Role of Dentists in the Treatment of Ectodermal Dysplasia

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Key Points
➢ Ectodermal dysplasia is rare genetic disorder. ➢ Mutation are x-linked. ➢ Use of implants and prosthesis as treatment for this condition.

Ectodermal dysplasia (ED) describes a heterogeneous group of rare congenital diseases, which is caused by genetic disorders during the embryonic development phase in which the affected structures are of ectodermal origin, at least one of these changes occurring in teeth, nails, hair or sweat glands.[1, 2]

Distinct classification are attributed to ectodermal dysplasia over time.[3-6] There are various types of EDs[7] and therefore the genetic mutations that give rise to them are numerous[3] there's a broad spectrum of clinical presentation for the individuals affected with this condition.[8] Among all the clinical indications of EDs, some are observed, since very early, in the child's oral cavity, with several characteristic and symptoms.[9]

Dentists, especially paediatric dentists have privileged and extremely important role in the diagnosis and treatment of oral manifestations of disease, due to the close contact established with children from an early age. In this sense, it is must need to know and recognize its possible orofacial manifestations,[10] as well as the available and most appropriate clinical approaches for each child.

Ectodermal dysplasia:

During the third week of fetal life, the human embryo is already formed by three layers of cells: ectoderm, mesoderm and endoderm, from which the all the tissue and organ of the fetus develop. The outmost layer of the embryonic leaflet, referred as ectoderm, is responsible for the origin of several structure such as: epidermis and its attachments (hair and nails), the sensory epithelia of the sensory organs, the central and peripheral nervous system ,the dental enamel and several glands including sebaceous sweat,[4] lacrimal, mucous and salivary glands.[11] When genetic disorder occur at this stage of embryonic development, that lead to an abnormal structure and occasionally, structure derived from the mesoderm, a condition called ectodermal dysplasia is observed[12,13]

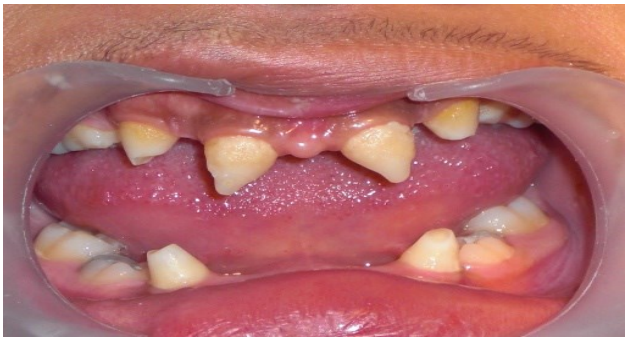
Genetics:

ED is mainly because of maldevelopment of ectodermal derivatives in the embryonic life. X-linked hypo hidrotic ED has been mapped to the Xq-12-q13.1 band.[18,20-23] We found that 60% to 80% of women have some extent of hypodontia. Prenatal testing would be possible for pregnant at high risk if pathogenic variants in the family were known.[18,24] there was mutation of EDA gene c.911A → G. Also in Y304 mediated, trimerization of EDA and cysteine replacement occurred to stop protein secretion. A pregnancy with two male sex twins was shown on ultrasonographic examination without dental germs.[25] Mutation in PKP 1 gene also causes ED and symptom like generalized skin fragility, alopecia, nail dystrophy, planter palmar dystrophy and painfully fissures.[26] mutation in Val472Glyfs 28 was also observed that causes premature protein termination.[27] Most common cause of HED is the pathogenic varianys of the EDA gene in Xq13.1. encodes for ectodysplasin which is a molecule that plays role in epithelial-mesenchymal communication during skin development process. Other gene like EDAR, EDARADDD and WNT10A are also linked with dysplasia. Where they will show recessive or dominant inheritance pattern.[19,28-30] Studies have shown that the hypo hidrotic subtype is X-linked. Pathogenic variants in the EDA gene have been found to exist in Xq13.1, EDAR, EDARADD and WNT10A.

Oral manifestation:

The most common oral appearance includes anodontia, retained teeth deficient alveolar, growth, prominent lips.[14,15,28,32,33] Central incisors, canines and

first molar in maxilla and canines, first premolar and molar in the mandible show the highest percentage: there is less probability in the anterior teeth.[14] In case of absence of teeth poor development of the alveolar bone occurs. In some cases, cleft lip and palate are also observed.[34] While considering the shape and size of teeth they are small or conical and bulbous or taurodontic respectively. Large shapes between teeth are also observed enamel is very suspeptible to decay and mechanical damage. Poorly nuristed mucosa proaky voice and difficulty in swallowing may also occur.[17] Defects in the number of teeth is a very common problem in these dysplasia's. Third molars usually do not develop in 25% of the world's population. The number of teeth can range from more than a dozen, through several, or the lack of all permanent teeth and even, in some cases the lack of both dentitions is present.[18,31,35,36] It is found that most frequent missing teeth are the upper lateral incisors , upper and lower premolars.[37]



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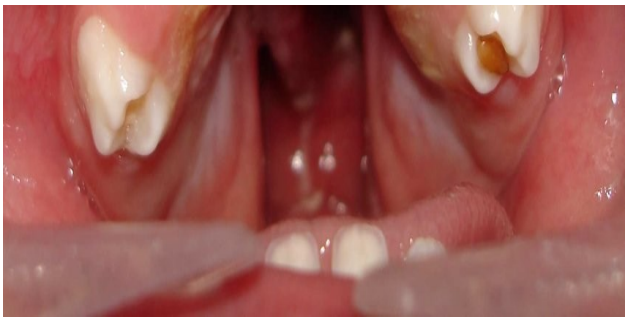

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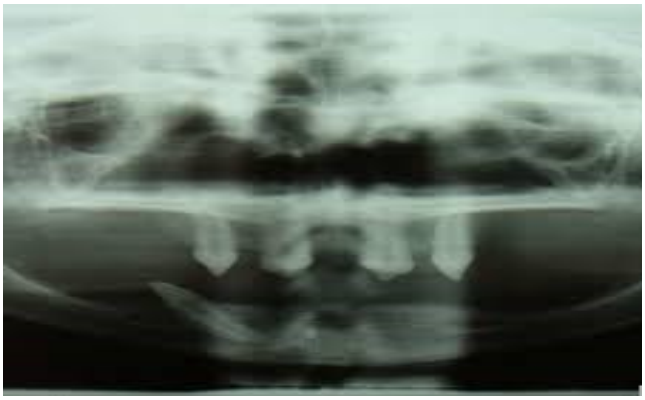

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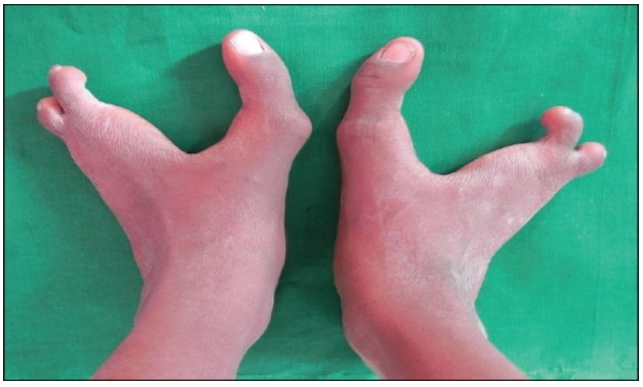

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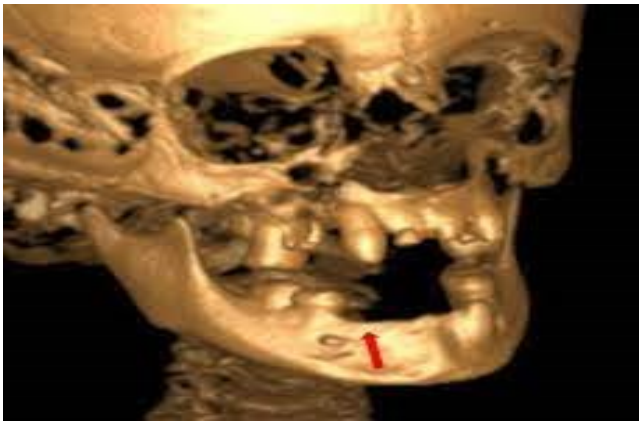

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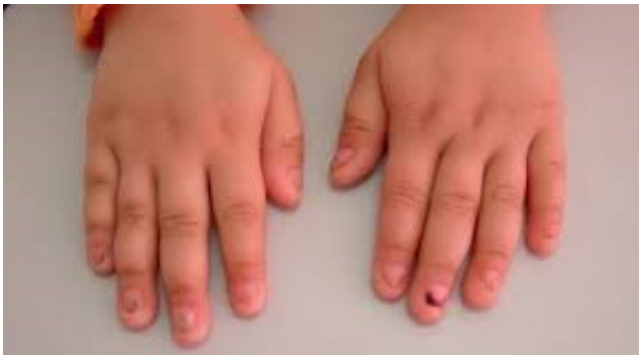

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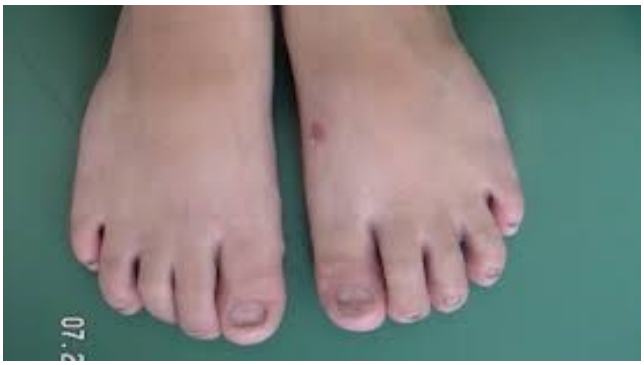

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Diagnosis and Treatment of ED:

ED is also described as genetically transmitted rare multisystem disorder. Mutations, autosomal dominant, autosomal recessive and X-linked recessive basic mode of inheritance. The medical history and pedigree of the families has confirmed the hereditary nature of the disorder. As it is difficult to find out accurate type so clinical diagnosis of ED is also difficult most specifically when there is not any collaboration between the patient a different medical specialist.

Steiner's cephalometric analyses have proven helpful to detect the presence of maxillary reduction, labial retrusion, chin prominence, nasolabial reinforcement, facial height reduction, and facial concavity. Partial or total dental aging could distrupt bone growth in the chin.

Due to absence of partial bone growth dental implant cannot be used in children.[41-43] They can be placed only after the bones of the jaw have completed their growth. Discrepancies between the alveolus and the implant are generally due to dentoalveolar growth. The placement of dental implants during the growth period will impede jaw growth and the movement of the teeth into their natural positions within the oral cavity. The earliest recommended ages for dental implants are as follows: at least 15 years for females and 17 years for males.

As the median sutures of mandible are closed up to age of six-year implants can be replaced in the anterior region of the mandible for the support of over denture in case of adult patients when the dentoalveolar growth is insuffient dentures can be placed via zygomatic fixation for the support of denture in maxilla for oral rehabilitation for zygomatic surgery is a save alternative to conventional modalities but the surgeon must know that it is difficult and not risk free procedure. A highly experienced surgeon must be consulted for successful outcome.[40]

Role of dentists:

Before the treatment is started, the Dentist must have complete knowledge of ED and have experience about main signs and symptoms. It is expected that molecular diagnosis will more affordable and easy in future. A dentist must read previous publications to have knowledge about prevalence characteristics variabilities of clinical features of ED and this information has to be delivered to patients with ED that can improve the diagnosis and help in clinical management of this disorder. [28,39]

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