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Enamel hypoplasia in children with down syndrome

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Abstract

Background: Dental anomalies introduce significant functional problem in both jaws and can be grouped based on the stage of dental development at which each anomaly is predicted to originate. Enamel hypoplasia, one of the dental anomalies, is a condition where a disturbance of enamel matrix secretion, defective calcification or defective maturation causes tooth crown surface defect. Various studies show the prevalence of enamel hypoplasia was around 30-40% and a rise in children with Down syndrome.

Objectives: This study aims to inform the readers about the correlation between enamel hypoplasia in children with Down syndrome.

Discussion: Enamel hypoplasia occurrence is related to several factors, such as hereditary and environmental. Studies showed, there is a link between mental retardation and enamel defects as a result of the correlation between the development of enamel and the development of the brain, which inhibits tissue growth and some degradation of the odontoblast which is required to induce pre-ameloblasts to become ameloblasts. It leads to the decrease of secrete enamel matrix and causes the disease. Enamel hypoplasia could also impact the quality of life in Down syndrome children which can aggravate their condition.

Conclusions: Studies have suggested associations with why Down syndrome could lead to enamel hypoplasia, even though there should be more comprehensive studies to establish a more definitive correlation between enamel hypoplasia and specific developmental characteristics in children with Down syndrome.

Keywords: Down syndrome; Enamel hypoplasia; Children; Dental anomalies; Developmental defect

1. Introduction

Dental anomalies introduce significant functional problems in both jaws. Therefore, a thorough survey of the factors involved in their development is important. Specific genetic factors have been reported responsible for developing dental anomalies in each jaw. In addition, disturbances created during tooth development may produce variations in the number of teeth (agenesis/supernumerary teeth), their size and shape, and tooth bud position, which can affect both permanent and deciduous dentition of both jaws. Dental anomalies can be grouped into those associated with teeth' number, size, shape, and structure. This system also groups anomalies based on the stage of dental development at which each anomaly is predicted to originate (Anggraini, *et al.,* 2019; Tunis, *et al.,* 2021). Dental anomalies occur with an incidence five times greater in DS individuals than in the general population. One of the dental anomalies is enamel hypoplasia. Enamel hypoplasia is a tooth crown surface defect caused by a disturbance of enamel matrix secretion, defective calcification or defective maturation (Areias, *et al.,* 2015). Research held by Miyamoto, *et al.*, (2023) shows the prevalence of EH in primary teeth among 1-year-old Japanese children was 38.5%, while a study by Makieh, *et al.,* (2022)

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in Syria shows the percentage of EH there is 34% in healthy children and 45% in children with Down syndrome. The enamel hypoplasia in people with Down syndrome results from a reduced amount of enamel matrix and reduced opacity of the enamel layer. The enamel hypoplasia in people with Down syndrome can be associated with abnormal blood supply to the embryonic jaw, which inhibits tissue growth and causes some degeneration of the odontoblasts, which are involved in the formation of the dentin; dentin is required to induce pre-ameloblasts to become ameloblasts, which secrete enamel matrix (Anggraini, *et al.,* 2019; Kanchan, *et al.,* 2015; Makieh, *et al.,* 2022; Miyamoto, *et al.,* 2023).

Nonetheless, besides enamel hypoplasia there are also few dental anomalies found in people with Down syndrome as follows anomalies of tooth number, namely, hypodontia and supernumerary conditions; anomalies of tooth size, namely, microdontia from permanent teeth; anomalies of tooth shape, such as short root, taurodontia, talon cusp, fusion, and gemination; as well as anomalies of tooth structure, such as enamel hypoplasia and enamel hypocalcification. Besides these abnormalities, tooth eruption in people with Down syndrome may occur in asymmetric sequences and may be delayed for 2 to 3 years compared to healthy individuals. The other studies found an association between mental retardation and enamel defects as a result of the correlation between the development of enamel and the development of the brain in as much as certain systemic disruptions interfering with neurological development may also alter the development of tooth germs. Damaged enamel cannot recover from the injury. Therefore, it may give information on the timing and nature of insults potentially affecting other ectodermally derived structures, such as the brain (Anggraini, *et al.,* 2019; Modrić, *et al*., 2016).

This study was conducted to identify correlations between enamel hypoplasia and developmental characteristics in children with Down syndrome.

AIMS

This study aims to inform the readers about the correlation between enamel hypoplasia in children with Down syndrome and its effect on oral health. Article searches were conducted using databases of *Pubmed, Scopus, ScienceDirect* and *Google Scholar* with the following keywords: *Down syndrome, enamel hypoplasia, children, dental anomalies,* and *developmental defect.*

2. Discussion

Down Syndrome (DS) is the most common chromosomal abnormality that was first discovered in humans. There are three distinct types of Down syndrome: (1) Trisomy 21, which constitutes the majority of cases, occurs when there's an extra copy of chromosome 21; (2) Mosaicism, accounting for 2–5% of cases, involves a mixture of cells containing either 46 or 47 chromosomes. The characteristics exhibited by the affected individual depend on the number of cells involved and their chromosomal composition. (3) Translocation type occurs when a segment of chromosome 21 detaches and attaches itself to another chromosome, resulting in a total of 47 chromosomes genetically. The incidence of trisomy 21 correlates strongly with increasing age, it makes young mothers have a low probability of having trisomy 21 children. Still, the risk increases rapidly after the age of 35 years. The condition is observed in 1 in 1,550 live births among women under 20 years old, contrasting with a higher incidence of 1 in 25 live births among women over the age of 45 (Desingu, *et al*., 2019; Koch, *et al.,* 2017; Radhi & El-Samarrai, 2015). DS can be found worldwide, with an incidence of 1 per 374 live births. The estimated incidence was about 8 million people. In 2002, the prevalence of children and adolescents (0-19 years) was 10.3% of 10,000 live births. According to the World Health Organization (WHO), the estimated incidence was 1 in 1,000 up to 1 in 1,100 births. About 3000 to 5000 babies were born with DS every year. Data from National Basic Health Research (Riskesdas) 2018 showed the percentage of children in Indonesia who were born with an abnormality is 0.41%. (Annastia, *et al*., 2019; Kemenkes, 2019 ; Makieh, *et al*., 2022). DS itself might occur in all races, and the growth of a child with DS might show abnormality, slowness and barriers. Among the causes of DS, advanced maternal age has been identified as the main one due to the formation of gametes during the intrauterine phase and their interruption in meiosis I, causing these oocytes to age along with the woman, as they will only mature from puberty. This ageing would destroy chromosomal fibres and deteriorate the centromere, causing an inability to separate chromosomes during anaphase I of meiosis. Another cause may be the lack of segregation in gametogenesis due to advanced paternal age (greater than 55 years). Inadequate consumption of alcohol, cigarettes, chemical substances, oral contraceptives, genetic inheritance, history of abortions and environmental agents, for example, radiation, are other causes that can cause genetic errors (Cristhiane Olívia Ferreira do Amaral, *et al*., 2023).

Patients with DS have a wide array of signs and symptoms like intellectual and developmental disabilities or neurological features, congenital heart defects, gastrointestinal (GI) abnormalities, characteristic facial features, altered immune system and other abnormalities. Furthermore, DS children have several orofacial features that can be used to diagnose it, which are a highly vaulted and narrow palate, large and thick lips, and cracks and fissures tongue that

develop gradually. Patients with DS also face many oral health problems as followed, such as smaller-sized mouth, macroglossia, and teeth anomalies such as microdontia, conical shape, hypodontia, taurodontia, and enamel defects such as enamel hypoplasia or enamel hypocalcification. Moreover, these individuals tend to have high rates of periodontal disease, dental caries, missing teeth, prolonged retention of primary teeth and malocclusion because their limitation aggravates their oral health condition (Akhtar & Bokhari, 2023 ; Elrefadi, *et al*., 2018).

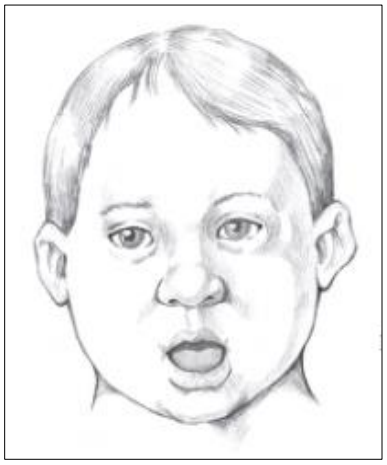

Figure 1 Clinical signs of a child with Down syndrome

There was a study that stated that severe illness or prolonged fevers could lead to hypoplasia and hypocalcification. Yet, there is no follow-up study in this relation. Other studies found an association between mental retardation and enamel defects as a result of the correlation between the development of enamel and the development of the brain. That is mainly because certain systemic disruptions interfering with neurological development may also alter the development of tooth germs. (Annastia, *et al*., 2019; Elrefadi, *et al*., 2018). Concerning the enamel defect among DS, almost 50% of persons with DS exhibit three or more dental anomalies, where 20% of them suffer enamel hypoplasia. Enamel defects occur more often on the surface of the buccal tooth than on other surfaces. The impact of growth and development disorders in children might be reflected in the teeth (Annastia, *et al*., 2019; Radhi & El-Samarrai, 2015).

Research held in Massachusetts showed that a common structural anomaly in people with Down syndrome is enamel hypoplasia, which is 32%. Generally, enamel hypoplasia is the quantitative reduction of the enamel resulting from alternations in the matrix formation stage. It actually has a high prevalence in children from developing countries, children with chronic or acute malnutrition, and children with very low birth weight. It occurs if matrix formation is affected and may manifest as pitting, grooving or even total absence of enamel. Hypomineralisation results when maturation is disturbed and manifests in opaque or chalky areas on normally contoured enamel surfaces. Thus, disturbance either in matrix formation or calcification can occur, depending chiefly on the tooth formation stage at the time of injury. Enamel hypoplasia appears as a surface defect resulting from reduced enamel thickness. Hypoplasia can occur in the form of pits – single or multiple, shallow or deep, or scattered or arranged in horizontal rows; or grooves – single or multiple, narrow or wide, or evident as a partial or complete absence of enamel over a considerable area of the tooth crown. The enamel of reduced thickness may be translucent or opaque (Anggraini, *et al.,* 2019; Koch, *et al.,* 2017).

Enamel hypoplasia or hypo mineralization may be caused by hereditary factors and environmental factors that include systemic factors such as nutritional factors, exanthematous diseases like measles and chicken pox, congenital syphilis, hypocalcemia, birth injury or premature birth, fluoride ingestion or idiopathic causes, and local factors such as infection or trauma from a deciduous tooth. The enamel hypoplasia in people with DS can be associated with abnormal blood supply to the embryonic jaw, which inhibits tissue growth and causes some degeneration of the odontoblasts, which are involved in the formation of the dentin; dentin is required to induce pre-ameloblasts to become ameloblasts, which secrete enamel matrix. A study showed the linkage between the frequency of enamel hypoplasia and chronological teeth calcification. The upper incisor is the first tooth to be calcificated at the third to fourth months prenatal. The amelogenesis of the primary incisor is almost complete at birth, but not the canines and molar teeth. Theoretically, if the disorder occurred in the 12th week of prenatal, the hypoplasia lesion will appear on the central incisor of the upper and lower jaws. Accordingly, if the disorder occurs at the 16th week, enamel hypoplasia will appear on the posterior teeth because that is when the enamel matrix forms on the anterior and posterior primary teeth (Annastia, *et al*., 2019).

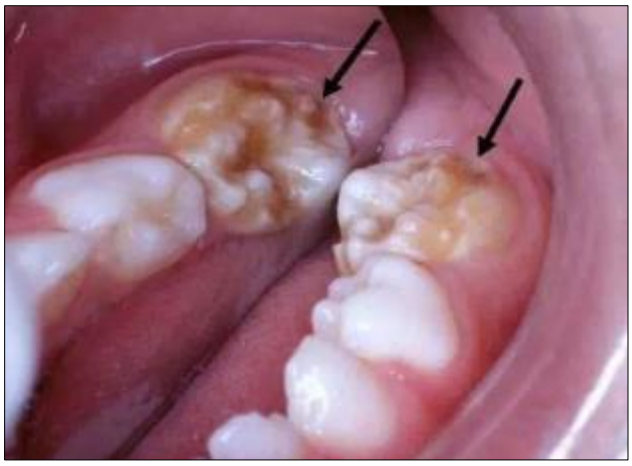

Figure 2 Enamel hypoplasia

Enamel hypoplasia could negatively impact the quality of life. This is because it could cause discolourations, involving discolouration of the teeth either extrinsically (superficially, either on the surface of the tooth or on the acquired pellicle) or intrinsically (possibly arising during the development of different tooth appearance and degree of severity), and can limit speaking, particularly in children with enamel hypoplasia and caries. Further, in as much as of these surface irregularities, hypoplastic teeth can have the following dental problems: more sensitive to heat or cold or pain, more prone to wearing down from grinding or "tooth to tooth contact", more susceptible to an "acid attack" from the sugars in our foods and drinks, more susceptible to trapping plaque and bacteria, and more prone to tooth decay. It makes enamel hypoplasia one of the predisposing factors for early childhood caries (ECC) and erosion. Thus, primary dentition with incomplete enamel calcification on pits and fissures provides a suitable site for the adhesion and colonization of cariogenic bacteria. Consequently, ECC will develop more rapidly on altered tooth surfaces (Annastia, *et al*., 2019; Folayan, *et al.,* 2018; Nota, *et al.,* 2020).

3. Conclusion

Children with Down syndrome exhibit a greater prevalence of oral and dental anomalies than their peers without the condition. Studies have suggested associations between enamel hypoplasia and Down syndrome, which is because certain systemic disruptions interfering with neurological development may alter the development of tooth germs. There should be a proactive approach and early diagnosis to ensure that dental care is well-prepared and tailored to their specific needs. However, more comprehensive studies are needed to establish a more definitive correlation between enamel hypoplasia and specific developmental characteristics in children with Down syndrome.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

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