REHABILITATION OF ANHIDROTIC ECTODERMAL DYSPLASIA: A CASE REPORT

Dr. Rooposhi Saha
Sr. Lecturer
Deptt. of Pedodontics, I.T.S Muradnagar

Introduction

Ectodermal dysplasia consist of clinical and genetic group of heterogeneous disorders, characterized by absence, incomplete or delayed formation of one or more appendages derived from epidermal tissues or oral ectoderm, during embryogenesis. **(Hollbrook, 1988)**

Ectodermal dysplasias were first described by Thurnam in 1948 and later in the 19th century by Darwin. Ectodermal dysplasia was assigned to the X chromosome in 1921 by Thadani, who later reported that carrier females could manifest signs of the condition. Their incidence is relatively rare (1/100000 births) and is characterized by the abnormal development of the ectodermal derivaties **(Clarke, 1987).**

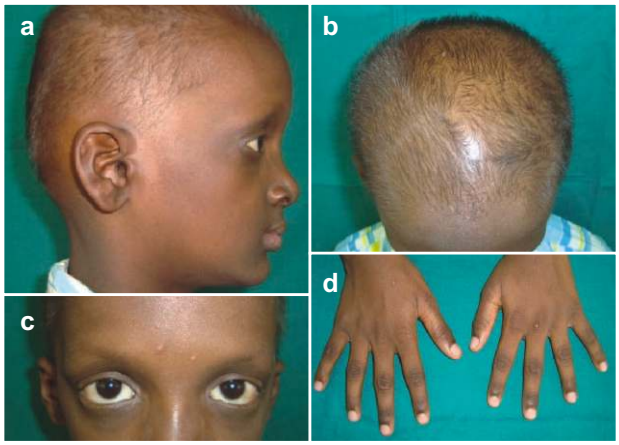
The condition is thought to occur in approximately 1 of 1, 00, 000 live births. Freire Maia and Pinherio described 117 varieties of ED with multiple combinations of abnormal ectodermally derived structures. Clinically ED may be divided into two broad categories the x-linked Hypohidrotic form, characterized by the classical triad of hypodontia, hypohidrosis and hypotrichosis and characterized by dysrmorphic facial features is also termed as Christ-Siemens Tourine syndrome.

The hypohidrotic form of ED usually spares the sweat glands, can affect the teeth, hair, nails 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. **(Pigno et al, 1996)**

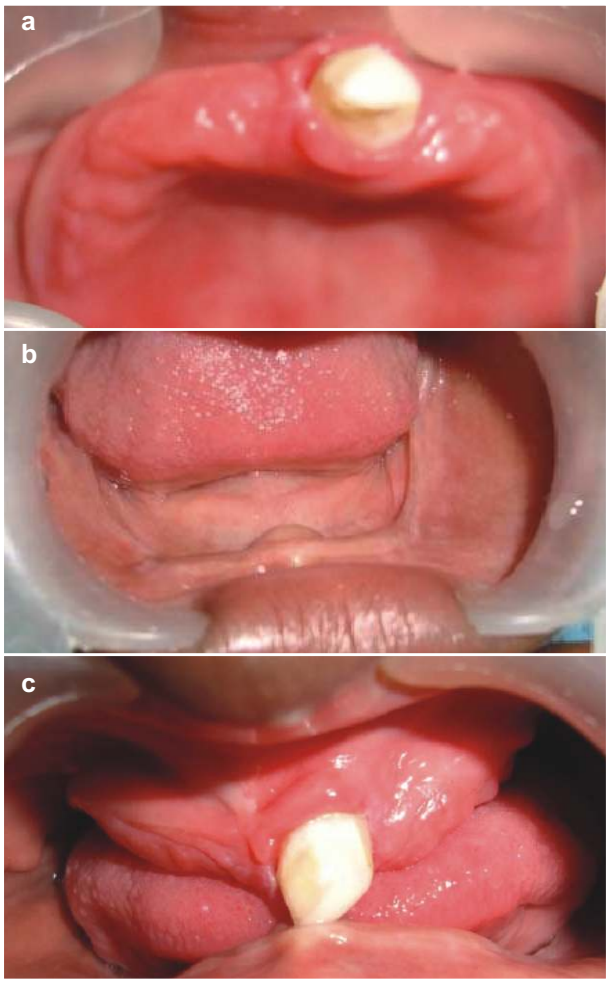
Case report

A male patient name Vishwa, age 8 years *(Figure 1)* reported with the chief complaint of difficulty in wearing of his upper denture. In the history of present illness it was recorded that there was a congenital absence of teeth and a single tooth was seen erupting past 3 months. Nothing relevant could be recorded from the medical, prenatal, natal and post natal history except that the child was unable to tolerate higher temperatures and did not have any sweating. Full upper and lower dentures were made for him when he was 5yrs of age.

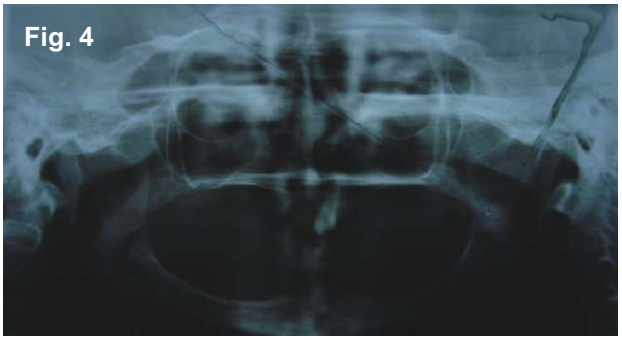
Extra-oral examination presented typical features like scanty hair on scalp and eyebrows, absence of eyelashes and everted lips. Other ectodermal structures like the nails were been to be normal. (Figure 2 a, b, c, d)



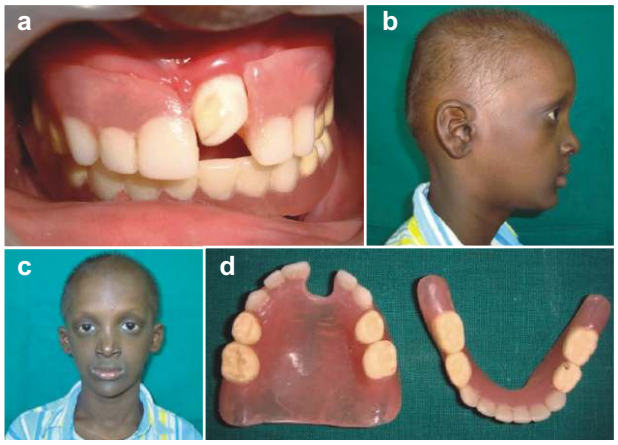
Local examination of the soft tissues did not reveal any abnormality. Hard tissues examination revealed complete absence of all teeth and only one erupting upper central incisor. (Figure 3 a, b, c)



Investigations were conducted and OPG *(Figure 4)* revealed congenital absence of all teeth except for an erupting maxillary central incisor. IOPAR of the incisor revealed a tooth with open apex. *(Figure 5)*



There were no systemic complains on presentation hence oral hygiene instructions were give along with maintenance of adequate hydration. Composite build up of 21 not attempted as the tooth was still in its erupting phase, the exposed dentin on the tooth was covered using glass ionomer cement lining. Selective grinding of the dentures was done to make space for the erupting tooth. Recall visits at 3 months interval were planned. *(Figure 6 a, b, c, d)*



Discussion

Ectodermal dysplasia is a hereditary disorder that occurs as a consequence of disturbances in the ectoderm of the developing embryo. The triad of nail dystrophy, alopecia or hypotrichosis and palmoplantar hyperkeratosis is usually accompanied by a lack of sweat glands and a partial or complete absence of primary and/or permanent dentition. Since the oral rehabilitation of these cases is often difficult; particularly in pediatric patients, treatment should be administered by a multidisciplinary team involving pediatric dentistry, orthodontics, prosthodontics and oral-maxillofacial surgery.

Oral rehabilitation of the ectodermal dysplasia patient is necessary to improve both the sagittal and vertical skeletal relationship during craniofacial growth and development as well as to provide improvements in esthetics, speech, and masticatory efficiency **(Tarzan et al, 2005)**. Although removable prostheses are the most common treatment method, dental implants are also considered to be a treatment option. Dental implants combined with implant-supported dentures for adolescents over 12 years of age are recommended as a treatment choice in literature. In situations where implant therapy is indicated, the main problem is insufficient bone; if bone atrophy progresses in these already alveolar-deficient patients, implant placement may not be possible without bone grafting **(Imizalioglu et al, 2002)**.

Conversely, implantation reconstruction surgery is subject to a greater risk of failure compared to more conservative prosthetic treatment, besides its psychological aspects particularly in young children.

Early prosthetic treatment leads to significant improvements in appearance, speech, and masticatory function. Although dentures are poor alternatives to healthy dentition, they create conditions for the maintenance of a normal, satisfactory diet for the child. This is very important, considering that the establishment of lifelong dietary patterns occurs during childhood **(Mehmet Bani et al, 2010)**.

Dental prostheses may also improve the tone of the muscles of mastication and may compensate for the reduced vertical dimension **(Ithhagarun et al, 1997).** Difficulty with mastication has been referred to as a major problem arising from loss of teeth **(Viera et al, 2007)**. As seen in the reported case, the facial profile and expression improved significantly with complete dentures; in addition, mastication and dietary patterns also improved.

Differential diagnosis for such cases can include namely:

- Ellis Van Crevald syndrome
- Witkop syndrome
- Incontenentia pigmenti

Ellis Van Crevald syndrome is associated with a normal sweat gland function, bilateral polydactyly and clinically there is fusion of the upper lip to the labial gingival, all these features were absent in the present case, hence the presence of this syndrome can be ruled out.

Witkop syndrome is caused by the gene MSX1 in which hair are not affected and teeth are not so severely involved, hence it can be ruled out.

Incontenentia pigmenti is mainly found in females as it is lethal for males. It is associated with delayed tooth eruption and presence of macules all over the body at the time of birth.

The following factors are to be considered while attending to a patient with such a disorder.

- growth and development,

- behavioral management,
- fabrication of a prosthesis,
- modification of existing teeth,
- motivation of the patient and parent
- long-term follow-up **(Nowak AJ, 1988)**

Other treatment modalities commonly used for treating patients with such a rare disorder include individual crown restorations, direct composite restorations**,** fixed prosthodontic treatment **(Pigno et al, 1996)** and periodic recalls for prosthesis modification or replacement. Salivary substitutes can also be included since mucosal dryness is a common finding with such patients.

Conclusion

The clinical manifestations of ectodermal dysplasia cause considerable social problems in individuals affected by the condition. In this case report, the prosthetic rehabilitation of the young boy with anhidrotic ectodermal dysplasia associated with severe anodontia was described. Since oligodontia or complete anodontia leads to atrophy of the alveolar bone, prosthetic treatment is of great value to these patients from functional, psychological, and psychosocial standpoints.

References

1. Tarjan I, Gabris K, Rozsa N. Early prosthetic treatment of patients with ectodermal dysplasia: a clinical report. *J Prosthet Dent* 2005; 93: 419-424.
2. Imirzalioglu P, Uckan S, Haydar SG. Surgical and prosthodontic treatment alternatives for children and adolescents with ectodermal dysplasia: a clinical report. *J Prosthet Dent* 2002; 88: 569-572.
3. Itthagarun A, King NM. Ectodermal dysplasia: a review and case report. *Quintessence Int* 1997;28: 595-602.
4. Vieira KA, Teixeira MS, Guirado CG, Gaviao MB. Prosthodontic treatment of hypohidrotic ectodermal dysplasia with complete anodontia: case report. *Quintessence Int* 2007; 38: 75-80.
5. Mehmet Bani, Ali Melih Tezkirecioglu, Nese Akal, Tamer Tuzuner; Ectodermal Dysplasia with Anodontia: A Report of Two Cases; Eur J Dent 2010;4:215-222
6. Holbrook KA, Structural abnormalities of the epidermally derived appendages in skin from patients with ectodermal dysplasia: insight into developmental errors; Birth Defects Orig Artic Ser. 1988; 24(2):15-44
7. Clarke A, Hypohidrotic ectodermal dysplasia, J Med Genet. 1987 Nov; 24(11):659-63
8. Pigno MA, Blackman RB, Cronin RJ Jr, Cavazos E; Prosthodontic management of ectodermal dysplasia: a review of the literature; J Prosthet Dent. 1996 Nov; 76(5): 541-5
9. Nowak AJ; Dental treatment for patients with ectodermal dysplasias; Birth Defects Orig Artic Ser. 1988;24(2):243-52

THE 98th FDI ANNUAL WORLD DENTAL CONGRESS-A GREAT SUCCESS

The 98th Annual World Dental Congress(AWC) was held at the "JOY CITY" of Salvador da Bahia, Brazil on 1-5 September 2010. The AWC is one of the biggest events on the international dental calendar, giving all of its members the opportunity to discover state-of-the art technology and overall progress in the dental profession and a broad view of dentistry worldwide.

The theme of the 2010 Scientific Programme was "**Oral Health for All: Localchallenges, Global solutions**". The main goal of this programme was to improve the oral health condition globally using scientifically sound strategies. The scientific programme included150 scientificlectures, symposiums, forumsand pre congress courses by 102 speakers about half from around the world and half from Latin America.

The World Dental Parliament gathered around 350 representatives from FDI members associations to establish the organisation's strategic direction and adopt policy statements that influence the world of oral health. Over 6000 Dental professionals from across the world attended the congress.

Apart from the highly successful Scientific Programme and the Trade fair` which included the latest in dental materials ,equipments and technology, The Brazilian culture composing of all its myths ,legends ,dances, and music was presented in a vibrant and magical night at the "Concha Acustica do Teatro Castro Alves" during the inaugural ceremony and the energy and sensations of Salvador"The joy city" was experienced during the beautiful "SALVODOR NIGHT".

Figure Legends

Fig. 1 : Chairman Organizing Committee Apdc Manila with Dr. U.S. Krishna Nayak

Fig. 2 : Dr Roberto Vianna-president Fdi with Dr. U.S. Krishna Nayak

Fig. 3 : Fdi President Elect with Dr. U.S. Krishna Nayak

Fig. 4 : With President Fdi, Apdc And Other Office Bearers with Dr. U.S. Krishna Nayak


Fig. 1


Fig. 2


Fig. 3


Fig. 4