Clinical management of amelogenesis imperfecta in primary dentition

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Abstract

Amelogenesis imperfecta patients suffered common clinical problems of poor esthetics, teeth sensitivity, and loss of occlusal vertical dimension. Amelogenesis imperfecta is a group of inherited disorders primarily affecting dental enamel. Variants of amelogenesis imperfecta generally classified hypoplastic, hypocalcified, or hypomaturation types based on the primary enamel defects.

The mildest problems were found in the pitted hypoplastic type whereas the most severe problems were encountered in the hypocalcified type amelogenesis imperfecta.

Management stragies include composite resin veneer and jacket crowns for anterior teeth as well as steel crowns for posterior teeth. Knowledge of the clinical features and dental complications of each variants if amelogenesis impecta helps in the diagnosis of the condition and allows institution of early preventive measures. The objective of this paper is to provide a review of the current concepts of the wide spectrum of etiological factors involved in the pathogenesis of this significance clinical entity in the primary dentition.

Introduction

Enamel hypoplasia may be defined as a deficiency in enamel formation manifesting clinically as grooves or pits, as well as partial or lack of surface enamel.

Clinically significance of enamel hypoplasia includes poor esthetics, tooth sensitivity, malocclusion, as well as predisposition to dental caries. (fig. 1)

Of further clinically importance is the fact that enamel hypoplasia may provide diagnostic clues related to genetic influences and systemic diseases as well as local insults occurring during the long time span of dental development.

The primary dentition may provide unique information related to in utero development as it commences mineralization in utero.

As the permanent dentition, enamel hypoplasia in the primary dentition may be inherited as genetic disease involving only the teeth, amelogenesis imperfecta, or as oral manifestation of many inherited systemic disease and dysmorphic syndrome.[1]

More commonly, however ,enamel hypoplasia in primary teeth results from acquired environmental factors encounters during the period of enamel formation.

At least three different clinical variations of amelogenesis imperfecta are observed : the hypocalcified type, the hypomaturation type, and the hypocalcified type.[2]

Burzynski, Gougalez, and Snawder have suggest several modes of inheritance for amelogenesis imperfecta.[10,11] Chaudhry and others, have reported that a few children have been observed to demonstrate enamel dysplasia without hereditary background.[3]

The defective tooth structure is limited to the enamel. On radiography examination the pulpal outline appear to be normal, and the root morphology is not unlike that of normal teeth. (fig. 16) The difference in the appearance and quality of the enamel is thought to be attributable to the state of enamel development at the time the defect occurs. In the hypoplastic type the enamel matrix appear to be imperfectly formed. Although calcification subsequently occurs in the matrix and the enamel is hard, it is defective in amount and has roughened, pitted surface. (fig. 5,8) The hypocalcified type, matrix formation appear to be of normal thickness, but calcification is deficient and the enamel is soft. In both of these more common types of defect the enamel becomes stained because of roughness of the surface and the increased permeability. (fig. 6)

In still another variation of amelogenesis imperfecta there is thin, smooth covering of brownish yellow enamel. In this type the enamel does not seem excessively susceptible to abrasion or caries. (fig. 1)

Prevalence

It is a heterogeneous group of disorders, with reported prevalence rates raning from 1 in 718, 1 in 4000, 1 in 8000 and 1 in 14,000.[4]

Clinical presentation

Amelogenesis imperfecta has been conveniently classified into hypoplastic, hypocalcification and hypomature types of the enamel, although distinction between these types may have little ultrastructual basis.

A defect is described as localized when only one tooth is affected and generalized when there is a symmetrical disturbance on teeth of the same type on both left and right sides. Approximately 100 etiological agents have been reported to causes developmental defects of enamel.[5]

Developmental defects of enamel can be classified according to their clinical appearance: discoloration, hypominerlization, hypoplasia.

Phenotypes

Phenotypes range from hypoplastic thin enamel with spacing between adjacent teeth, to varying degrees of hypomimeralization poorly formed enamel with altered colour and translucency, although in many cases both hypoplasia and hypomineralization are seen together.

The color of the enamel reflects the degree of hypomineralization in general.

In X-linked amelogenesis imperfecta, females exhibit vertical bands of altered enamel manifesting. These bands may be thinner and altered in color because of hypomineralization.[6,12] In such families there will be no male to male transmission, whereas the heterozygous females may pass on the trait to children of other sex. In some forms of amelogenesis imperfecta the teeth fail to erupt, presumably due to a disturbance of the enamel organ, and they undergo replacement resorption of their crowns. In other forms, a skeletal anterior open bite is seen.[7]

Hypoplastic forms

- Thin enamel
- Account for the majority of cases
- Lack of contact points between teeth in thin enamel type
- Enamel may be rough,smooth,or randomly pitted (fig. 5,6)
- Female carriers of X-linked forms manifest with vertical banding of normal and abnormal enamel.
- Teeth are delayed in eruption
- Unerupted teeth may undergo replacement resorption
- Anterior open bite associated with 60% of cases (fig. 18,19,20,21)

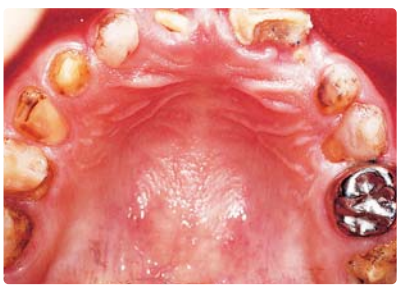
Hypomineralized forms

- Normal thickness of enamel initially
- Dark yellow to brown in colour (fig. 1,2,3,4)
- Enamel softer than normal, tends to chip and can be penetrated with an explorer. In several forms the enamel may be scraped away with scaler (fig. 7)
- Teeth erupt with normal thickness but enamel is soon lost, exposing rough highly sensitive dentine
- Large masses of supra gingival calculus are present (fig. 4,13,14)
- Radiographically, difficult to distinguish between enamel and dentin and may appear moth eaten in severe cases (fig. 15)

Management

In general, the aims of management are to treat pathology and pain, provide adequate esthetic appeal, maintain occlusal function and maintain the vertical dimension.

- Genetic counselling
- Preservation of molar teeth with full coverage restorations to maintain vertical dimension. (fig. 10,11)
- Over dentures may be an option in cases with small hypoplastic teeth.
- Stainless steel crowns or gold onlays on molars (fig. 8)
- Care is required when trial fitting crowns, because defective enamel can be easily scraped or flaked off the tooth (fig. 12,13)
- Composite resin veeners over anterior teeth for esthetics. It is possible to successfully bond composite to hypoplastic and hypomineralized enamel (fig. 9,17)
- Orthodontic correct anterior open bite in hypoplastic forms
- Delay definitive treatment with porcelain and precious metals until late adolescence (fig. 17)
- Adequate margins may be difficult to achieve because of the poor quality of the enamel



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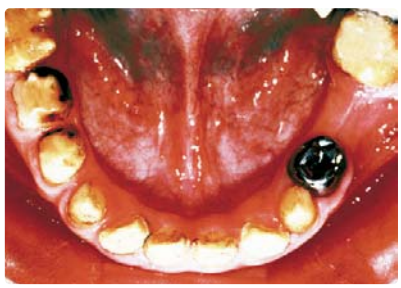

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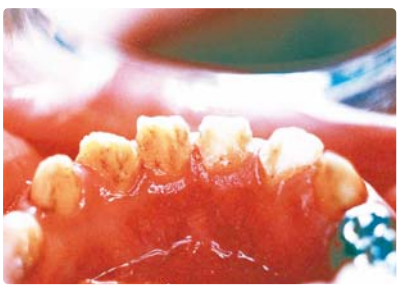

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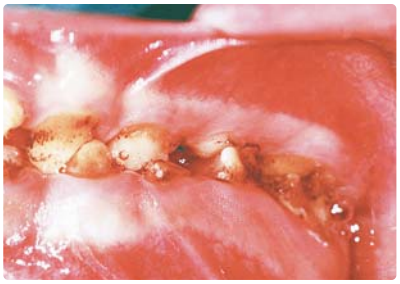

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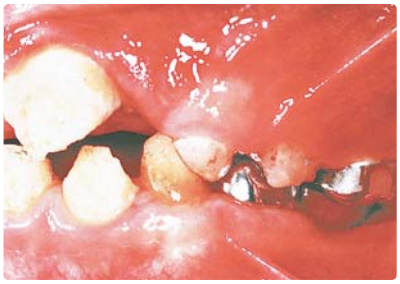

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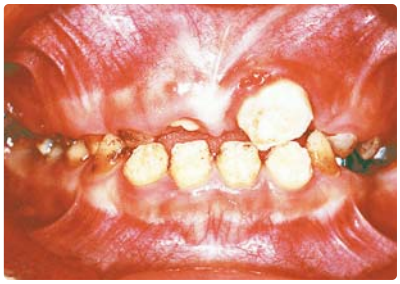

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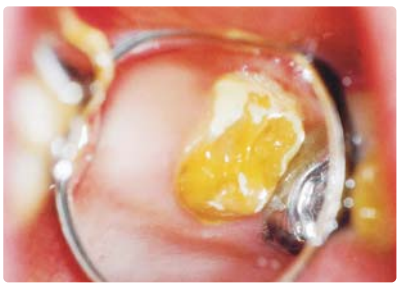

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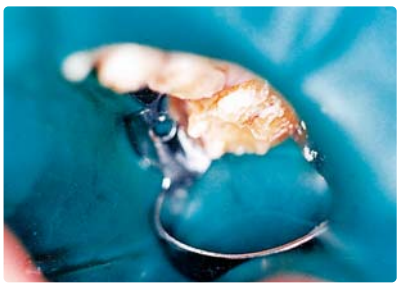

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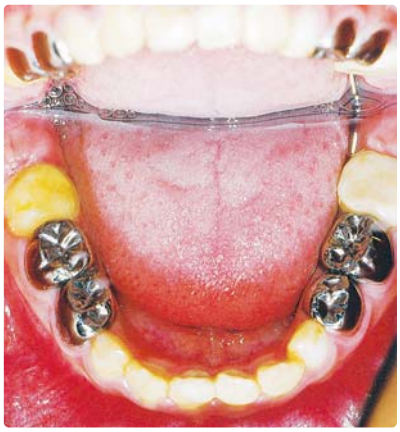

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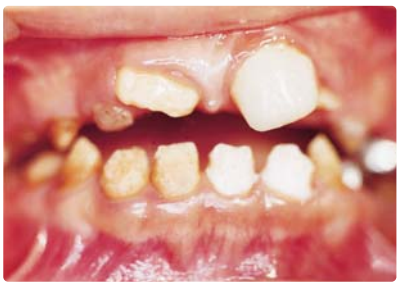

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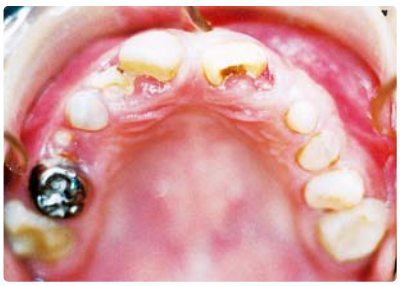

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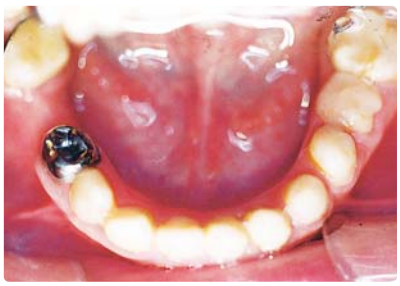

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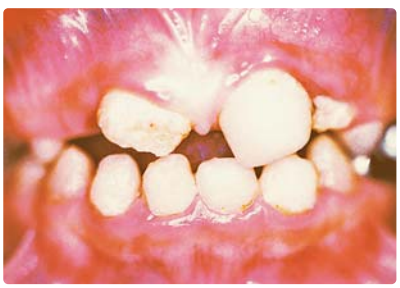

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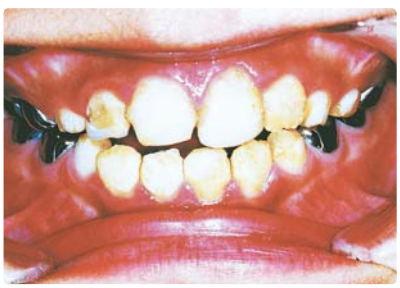

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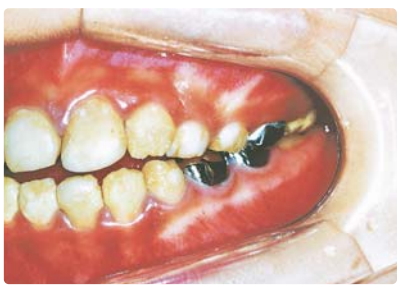

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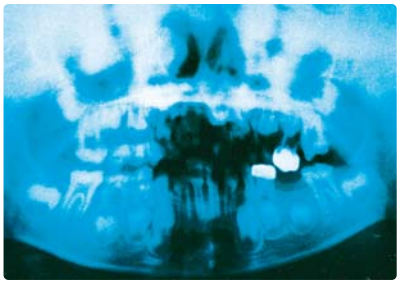
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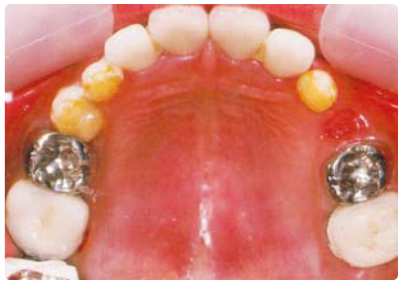
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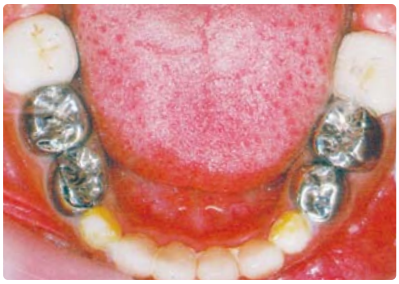
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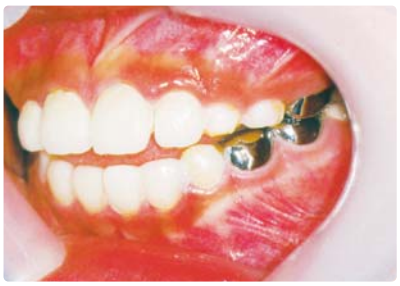
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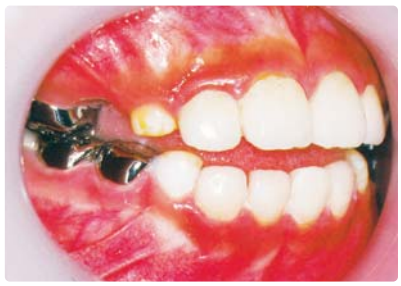
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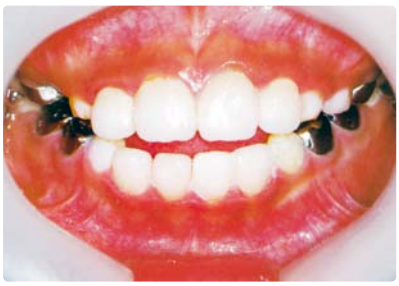
fig. 21

Disscussion

Several aspects of amelogenesis imperfecta are requried to improve the understanding of this condition.

Molecular studies of the genetic aspects of the disease would provide importance insight into its pathogenesis.

Comparative biochemical, clinical, and electron microscopic studies of affected teeth from different variants of amelogenesis imperfecta would lead to better understanding of the differences in defects found in each type.[8] Furthermore, while previous prevalence studies have provided useful information. Further epidemiological studies of other populations racial groups are necessary. In these studies, improved diagnostic criteria based on current understanding of phenotype expressions of the different variants may provide more accurate figures of prevalence.[9]

In spite of significant research advances into dental development and enamel formation in recent years, the pathogenetic mechanism of developmental enamel defects remain poorly understood. It is likely, however, that advances in cell and

molecular biology will rapidly improves our understanding of this complex field in the future. The development of molecular probes for enamel proteins will enable accurate biochemical analysis of abnormal enamel and may aid in the diagnosis of many inherited and acquired types of enamel hypoplasia.[16] Finally, properly controlled long term clinical studies of children with systemic and local enamel anomalies are required to identify the clinical complications associated with this common clinical entity.

Summary

Although the biochemical basis of enamel hypoplasia is not completely understood, it is likely that enamel defects are related to alteration of the enamel matrix proteins during amelogenesis. These proteins consisting of amelogenins, which are present in the early stages in enamel formation, have major roles in the structural organization and mineralization of developing enamel.[13,14]

In recent investigaton of teeth obtained from a patient with the hypomaturation type of amelogenesis imperfecta, excessive amelogenins was detected in the hypoplastic enamel, confirming that the primary defect in this condition was abnormality in maturation process.[15]

Furthermore, environmental insults such as high levels of fluoride have been shown to inhibit the secretion of enamel matrix proteins, as well as causing abnormal retention of amelogenins during the early maturation stage of enamel formation. This may interfere with crystallite growth and delineation.[16]

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