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Hypohidrotic Ectodermal Dysplasia in two Siblings with Missing Teeth: A Dental Perspective

Abstract:

Hypohidrotic ectodermal dysplasia is a rare congenital disease that affects several ectodermal structures. The condition is usually transmitted as an x-linked recessive trait, in which gene is carried by the females and manifested in males. Manifestations of the disease differ in severity and involve teeth, skin, hair, nails and sweat and sebaceous gland. Ectodermal dysplasia is usually a difficult condition to manage. Prosthodontically, because of the typical oral deficiencies, and afflicted individuals are quite young to receive extensive prosthodontic treatment, which restores their appearance and helps them, for the development of positive self-image. This case report describes the management of two siblings having features of ectodermal dysplasia with the help of over denture with copings on existing teeth in upper arch, fixed partial denture and tooth supported full denture in lower arch.

Key-words: Ectodermal dysplasia, Hypodontia, Prosthetic Management

Introduction:

Ectodermal dysplasia is the term used to describe a group of rare, inherited disorder characterized by dysplasia of tissues of ectodermal origin-primarily nail, teeth and skin and occasionally, dysplasia of mesodermally derived tissues.[1] The condition is thought to occur in approximately 1 of 1,00,000 live births.[1],[2] Freire Maia and Pinherio[6] described 117 varieties of ED with multiple combination of abnormal ectodermally derived structures. Clinically ED may be divided into two broad categories the x-linked Hypohidrotic form, characterized by the classical triad of hypodontia, hypohidrosis and hypotrichosis and characterized by dysmorphic facial features is also termed as Christ-Siemens Tourine Syndrome.[3],[4]

The hypohidrotic form of ED usually spares the sweat glands, can affect the teeth, hair, nail and is inherited as an autosomal trait. This was described by Cloustan in 1929 and Lowrey et al in 1966, as an autosornal dominant, which is found in Canadian families of French descent.[4] ED is usually a difficult condition to manage prosthodontically, because of the typical oral deficiencies and afflicted individuals are quite young to receive extensive prosthodontic treatment, which restores their appearance, for the development of positive self-image. [5],[6]

This case report present the management of two siblings having features of ectodermal dysplasia essentially emphasizes on a different approach of prosthetic management of appearance, functionality of treatment in the form of denture provided.

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Case Report:

Two males aged 13Yrs (Figure1) and 18 Yrs.(Figure 2) respectively, reported with the complaint of absence of multiple teeth since childhood, difficulty in eating properly. Both belonging to same family and are siblings. They exhibited the classical features of ED: hypodontia, hypohidrosis, hypotrichosis, prominent forehead, saddle nose and everted lips

Profile view of two siblings, showing features of Ectodermal Dysplasia.

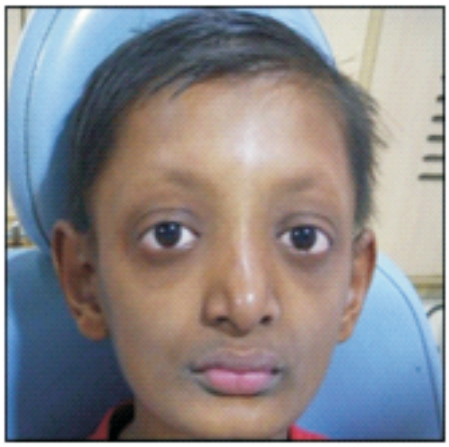

Figure1

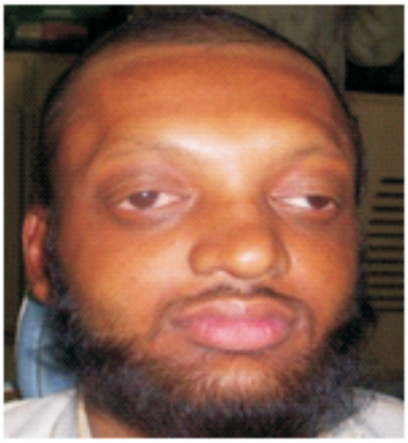

Figure 2

Intra oral examination of younger brother (Case 1) shows three teeth in upper anterior region and two teeth in lower anterior region and all are conical in shape[Figure-3].Intra oral examination of elder brother (Case 2) shows 5 teeth in upper arch and only one teeth in lower arch. [Figure 4]. Upper anterior teeth are conical shaped. They exhibited aplasia of alveolar bone in the edentulous area. These are the only teeth present in both cases and other permanent teeth buds are missing.

Photographs of intra oral examinations showing multiple missing teeth.

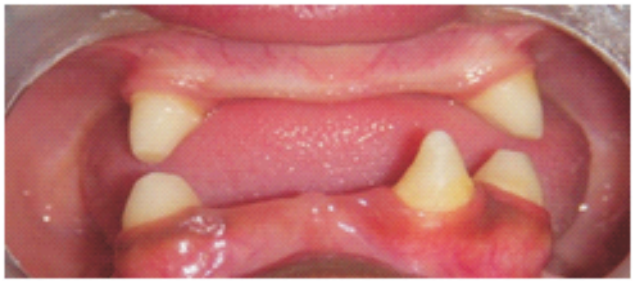

Figure 3 (Case 1)

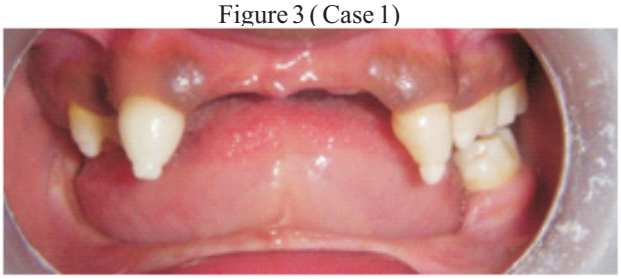

Figure 4 (Case 2)

OPG of Case1 [Figure 5] shows three permanent teeth present in upper arch in anterior region, two teeth present in lower arch and all are conical shaped. Other permanent tooth buds are missing. OPG of Case 2 [Figure-6] revealed, Maxillary teeth are 5 in number, in upper arch . Lower arch shows one teeth in left posterior region. All other teeth buds are absent.

OPG showing multiple missing teeth

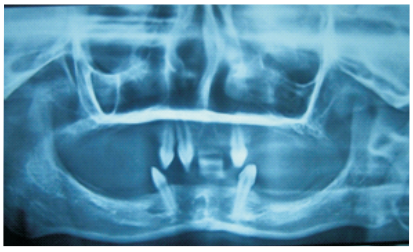

Figure 5 (Case 1)

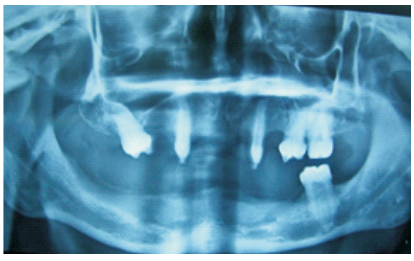

Figure 6 (Case 2)

Case 1 was treated by Metal coping supported over denture in both the arches, because the objective was to preserve the remaining dentition, to restore function and esthetics and to allow certain modification to be made to meet the needs of the

developing stomatognathic system. Teeth preparation was done in upper and lower teeth and metal coping was placed. Impression was taken of both the arches for fabrication of teeth supported over denture[Figure-7].in case two, as age is 18 year and 5 teeth present in upper arch, two on left side and three on right side with sufficient tooth structure so fixed partial denture was given. In lower arch only one tooth present in left posterior region so tooth supported over denture was fabricated.[Figure-8] Recall appointments were scheduled at 24 hours, 72 hours, 2 weeks, 4 weeks and then every 3 months for the first year and every 6 months for the second year. Written oral hygiene instructions were given and explained to the patient

Post treatment photographs

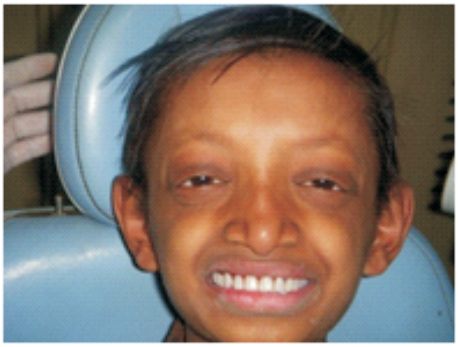

Figure 7 (Case 1)

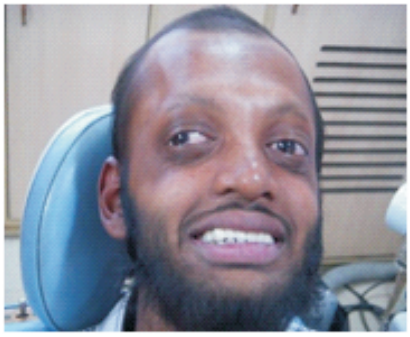

Figure 8 (Case 2)

Discussion:

Nowak stated that "treating the pediatric patient with ED requires the clinician to be knowledgeable in growth and development, behavioural 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". According to Nowak, a series of introductory visits may be needed before treatment commences, to attain the required patient trust.[7]

Removable prosthodontics is the most frequently reported treatment modality for the dental management of ED. Because anodontia or hypodontia is typical in individuals with this condition, complete dentures, partial dentures, or over dentures are often part of the treatment provided. Although complete dentures can provide an acceptable esthetic and functional result, underdevelopment of the edentulous alveolar ridges in individuals with ED can compromise denture retention and stability.

When there are teeth present for support over dentures are a desirable treatment option for these patients. Cram provided an excellent overview of the advantages of conventional overdentures as opposed to complete dentures. One important advantage is that overdentures preserve alveolar bone. Van Waas et al. verified this claim with a well-designed, randomized controlled clinical trial. The trial compared average rnandibular bone reduction in 74 patients treated randomly with either an immediate overdenture on two mandibular canines or an immediate complete denture. There was a significant reduction in alveolar bone loss in the overdenture patients after 2 years. Preservation of alveolar bone is imperative in individuals with ED because they must depend on the alveolar ridges for prosthesis support from an early age. If an overdenture is fabricated, retention can be augmented by various attachments anchored to the available teeth.[6]

Fixed prosthodontic treatment is seldom used exclusively in the treatment of ED, primarily because many afflicted individuals have a minimal number of teeth. In addition, ED patients are often quite young when they are first treated, and fixed partial dentures (FPD) with rigid connectors should be avoided in young, actively growing patients. This is because rigid fixed partial dentures could interfere with jaw growth, especially if the prosthesis crosses the midline. Hogeboom presented a case that dramatically demonstrated the occurrence of jaw growth in an individual treated for ED in which the two segments of a detachable fixed prosthesis separated at the midline because of transverse jaw growth(6). In our case elder brother having sufficient number of teeth so fixed partial denture is planned in upper arch.

Periodic recalls of young ED patients are also important because prosthesis modification or replacement will be needed as a result of continuing growth and development. In addition to adjustments related to fit, occlusion of prosthesis must be monitored for ages because of jaw growth. Other problems related to removable prostheses are speech difficulties, dietary limitations, and loss of the prosthesis.[7]

Conclusion:

A proper family history of patient with ectodermal dysplasia along with assessment of the socioeconomic status of the family is very essential for making a proper treatment plan. Children with ectodermal dysplasia may present a shy and self-withdrawn behaviour due to their esthetic and functional limitations. Oral rehabilitation of such children at the earliest with removable overdentures is important to help him improve his esthetics, masticatory function and phonetics. This further helps to develop a positive self-esteem and goes a long way to restore social life.

References:

1. Dhanrajani PJ, Jiffiy AO. Management of Ectodennal Dysplasia. A Literature Review. Dental Update 1998;25:73-5.
2. Hodges J, Sarnantha, Harley KE. Witkop tooth and nail syndrome: Report of two cases in a family. Int J Paediatric Dentistry 1999;9:207-11.
3. Kupietzky K, Milton H. Hypohidrotic ectodermal dysplasia: Characteristics and treatment. Quintessence Int 1995;26:285-91
4. Richard AS, Karin V, Gerard K, Caries B, Kournjiarn J. Placement of an endosseous implant in a growing child with ecto dermal dysplsia. Oral Surg, Oral Med, Oral Pathol 1993;75:669-73.
5. Shaw RM. Prosthetic management of hypohidrotic ectodermal dysplasia with anodoutia. Case report. Aust Dent J 1990;35;113-6.
6. Piegno MA, Blackrnan RB, Cronin RJ, Cavazos E. Prosthodontic management of ectodermal dysplasia: A review if the literature. J Prosthet Dent 1996;76:541-5.
7. Nowak AJ, Dental Treatment for patients with Ectodermal dysplasia. Birth Defects 1988;24:243-52.
8. Ellis RK, Donly KJ, Wild TW. Indirect composite resins crowns on an esthetic approach to treating Ectodermal dysplasia: A case report. Quintessence Int 1992;22:727-9.
9. Till MJ, Marques AP, Ectodermal Dysplasia: Treatment considerations and case report. Northwest Dent 1992;71:25-8
10. Goepferd SJ, Carroll CE, Hypohidrotic ectodermal dysplasia: A unique approach to esthetic and prosthetic management. J Am Dent Assoc 1981;102:867-9