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REVIEW ARTICLE

Anodontia: Types, possible etiologic, and genetic background in both syndromic and non-syndromic aspect

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Abstract

Background: Anodontia is a congenital absence of nearly all permanent teeth with retention of many deciduous teeth. Developmentally missing teeth may also be the result of numerous independent pathological mechanisms that affect the orderly formation of the dental lamina, failure of the tooth germ to develop at the optimal time, the lack of necessary space imposed by a malformed jaw, and a genetically determined disproportion between the tooth mass and the jaw size. **Aim:** This review article tells about syndromes that the anodontia associated with and its etiology and its genetic background, anodontia can be isolated anodontia or due to other anomalies as described in the flowchart given. **Conclusion:** Although the etiology of a single missing tooth is unknown, a familial tendency for this defect is present in many. Graber reported the accumulating evidence that it is actually the result of one or more point mutation in a closely linked polygenic system, most often transmitted in an autosomal dominant pattern with incomplete penetrance and variable expressivity. Before treatment of anodontia, we should know the possible background and etiology. **Clinical Significance:** Wide knowledge of etiology and genetic background before commencement of treatment will help in diagnosis and treatment planning.

Keywords: Anodontia, Etiology, Flowchart of anodontia, Genetic background, Related syndromes

Anodontia

Anodontia is a rare genetic disorder which is known to have the congenital absence of all primary or permanent teeth. It is frequently associated with generalized disturbances like hereditary ectodermal dysplasia (ED).[1]

Hypodontia

Hypodontia is a frequently noticed dental anomaly and it is defined as the developmental absence of one or more teeth other than the third molars. It rarely occurs in primary and the most commonly affected are the adult second premolars and the upper lateral incisors.

Oligodontia

Oligodontia is a rare genetic disorder which represents the congenital absence of more than 6 teeth in primary, permanent, or both dentitions.

Subdivision

Complete absence of teeth or only some absence of teeth are the subdivisions of anodontia.[1] Partial anodontia is further divided, depending on how many teeth you have:

- Hypodontia – when one to six permanent teeth are missing
- Oligodontia – when more than six (but not all) permanent teeth are missing.

Etiology

Anodontia is an genetic defect. The defect is due to dental lamina obstruction at period of embryogenesis due to local, systemic, and genetic factors.[2] The exact genes involved are unknown. Genes involved in this are EDA, EDAR, and EDARADD genes. WNTIOA is a major gene.[2]

The pathological mechanisms such as lack of development of tooth germ at the given time period, the lack of adequate space due to deformed jaw, and a genetically predetermined disproportion between the tooth mass and the jaw size may also be the result of developmentally missing teeth.[3] The familial tendency is noticed in this condition.[4] Graber reported congenital missing of teeth due to one or more point mutations in a closely linked polygenic system.[4] Variation are seen in familial occurrence which says that apart from genetic, epigenetic, and environmental factors are also involved.[5] These above finding concludes the multifactorial etiology for this condition.

Association with Syndromes

Mostly anodontia is associated with ED. Anodontia is often associated with a syndrome and never occurs in isolation, this is likely due to an unknown genetic mutation.

Denominations

Denominations are there for this anomaly, namely: Partial anodontia, hypodontia, oligodontia, the congenital absence, anodontia, and bilateral aplasia.[1]

Factors Affected Due to Anodontia

Anodontia may contribute to dysfunction during mastication, impairment in speech, esthetic problems, and malocclusion (Shapiro and Farrington, 1983).

Sequence of Teeth Involved

Absence of lateral incisors represents a common type. The most common missing teeth are found to be the third molars >second premolars >maxillary and the mandibular central and lateral incisor and it can be either unilateral or bilateral.[4]

Prevalence

It is studied that hypodontia is more common than supernumerary teeth. Anodontia is noticed less than the hypodontia which seen as 0.1–0.7% in primary teeth and 3–7.5% in permanent teeth.[1] Prevalence is 2–8% of general population (including third molar). Clinically, hypodontia in the permanent dentition without inclusion of third molar is seen in 3–10% of the population.[4]

Racial Difference

Hypodontia is more often found in Asians and Native Americans.[5] Racial differences have been recorded. The mandibular second premolars and the maxillary lateral incisors are the mostly noticed missing teeth on Caucasians group and the mandibular incisor is the mostly noticed as missing tooth in Asian groups.[4]

Sex difference

The prevalence rates are higher in females compared to the males (3:2, respectively).

Time of Diagnosis

Anodontia is usually diagnosed when baby does not start developing teeth by the time they are about 13 months old.[6] Or it may be diagnosed if a child does not start developing permanent teeth by age 10.[2]

The Differential Diagnosis

For hypodontia most often includes ED which results in the absence of teeth.

Treatment Option

Hypodontia can easily treate by orthodontics if it is mild and when the changes related to it also mild.

If it is severe, prosthetic rehabilitation has to be undertaken.

Only on requirement treatment should be undertaken. In patients with severe anomalies prosthetic rehabilitations can be done. The treatment has effect on psychology of patient which boosts self-confidence apart from improving speech and masticatory function.[7]

The followings are the possible anomalies associated [Flow Chart 1].

Hypophosphatasia

Hypophosphatasia is a rare genetic disorder caused by mutation of ALPL gene which causes defect in mineralization of bone and teeth due to tissue non-specific alkaline phosphatase deficiency which is characterized by the abnormal development of bones and teeth and lead to premature tooth loss. Minerals such as calcium and phosphorus are taken up by teeth and bones.[8]

Dental Effects

The earliest manifestation of the disease may be the loosening and premature loss of deciduous teeth, mostly incisor.

Dental radiographs shows hypocalcification of the teeth with presence of large pulp chambers as well as alveolar bone loss.

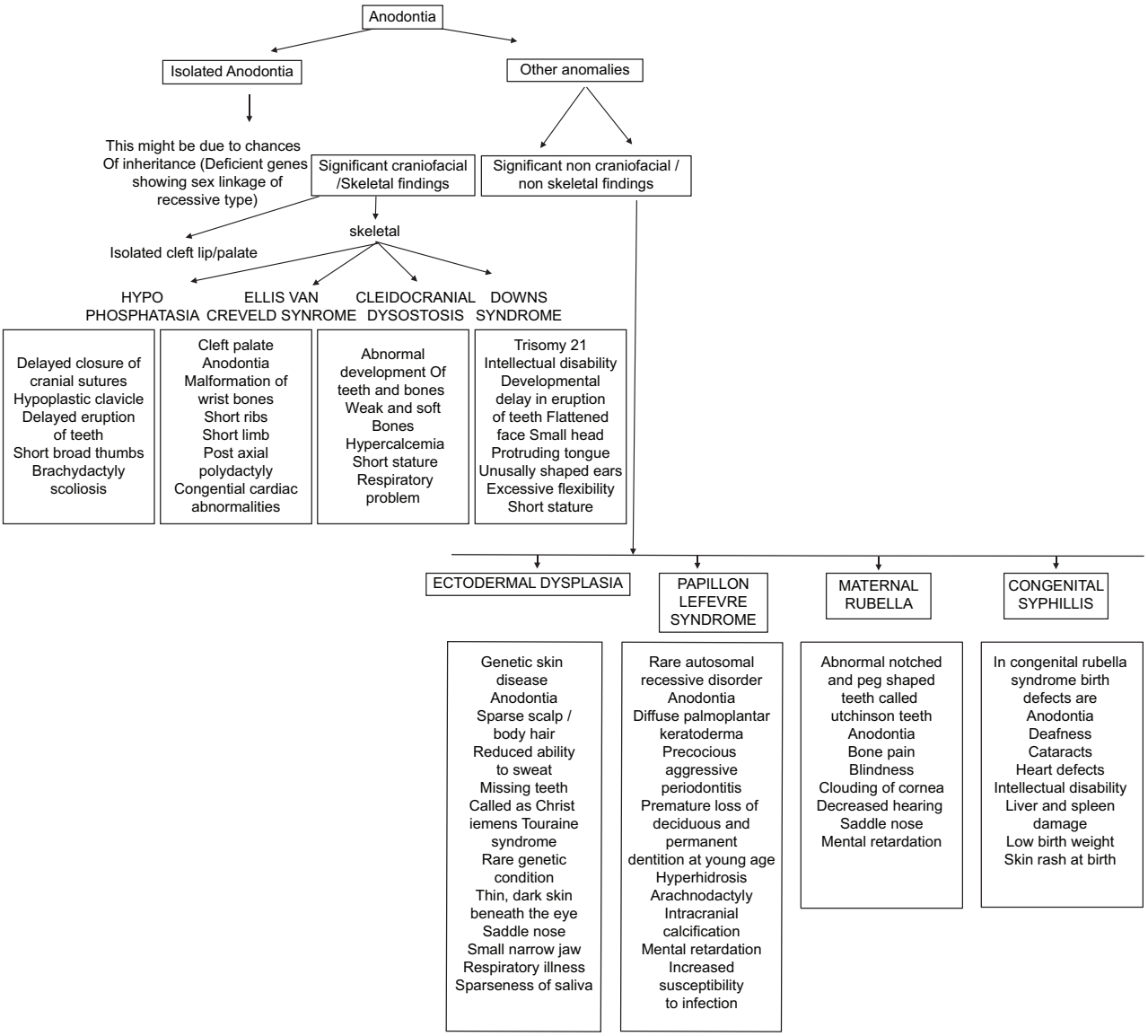
Histological examination shows decrease in cementum which can also be the reason for early exfoliation of teeth.

ED

The EDs are ectodermal disorder affecting skin, hair, teeth, nails, and sweat glands which are an hereditary disease involves two or more tissue of ectoderm. Oral manifestations of ED occur in teeth. The tooth is affected in the developmental stage and leads to hypodontia and anodontia and those present will be in conical in shape.

Dental Aspects

ED apart from its oral manifestations this rarely needs management. LA is safe, a special care is needed in hypo hidrotic forms. Teeth sometimes in extreme cases the both dentition get affected which fail to form leading to hypoplasia of jaws too. The enamel is thin. If the teeth are lost a prematurely senile (nutcracker) profile with protuberant lips is seen. Cleft lip and palate is seen. Xerostomia occurs as salivary gland fail to form. Preventive dental care for avoiding dental caries is particularly important.[9]



Flow Chart 1: Possible etiological and genetic causes of anodontia

Ellis-Van Creveld (EVC) Syndrome

General aspects

EVC syndrome is an inherited autosomal recessive skeletal rare genetic disorder characterized by short limb, short ribs dwarfism, additional fingers and/or toes (Postaxial polydactyly), dysplastic fingernails and, in over half of the cases, congenital heart defects. Motor development and intelligence are normal.

Etiology

EVC syndrome due to mutation of number four chromosomes called EVC and EVC2 which results in the production of abnormally small EVC and EVC2 proteins. Abnormal development of hair, nails, and teeth which is an ectodermal abnormality is seen.

Cleidocranial dysostosis

- Cleidocranial dysplasia (CCD) is a birth defect that mainly affects the development of the bones and teeth. CCD has an autosomal dominant inheritance and mutation occurs in RUNX2 gene. It can be inherited from an affected parent or occur due to a new mutation in the RUNX2 gene
- Dental features: The mandible is prognathic due to hypoplastic maxilla (micrognathia) and other facial bones
- This condition includes supernumerary teeth along with permanent dentition. These supernumerary teeth will lead to crowding in an already underdeveloped jaw unless until it is removed. Hence, the supernumeraries will probably need to be removed to make space for the permanent teeth. Teeth may also be displaced. Cementum formation may be deficient

- Prolonged retention of deciduous teeth and subsequent delay in the eruption of permanent teeth. Failure of eruption of permanent teeth (Anodontia).

Downs Syndrome

Down syndrome is a genetic disorder caused by abnormal cell division results in an extra full or partial copy of chromosome 21 called as trisomy 21. Which leads the developmental changes, mental delays, and physical changes which are a feature of down syndrome.

Dental features

DS children have characteristic orofacial features that are most common such as mouth breathing, open bite, macroglossia, fissured lips and tongue, Protruding tongue, angular cheilitis, delayed eruption of teeth, missing and malformed teeth, oligodontia, supernumerary teeth, microdontia, crowding, malocclusion, bruxism, and low level of caries.[10]

Papillon–lefevre Syndrome (PLS)

PLS is an extremely rare genetic disorder with an inherited autosomal recessive pattern, which mainly affects and symptom is noticed at approximately 1–5 years of age. It is due to mutation of the CTSC gene which induces production of an enzyme known as cathepsin C which is a major coordinator for inducing many serine proteinases in immune/inflammatory cells.

Dental aspects

PLS is characterized by the development of dry scaly patches on the skin which is a diffuse keratoderma of the palms and the soles along with precocious aggressive periodontitis. The primary (deciduous) teeth often undergo premature loss at an early five age. Without treatment, most of the secondary (permanent) teeth may also be lost at an early age of adolescent. The mouth may become inflamed (stomatitis), lymph adenopathy is seen, due to defect in immune cell system the pockets of gums causing susceptibility to infections. Thus, leads to a notable bad breath, Chewing gets painful due to swelling of gums and infection. Degeneration of supporting structures such as atrophy of the alveolar ridge is seen. Frequent pus-producing (pyogenic) skin infections, nail dystrophy, and hyperhidrosis seen.

Rubella Syndrome

Congenital rubella syndrome (CRS) has a low occurrence rate and it is reduced to 1/100 000 live-births. The togavirus family of microorganisms is responsible for this disease usually in the first 3 months of pregnancy. Fetal development is affected as soon as the microorganism make entry into fetus.[11]

General and dental aspects

Patients with CRS are considered as special patients with extreme dental problems usually due to lacking skills in performing tasks and due to care taker who consider dental problem secondary to the systemic problem occurred due to the disease which leads to failure in timely oral disease prevention. It assumed as prolonged effect of the virus on ameloblasts which affects the teeth during development leading to hypoplastic enamel in primary teeth and erupting permanent teeth.[12]

Congenital Syphilis

Syphilis is known as sexually transmitted disease which targets only humans as it acquired by sexual contact, vertical transmission from mother to fetus and by blood transfusion from infected person which is caused by microorganism called treponema palladium.

Dental aspects

Dental manifestations of congenital syphilis are more common. Congenital syphilis can affect several organs and disrupts the system and tissues such as bones and skin. Hutchinson's incisors and Fournier's molars, also known as mulberry molars, are the two types of most common dental defects associated with congenital syphilis.[13]

A flowchart is drawn which gives an idea of genetic background of syndromic anodontia [Flow Chart 1]

Non-syndromic Anodontia/Hypodontia/Oligodontia

Genes involved in non-syndromic oligodontia are known to be MSX1 and PAX9 genes. During physical examination, there was no abnormality in either hairs or nails, perspiration was normal and no congenital clefts of lip or palate was seen. Oligodontia is seen more common in non-syndromic or familial form than syndromic form.[14,15]

Hypodontia/oligodontia has been identified in association with other dental abnormalities such as inclusion, microdontia, and enamel hypo mineralization. The most frequent was the association of hypodontia with enamel hypo mineralization.

Conclusion

Anodontia can be an isolated or associated with syndrome, whatever a thorough knowledge of cause before the start of treatment is essential. Hence, by the above article, we could understand the possible associated syndromes with anodontia in general.

References

1. Vahid-Dastjerdi E, Borzabadi-Farahani A, Mahdian M, Amini N. Non-syndromic hypodontia in an Iranian orthodontic

population. J Oral Sci 2010;52:455-61.
2. Larmour CJ, Mossey PA, Thind BS, Forgie AH, Stirrups DA. Hypodonhia--A retrospective review of prevalence and etiology. Part I. Quintessence Int 2005;36:263-70.
3. Nohl F, Cole B, Hobson R, Jepson N, Meechan J, Wright M. The management of hypodontia: Present and future. Dent Update 2008;35:79-90.
4. Byahatti SM. The concomitant occurrence of hypodontia and microdontia in a single case. J Clin Diagn Res 2010;4:3632-8.
5. Parkin N, Elcock C, Smith RN, Griffin RC, Brook AH. The aetiology of hypodontia: The prevalence, severity and location of hypodontia within families. Arch Oral Biol 2009;54:S52-6.
6. Rakhshan V. Congenitally missing teeth (hypodontia): A review of the literature concerning the etiology, prevalence, risk factors, patterns and treatment. Dent Res J 2015;12:1.
7. Dali M, Singh R, Naulakha D. Idiopathic nonsyndromic tooth agenesis: A report of rare three. J Interdiscip Dent 2012;2:190.
8. Whyte MP. Hypophosphatasia aetiology, nosology, pathogenesis, diagnosis and treatment. Nat Rev Endocrinol 2016;12:233.
9. Cameron AC, Widmer RP. Handbook of Pediatric Dentistry E-Book. Amsterdam, Netherlands: Elsevier Health Sciences; 2013.
10. Asokan S, Muthu MS, Sivakumar N. Oral findings of down syndrome children in Chennai city, India. Indian J Dent Res 2008;19:230.
11. Ahuja R, Shigli AL, Thakur G, Jain U. Dental manifestations of congenital rubella syndrome. Case Rep 2015;2015:bcr201520938220.
12. Chakravarti A, Jain M. Rubella prevalence and its transmission in children. Indian J Pathol Microbiol 2006;49:54-6.
13. Stamm LV, Mudrak B. Old foes, new challenges: Syphilis, cholera and TB. Future Microbiol 2013;8:177-89.
14. Tangade P, Batra M. Non syndromic oligodontia: Case report. Ethiopian J Health Sci 2012;22:219-21.
15. Tavajohi-Kermani H, Kapur R, Sciote JJ. Tooth agenesis and craniofacial morphology in an orthodontic population. Am J Orthod Dentofacial Orthop 2002;122:39-47.