

## ECTODERMAL DYSPLASIA SYNDROME WITH CLEFT PALATE, HYPODONTIA, METATARSUS ADDUCTUS AND IMPERFORATE ANUS: A NEW SYNDROME?

<sup>1</sup>UMAIR KHAN, BDS, C-Radiology (UK), MSc Oral Medicine (EDI-UCL, UK), MIAOMT (USA), MBSOM (UK)

<sup>2</sup>RABIA AZIZ, BDS

<sup>3</sup>GHAZALA HASSAN, BDS

<sup>4</sup>TAHIRA ZAFAR, BDS

<sup>5</sup>SAHAR IQRAR, BDS

### ABSTRACT

*Ectodermal dysplasia constitute a large group of rare, heterogenous (under clinical and genetic aspects), congenital / hereditary disorders characterized by a constellation of findings involving a primary defect (hypoplasia or aplasia) in at least two embryonic ectodermal-derived tissues including the teeth, skin, appendageal structures, hair, nails, nerve cells, eccrine glands, sebaceous glands and parts of the eye (conjunctiva), ear, and certain other structures. More than 192 distinct disorders have been described. The most common Ectodermal dysplasias are X-linked recessive hypohidrotic / anhidrotic type known as Christ-Siemens Touraine syndrome with the gene mapping to Xq12-q13 and hydrotic type known as Clouston's syndrome. Several ED syndromes may manifest in association with midfacial defects, mainly cleft palate and / or lip. Hypodontia of the primary and permanent dentition is the most common oral finding. This study presents four cases of the same family, two suffering from Ectodermal dysplasia along with hypodontia and cleft palate, one of which also presents metatarsus adductus and imperforate anus (proband) and the remaining two out of four with hypodontia in the absence of ectodermal dysplasia, one of which also presents metatarsus adductus (elder sibling) with imperforate anus in the other (youngest sibling). Oral and Maxillofacial Physicians & Dentists can be the first to diagnose ectodermal dysplasia due to the presence of specific facio-oral features and absence of teeth respectively.*

**Key Words:** Dracula teeth, Anodontia, Hay Wells syndrome, Ectrodactyly ectodermal dysplasia clefting syndrome, cleft palate.

### INTRODUCTION

Thurman in 1848<sup>1</sup>, first reported 2 male first cousins and their maternal grandmother with a hereditary syndrome associated with sparse hair, missing teeth and dry skin. However the term "Ectodermal Dysplasia" was coined by Weech in 1929<sup>2</sup> and described three essential features; (a) tissues affected

are principally of ectodermal origin, (b) the defects are developmental anomalies and (c) the hereditary tendencies are strongly developed. The triad of nail dystrophy (onychodysplasia); scanty, fine light hair on the scalp and eyebrows (trichodysplasia) and palmo-plantar hyperkeratosis (palmo-plantar keratoderma) is usually accompanied by lack or absence of sweat glands (Hypohidrosis or anhidrosis) and partial or complete absence of primary and/or permanent dentition (hypodontia/oligodontia or anodontia).<sup>3,4,5,6</sup> Ectodermal dysplasia might be inherited in any form of several genetic patterns including autosomal-dominant, autosomal-recessive, and X-linked modes.<sup>7</sup> Although more than 170 different clinical subtypes of ectodermal dysplasia have been identified, these disorders are considered to be relatively rare with an estimated incidence of 1 case per 100,000<sup>8</sup>, with a mortality rate

<sup>1</sup> Assistant Professor and Head of Oral Medicine Department, Fatima Memorial Hospital College of Dentistry, Lahore

<sup>2</sup> FCPS trainee in Orthodontics & Dental Surgeon in "Cleft Centre" Children's Hospital, Lahore.

<sup>3,5</sup> Demonstrators Oral Medicine Department, FMH.

<sup>4</sup> House Officer Oral Medicine Department, FMH.

**Correspondence:** Dr Umair Khan, Assistant Professor and HOD, Fatima Memorial Hospital College of Medicine and Dentistry, Shadman 54000, Lahore, Pakistan,

Email: dr.u.khan.1@hotmail.com,

Contact number: Cell: 0300 9638625

**Received for Publication:** July 2, 2013

**Accepted:** July 18, 2013

of 28% in males up to 3 years of age.<sup>9</sup> Largely, there are 2 major types of Ectodermal dysplasia depending on the number and functionality of the sweat glands: (1) Hypo-hidrotic or anhidrotic (Christ-Siemens-Touraine syndrome) in which sweat glands are either absent or significantly reduced in number; (2) Hidrotic (Clouston's syndrome) in which sweat glands are normal. Dentition and hair are involved similarly in both types but hereditary patterns of nails and sweat glands involvement are different.<sup>10,11</sup> Hypohidrotic ectodermal dysplasia as the most common type seems to show an X-linked inheritance pattern with the gene mapping to Xq12-q13; therefore, males are more susceptible than females. Hidrotic type is inherited in an autosomal dominant pattern.<sup>12</sup> The first classification system for Ectodermal dysplasia was proposed by Pinheiro and Freire-Maia in 1982.<sup>13</sup> The patients are classified into subgroups based on the presence or absence of the following: (1) trichodysplasia, (2) dental abnormalities, (3) onychodysplasia, and (4) dyshidrosis. Overall, EDs were classified into either group A disorders, which were manifested by defects in at least two of four classic ectodermal structures as defined above (1 to 4), with or without other defects, and group B disorders were manifested by a defect in one classic ectodermal structure, in combination with a defect in one other non-classical ectodermal structure, i.e., ears, lips, and dermatoglyphics. A condition characterized by only ectodermal signs is called a pure ectodermal dysplasia; if it combines ectodermal signs and other malformations, it is termed as ectodermal dysplasia malformation syndrome or an ED syndrome.<sup>14</sup> Generally the embryonic ectodermal-derived malformations in individuals suffering from Ectodermal dysplasia includes frontal bossing (protuberant forehead)<sup>15</sup>; malar hypoplasia/midface hypoplasia (poorly developed malar bones giving the appearance of maxillary micrognathia)<sup>15</sup>; depressed nasal bridge (broad nose)<sup>15</sup>; protuberant sometimes everted lips; prominent chin<sup>15</sup>; palmo-plantar dryness<sup>16,17</sup>; hair manifestations (trichodysplasia/dry, thin, fragile, hypo-pigmented/fair, sparse, scanty, kinky and silky hair with sparse or even absent eyelashes, eyebrows and/or scalp hair)<sup>17</sup>; skin manifestations (soft, warm, dry, scaly/keratotic or smooth, thin skin with sometimes peri-ocular fine wrinkling and hyper-pigmentation)<sup>15,18</sup>; eccrine manifestations (anhidrosis/hypohidrosis/absence or decreased number of sweat and sebaceous glands or their non-function resulting in elevated body temperature<sup>19</sup> and giving the hair an unruly appearance and some of the above mentioned skin manifestations<sup>15</sup>); nail manifestations (Onychodysplasia/small, thick or thin, brittle,

discoloured, cracked and/or ridged nails)<sup>20,21</sup>; oro-facial manifestations (hypodontia/oligodontia or anodontia, conical/tapered/peg shaped or pointed deciduous and/or permanent teeth sometimes referred to as "Dracula teeth" because of the frequent involvement of maxillary anterior teeth particularly canines<sup>15</sup>, cleft palate/lip<sup>22</sup>, lack of alveolar ridge development due to lack of tooth bud formation, decreased lower vertical occlusal height<sup>15</sup>, xerostomia<sup>23</sup>, malformed molars/reduced sized molars<sup>24</sup> and/or taurodontism<sup>25</sup> and old age appearance<sup>26,27,28</sup>). Diagnosis of Ectodermal dysplasia is often difficult since any ectodermal derivative may be involved in varying degrees. The number of ectodermal dysplasia syndromes has increased to more than 192.<sup>29</sup> Despite the different genetic causes, their presentations may be very similar. Diagnosis is usually made by clinical observation, often with the assistance of family medical histories. Numerous types of ED syndromes have been described and several classifications exist. However certain diagnostic tests and investigations might be helpful in aiding diagnosis. These include: (a) radiographic investigations; a dental panoramic tomograph/DPT or OPG can be done to rule out hypodontia or oligodontia or anodontia of deciduous and/or permanent teeth as well as dental morphological abnormalities such as taurodontism, x-rays of hands and feet may reveal specific skeletal defects such as ectrodactyly seen in "Ectrodactyly ectodermal dysplasia syndrome with clefting/EEC", (b) sweat pore count<sup>29</sup>; by using yellow starch-iodine powder applied to palmar or dorsal skin. In unaffected persons, sweating turns the powder to deep purple, allowing visualization of sweat pores in the form of mosaic pattern following the streaks of Blaschko's lines.<sup>30</sup> Sweat pores are poorly visualized in affected children<sup>31</sup>, (c) skin biopsy; hypothenar eminence is the most reliable biopsy site to demonstrate an absence or hypoplasia of sweat glands.<sup>21</sup> Histopathology may reveal a reduction in the number of sweat glands, hair follicles and sebaceous glands associated with different ectodermal dysplasias. The epidermis is thin and flattened. Eccrine sweat glands are few or poorly developed or are rudimentary<sup>32</sup>, (d) pre-natal diagnosis; fetal skin biopsy and chorionic villus sampling at the 10<sup>th</sup> week of gestation can be done for some EDs.<sup>21</sup>

## CASE REPORT

### PROBAND (4 YEARS)

A 4 years old female patient was referred to the Oral and Maxillofacial Medicine Department of Fatima

Memorial Hospital College of Dentistry, Lahore, Pakistan with the complaint of difficulty in mastication and speech. The parents of the patient were also concerned about severe partial absence of deciduous teeth. History revealed presence of features of ectodermal dysplasia with variable clinical expressivity in the family as two other siblings seemed affected along with the mother. General examination of the patient revealed thin, fine, dry, sparse and fragile hair (Fig 1). The nails of the patient were thin and brittle. Skin was dry, coarse and slightly warm as the patient had slightly elevated body temperature due to mild hypohidrosis. She had a shrill voice which seemed non-significant. Extra oral examination revealed frontal bossing, depressed nasal bridge, malar/midface hypoplasia with relative mandibular prognathism. The chin of the patient was prominent with protuberant lips (Fig 1).



Fig 1: Extra oral features of ectodermal dysplasia including trichodysplasia, frontal bossing, malar hypoplasia, mandibular prognathism, prominent chin and protuberant everted lower lip

Intra oral examination revealed partial anodontia or oligodontia/severe hypodontia with conical/tapering hypoplastic deciduous right and left maxillary lateral incisors, left deciduous maxillary canine and right deciduous mandibular lateral incisor, lack of alveolar ridge development due to lack of tooth bud formation (Figs 2 & 4), decreased lower vertical occlusal height and scar mark of treated isolated cleft palate (Fig 3). Radiographic examination revealed complete root resorption of deciduous right and left maxillary lateral incisors, left maxillary canine and right mandibular lateral incisor. The permanent max-

illary canines were malformed and conical in shape (Dracula teeth). All other permanent teeth were absent. The vertical height was reduced due to the absence of teeth with thin, less developed alveolar ridges (Fig 4). She had median tilting of the feet (metatarsus adductus) which was not treated as the Orthopaedics and the Paediatric surgeons were of the opinion that this abnormality is not severe in the proband and she is able to walk properly as surgery might bring more harm than the condition she is already in. She also had congenital colorectal abnormality in the form of imperforate anus which was surgically treated at birth.



Fig 2: Intra oral features including oligodontia or severe hypodontia, conical/tapered teeth and thin alveolar and residual ridges



Fig 3: Scar mark of treated cleft palate of the proband ELDER SIBLING (7 YEARS)

The elder sibling had no significant features of ectodermal dysplasia (Figs 6 & 7) apart from hypodontia (Fig 8). He had the history of urinary incontinence which seemed non-significant. But similar to his sister he also had metatarsus adductus (median tilting of the feet) for which he had undergone treatment twice (Fig 9).

Radiological examination revealed hypodontia as the tooth buds of all the maxillary and mandibular 2nd





Fig 4: Orthopantomograph shows severe hypodontia or oligodontia of the deciduous and permanent teeth with pointed tapered canines (Dracula teeth)



Fig 5: Mild Metatarsus adductus of the proband

and 3rd molars were congenitally missing. Left mandibular first molar was impacted. No other radiographic sign was visible relative to Ectodermal dysplasia (Fig 10).

#### YOUNGEST SIBLING (1.5 YEARS)

The youngest sibling (18 months old boy) had frontal bossing, depressed nasal bridge, mild malar hypoplasia, mandibular prognathism and protuberant lips (Fig 11) along with cleft palate (Fig 12). The hair of the patient were scarce, dry, brittle and thin (Fig 11). He also had a rectal surgery done at birth for imperforate anus similar to his sister.

Intra orally only right and left maxillary deciduous canines were present which were hypoplastic and conical/tapered (Fig 12). Radiological evaluation of the



Fig 6: No extra oral features of ectodermal dysplasia as evident from the front facial profile of the elder male sibling.



Fig 7: No extra oral features of ectodermal dysplasia as evident from lateral facial profile of the elder male sibling.



Fig 8: Hypodontia in elder sibling

youngest sibling could not be done as his mother was reluctant for undergoing radiographic exposure (orthopantomograph/OPG) of her son because of the risk of harmful effects of x-ray exposure.

#### MOTHER

The mother had moderate hypodontia with congenitally missing right permanent maxillary and man-



Fig 9: Twice treated metatarsus adductus of the elder male sibling



Fig 10: Dental panoramic tomograph (DPT) of the elder sibling revealing hypodontia



Fig 11: Lateral facial profile of the youngest male sibling showing most of the extra oral features of ED



Fig 12: Tapered/conical hypoplastic teeth of the youngest male sibling with sutures on the palate because of the treatment of cleft palate

dibular 2<sup>nd</sup> pre-molars, left permanent maxillary and mandibular 2<sup>nd</sup> molars and all the permanent 3<sup>rd</sup> molars. She had the extraction done for the right permanent maxillary 2<sup>nd</sup> molar which could explain the mild radiolucency at the extraction site along with reduced height of alveolar ridge which is not seen for the other missing teeth (Fig 13). The justification of the above mentioned congenitally missing teeth could be further explained through the spacing of her anterior teeth (U/L) and the resulting malocclusion in the form of cross bite of her right maxillary teeth and edge to edge bite in her left posterior teeth (Fig 14).

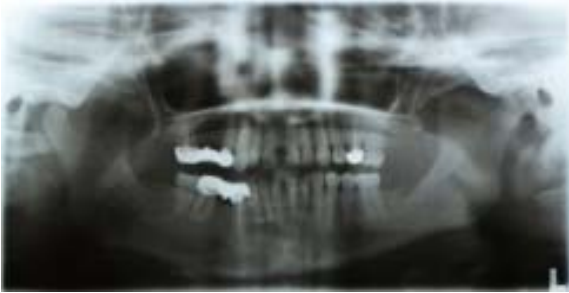


Fig 13: Orthopantomograph showing hypodontia of the mother's dentition



Fig 14: Generalized spacing and malocclusion relative to hypodontia of the mother's dentition

## DISCUSSION

Ectodermal dysplasia is a rare genetic entity in which two or more structures derived from embryonic ectoderm are affected i.e. skin, hair, nails, nerve cells, sweat glands, dentition and parts of eye and ear as well. Hypodontia is an important intraoral finding that may aid in the diagnosis of ectodermal dysplasia. Mid facial defects mainly cleft palate and/or lip maybe found associated with several ED syndromes. Among them two are quite distinctively being reported in literature. These include "Ectrodactyly ectodermal dysplasia with clefting" and "Ankyloblepharon ectodermal dysplasia clefting syndrome" also known as Hay-Well's syndrome. In "Ectrodactyly ectodermal dysplasia with clefting" sometimes being abbreviated as EEC Syndrome (the term first coined by Rudiger in 1970)<sup>33</sup>, there are complex pleiotrophic multiple congenital anomalies/dysplasias in which any of the cardinal features can be present in variable expressions. These features include ectrodactyly (deformities of hands and feet) classically presenting in the form of absent or abnormal (rudimentary) first and second fingers with syndactyly (webbing of the fingers) between the last two fingers; ectodermal dysplasia (anomalies of hair, teeth, nails, nasolacrimal ducts,

sweat glands etc); and cleft palate/lip. Other less common findings include microcephaly, mental retardation deafness and genitourinary anomalies.<sup>34</sup> Hay-Wells syndrome, also known as AEC syndrome (Ankyloblepharon ectodermal dysplasia clefting syndrome), is a rare genetic disorder, initially described by Hay and Wells in 1976<sup>35</sup> in 7 individuals from four families in multiple and complex malformations in the form of ankyloblepharon filiforme adnatum (tissue strands joining the upper and lower eyelid margins); features of ectodermal dysplasia and cleft palate and/or lip, were associated with patterns of autosomal dominant inheritance of varying degrees of penetrance. This report also presents patients suffering from cleft palate, features of ectodermal dysplasia including hypodontia and remarkable additional features of metatarsus adductus and imperforate anus in a particular mode of inheritance.

Metatarsus adductus also known as metatarsus varus/ pigeon toe/ intoeing/ false club foot is a foot deformity in which the front part of the foot is pointed inwards (medially) especially during walking.<sup>36</sup> It is most common in infants and children under 2 years of age and when not the result of simple muscle weakness, may arise due to abnormal infants' position inside mother's womb, either due to the breech position of the foetus (baby's bottom pointed down in the womb) or due to oligohydramnios in which insufficient amniotic fluid is being produced by the mother.<sup>37</sup> There may be a positive family history of the condition as well.<sup>38</sup> Metatarsus adductus though a fairly common problem, is not found associated with classical ectodermal dysplasia and/or ectodermal dysplasia syndromes associated with palate clefting. Hence in this case report, the presence of such features along with imperforate anus associated with ectodermal dysplasia in the form of a syndrome demands a new name. Metatarsus adductus is sometimes associated with another developmental anomaly called developmental dysplasia of the hip (DDH) in which the thigh bone slips out of the hip socket. Metatarsus adductus should be differentially diagnosed from a number of similar anomalies such as club foot (congenital talipes equinovarus), femoral anteversion, calcaneovalgus foot and congenital vertical talus (CVT/Rocker-bottom foot).<sup>36,37</sup>

Imperforate anus or anorectal malformations (ARMs) are birth defects in which the rectum is malformed (opening to the anus is missing or blocked). Anorectal malformations are a spectrum of different



congenital anomalies in males and females, that varies from fairly minor lesions to complex anomalies.<sup>39</sup> Imperforate anus may occur in several forms in relation to the type of associated fistula (A) A low lesion, in which the rectum may end in a blind pouch that does not connect with the colon or there may be narrowing (stenosis) of the anus or no anus. (B) A high lesion, in which the rectum may have openings to the urethra, bladder, base of the penis or scrotum in boys, or vagina in girls, (C) A persistent cloaca, in which the rectum, vagina and urinary tract are joined into a single channel.<sup>39</sup>

Oral rehabilitation of patients suffering from ectodermal dysplasia is necessary to improve the sagittal and vertical relationships during craniofacial growth and development as well as aesthetics, speech and masticatory efficiency.<sup>10</sup> Prosthodontic management of ED patients include fabrication of complete or partial removable prosthesis, overdentures with or without tooth preparations or overdentures with or without attachments, long span fixed partial denture prosthesis and dental implants.<sup>40,41,42,43</sup> Although removable prosthesis are the most common treatment method, implant supported dentures are also suggested to be one of the reconstruction modality for adolescent over 12 years. In situations where implant surgery is indicated, the main problem is insufficient bone therefore if the bone atrophy progresses in these already alveolar deficient patients, implant placement may be impossible without bone grafting.<sup>26</sup>

The placement of implants in growing children is not recommended particularly in the maxilla, where the implants can be submerged by the downward growth of inverting tissues. Similarly, early implant placement in growing children may cause cosmetic problems because implants act similar to ankylosed teeth. However implant placement can be considered in the anterior mandible as the growth of anterior mandible is completed by 3 years of age. An important factor is that it becomes necessary to remake the prosthesis to accommodate changes in occlusal plane as person grows.<sup>44</sup> For example, implant over structures may not be in occlusion with opposing teeth resulting in prosthetic infra occlusion and even adjacent teeth may tilt into the space. Hence removable prosthesis is the widely suggested treatment modality for young patients due to rapid growth of the jaws, however the underdeveloped ridges and xerostomia may still affect the retention and stability.<sup>45</sup> Therefore while fabricating dentures for these patients, care should be taken to obtain a wider distribution of

occlusal loads by extending the denture base as much as possible. Although the atypical anterior conical teeth may not be suitable for removable partial denture stability, however they may be used as abutments for overdentures.<sup>46,47,48</sup> Early prosthetic treatment is generally recommended from the age of 5 years. With regard to child co-operation, dentures may/should also be fabricated as early as 3 to 4 years of age because anodontia or oligodontia leads to atrophy of alveolar ridges, reduced vertical dimension, prominent chin and class III intermaxillary relationship.<sup>45</sup> Positive effects include more self confidence, facial esthetics, speech and masticatory function improvement (by the improvement of tone of muscles of mastication).<sup>45</sup>

## CONCLUSION

Ectodermal dysplasia itself being a rare genetic anomaly, is not found associated with metatarsus adductus and imperforate anus in a particular significant pattern of inheritance as seen in this case report in the form of ectodermal dysplasia syndrome, which makes it even far more rare. Therefore a new name is suggested for this meticulous type of ectodermal dysplasia i.e. HCMI Ectodermal dysplasia syndrome/HCMI-ED syndrome (Hypodontia, cleft palate, metatarsus adductus, imperforate anus associated with ectodermal dysplasia syndrome). Because it is yet not being reported in a syndromic form having such manifestations, therefore genetic evaluation of the patient along with the family is needed for the further justification of a request a new name for this atypical condition.

## REFERENCES

- 1 Thurnam J. Two cases in which the skin, hair and teeth were very imperfectly developed. *Med Chir Trans.* 1848; 31: 71-82.
- 2 Weech AA. Hereditary ectodermal dysplasia (congenital ectodermal defect) *Am J DisChild.* 1929; 37: 766-90.
- 3 Tarjan I, Gabris K, Rozsa N. Early prosthetic treatment of patients with ectodermal dysplasia: a clinical report. *J Prosthet Dent* 2005; 93: 419-24.
- 4 Vieira KA, Teixeira MS, Guirado CG, Gavião MB. Prosthodontic treatment of hypohidrotic ectodermal dysplasia with complete anodontia: case report. *Quintessence Int* 2007; 38: 75-80.
- 5 Abadi B, Herren C. Clinical treatment of ectodermal dysplasia: a case report. *Quintessence Int* 2001; 32: 743-45.
- 6 Yavuz Ý, Ülkü SZ, Ünlü G, Kama JD, Kaya S, Adýgüzel O, Kaya FA, Tümen EC, Zortuk M, Bahsi E, Arslanođlu Z. Ectodermal dysplasia: clinical diagnosis. *Int Dent Med Disorders* 2008; 1: 1-10.
- 7 Hickey AJ, Vergo TJ Jr. Prosthetic treatments for patients with ectodermal dysplasia. *J Prosthet Dent* 2001; 86: 364-68.

- 8 Vieira KA, Teixeira MS, Guirado CG, Gavião MB. Prosthodontic treatment of hypohidrotic ectodermal dysplasia with complete anodontia: case report. *Quintessence Int* 2007; 38: 75-80.
- 9 Nunn JH, Carter NE, Gillgrass TJ, Hobson RS, Jepson NJ, Meehan JG, et al. The interdisciplinary management of hypodontia: background and role of paediatric dentistry. *Br Dent J* 2003; 194: 245-51.
- 10 Tarjan I, Gabris K, Rozsa N. Early prosthetic treatment of patients with ectodermal dysplasia: a clinical report. *J Prosthet Dent* 2005; 93: 419-24.
- 11 Nunn JH, Carter NE, Gillgrass TJ, Hobson RS, Jepson NJ, Meehan JG, et al. The interdisciplinary management of hypodontia: background and role of paediatric dentistry. *Br Dent J* 2003; 194: 245-51.
- 12 Cambiaghi S, Restano L, Paakkonen K, Caputo R, Kere J. Clinical findings in mosaic carriers of hypohidrotic ectodermal dysplasia. *Arch Dermatol* 2000; 136: 217-24.
- 13 Pinheiro M, Freire-Maia N. The ectodermal dysplasias. *Arch Dermatol*. 1982; 118: 215-16.
- 14 Pinheiro M, Freire-Maia N. Ectodermal Dysplasias: A clinical classification and a causal review. *Am J Med Genet*. 1994; 53: 153-62.
- 15 Somayeh Hekmatfar, Karim Jafari, Raziye Meshki, Samaneh Badakhsh. *J Dent Res Dent Clin Dent Prospect* 2012; 6(3): 108-12.
- 16 Mehmet Bania, Ali Melih Tezkirecioglu, Nese Akalc, Tamer Tuzuner. *Eur J Dent* 2010; 4: 215-22.
- 17 Ogunrinde GO, Zubair RO, Ajike SO, Ige SO. *Niger J Clin Pract*. 2012 Jan-Mar; 15(1): 98-100.
- 18 Rajendran R. Diseases of skin. In: Shafer WG, Hine MK, Levy BM, Rajendran R, Sivapathasundaram editors. A text book of oral pathology. 5<sup>th</sup> ed. New Delhi: Elsevier Publications; 2006. p. 1099-102.
- 19 Imirzalioglu P, Uckan S, Haydar SG. Surgical & prosthodontic treatment alternatives for children & adolescents with ectodermal dysplasia: a clinical report. *J Prosthet Dent* 2002; 88: 569-72.
- 20 Tarjan I, Gabris K, Rozsa N. Early prosthetic treatment of patients with ectodermal dysplasia: a clinical report. *J Prosthet Dent* 2005; 93: 419-24.
- 21 Nallanchakravarthy S. Oral rehabilitation of a patient with ectodermal dysplasia with prosthodontics treatment. *Indian J Dermatol*. 2013 May; 58(3): 241.
- 22 Shafer, Hine, Levy Chapter 19-Diseases of the skin. 5<sup>th</sup> ed. New Delhi: Elsevier publications; 2006. Shafer's Textbook of Oral Pathology; pp. 1099-102.
- 23 Lo Muzio L, Bucci P, Carile F, Riccitello F, Scotti C, Cocia E, et al. Prosthetic rehabilitation of a child affected from anhydrotic ectodermal dysplasia: a case report. *J Contemporary Dent Pract* 2005; 6: 120-26.
- 24 Kearns G, Sharma A, Perrott D, Schmidt B, Kaban L, Vargervik K. Placement of endosseous implant in children and adolescent with hereditary ectodermal dysplasia. *Oral Surg Oral Med Oral Pathol Oral Radiol Endontol* 1999; 88: 5-10.
- 25 Anuroopa A, Abdullah J, Lovely M. Oral rehabilitation of a young patient with hypohidrotic ectodermal dysplasia: a clinical report. *Contemporary Clin dent*. 2012 Apr; 3 (suppl 1): S33-S36.
- 26 Imirzalioglu P, Uckan S, Haydar SG. Surgical & prosthodontic treatment alternatives for children & adolescents with ectodermal dysplasia: a clinical report. *J Prosthet Dent* 2002; 88: 569-72.
- 27 Lo Muzio L, Bucci P, Carile F, Riccitello F, Scotti C, Cocia E, et al. Prosthetic rehabilitation of a child affected from anhydrotic ectodermal dysplasia: a case report. *J Contemporary Dent Pract* 2005; 6: 120-26.
- 28 Adiguzel O, Kaya S, Yavuz I, Atakul F. Oral findings of ectodermal dysplasia and literature review. *Int Dent Med Disorders* 2008; 1: 43-49.
- 29 Tso MS, Crawford PJ, Miller J. Hypodontia, ectodermal dysplasia and sweat pore count. *Br Dent J*. 1985 Jan 19; 158(2): 56-60.
- 30 Cambiaghi S, Restano L, Paakkonen K, Caputo R, Kere J. Clinical findings in mosaic carriers of hypohidrotic ectodermal dysplasia. *Arch Dermatol* 2000; 136: 217-24.
- 31 Rouse C, Siegfried E, Breer W, Nahass G. Hair and Sweat Glands in Families with Hypohidrotic Ectodermal Dysplasia. Further Characterization. *Arch Dermatol* 2004; 140: 850-55.
- 32 Seth V. Anhydrotic ectodermal dysplasia. Report of two cases. *Indian J Pediatr* 1971; 38: 70-72.
- 33 Rudiger RA, Haase W, Passarge E. Association of ectrodactyly, ectodermal dysplasia and cleft palate. *Am J Dis Child*. 1970; 120: 160-63.
- 34 Rollnick BR, Hoo JJ. Genitourinary anomalies are a component manifestation in the ectodermal dysplasia, ectrodactyly, cleft lip/palate (EEC) syndrome. *Am J Med Genet* 1988; 29: 131-36.
- 35 Hay RJ, Wells RS. The syndrome of ankyloblepharon, ectodermal defects, and cleft lip and palate: an autosomal dominant condition. *Br J Dermatol* 200; 94: 277-89.
- 36 Jackson, JF, Stricker SJ Review of Common Congenital Foot Deformities. *International Pediatrics / Vol. 18/No. 3/2003*.
- 37 Craig CL, Goldberg MJ, Foot and Leg Problems. *Pediatric in Review Vol. 14/ No.10, 10/1993*.
- 38 Scherl, SA, Common Lower Extremity Problems in Children. *Pediatric in Review, Vol. 25/No. 2, 2/2004*.
- 39 Gore AI, Spencer PS, The Newborn Foot. *American Family Physician* 2004; 69: 865-72.
- 40 Lakomski J, Kobiela A, Trzeciak WH. Correcting facial dysmorphism in a patient with anhydrotic ectodermal dysplasia: A clinical report. *J Prosthet Dent* 1998; 80: 524-26.
- 41 Escobar V, Epker BN. Alveolar bone growth in response to endosteal implants in two patients with ectodermal dysplasia. *Int J Oral Maxillofac Surg*. 1988; 27: 455-57.
- 42 Penarrocha-Diago M, Uribe-Orion R, Rambla-Ferrer J, Guarinos-Carbo J. Fixed rehabilitation of a patient of hypohidrotic ectodermal dysplasia using zygomatic-implants. *Oral Surg Oral Med Oral Pathol Oral Radiol Endontol* 2004; 98: 161-65.
- 43 PAe A, Kim K, Kim HS, Kwon KR. Over denture restoration in growing patient with hypohidrotic Ectodermal Dysplasia- A clinical report. *Quintessence Int*. 2011; 42: 235-38.
- 44 Rad AS, Siadat H, Monzavi A, Mangoli AA. Full mouth rehabilitation of hypohidrotic Ectodermal Dysplasia patient with dental implants: A clinical report. *J Prosthodont* 2007; 16: 209-13.
- 45 Ramos V, Giebink DL, Fisher JG, Christensen LC. Complete dentures for a child with hypohidrotic ectodermal dysplasia: a clinical report. *J Prosthet Dent* 1995; 74: 329-31.
- 46 Dyson JE. Prosthodontics for children. In: Wei SHY, ed *Pediatric Dentistry and Orthodontics: Total Patient Care*. Philadelphia: Lea & Febiger; 1988: 259-74.
- 47 Abadi BJ, Kimmel NA, Falace DA. Modified overdentures for the management of oligodontia and developmental defects. *ASDC J Dent Child* 1982; 49: 123-26.
- 48 O'Dwyer MR, Renner RP, Ferguson FS. Overdenture treatment – One aspect of the team approach for the EEC syndrome patient. *J Pedod* 1984; 8: 192-205.