THE KEYS TO DIFFERENTIATE LOCAL, SYSTEMIC AND HEREDITARY ENAMEL HYPOPLASIAS

Alberto Consolaro[1,2]

ABSTRACT:

An accurate diagnosis of enamel hypoplasia, as well as its cause, prognosis and treatment, is difficult to establish because of the several misconceptions about its origin, nature and treatment at the time of diagnosis. There are three key lines of thought to be followed to build the basis of an accurate differential diagnosis between types of enamel hypoplasia and to define an accurate prognosis in their treatment plans: 1) if it is localized enamel hypoplasia, it is not bilateral, and if it happens to be bilateral, it is not symmetrical; 2) if it is enamel hypoplasia caused by systemic causes, it is bilateral and symmetrical, but does not affect all teeth; and 3) if it is hereditary enamel hypoplasia, called hereditary amelogenesis imperfecta, it is bilateral, symmetrical and affects all teeth without any difference.

KEYWORDS:

Enamel hypoplasia. Turner's tooth. Dental trauma. Dental anomalies.

1. Universidade de São Paulo, Faculdade de Odontologia de Bauru (Bauru/SP, Brazil).

1. Universidade de São Paulo, Faculdade de Odontologia de Ribeirão Preto, Programa de Pós-graduação (Ribeirão Preto/SP, Brazil).

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INTRODUCTION

Enamel formation may be affected by errors in ameloblast functioning, as these cells are highly sensitive to metabolism. When the development of an organ or tissue is incomplete or restricted, the term hypoplasia is used, even when that organ or tissue still performs its function partially or totally.

Enamel hypoplasia may present as a single demarcated area of white discoloration with a smooth surface on a single tooth, or as yellow or brown discoloration areas with or without a smooth surface (Figs. 1, 2 and 3). In several cases, in addition to these whitish and brownish discolorations, the tooth has surface defects, such as pits, bands, grooves, fissures, roughness and erosion with loss of enamel structure (Fig. 2).

As its cause may be localized, it may affect a single tooth, but it may also be systemic and even hereditary, in which case it may affect several teeth, or even all the patient's teeth, bilaterally in the form of superficial enamel discoloration and defects.

The diagnosis of enamel hypoplasia may reflect several imprecisions about origin, nature and treatment. To establish an accurate differential diagnosis between types of enamel hypoplasia, this study describes three lines of thought. They are here called key factors, and should be followed by dentists to define an accurate prognosis in their treatment plans.

WHAT ARE THE CAUSES?

Enamel formation errors may be:

1. Inherited or transmitted by genetic errors; or

2. Acquired by the action of causes that are external to dental development.

Acquired causes may be: a) systemic; or b) localized.

Systemic acquired causes: The most prominent are febrile exanthematous diseases and other clinical conditions that keep children hospitalized or sick for long periods (Figs. 4 and 5).

Systemic causes are those that affect the whole body and the teeth. Dental fluorosis is the result of excess fluoride ingestion and accumulation in the body. Fluoride toxicity affects ameloblasts metabolically and results in the formation of defective or hypoplastic enamel. Therefore, it may be concluded that dental fluorosis is systemic.

Antibiotics, particularly tetracycline, change enamel color, but do not affect its resistance and structure. Antibiotics are associated with enamel hypoplasia when prescribed to treat serious systemic clinical conditions that cause enamel hypoplasia. Antibiotics are not toxic to ameloblasts and do not affect enamel formation, but change the color of teeth without affecting its structure or resistance.

Localized acquired causes: The most frequent are trauma and periapical and furcation lesions of deciduous teeth (Figs. 1, 2 and 3). In both cases, the cause affects permanent teeth directly while enamel is still being formed.

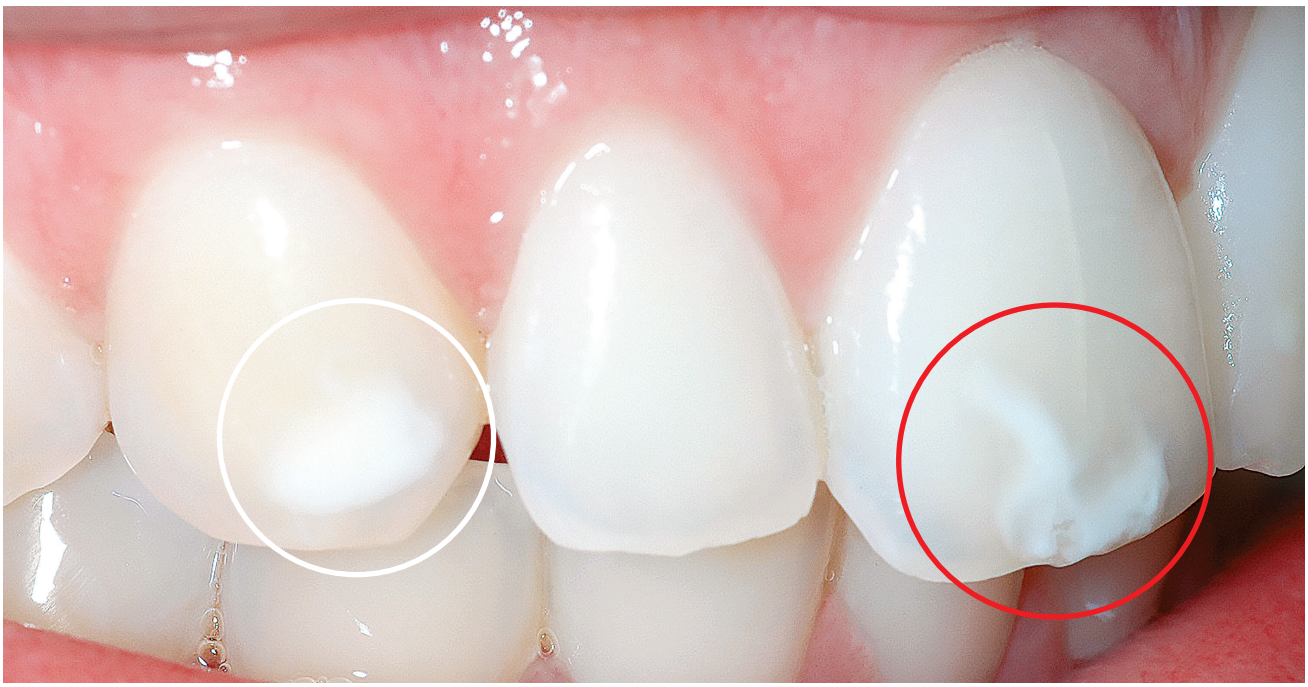

Figure 1:
Localized enamel hypoplasia in form of white discoloration with preserved surface on canine, and abnormal surface on central incisor, associated with tooth trauma.

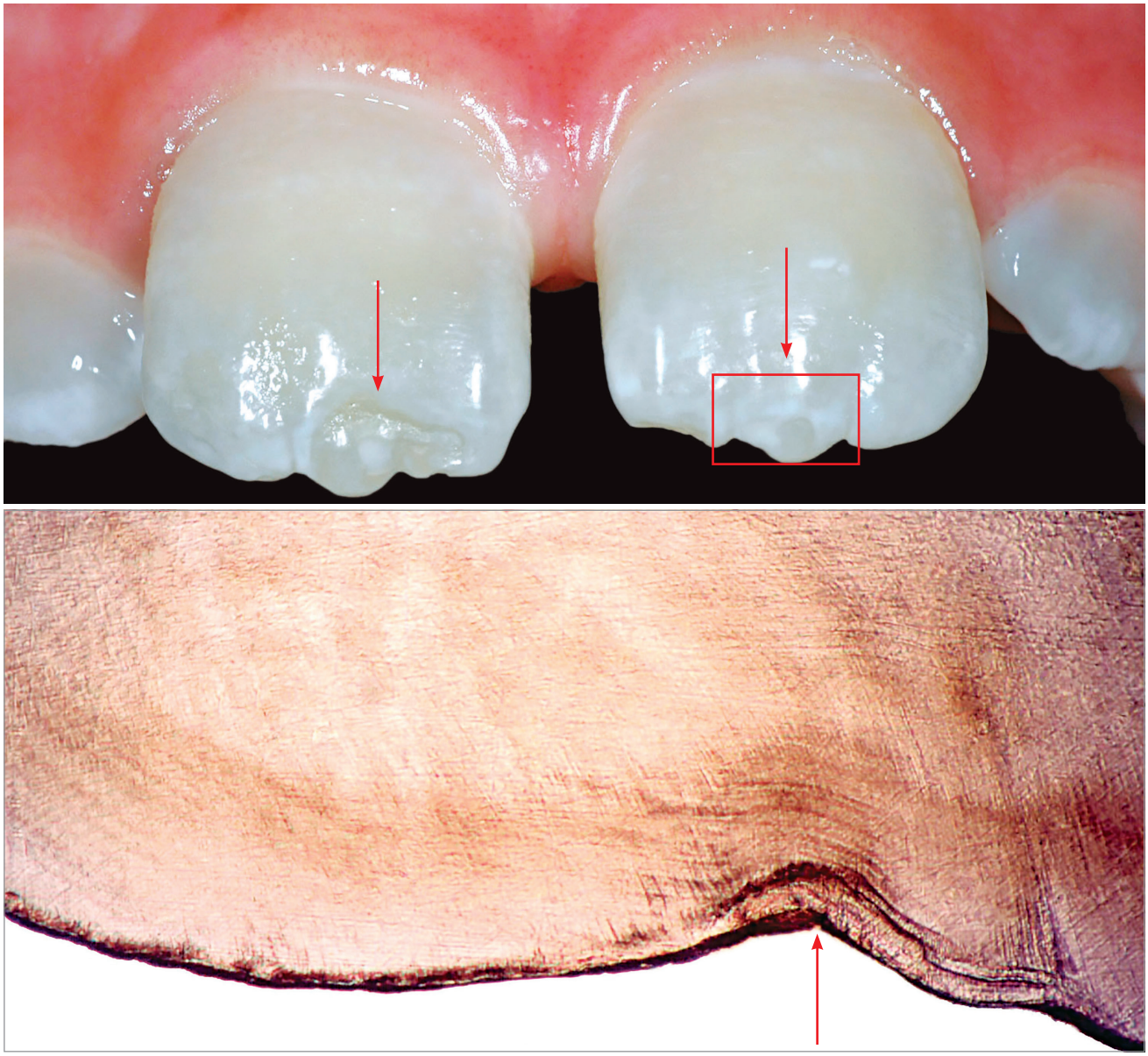

Figure 2:
Enamel hypoplasia in form of discolorations, pits and grooves on permanent maxillary central incisors. Microscopic view of prisms and incremental lines, known as striae of Retzius, are disorganized, especially in areas with enamel loss (arrows).

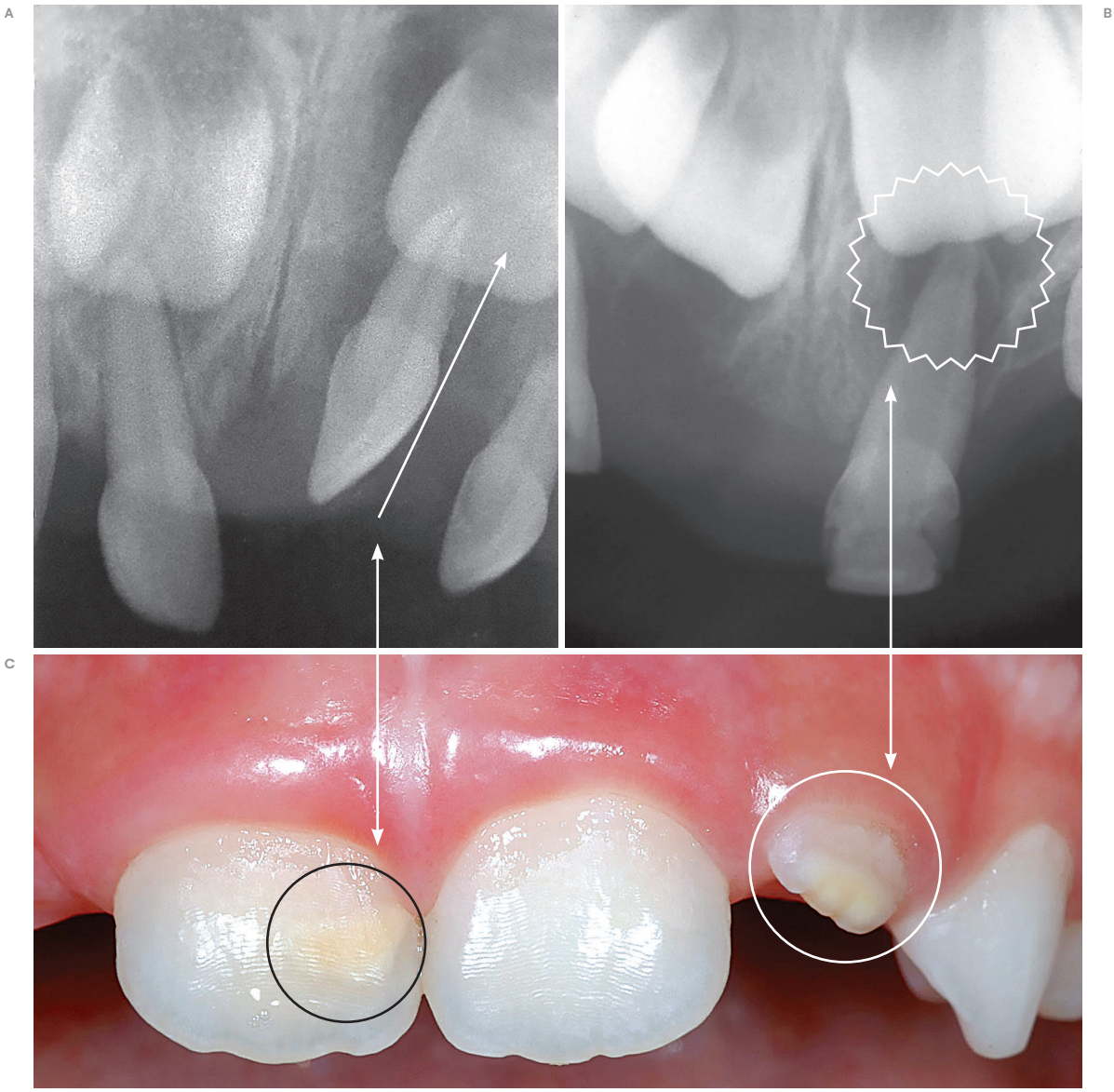

Figure 3:
Two main causes of localized enamel hypoplasia. (**A**) tooth trauma and intrusion of deciduous tooth, slipping over permanent tooth germ; (**B**) chronic inflammatory periapical lesion of deciduous tooth affecting permanent tooth germ by contiguity. Modified amelogenesis produced white or yellowish discolorations.

IN THE OFFICE: IS IT LOCALIZED, SYSTEMIC OR HEREDITARY? HOW TO RECOGNIZE THE DIFFERENCES

Here are the three key factors for the safe diagnosis of enamel hypoplasia and its nature:

1st key factor:

If it is localized, it is not bilateral, and, if it happens to be bilateral, it is not symmetrical.

Local causes do not affect the same site on both sides simultaneously (Figs. 1, 2 and 3). Keep in mind that the main localized causes are trauma to deciduous teeth or inflammatory periapical or furcation lesions of deciduous teeth associated with pulpal necrosis (Fig. 3). In trauma due to deciduous tooth intrusion, the apex touches and disorganized, and even necrotizes, the ameloblasts that were producing enamel at that time.

In inflammatory periapical or furcation lesions of deciduous teeth with pulpal necrosis, bacterial byproducts, such as lipopolysaccharides and microbial enzymes, necrotize ameloblasts and block their synthesis in that region. While these byproducts are not removed from the site by endodontic procedures or deciduous tooth extraction, enamel production remains blocked or defective in that area.

When trauma and contamination are eliminated, ameloblasts in the areas neighboring the permanent tooth germ proliferate and resume the formation of the enamel-producing cell layer and the synthesis of the enamel matrix in that area, which now has an irregular structure and is discolored. This process may generate discrete white, yellowish areas and even pits, grooves, and other defects on the surface of the tooth, which will be seen and diagnosed only after the tooth erupts. Enamel surface may be normal, smooth and bright despite the discolorations induced by hypoplasia.

Enamel hypoplasia of permanent teeth associated with inflammatory periapical and furcation lesions of deciduous teeth was first described by Turner[1] in 1912, and is most often called Turner's tooth.

Surgical handling of dental germs during amelogenesis (Fig. 3) may induce localized enamel hypoplasia in cases of:

a) deciduous tooth extraction followed by vigorous alveolar curettage;
b) removal of cysts, especially inflammatory follicular cysts and dentigerous cysts; and
c) surgical removal of odontogenic tumors, particularly odontomas, common in the first decade of life.

Another local and direct cause is radiotherapy of malignant tumors in the maxillae of children, which may result in lesions to the

ameloblasts of the teeth in the region when still in the phase of amelogenesis, and thus lead to enamel hypoplasia.

2nd key factor:

If it is systemic enamel hypoplasia, it is bilateral and symmetrical, but does not affect all teeth.

Only the teeth whose enamel was under formation at the time when the disease was active will be discolored, with irregular or defective surfaces (Figs. 4 and 5). Some bands or segments of the enamel that were under formation at that time will stand out against the enamel already formed up to that moment, which has normal color and structure (Fig. 5).

It is possible to define the time when it occurred!

Over two thirds of the cases of enamel hypoplasia due to systemic causes occur in the first 10 months of postnatal life[2,3] (Figs. 4 and 5). Because of that, these cases affect:

- the middle and cervical thirds of maxillary and mandibular incisors, except the maxillary lateral incisor;
- the tip of canine cusps; and
- the surface on the occlusal third of maxillary and mandibular first molars.

A pathognomonic sing to clinch the diagnosis!

The first molars of these patients may have a pathognomonic sign of systemic enamel hypoplasia induced postnatally:

- the tip of the mesiolingual cusp is preserved, with the presence of preserved enamel (Fig. 5).

At the time of eruption, it is the only healthy area of enamel on the permanent first molars and is indicative that the systemic change occurred after the child was born. The other one third of the cases of systemic enamel hypoplasia occur between the 11th and 34th month of life. In these cases, enamel hypoplasia is found in:

- maxillary lateral incisors, particularly in the incisal third; and
- premolars, at any point of its crown.

In 2% of the cases of systemic enamel hypoplasia, the cause is active between the 35th and 80th month of life and affects, primarily:

- The second molars.

According to the period of enamel formation, the time of life when the systemic change occurred may be determined, as showed in Figure 4.

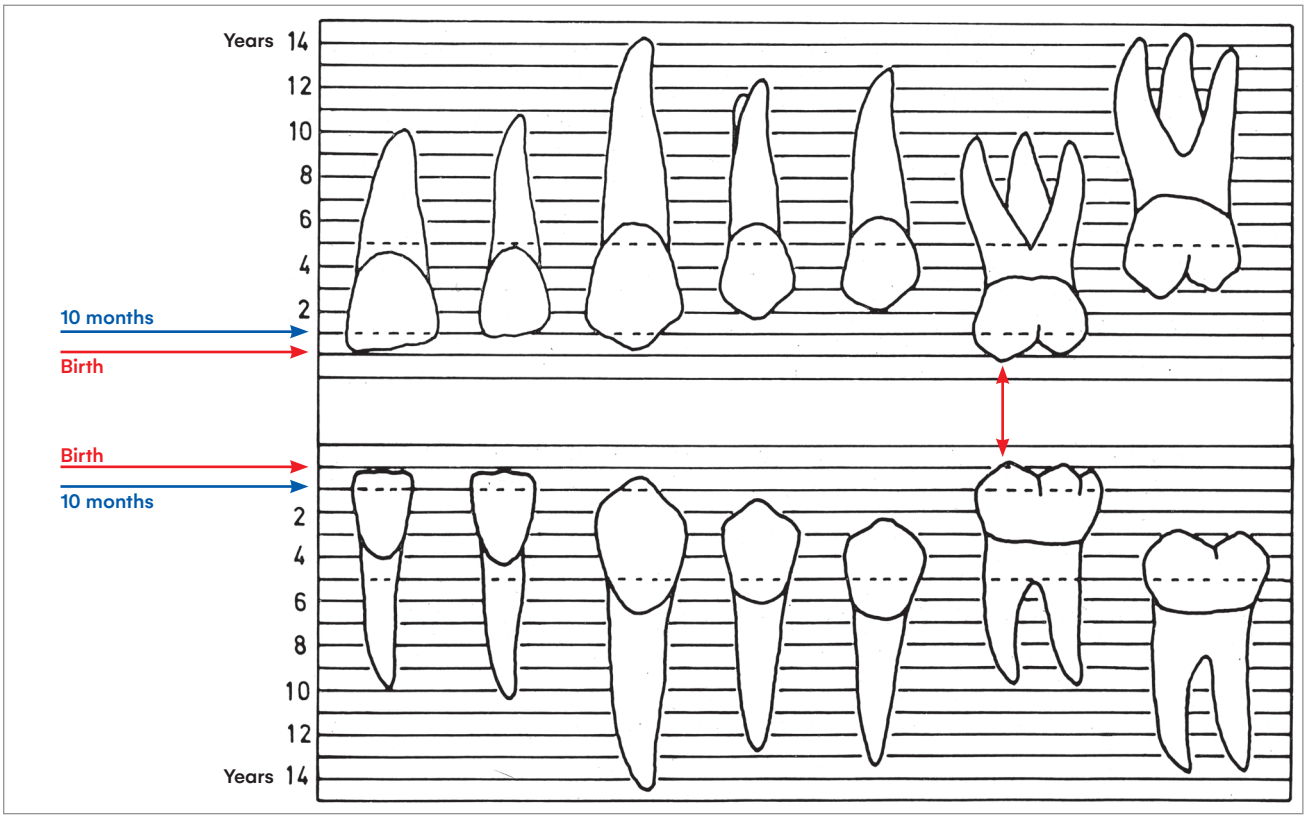

Figure 4:
At birth (red), only tip of mesiolingual cusp has enamel already formed. Enamel formed up to 10th month of postnatal life, time when 2/3 of systemic enamel hypoplasia occur (chronology of permanent tooth development by Massler, Schour and Poncher, 1941).

An important and intriguing fact: despite the severity of the clinical condition in a systemic change, not all children with systemic diseases will develop enamel hypoplasia.

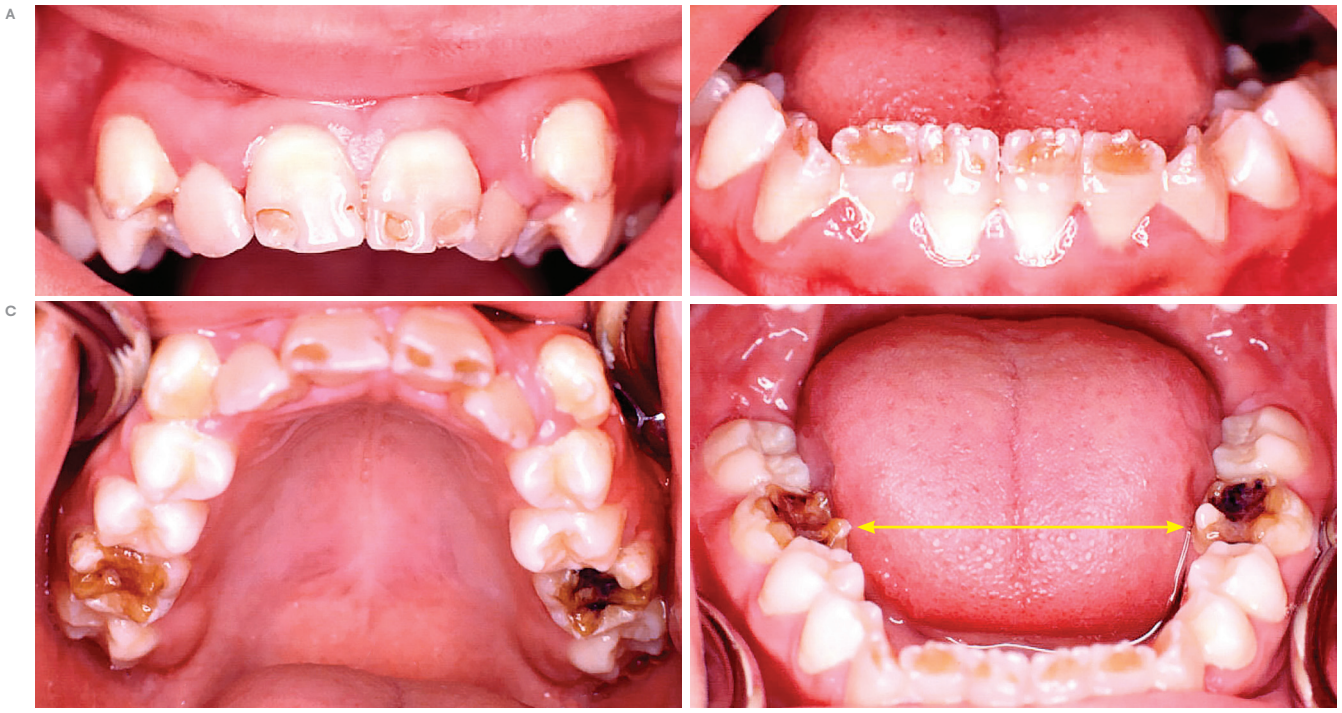

Figure 5:
Irregular symmetrical bilateral lesions on enamel in patient with systemic enamel hypoplasia, which may affect teeth after birth and up to 10th month of life; arrow indicates tip of mesiolingual tip with normal enamel formation; correlation made by comparing sites and teeth affected by hypoplastic lesions with data in Figure 4.

Over 100 systemic causes of enamel hypoplasia have already been described, but few are accurately diagnosed. When faced with a case of enamel hypoplasia and a suspicion of a systemic cause, the possibilities below should be gradually investigated while taking the patient's history:

- Exanthematous diseases, such as rubella, measles, varicella (chicken pox), scarlet fever, dengue and zika;
- Prolonged hospitalizations that indicate severe trauma or infections;
- Hypocalcemia associated with parathyroid dysfunction, particularly rickets;
- Vitamin A and C deficiencies;
- Excess fluoride: ingestion affects ameloblasts metabolically, and fluoride toxicity promotes the formation of defective or hypoplastic enamel. Dental fluorosis has a systemic nature and does not affect only the teeth. Its clinical dental presentation is peculiar and has specific characteristics of enamel hypoplasia – dental fluorosis is a specific enamel hypoplasia; and
- Congenital syphilis: Treponema pallidum in patients born with this disease may be in the tooth germ among the ameloblasts, affecting enamel production and leading to a very specific type of systemic enamel hypoplasia, known as Hutchinson's incisors or as barrel shaped teeth, when it affects maxillary teeth, or as Moon's molars, also known as mulberry molars when it affects molars.

3rd key factor:

If it is hereditary enamel hypoplasia, called hereditary amelogenesis imperfecta, it is bilateral, symmetrical and affects all teeth without any difference.

Genetic changes do not depend on the time of enamel formation in a specific tooth, and enamel will be abnormal in all teeth.

Hereditary amelogenesis imperfecta is a genetic enamel hypoplasia that is inherited in an autosomal dominant, recessive or X-linked pattern. It affects only the enamel, with no repercussions on other dental tissues, and compromises both deciduous and permanent teeth.

Amelogenesis may be divided into three phases:

a) formation, characterized by deposition of organic enamel matrix.
b) mineralization or calcification, characterized by the integration of mineral salts to the preformed organic matrix; and
c) maturation, characterized by the reduction of liquids and of the protein component of enamel to refine its crystalline structure.

Hereditary amelogenesis imperfecta occurs due to genic defects that may affect only one of these

three phases, which characterize it as a disease with three different clinical types of manifestation: hypoplastic, hypocalcified and hypomature. This disease affects only enamel, and the other mineralized tissues are normal.

A definitive diagnosis should be established by associating family history, clinical examination and imaging studies, including of tooth germs. An early diagnosis may lead to more favorable esthetic and functional corrections when associated with family and genetic guidance and counseling of these patients.

In some cases, without a known family history or no other occurrences in the family, the differential diagnosis should include systemic enamel hypoplasia, as described above, in which hypoplastic enamel has grooves, lines or bands in a symmetric and bilateral pattern, compromising only a segment of enamel. This is explained by the fact that this change occurs only during the time when the systemic cause was acting upon ameloblast metabolism. In hereditary amelogenesis imperfecta, all the enamel of all teeth is compromised, regardless of when they were formed.

FINAL CONSIDERATIONS: THERAPEUTIC IMPLICATIONS

In localized and systemic enamel hypoplasia, enamel in neighboring teeth is normal and should be included in the esthetic and functional treatment planning. Dentin, pulp, cement, gingiva and periodontium are also normal (Figs. 1, 2 and 3).

Ameloblasts resume the production of translucent enamel with a well-organized crystalline structure after some days, weeks or months of the activity of the localized cause, or of the systemic changes that compromised enamel formation, (Figs. 1, 2 and 3). As an effect of this transient change on amelogenesis, teeth have discolorations, pits, grooves, lines or bands without enamel or with defective enamel on its crown, which contrast with the areas of normal enamel when the tooth erupts.

In hereditary enamel hypoplasia, specifically called hereditary amelogenesis imperfecta, neighboring enamel is not normal and usually does not react well to esthetic and functional procedures, even when it appears to be normal. Therefore, these procedures should not include abnormal enamel, and should count only on normal dental tissues, such as dentin, cementum and pulp, to reestablish function and esthetics.

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Contact address: Alberto Consolaro
consolaro@uol.com.br

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