

Original Article

Ectodermal Dysplasia and Malocclusion – Retrospective study

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Abstract

Aim: The aim of this article is to find out the malocclusion and cranio-maxillofacial deformation in patients with ectodermal dysplasia.

Methods: A total of 10 patients (6 males and 4 females, aged 5–26 years) with ectodermal dysplasia underwent clinical examination and treatment.

Results: Most of the patients had a short face with an abnormal facial concavity, a maxillary retrognathism, mild mandible prognathism, with thin or scanty hair, very smooth, dried out skin. Depending on age and malocclusion, patients were treated with prosthodontic and orthodontic approaches or implant treatment.

Conclusion: Aesthetic dental interventions in patients with ED assist in improvement of a positive self - image and on the whole oral health is maintained.

Key Words: Ectodermal Dysplasia, Oligodontia, Anodontia, Malocclusion

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Introduction

The ectodermal dysplasias (EDs) contain a large, heterogeneous group of inherited disorders that are defined by major defects in the development of two or more tissues derived from developing ectoderm¹.

ED patients show the clinical traits like hypotrichosis, hypohidrosis, and cranial abnormality. Patients frequently have markedly small face because of frontal bossing, and a low nasal bridge. The absence of sweat glands results in very smooth, dry skin and/or hyperkeratosis of hands and feet. Oral effects may be noticeable as anodontia, hypodontia, conical teeth, and lack of alveolar ridge development².

ED is considered a reasonably uncommon disorder, with a reported incidence of 1 in 10,000 to 1 in 100,000 births². Many genetic material defects can cause ectodermal dysplasia². The most common form of ectodermal dysplasia frequently affects men. Other forms of the disease affect men and women uniformly². Dental treatment of the clinical manifestations of ED can have intense impact on these patients, because the ability to appear and behave like their peers is vital to their psychosomatic development². The article shows that management is not only required to develop patient's functional and esthetics, but also considerably increases their self confidence and self-respect². Depending on their age and abnormality, periodontal therapy, caries management, and prosthodontic or orthodontic treatment were assessed^{3,4,5}.

Materials and Methods

This retrospective study was carried out over the period of two years on 10 patients (6 males, 4 females, aged 5–26 years) with a diagnosis of ED, in the Department Of Orthodontics, Chettinad Dental College and Research Institute. Clinical and radiographical examination for diagnosis and therapy were conducted and it included the skull, face, hair, jaws, teeth, nails, skin, and sweat glands⁶.

Results

All major symptoms of ED were studied, such as thin hair (trichodysplasia) (Fig 1), smooth skin (hypohidrosis), unusual finger and toe nails, skull and facial abnormalities and the family history of the patients were considered. The hair is usually light colored and scanty. Hypotrichosis and partial or total alopecia are commonly reported. Body hair follicles are frequently scanty or missing⁷. Some patients had dystrophic, hypertrophic, unusually keratinised, thickened, discolored, striated, split or fragmented nails. The epidermis was dry, fine and smooth (Fig 2), hypopigmented, with patches of hyperkeratosis or eczematous⁷. Oligodontia (Fig 3) or anodontia are very frequent features, but undeveloped or conical teeth and enamel dysplasia may be noticed⁸ (Table 1).

Steiner analysis was used to determine the dento-facial abnormalities. The mean value of Steiner's analysis is as follows: SNA Angle (Sella Nasion and Point B Angle) (76.60), SNB Angle (Sella Nasion and Point B Angle) (820), ANB Angle (Point A to Nasion to Point B)

(-5.50), SND Angle (Sella – Nasion to Point D) (82.30) and SN to Go-Gn Angle (Sella – Nasion to Gonion – Gnathion) (29.60) which is depicted in the tabulation (Table 2). Steiner analysis revealed a mean SNA of 76.60 which shows an incidence of maxillary retrognathism. Mean value of 820 for SNB shows a relatively normal position of mandible. Additional results found were facial concavity which was reflected in ANB value of -5.50, however increased SND value of 82.30 shows chin prominence. Retrognathism is more prominent in the maxilla. Skeletal class III patients usually show high angles and large gonial angles on the mandible; however; these patients showed low angles of 29.60 because of missing teeth.

Discussion

Steiner analysis revealed a low mandibular plane angle and facial concavity in five of the affected patients (patients no 3,4,5,6,9) which are enlisted in table 1. Most of the patients had maxillary retrognathism and mandibular prognathism. Skeletal class III patients typically show high mandibular plane angles on the mandible, however; our patients showed low angles as of missing multiple teeth. As an outcome of retrognathism of the maxilla and mandible, the soft tissues also showed retrusion.

Ten patients in this study had hypodontia. Four patients had fewer than 10 teeth, and six patients had more than 10 teeth (Table 1). ED usually affects the hair, teeth, nails, and/or skin of the patients⁹. Most of our ED patients had partial or complete lack of certain sweat glands, causing lack or less sweating, heat intolerance, and sometimes fever; abnormally sparse hair (hypotrichosis) and absence of teeth (anodontia) and abnormality of certain teeth¹⁰. Our patients with ED also had typical facial abnormalities with prominent forehead, sunken nasal bridge (so called "saddle nose") abnormally thick lips, and a large chin¹¹. The skin of most of the patients was abnormally thin, dry, and soft with abnormal lack of pigmentation (hypopigmentation)¹¹. The affected infants and children exhibited underdevelopment (hypoplasia) or absence (aplasia) of mucous glands in the respiratory and gastrointestinal tracts and in a few cases there were decreased function of immune system, causing an increased susceptibility to certain infections and/or allergic conditions¹². Several affected children experienced recurrent attacks of wheezing and breathlessness (asthma)¹². Certain children with this disease had difficulty in controlling fevers. Affected adults were not capable to tolerate a humid environment and require unique actions to maintain a normal body temperature¹².

Table 1: Clinical summary of ED subjects evaluated

PATIENT NO's	AGE (Years)	SEX	NO. OF TEETH PRESENT	PROSTHETIC TREATMENT	PEG SHAPED CONICAL TEETH
1	5	Male	6	Removable Denture	Yes
2	9	Male	11	No Denture	Yes
3	17	Female	8	Removable Denture	Yes
4	21	Female	3	Fixed Denture	No
5	26	Female	24	Fixed Denture	Yes
6	18	Male	17	Removable Denture	Yes
7	12	Female	26	Removable Denture	Yes
8	6	Male	14	No Denture	No
9	21	Male	9	Removable Denture	Yes
10	15	Male	15	Fixed Denture	Yes

Table 2: Summary of the results of Steiner cephalometric analysis performed on severely affected patients.

Cephalometric parameters	Normal values	Patient 3	Patient 4	Patient 5	Patient 6	Patient 9
SNA	82 ± 2°	74°	72°	79°	75°	77°
SNB	80 ± 2°	80°	84°	87°	81°	80°
ANB	2°	6°	12°	8°	6°	3°
SND	76°	81°	87°	90°	84°	81°
SN to Go-Gn	32°	35°	38°	33°	29°	31°
Skeletal relation	I	III	III	III	III	III

In some patients hypoplasticity or absence of oral and mucous gland and even salivary glands were noted¹³. The absence of the salivary glands may lead to dryness of mouth (xerostomia). Dysphagia was also seen¹³. Malformation of the enamel was seen in all patients, resulting in dental caries and altered contour of the teeth, leading to pegged shaped appearance and accessory cusps^{14,15}. The nails of all the affected patients were normal. The hypodontia vary in each case, but usually 5 to 7 permanent teeth (Table 1) are present, the teeth are slightly smaller than normal, and the eruption was frequently delayed for all patients.



Fig 1: ED patient with sparse hair



Fig 2: Smooth skin and patches of hyperkeratosis



Fig 3: ED case with oligodontia

Conclusion

Dental management is required in patients with ED and a few children should wear dentures as early as 2 years of age^{15,16}. It is essential to visit dentist opinion for

safeguarding the alveolar ridge for later dental intervention. Various sets of denture replacements are regularly considered necessary as the child grows, and dental implants may be an option in adolescence, once the jaws are completely developed¹⁶. Prosthetic teeth are implanted in adults for mastication and speech^{16,17}. Importantly, aesthetic dental interventions in patients with ED assist in improvement of a positive self - image and on the whole oral health is maintained¹⁷.

Conflict of Interest

The authors declare no conflict of interest.

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Are smartphones smart enough to detect skin cancer?

In Caucasians, skin cancers (melanoma and non-melanomatous cancers) are among the commonest types of cancer. Most of them, especially non-melanomatous lesions, are curable if they are detected early. Several commercially available smartphone applications ("UMSkinCheck", "Mole Detective") have been designed based on well-known clinical algorithms to detect these lesions early. But how effective are these in practice? Several studies have examined this question and their conclusions are similar: these applications can accurately pickup early lesions (all types) only in 81% of cases and may altogether miss melanoma (a particularly aggressive skin cancer) in nearly 30% of cases. So using them alone without consulting a dermatologist to decipher a suspicious skin lesion is a bad idea as it may lead to delayed detection and treatment. So, smart phones are not there yet, when it comes to skin cancer. Calling something "smart", does not make it one! (Whiteman, H. (2014, November 19). "Skin cancer: how effective are smartphone apps for early detection?" <http://www.medicalnewstoday.com/articles/285751.php>.)

- Dr. K. Ramesh Rao