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SYNDROMES ASSOCIATED WITH DEVELOPMENTAL ANOMALIES OF THE TEETH-A COMPLETE LITERATURE REVIEW

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ABSTRACT Dental anomalies are common congenital malformations which can occur as isolated findings or part of a syndrome. This review focuses on genetic causes of abnormal tooth development and implications of these abnormalities for clinical care. These includes abnormalities due to shape size and structure of the tooth such as Microdontia, Macrodontia, Amelogenesis Imperfecta ,Anodontia, Talons cusp, Dentin dysplasia, Dentinogenesis Imperfecta.This is followed by discussion of syndromes associated with the the developmental disturbances of the tooth such as size,shape,number and structure of the tooth . Next, we review the clinical conditions abnormalities and treatment for the syndromes with abnormalities in the tooth size, shape, and form.

KEYWORDS : Anomalies Of Teeth, Developmental Defects, Syndromes

INTRODUCTION:

Developmental disturbance of teeth is a highly complex biological process, when there is a disturbance in the developmental processes they results in the development disturbance of teeth which cause a change in size, shape, number and the structure of the tooth.[1]

SYNDROME:

A syndrome is defined as a group of symptoms corresponding to a specific condition that is associated with either shape, size, number or structure of teeth.

Orofacial Syndrome:

Oro-facial syndrome is a group of condition which affects the development of oral cavity (tongue, teeth and jaw).There are symptoms present for development disturbances of teeth that results in abnormal growth of tooth.

Incidence:

Oro-facial syndrome has an occurrence rate of 1 in 50,000 to 250,000 newborns. There are several types of oro-facial syndromes but type 1 oro-facial syndrome is the most common type. The other forms of oro-facial syndrome are very rare that they occur only in very few numbers.

Developmental Disturbances In Size Of Teeth

- Microdontia
- Macrodontia

Developmental Disturbances In Number Of Teeth

- Anodontia
- Supernumerary teeth
- Predeciduous dentition

Developmental Disturbances Of Shape Of Teeth

- Dens Invaginatus
- Taurodontism
- Talons cusp

Developmental disturbances in structure of teeth

- Amelogenesis imperfecta
- Dentinogenesis imperfecta
- Dentin dysplasia

Developmental Disturbances In Size Of The Tooth:

Microdontia:

Microdontia refers to the reduction in the tooth size than that of the normal size and the syndrome associated with microdontia is [2]

Williams Syndrome:

- Williams syndrome is caused by the deletion of genes on chromosome 7q11.23 which codes for elastin.
- It affects one in ten thousand individuals
- Individuals who are affected by this syndrome suffer conditions like that of a disease or seem to be dead when they are exposed to anesthesia or sedatives due to abnormalities in the cardiovascular system.[3]

Macrodontia:

Macrodontia refers to the increase in the tooth size than that of the normal size and the syndrome associated with macrodontia is [2]

Otodental Syndrome:

Otodental syndrome is a dental abnormality associated with the sensory neural high-frequency hearing loss. It is mostly presented with globe shaped posterior teeth – globodontia. Though etiology is not specific It might be caused due to defect in dental papilla due to hereditary enzyme derangement. It is highly associated with clinical features such as generalised macrodontia, combination of hypodontia and hyperdontia. Premolars are most commonly affected , there is a significant delay in mineralization of premolars which leads to it's absence in the maxilla and delayed eruption in the mandible[4]

Developmental Disturbance In Structure Of The Tooth:
Amelogenesis Imperfecta:
Amelogenesis imperfecta is a defect that in the enamel of tooth it that occurs due to irregular or uncommon inheritance pattern. It is differentiated into three main types

They are :
- Hypoplastic
- Hypocalcified
- Hypomaturation

The classifications is based upon the clinical abnormalities that takes place during enamel formation.[2]
The syndrome associated with amelogenesis imperfecta is

Lubinsky Syndrome:
Keppen-Lubinsky syndrome is causedby a delayed development that results in large prominent eyes, a narrow nasal bridge, rented upper lips, a high palatal arch, an open mouth, tightly appearing skin, an old aged appearance and lipodystrophy.

Etiology :
Keppen-Lubinsky syndrome occurs due to mutations occuring in the inwardly rectifying K+ channel encoded by KCNJ6 gene.

Clinical Conditions :
The typical oral features observed in lubinsky syndrome are significant enamel hypoplasia, delayed or absent tooth eruption, ectopic eruption route, pulpal and gingival calcification in primary and permanent dentition.[5]

DENTINOGENESIS IMPERFECTA
It is an autosomal dominant condition that's seen in both deciduous and permanent dentition. The teeth appear from grey to yellowish brown colour,the affected tooth has broad enamel with constriction near the cervical area giving it tulip shaped. In radiograph the affected tooth seems to be lacking pulp chamber and root canal.[2]

ETIOLOGY:
This occurs due to break of enamel and the exposure of the dentin and this occurs due to a defective gene in the chromosome 4 that encodes sialophosphoprotein.

TYPES:
There are two types of Dentinogenesis Imperfecta:
- Dentinogenesis Imperfecta 1 (with opalescent dentin) which corresponds to Dentinogenesis imperfecta of Shield's classification.
- Dentinogenesis Imperfecta 2 (Bradywine type) corresponds to Dentinogenesis Imperfecta 3 of Shield's classification.[2]

The syndrome associated with Dentinogenesis Imperfecta is

Ehlers-Danlos Syndrome:
EHLERS-DANLOS syndrome is a group of rare inherited conditions in which the connective tissues are affected.

Clinical Conditions:
Connective tissues provide clinical findings in the skin, tendons, ligaments, blood vessels, internal organs and bones.

TREATMENT:
Treatment is provided based on the severity of the disease. Treatment helps in the management of symptoms and monitoring of the complications. Treatment includes drugs, physiotherapy and surgery.

Therapies:
Physical therapy is given.

Medication:
Analgesic along with Non steroidal anti- inflammatory drugs (NSAIDs) are administered.[6]

DENTIN DYSPLASIA
Dentin dysplasia is a autosomal dominant hereditary disease, developmental disturbance of dentin formation characterized by normal enamel and it is associated with formation of abnormal pulpal structure.

This condition has been referred to as (type I) radicular dentin dysplasia and (type II)coronal dentin dysplasia [2]

Clinical Features
Type I dentin dysplasia:
In this condition slight amber translucency appears in the teeth. Extreme mobility are seen in type 1 dentin dysplasia and are commonly exfoliated prematurely or abnormal short roots are resulting after the minor trauma. In this type both dentitions are affected.

Type II dentin dysplasia:
The clinical appearance of the permanent dentition is normal. However, the primary dentition have the yellow, brown, or bluish-grey opalescent appearance as seen in dentinogenesis imperfecta.[2]

Goldblatt Syndrome:
- Goldblatt syndrome or odontochondrodysplasia is a rare primary bone dysplasia characterized by the association of generalized joint laxity ,spondylometaphyseal dysplasia, and dentinogenesis imperfecta. Main skeletal abnormalities comprise short stature, narrow chest and brachydactyly.[7]
- Goldblatt in 1991 described a single child with an unusual combination of radiographic and clinical findings. Dentinogenesis imperfecta (DI) is relatively have a specific sign for osteogenesis Imperfecta (OI), Goldblatt noted that their patient with white sclerae and no fractures or Wormian bones. A year later this conclusion was supported by the identification of two further cases early childhood.The signs that are present in the newborn period are not entirely specific and the differential diagnosis includes spondylometaphyseal dysplasia (SMD) sedaghatian type or platyspondyle lethal dysplasia (PSLD)Torrance type.[8]

Developmental Disturbances In Number Of Tooth:
Anadontia
- Anodontia is a congenital absence of teeth rare condition which involve in both permanent and deciduous dentition.Ectodermal dysplasia is developmental disorder which common and frequently associated with this condition .
- True partial anodontia is the rare condition which involves one or more teeth. Hereditary ectodermal dysplasia may be associated with anodontia, and in these condition the few teeth that are present may have deformed or misshapen, frequently shows cone-shaped .[2]

ECTODERMAL DYSPLASIA:
- Ectodermal dysplasia(ED) is a rare hereditary disorder . It is a genetic disorder which was autosomal recessive and it was first described by Thurnam in 1848.
- It is the disorder the affecting the development or function of the teeth, hair, nails, sweat glands and congenital heart failure. Hypohidrotic ectodermal dysplasia is being transmitted as a X-linked recessive trait.[2]

According to Freire-Maia's classification, EDs are divided into two groups:

Group A which consists of all the entities with defects in two or more of the standard structures

Group B comprises of those with disturbances in only one of these structures plus another ectodermal defect.[9]

Clinical Conditions:

- Ectodermal dysplasia is characterized by various ectodermal disturbances which involve nails and teeth and sometimes congenital heart diseases also, the nail shows hypoplastic and koilonychia.
- The limbs are shortened and thickened in this anomaly. The hands are affected by bilateral polyductyly [2]

Supernumerary Teeth:

- Supernumerary teeth is an additional tooth in deciduous or permanent dentition and it can be noticed in any region of arch i.e molars,premolars or anterior teeth.It is developed from a third tooth bud arising from the dental lamina.
- The incidence for supernumerary tooth is 2.1% for permanent dentition and 0.8% for deciduous dentition. gardner syndrome,cleidocranial dysplasia are the common syndrome associated with this is Gardner syndrome.[2]

Gardner Syndrome:

- Gardner syndrome is due to a single pleiotropic gene ,an autosomal dominant disease characterized by numerous adenomatous polyps lining the intestinal mucosal surface. It is the variant of familial adenomatous polyposis (FAP)
- This syndrome Consists of multiple polyposis of the intestine, osteomas of theBones such as skull and jaws, multiple epidermoid or sebaceous cysts of the skin, particularly on the scalp and the impacted supernumerary and permanent teeth [10]

Clinical Conditions

- Gardner syndrome may be present with skull, osteomas of the mandible and epidermal cysts, or fibromatosis.
- Desmoid tumors, which occur in the chest, abdominal region and upper and lower extremities, affect the tissues that comprise the tendons and ligamentsThe key features of Gardner syndrome is the development of intestinal polyposis and colorectal adenocarcinoma [2]

Predeciduous Teeth:

- The predeciduous teeth is defined as epithelial structures present on the gingiva over the alveolar ridge, which can be easily removed.some infants are born with this structures which appear to be erupted teeth which occurs in the mandibular incisor area. It is present at the time of birth so it is called as natal teeth.[2]
- It can be also described as by the terminologies that are Dentitia praecox, dens connatalis, congenital teeth, foetal teeth, infancy teeth.[11]

HALLERMAN STERRIF SYNDROME:

- Hallermann-Streiff syndrome (HSS) is named after German ophthalmologist Wilhelm Hallermann and Italian–Swiss ophthalmologist Enrico Bernardo Streiff, a rare congenital disorder which is characterized by malformations of craniofa cial region,eye abnormalities, hypotrichosis, dental defects
- The etiology of Hallermann-Streiff syndrome is still unknown and it is widely believed to be caused by mutation which is called as de novo dominant mutation.In craniofacial features include a brachycephaly which is short and broad head with an unusually prominent forehead ,hypoplastic mandible (small underdeveloped lower jaw), Mouth with narrow,highly arched roof and a thin tapering nose.[12]

Development Disturbances In Shape Of Tooth:

Taurodontism:

- Taurodontism refers to bull-like teeth, its usage is derived from similarity of teeth of cud-chewing animals.[2]

RAPP – Hodgkin Syndrome:

- Rapp–Hodgkin syndrome is an inherited as an autosomal dominant trait with variable expression.This condition has significant clinical overlap with both the EEC and AEC syndromes, and they are all allelic disorders and they associated with mutations of p63 gene.Rapp-Hodgkin syndrome is a rare disease which affect your hair, nails, skin, sweat glands, and teeth.
- It have signs and symptoms which overlap extensively with those of ankyloblepharon-ectodermal defects-cleft lip/palate (AEC) syndrome, also called Hay-Wells syndrome.
- These include missing teeth, wide gaps between teeth, cone-shaped teeth, and thin enamel (the hard outer covering of the teeth).[13]

TALON CUSP:

- The talon cusp, projects lingually from cingulum area of maxillary and mandibular permanent incisor and resembles eagle feet.[2]

Rubinstein Taybi Syndrome:

- Talon cusp is a developmental anomaly that is generally noted to as accessory cusp. It is seen as projection of tooth structure from cervical third of the lingual aspect, extending at least to half the crown length. It occurs due to alteration of the inner enamel epithelium during tooth development.
- The anomaly is prevalent in about 1–6 percent of the population, that associated with considerable ethnic variation.
- Talon cusps sometimes associated with few syndromes. Usually in maxillary incisors, and rarely on mandibular.
- Even it also occurrence in the facial aspect of mandibular incisors, with only up to 20 cases having been reported .
- However, none of them associated with fusion.
- The case report highlights the extreme rarity of such phenomenon and discusses clinical features and management.[14]

DENS INVAGINATES:

- Dens invaginates arises as a result of invagination in the surface of tooth crown before calcification.
- Permanent maxillary lateral incisor are commonly involved sometimes,maxillary central incisors are also involved[2]

Mohr's Syndrome:

Mohr's syndrome is a rare genetic disorder of oral cavity, facial and digits anomalies. Orofacial digital syndrome of type II, also called the "Mohr syndrome" is very rare subtype which has been reported scarcely in Asia.It is an autosomal recessive inherited disease which is characterized by median cleft lip, poly lobed tongue, absence of medial incisors.It is a very rare disorder with incidence of 1/1000000 worldwide. It has been reported scarcely in Asia especially in Japanese patients.[15]

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