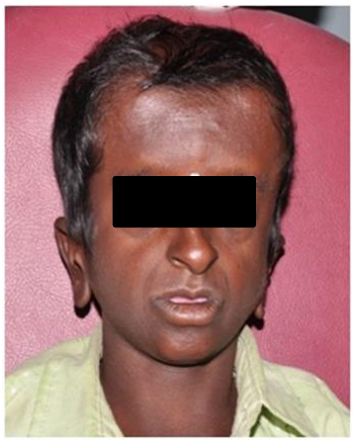

Fig 9: CASE 2 Pre treatment extraoral frontal view.

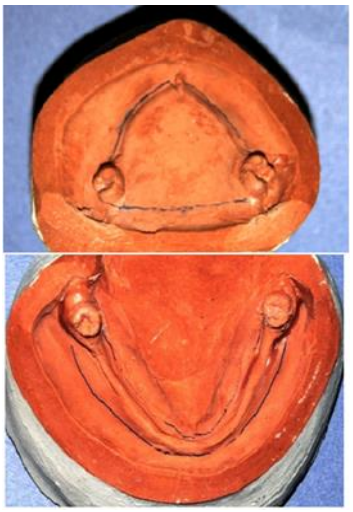

Fig 10: Maxillary and mandibular master cast.

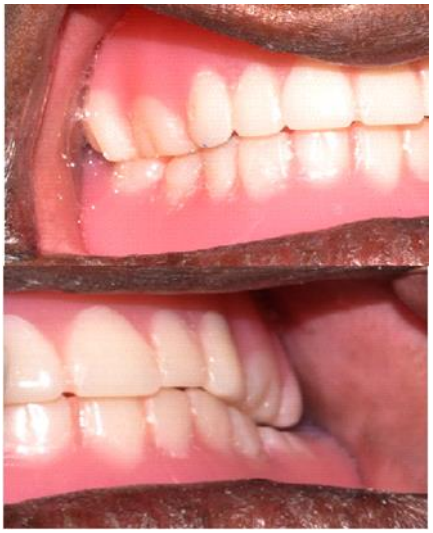

Fig 11: Intraoral view of complete denture in occlusion.

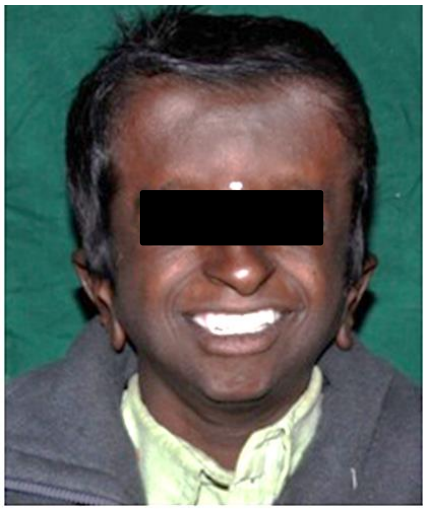

Fig 12: Post treatment extraoral frontal view.

Hypohidrotic (anhidrotic), Hidrotic (Clouston'syndrome), Ectrodactyly-ectodermal dysplasia-cleft syndrome (EEC), Rapp- Hodgkin Syndrome, Hay-Wells syndrome or ankyloblepharon ectodermal dysplasia [5]. It is basically of two type i.e. hypohidrotic also known as Christ-Siemens-Touraine syndrome and hidrotic ectodermal dysplasia also known as Clouston syndrome [1]. Hypohidrotic ectodermal dysplasia show x-linked recessive type of inheritance, this pattern of inheritance was described by "Darwin" for the first time, and hidrotic type shows autosomal dominant pattern of inheritance [7].

PROSTHODONTIC MANAGEMENT

Most individuals who present with ectodermal dysplasia are young and still in a stage of continuous growth, which makes treatment planning even more challenging [6]. Prosthodontic management of ectodermal dysplasia requires a sound knowledge to handle special problem associated with treatment like poor bony support, fragile and thin mucosa, underdeveloped ridge and behavioral problems. A multidisciplinary team approach is recommended for optimal dental management of the condition [7]. Even though it is important to provide early treatment, it should be remembered that any prosthesis made for young patient must be closely monitored. Also a "tell show do" approach is recommended for young ectodermal dysplasia patient.

Treatment generally includes a removable denture including removable partial denture, overdenture, overlay denture, complete denture, and fixed denture and implant retained prosthesis. Nowak stated that "treating the pediatric patient with ectodermal dysplasia requires the clinician to be knowledgeable in growth and development, behavioral management, techniques in the fabrication of a prosthesis, the modification of existing teeth utilizing composite resins, the ability to motivate the patient and parent in the use of the prosthesis, and the long-term follow-up for the modification and/or replacement of the prosthesis."

If the treating dentist is not knowledgeable in one or more of these areas, he should obtain consultations or refer treatment when needed.

CLINICAL REPORT

CASE 1

A 9 year old male with thin physique and normal intellect reported to the department of Prosthodontics, MIDSR Latur with the chief complaint of difficulty in mastication, speech and poor esthetic. On extra oral examination it was found that patient had reduced number of scalp and skin hair (hypotrichosis), reduced sweat secretion (hypohidrosis). Depressed malar process (sunken cheek), and nasal bridges (saddle nose) everted lip and prognathic mandible, absence of eyebrows and eye lashes, and a typical old man facies with sacling of skin [Fig-1].

Intraoral examination reveals completely edentulous mandibular arch and partially edentulous maxillary arch with only four maxillary molars were present [Fig-2]. His OPG reveals absence of permanent tooth bud with reduced density of bone and poor bony support to the existing natural teeth [Fig-3]. Based on the findings of reduced number of teeth and classical features a diagnosis of hypohidrotic ectodermal dysplasia was made. Radiographic examination confirmed the clinical diagnosis. Cephalometric radiograph analysis revealed a reduced vertical dimension of occlusion of the lower third of the face. The facial height index score was (95%) as compared to the ideal score of (80%) which indicated reduced vertical dimension [Fig-4]. This child was second male affected in the family. The same features were present in one of his maternal cousin. Considering the age of the child overlay denture was planned for maxillary arch and conventional complete denture was planned for mandibular arch as the existing teeth were poorly supported by bone which was revealed in the RVG. Implant and FPD is contraindicated before the growth is complete as fixed restoration tends to interrupt the growth of stomatognathic system.

CLINICAL PROCEDURE

Maxillary primary impression made with irreversible hydrocolloid and a snap compound impression was made for mandibular arch. Custom trays were fabricated using auto ploymerized acrylic resin. Border moulding was done and secondary impression was made with light body

[Fig-5]. Facebow record was made (spring bow whipmix) and maxillamandibular relation was recorded [Fig-6]. The sealed jaw relation record was transferred to semi adjustable articulator (Hanau wide vue series). Teeth selection and arrangement was done. Smallest teeth size was chosen which mimic the shape of primary teeth to achieve an age appropriate appearance of child. The occlusion and position of the prosthetic teeth were evaluated intraorally and necessary corrections were made before processing the waxed denture. The complete denture was processed under injection moulding technique and insertion was done [Fig-7]. Cephalometric facial height index score was evaluated after denture insertion which showed approximately normal values (79%) [Fig-8]. The vertical height was restored successfully.

CASE 2

A 15 year old male with his father reported to the department of Prosthodontics, MIDSR Latur with the chief complaint of difficulty in eating and speech. On extra oral examination it was found that patient had scanty eyebrows and eye lashes, saddle nose, diminished lower facial height, protuberant lips, frontal bossing, dry cheeks, hypohidrosis, pulsating scalp with no ossification. There was overall reduction of lower half facial height [Fig-9].

Intraoral examination reveals partial edentulous maxillary and mandibular arch with all permanent first molar present**.** Maxillary and mandibular arch size was smaller with flat ridge in class I ridge relationship. Oral mucosa was dry and sticky with reduced saliva. Radiographic examination reveals absence of permanent tooth bud with severe bone resorption in maxillary and mandibular arch. Bone support for the all first molars was good.

Primary impression made with putty consistency addition silicone impression material (3M ESPE). Custom trays were fabricated using auto ploymerized acrylic resin. Border moulding was done using low fusing impression compound and secondary impression was made with light body addition silicone. Impression was poured in Type IV die stone and master cast was fabricated [Fig-10]. Maxillo mandibular relation was recorded using Niswonger's vertical check bite method. Smallest teeth size was chosen to achieve an age appropriate appearance of child. Try in was done for evaluation of occlusion, phonetics and esthetics. Overlay complete denture was processed under compression moulding technique and insertion was done [Fig-11].

Instructions were given to the patient and parent about use and maintenance of denture. The patient was kept on recall appointments for three weeks for inspection and adjustment purposes. The patient got accommodated to denture early with considerable improvement in phonetics and masticatory functions [Fig-12]. The patient and parent were explained about future relining or remaking procedures.

DISCUSSION

Prosthodontic rehabilitation of two patients with ectodermal dysplasia has been described. Patients with ectodermal dysplasia, especially children, require interdisciplinary planning and treatment to regain esthetics and function of the stomatognathic system [4]. Removable prosthesis including complete or partial denture and implant is primary treatment alternative for patient with partial or complete anodontia [3]. Oligodontia or anodontia is often characterized by underdeveloped alveolar ridges which adversely affect the less bony support available for complete denture as well as implant placement [9]. It is commonly agreed that osseo-integrated implants should be not placed before cessation of growth. Implant and fixed restoration tends to interrupt the growth of alveolus. There are several reports of submerged implants following the growth of alveolar bone. Use of implants in young children should be considered carefully, taking into account the dental and skeleton maturation as compared to the chronologic age of the patient. In both of the above cases, implant therapy was not the treatment choice due to ongoing growth and development and insufficient alveolar bone support [11]. Reduction in growth leads to severe loss of vertical dimension. It is important to rehabilitate the young by restoring lost vertical dimension as well as masticatory function with pleasant smile and improvement in self-esteem. Early prosthetic treatment is generally recommended from the age of 5 years. The positive changes increase the self-confidence of the children.

The remaining maxillary posterior teeth, due to their poor bony support were not suitable as an abutment for overdenture as per radiographic findings. An overlay denture is less expensive, requires less chair time, and does not require any specialized tooth preparation giving all the advantages of overdenture like proprioception and maintenance of alveolar bone. One big advantage of overlay denture is that in future existing teeth can be utilized as an abutment for fixed restoration [8].

In young children behavioural problems encountered during treatment. This can be overcome by educating patient and the parent about importance of rehabilitation. The child must be made accustomed for procedure by establishing a friendly relationship, or by behaviour modification techniques like TELL SHOW DO, and MODELLING [10]. Fortunately, these patients were well educated and enthusiastic to get the treatment done. Periodic recall of young patient is very important due to frequent loosening of prosthesis as the patient grows.

CONCLUSION

The clinical manifestations of ectodermal dysplasia cause considerable social problems in individuals affected by the condition. Young children's with ectodermal dysplasia suffers not only from the difficulties in mastication, phonetics and esthetics but also poor self-esteem in the society. Early prosthodontic rehabilitation is required to improve and maintain normal physiological growth and development of stomatognathic system.

CONFLICT OF INTEREST

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

REFERENCES

1. Mortier K, Wackens G. Ectodermal Dysplasia anhidrotic. Orphanet Encyclopedia 2004; 9:1-6.
2. Priolo M, Lagane C. Ectodermal dysplasia: a new clinical-genetic classification. J Med Genet 2001; 38:579-85.
3. Ildiko T, Katalin G, and Noemi R. Early prosthetic treatment of patients with ectodermal dysplasia: A clinical report. J Prosthet Dent 2005; 93:419-24.
4. Alirez M, Arman T, Kian K, Alexandre AA, Kazunari T, Winston W, Chee L. Full mouth rehabilitation of a young patient with partial expression of ectodermal dysplasia: A clinical report. J Prosthet Dent. 2014;112:449-54
5. Varsha JM, Rucha V. Prosthetic management of an ectodermal dysplasia: A case report. People's journal of scientific research. 2010; 7: 3(2): 37-40.
6. Hickey AJ, Salter M. Prosthodontic and psychological factors in treating patients with congenital and craniofacial defects. J Prosthet Dent 2006; 95:392-6.
7. Hekmatfar S, Jafari K, Meshki R, Badakhsh S. Dental Management of Ectodermal Dysplasia: Two Clinical Case Reports. J Dent Res Dent Clin Dent Prospects. 2012 ;6(3):108-12
8. Erle S, Wilson R, Spuller L. Complete overlay denture for the pediatric patient: Case reports. Pediatric dentistry; 1988;9:10(3): 222-5.
9. Guckes AD, Scurria MS, King TS, McCarthy GR, Brahim JS. Prospective clinical trial of dental implants in persons with ectodermal dysplasia. J Prosthet Dent 2002; 88:21-5.
10. Mark A, Pigno RB, Blackman. Prosthodontic management of ectodermal dysplasia: A review of literature. J Prosthet Dent 1996; 76: 541-5.
11. Mehmet B, Ali MT, Nese A, Tamer T. Ectodermal dysplasia with anodontia: A report of two cases. Eur J Dent 2010; 4:215-22.