Journal of Applied Dental and Medical Sciences
NLM ID: 101671413 ISSN:2454-2288
Volume 2 Issue 1 January - March 2016

Case Report

Oral Rehabilitation Of Patient With Hypohidrotic Ectodermal Dysplasia By Increasing Vertical Dimension- A Case Report

Natasha Kale[1], Alok Patel[2], Chetan Bhat[3], Smita Patil[4]

[1,4]Post Graduate student, [2]Professor and Head of the Department, [3]Associate Professor, Department of Paediatric and Preventive Dentistry, Bharati Vidyapeeth Dental College Pune, India

ARTICLE INFO

Keywords:
hypohidrotic ectodermal dysplasia, reduced vertical dimension, hypodontia, oral rehabilitation..

ABSTRACT

Ectodermal dysplasia is rare congenital disease that affects the ectodermal structures including teeth, nails, hair, and sweat glands. We report an unusual case of hypohidrotic ectodermal dysplasia in a 7 year old female patient with extraoral findings of sparse hair, scanty eyebrow and eye lashes, depressed nasal bridge, frontal bossing. Intraoral finding showed hypodontia of deciduous and permanent teeth, reduced vertical dimension and marked resorption of maxillary and mandibular alveolar ridges. Oral rehabilitation was achieved by increasing vertical dimension and later with removable acrylic prosthesis that enhances facial esthetics, masticatory function, speech and self esteem of the patient.

Introduction

Ectodermal dysplasia represents a large group of conditions in which two or more ectodermally derived structures fail to develop, and was first described by Thurman.[1] It is classified as hypohidrotic ectodermal dysplasia also known as Christ-Siemens- Touraine syndrome (in which sweat glands are either absent or significantly reduced in number) and hidrotic ectodermal dysplasia also known as Clouston syndrome(in which sweat glands are normal).[2] Hypohydrotic ED can either be X linked recessive or autosomal recessive with male predilection and resultant defective formation of ectodysplasin protein. They are relatively rare with frequency 1:10,000 and 1: 1oo, ooo.[3]

It is characterised by sparse hair, scanty eyebrow and eye lashes, depressed nasal bridge, frontal bossing, reduced vertical facial height and depth and marked resorption of maxillary and mandibular alveolar ridges. The most striking feature in the oral cavity is oligidontia. Teeth in anterior region of maxilla and mandible are conical in shape, whereas posterior teeth exhibit a bird crown. The pre-ocular skin may show fine wrinkling with hyper pigmentation and mid face hypoplasia. Reduced salivary secretion, dry oral mucosa and horse voice qualityhave also been reported in this patients.[4]

Case report

A 7 year old girl reported to the Department of Pedodontics and Preventive Dentistry, Bharati, Vidyapeeth Dental College and Hospital with missing upper and lower front teeth as the chief complaint. Extra-oral examination revealed dry skin, pre-ocular wrinkling & dryness of skin (fig.1), depressed nasal bridge, frontal bossing (fig.2), sparse hair (fig.3), scanty eyebrow and eye lashes. Intra oral examination showed hypodontia of

* *Corresponding author:* ***Dr. Alok Patel,Professor and Head Department of Paediatric and Preventive Dentistry,,Bharati Vidyapeeth Dental Sciences,Pune,India***

the teeth. She had two peg shaped anterior teeth, second deciduous molar and first permanent molar in maxilla. Mandible had second deciduous molar and first permanent molar. Remaining teeth/tooth buds were congenitally missing and alveolar ridges were thin with reduced vertical bone height (fig.4). These finding matched typical features of hypohidrotic ectodermal dysplasia. The radiographic findings and sweat pore count test (fig.5) confirmed the clinical diagnosis. OPG showed two unerupted permanent central incisors and tooth buds of right lower second premolar, upper and lower second permanent molar(fig.6).

Detailed family history revealed that the patient's younger sister also suffered from hypodontia and sparse hair.

Upper two central incisor were grade III mobile, and about to exfoliate. Interocclusal rest space determined by measuring the difference in distance between the nose tip and the highest point on chin, i.e Vertical height, at rest was 3.4 mm and at occlusion it was 3 mm. Hence it was decided to increase vertical dimension of occlusion (VDO). Maxillary and Mandibular removable dentures were considered to be the treatment of choice. Thus to increase the VDO, metal crowns on 55, 65, 75 and 85 in combination with the removable partial denture were planned.

The two second deciduous molars in mandibular arch were prepared to receive stainless steel crowns to increase VDO by 1mm.(fig.7) Once the patient was comfortable with increased VDO in the mandibular arch, stainless steel crowns were given in upper second deciduous molars to increased 1mm VDO in the next visit.(fig.8) Impressions were made with irreversible hydrocolloid material (Alginate). Fabrication of baseplate and wax rims was carried out. Incisal edge of upper conical central incisor was visible in the mouth. Hence it was planned to create window for eruption upper central incisor. A centric relation was recorded at the increased vertical dimension of occlusion. The cast were mounted on a3 point articulator. Artificial teeth were chosen to achieve an age appropriate appearance and were arranged in wax for trial evaluation. C clasps were made as retainers. The occlusion and position of the teeth were checked intra-orally to evaluate the patient's tolerance to the increased occlusal vertical dimension (fig. 9) as well as esthetics, phonetics and occlusal harmony. The partial dentures were processed with heat activated acrylic resin, polished and inserted (fig. 10). Instructions were given for regular cleaning and maintenance to the patient and parents. The Patient was instructed to remove the partial dentures at night. Recalls were scheduled every 3 months to evaluate developing jaws and teeth eruption.

Discussion

Oral rehabilitation of patients with ectodermal dysplasia is necessary to improve sagittal and vertical skeletal relationship during craniofacial growth as well as speech and masticatory efficiency[5]. Its diagnosis is based on thorough family history, characteristic symptoms, that include hypodontia, anodontia or oligodontia with conically shaped teeth, hypohidrosis with concomitant hypertheremia caused by lack of sweat gland and hypotrichosis.

Sweat pore count test confirms the presence of hypohidrosis. Yellow starch and iodine powder is `applied to palmer or dorsal skin surfaces. In normal individuals sweat turns powder colour to deep purple, allowing visualization of the sweat pores, while in ectodermal dysplasia sweat pores are poorly visualized. Skin histopathology shows sparse appendages in the form of reduced number of sweat gland, hair follicles and sebaceous glands[6]

Fig. 1:pre-ocular wrinkling & dryness of skin

Fig. 2: lateral profile

Fig. 3: sparse hair

Fig. 4: maxillary & mandibular arch Interrelationship

Fig. 5: sweat pore count test

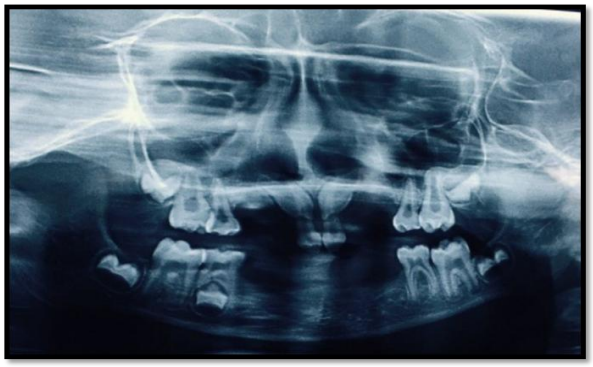

Fig. 6: OPG

Fig. 7: stainless crown on 75, 85

Fig. 8: stainless steel crown on 55, 65

Fig. 9 maxillary & mandibular interarch relationship with increased VDO Fig. 10: removable partial denture

The most common treatment plan is removable prosthesis. Implant supported denture is also suggested as the ideal reconstruction modality for adolescents over 12 years. When 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.[7] In addition to the psychological effects particularly in young children, implant surgery accompanied by higher risk of failure compared to that of more conservative prosthetic treatment.[4] Early implant placement in growing child may cause cosmetic problem because implant act similar to ankylosed teeth. Along with craniofacial growth implant over structure may not be in

occlusion with opposite teeth and even the adjacent teeth may tilt in to the space. Thus implant supported prosthesis may be less favourable.[7]

Early treatment with removable appliance can result in significant growth stimulation of the alveolar processes under the pressure of the interim prosthesis and can provide a better foundation for future implant placement. Hence In the present case we have delivered removable partial prosthesis to a patient.[8]

The patient had oligodontia with retained deciduous molar and absent mandibular anterior teeth which resulted in the collapsed vertical height. This loss of

VDO was compensated by restoring the molars in maxillary and mandibular arch with individual metal crowns. Individual crown restorations have no age restriction related to jaw growth, hence facilitating their placement in young individual.

An interocclusal rest space (IORS) of 2mm has been suggested as the physiological space and therefore an IORS of more than 2 mm indicates that the occlusal vertical dimension can be safely increased.[9.] The patient remains comfortable at the increased occlusal vertical dimension and early placement of the removable partial prosthesis was planned in order to maintain the relationship of maxilla to mandible and allow normal pattern of growth.

Retention and stability for the prostheses are difficult to obtain as patients with ectodermal dysplasia show dryness of oral mucosa and under developed maxillary tuberosities and alveolar ridges. Thus while fabricating prostheses for these patients care should be taken to obtain a wider distribution of occlusal loads by extending denture base as much as possible.[10]

Further future treatment will include incisal edge reshaping, modification of denture by relining or replacement of dentures according to alveolar growth and an eventually, placement of endosseous implants for additional support of denture.

CONCLUSION-

The clinical manifestations of ectodermal dysplasia not only affect the oral functions of patient but also cause considerable social problems for the individual as well as the for family. Management of these young edentulous patients to improve their function, speech, esthetics and psychosocial condition poses unique challenge to pedodontist. However a correct diagnosis and treatment by a multidisciplinary approach can be helpful to improve patient quality of life and instil within them a positive attitude.

REFERENCES

1) Nunn JH, Carter NE, Gillgrass TJ, Hobson RS, Jepson NJ, Meechan JG, et al. The interdisciplinary management of hypodontia: background and rolr of pediatric dentistry. Br Dent J. 2003; 194:245-51.
2) Mortier K, Wackens G. Ectodermal dysplasia anhidrotic. Orphanetencyclopedia. 2004:1-6.
3) Vallejo AP, Monje ELA, Garcia MG, Fernandez mm, Buylla FBMA. Treatment with removable prosthesis in hypohidrotic ectodermal dysplasia: a clinical case. Med oral patol oral cirbucal 2008; 13:119-23.
4) Lo Muzio L, Bucci P, Carile F, Riccitiello F, Scotti C, Coccia E, et al. Prosthetic rehabilitation of child affected from anhydrotic ectodermal dysplasia: Acase report. J Contemp Dent Pract 2005; 6:120-6.
5) Tarjan I, Gabris K. Rozsa N. Early prosthetic treatment of patients with ectodermal dysplasia: a clinical report. J Prosthet Dent 2005; 93:419-24.
6) Ramesh K, VinolaD, John JB. Hypohidrotic ectodermal dysplasia: diagnostic aids and a report of 5 cases. J Indian SocpedodPrev Dent 2010; 28:47-54.
7) Imirzalioglu P, UckanS ,Haydar SG . Surgical and Prosthodontic treatment alternatives for children and adolescents with ectodermal dysplasia : a clinical report. J Prosthet. Dent. 2002; 88: 569-72.

8) Rahul P, Abdulaziz Abdullah, Nidhi K, V. Madhav, S. Bhide. Ectodermal dysplasia: Dental management and complete denture therapy. World Appl. Sci. J.2012; 20(3): 423-28.
9) J Abduo, K Lyons. Clinical consideration for increasing occlusal vertical dimension: a review. Aus Dent J. 2012; 57: 2-10.
10) Dyson JE. Prosthodontics for children. In: Wei SHY, ed. Pediatric dentistry and orthodontics: Total patient care. Philadelphia: Lea &Febiger, 1988: 259-74.

Source of Support: Nil Conflict of Interest: None