Review Article

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Syndromes Presenting the Oral Manifestations: A Short Review

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ABSTRACT

Background: There are variety of syndromes which shows multiple manifestations in oral cavity. Awareness and familiarity of these syndrome and their oral manifestations is important as it will help in diagnosis and management of syndromes and dental diseases. In this review article, we are discussing the various syndromes with their oral manifestation.

Keywords: Syndromes, oral manifestation, knowledge.

INTRODUCTION

In the past some years, awful growth has been seen in researches which is related to genetics. These researches increases the awareness of variety of genetic disorders as well as various syndromes. There are many syndromes which affects various body parts or body organs. Out of theses, oral cavity is considered as one of the most common site, where various syndromes presents their manifestation in various types of dental diseases. It has been observed that some syndromes are specifically seen in children's, while some are predominant in adults. So, in this review article we will discuss about some syndromes which shows oral manifestations predominantly.

ZELLWEGER SYNDROME

Zellweger syndrome is known as autosomal recessive disorder. This syndrome is caused by mutation in various genes which is involved in biogenesis of peroxisome1. Incidence of zellweger syndrome is approximate[1]: 100,000 live births[2].

Patients suffering from Zellweger syndrome are characterized by various features. These includes craniofacial abnormalities and neurological abnormalities. The craniofacial abnormalities includes large fontanels, epicanthal folds, external ear deformities, high forehead, hypoplastic supraorbital ridges. Neurological abnormalities include abnormal moro reflex, epileptic seizures, severe hypotonia, psychomotor retardation[3,4,5] Oral manifestation: It includes malocclusion, congenitally missing teeth, delayed dental development, micrognathia and high arched palate.[6,7]

ROBINOW SYNDROME

Robinow syndrome was first described in the year 1969 by Robinow and their colleagues. Robinow syndrome may be autosomal recessive or autosomal dominant, that's why they are referred to as recessive robinow syndrome or dominant Robinow syndrome.[8,9] This syndrome affects multiple body

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parts like skeletal system, urogenital system, cardiovascular system, cranium as well as face. Various cardiovascular abnormalities includes ventricular as well as atrial septal defect, fallot tetralogy, aortic coarctation and tricuspid atresia. Various skeletal abnormalities includes short stature, rib fusion, vertebral malsegmantation, and hemivertrbrae[10,11].

Oral manifestation : Various oral manifestations shows by Robinow syndromes are gingival hyperplasia, dental crowding, cleft lip or cleft palate, malocclusion, hypodontia, short rooted teeth, narrow pulp chambers, deep bite, anterior as well as posterior cross bites. Various tongue anomalies seen in this syndromes are macroglossia or ankyloglossia or bifid tongue or shortened tongue[11,12,13].

CATCH 22 SYNDROME

This syndrome occurs because of chromosome 22 deletion. It has been seen that this syndrome occurs more in females as compared to males. Characteristic feature of these syndromes are low nasal bridge, malformed auricles, malar hypoplasia, prominent nose[14,15]. Oral manifestation : enamel hypoplasia, hypodontia, delayed eruption of permanent teeth, aberrant tooth shape, enamel hypomineralization, dental caries[16,17]

SECKEL SYNDROME

Seckel syndrome is first narrated by Helmut P.G. Seckel in the year 196018. This syndrome is extremely rare diseases. There are 3 subtypes of Seckel syndrome : Seckel I, seckel II, seckel III[19,20].

General characteristic feature includes postnatal growth retardation, mental retardation, microcephaly, bird-headed facial appearance.Various skeletal defects are scoliosis, hypoplaisa of the proximal radius and fibula, delayed bone age, elbow flexion contracture. This syndrome also shows some neurologic features. This includes large basal ganglia, seizures, arachnoid cysts, pachygyria and cerebellar vermis hypoplasia[21,22]. Oral manifestation: Enamel hypoplasia, cleft palate, hypodontia, high arched palate, dental crowding[23].

SILVER RUSSELL SYNDROME

Silver Russell syndrome is pre natal as well as post natal growth disorder of unknown etiology. Various characteristics features includes congenital asymmetrical body, psychomotor impairements, low birth weight, fifth finger clinodactyly and low set ears[24,25,26].

Oral manifestation : Hypodontia, delayed dental development, hypomineralization of tooth enamel in primary as well as permanent dentition, microdontia, high arched palate[27,28,29].

DUBOWITZ SYNDROME

In the year 1965,Victor Dubowitz described the Dubowitz syndrome for the first time. The characteristic feature of this syndrome includes various congenital anomalies, hematologic malignancies, growth failure, blood dyscrasias[30].

Oral manifestation : Dubowitz syndrome shows various oral manifestations. It includes microdontia, diastema, macrodontia, rotated lower incisors, malaligned teeth, microdontia, crowded teeth, conical crowns, cleft lip and cleft palate[31].

CRI DU CHAT SYNDROME

Cri du chat syndrome was described by Lejeune Et al in the year 196332. This syndrome was seen more in females as compared to males. A newborn baby having this syndrome shows a low birth weight. Patient having this syndrome shows slow growth because of difficulty in feeding.

Other complications associated with this syndrome are cyanotic crisis, asphyxia episodes , inspiratory stridor. In some cases, it has been observed that children affected with this syndrome shows absence of spleen or kidney, short metacarpals, transverse palmar creases[33,34].

Oral manifestation : protruding maxillary and mandibular retrognathism, enamel hypoplasia, root resorption, high palate, supernumerary teeth, macrodontia, transposed teeth, tooth agenesis, enamel opacities[34,35].

KABUKI SYNDROME

Kabuki syndrome is a known multiple congenital anomaly. The name of this syndrome is derived from the facial features of kabuki actors in Japanese theatre36,37.The kabuki syndromes shows various

features like high arched eyebrows, depressed nasal tip, shortness of fifth finger as well as toes. Mental retardation (mild to moderate), postnatal growth deficiency, dislocation of hip joints, deformed vertebrae or ribs[38,39,40].

Oral manifestation: High palate, screw driver shaped incisors, hypodontia, widely spaced teeth, conical incisors, ectopic upper permanent first molars, micrognathia[40,41].

CONCLUSION

In the present time, we have advanced as well as latest technologies in the field of medical sciences. It is important for every health professions to have adequate and in-depth knowledge of syndromes. If we have adequate knowledge, we can easily identify, diagnose, and helps in prevention as well as treatment of syndromes as well their manifestations in oral cavity.

CONFLICTS OF INTEREST

The authors declare they have no potential conflict of interests regarding this article.

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