Case Report

Prosthetic Management of an Ectodermal Dysplasia: A Case Report

J. Varsha Murthy, *Rucha Vaze

Department of Prosthodontics & *Periodontics, Peoples Dental Academy, People's Campus Bhanpur, Bhopal-462037.

Abstract:

Ectodermal dysplasia represents a group of inherited conditions in which two or more ectodermally derived anatomical structures fail to develop. Early dental intervention can improve patient's appearance, thereby minimizing the associated emotional and psychological problem in these patients. Treatment requires a team work by medical personal along with dental professionals.

Key Words: Prosthetic rehabilitation, Ectodermal Dysplasia, Genetic disorder, Syndrome.

Introduction:

Ectodermal dysplasia (ED) is defined by National foundation for ectodermal dysplasia as a genetic disorder in which there are congenital birth defects of two or more ectodermal structures (Hickey, 2001). Thurman published the first report of a patient with ED in 1848, but the term was not coined until 1929 by Weech. Female carriers outnumber affected men but females show little or no signs of the condition (Pigno et al, 1996)

The syndrome involves overlapping features, thereby complicating a definitive classification. Lamartine in 2003 has described various well defined ectodermal dysplasia as Hypohidrotic (anhidrotic), Hidrotic (Clouston'syndrome), Ectrodactyly-ectodermal dysplasia-cleft syndrome (EEC), Rapp- Hodgkin Syndrome, Hay-Wells syndrome or ankyloblepharon ectodermal dysplasia.Usually the ectodermal dysplasia is divided into two types based on the number and function of sweat glands (Viera et al, 2007) as mentioned below:

- **Hypohidrotic (anhidrotic) Ectodermal Dysplasia (Christ-Siemens- Tourine Syndrome)**

In this form sweat glands are absent or significantly decreased. This disorder is usually inherited as either autosomal dominant / recessive or X-linked recessive trait and the gene locus is X q13-q21. It is commonly X-linked recessive with full expression in males. Female carriers have a minimal expression. 60-70% of cases usually show manifestations restricted to minimal hypodontia, aplastic or hypoplastic mammary glands, impaired lacrimal gland function, glaucoma and increased susceptibility to allergic disorders such as asthma or eczema. Typical general mental development, frontal bossing with characteristics reduction in amount of hair (hypotrichosis), absence of sweat glands (anhidrosis) resulting in temperature elevation, absence of sebaceous glands (asteatosis) resulting in dry skin, depressed nasal bridge, protuberant lips, prominent supra orbital ridges , sunken cheeks, wrinkled hyperpigmented skin around the eyes and large low set ears (Crawford et al ,1991).

The oral manifestations include conical or peg shaped teeth, hypodontia (partial absence of teeth) or complete anodontia (complete absence of teeth) of both the deciduous and the permanent dentition with malformation of any teeth that may be present, generalized spacing, underdeveloped alveolar ridges and delayed eruption of permanent teeth. Even when complete anodontia exists the growth of the jaw is not impaired. This would imply that the development of the jaws except for the alveolar process is not dependent upon the presence of teeth. However, since the alveolar process does not develop in the absence of teeth, there is a reduction in the normal vertical dimension resulting in the protuberant lips. In addition, in oropharynx the defect may be manifested as a high palatal arch or even a cleft palate. The salivary glands including the intraoral accessory glands are sometimes hypoplastic in this disease. This result in xerostomia, and the protuberant lips may be dry and cracked.

- **Hidrotic Ectodermal Dysplasia (Clouston Syndrome)**

Here the clinical features include nail dystrophy, hair defects and palmoplantar dyskeratoris. The patients have normal facies, normal sweating and no specific defect is seen.

Corresponding Author: Dr. J.Varsha Murthy,
2, M.I.G. Peoples Dental Academy Campus, People's Campus Bhanpur, Bhopal-462037.
Phone No.: 9993234628, 9993956951
E-mail : getyuva@rediffmail.com

Case Report:

An 11 year old male patient reported in the Department of Prosthodontics with the chief complaint of lack of esthetics and difficulty in mastication. On general examination the patient presented with the classical triad of hypohidrosis, hypotrichosis and hypodontia. Apart from this, the patient presented with dry skin, depressed nasal bridge, protuberant lips, frontal bossing, sunken cheeks with prominent supraorbital ridges (Fig I) and palmoplantar dyskeratosis (Fig III).

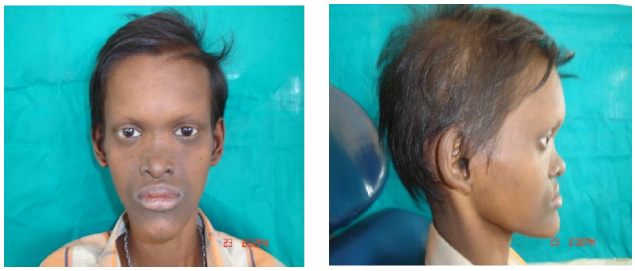

Fig.I: Frontal & Profile view showing hypotrichosis, depressed nasal bridge, protuberant lips, sunken cheeks & frontal bossing with prominent supraorbital ridges

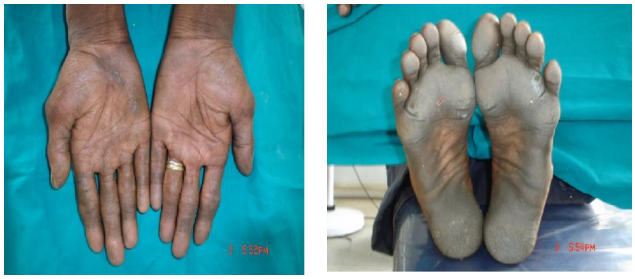

Fig.II: Palms & fect showing keratinisation.

Oral examination revealed (Fig. III) partial anodontia, hypoplastic peg shaped or conical teeth, generalized spacing and underdeveloped alveolar ridges.

The treatment option preferred was of a removable partial denture for the missing teeth. The carious teeth were restored and oral prophylaxis was performed. The peg shaped teeth were modified with composite resin which helped in enhanced retention of upper denture.

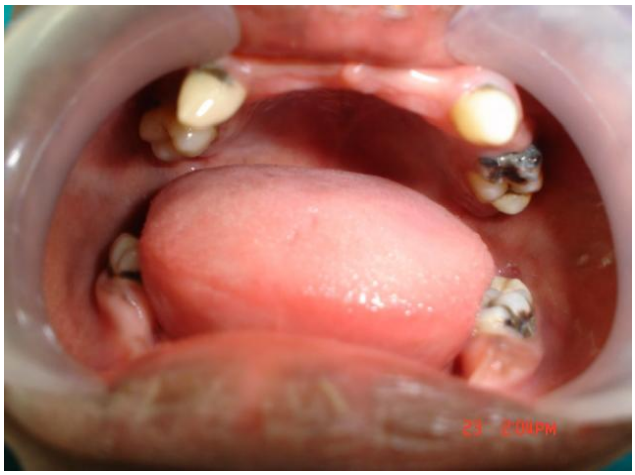

Fig.III: Intaoral view showing peg shaped teeth-hypodontia.

Alginate hydrocolloid (irreversible hydrocolloid) impression material was used for primary impression. This impression material was used because it is clean, biocompatible and pleasant for the patient. Its setting time is less so it causes less discomfort to the patient. It is an elastic material which is easy to remove from undercut area. Because both dentulous and edentulous areas had to be recorded accurately so dual impression was made (by border molding and final impression of the edentulous area and pick up impression by alginate for dentulous area) for both maxillary and mandibular arches (Fig.IV). After beading and boxing of the impressions, the cast were poured with improved die stone for greater strength and abrasion resistance (Fig. V). Jaw relation was done in the conventional manner. Teeth arrangement was done with resin teeth. After final try-in the waxed dentures were processed in a heat polymerized denture base resin. The complete dentures were delivered (Fig. VI) and the patient was instructed on the maintenance of oral hygiene and dentures. Recall appointments were done for adjustments.

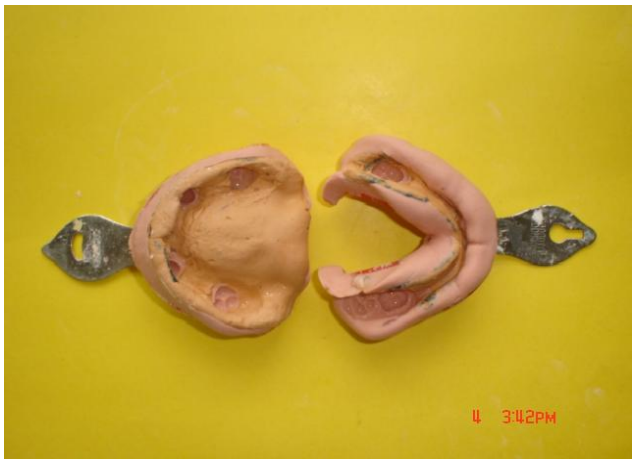

Fig.IV: Dual impressions of the maxillary and mandibular alveolar ridges.

Discussion:

Ectodermal dysplasia often involves overlapping features, thereby complicating a definitive classification. Some ectodermal dysplasia syndrome may be mild whereas others may be devastating. Diagnosis becomes difficult as the characteristic features are not obvious during birth, though during neonatal period there is extensive scaling of the skin

and unexplained pyrexia (Guckes et al, 1991). After diagnosis, the parents of the affected patient can be counselled regarding the treatment options available.

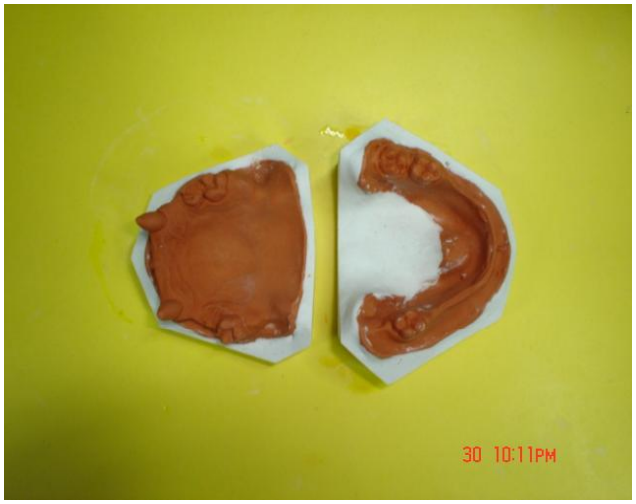

Fig.V: Final cast made up of die stone.

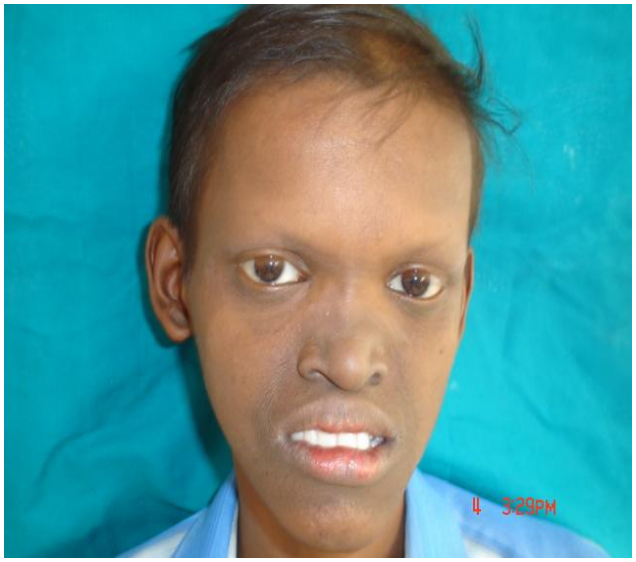

Fig.VI: Post operative appearance of the patient with maxillary and mandibular partial dentures.

An early age intervention helps to modify the intraoral prosthesis during growth spurts or rapid growth periods. Prosthetic intervention can be done with a child as young as 2 or 3 years if the child is cooperative (Hickey, 2001). This also allows the child to adjust with the prosthesis or appliance and develop normal appearance, speech, mastication and swallowing as well as temporomandibular joint function (Ellis, 1992). Apart from dental benefits, an early age intervention also provides psychosocial benefits. The unaesthetic appearance, poor self image, school/job related discrimination often accompanies ectodermal dysplasia syndrome which has a negative psychological effect on the patient. Thus management of the orofacial disfigurement provides the patient with some measure of confidence.

Treatment generally includes a removable and /or fixed partial denture, an overdenture, complete denture prosthesis or an implant retained prosthesis. In cases where there is associated cleft lip and palate the treatment may consist of intervention by a plastic surgeon and an oral and maxillofacial surgeon. In such cases a maxillofacial prosthesis may be indicated. In the present case prosthodontic management was done by removable partial denture. The option of complete denture was ruled out to preserve remaining alvealor ridges. Fixed prosthodontic management is seldom used for such type of patients because of minimal number of teeth present and often patient are young in which fixed partial denture with rigid connector will hamper active growth of dental arches. Options which could be considered in our patient were of overdenture after intentional root canal treatment of the existing teeth. But considering the increased number of appointments and disinterest of patient this option was also ruled out. The treatment option preferred was of a removable partial denture considering his present age. Moreover, considering the growth potential of our patient erupting tooth remained as potential overdenture abutment and an option for a fixed partial denture or implants in future.

As the child matures the removable prosthesis needs relining, rebasing or remaking to accommodate growth changes and maintain normal oral functions. When child reaches teenage years, orthodontic treatment may be indicated as better management of spacing may prepare the mouth for a fixed partial denture or implants in future.

In older patients, depending on pattern of missing teeth and the available bone support, osseointegrated implants can be used. If bone support is inadequate then bone grafting may be necessary. Overlay or overdenture may also be indicated as it involves retaining the tooth or tooth root after intentional root canal treatment.

Summary:

This clinical report describes the types, characteristic features and treatment options for a young male patient with ectodermal dysplasia. With proper care and prosthodontic treatment the patient

can enjoy a relatively normal life. The options for a definitive treatment plan may include fixed, removable or implant supported prosthesis, singly or in combination.

Bibliography:

1. Crawford PJ,Alred MJ,Clarke A: Clinical and radiographic dental findings in X- linked hypohidrotic ectodermal dysplasia. *Journal of Medical Genetics*, 1991; 28:181-185.
2. Ellis RK, Danly KJ, Wild TW: Indirect composite resin crowns as an aesthetic approach in treating ectodermal dysplasia – A case report. *Quintessence International*, 1992; 23:727-729.
3. Guckes AD, Brahim IS, Mccarthy GR, Rudy SF, Cooper LF: Using endosseous dental implants for patients with ectodermal dysplasia. *Journal of American Dental Association*, 1991; 122 (10):59-62.
4. Hickey AJ: Prosthetic treatment for patients with ectodermal dysplasia. *Journal of Prosthetic Dentistry*, 2001; 86:364-368.
5. Itthagarun A, King NM: Ectodermal dysplasia – A review and case report. *Quintessence International*, 1997; 28(9):595-603.
6. Lamartine J: Towards a new classification of ectodermal dysplasia. *Clinical and Experimental Dermatology*, 2003;28(4):351-359.
7. Pigno MA, Blacksman RB, Cronin RJ, Cavazos E: Prosthodontic management of ectodermal dysplasia – A review of literature. *Journal of Prosthetic Dentistry*, 1996; 76:541-545.
8. Ramos V, Giebink DL, Fisher JG, Christensen LC: Complete dentures for a child with hypohidrotic ectodermal dysplasia. *Journal of Prosthetic Dentistry*, 1995; 74:329-331.
9. Thurman J: Two cases in which the skin hair and teeth were very imperfectly developed. *Medico-chirurgical Transaction*, 1848; 31:71-82.
10. Vieira KA, Teixeira MS, Guirado CG, Gaviao MB: Prosthodontic treatment of hypohidrotic ectodermal dysplasia with complete anodontia: Case report. *Quintessence International*, 2007;38:75-80.
11. Weech AA: Hereditary ectodermal dysplasia (congenital ectodermal defect). *American Journal of Diseases of Children*, 1929;37:766-790.