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Amelogenesis Imperfecta in Primary Dentition- A Case of Full Mouth Rehabilitation

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Abstract

The most common anomalies of dental hard tissues include hereditary defects of enamel. Amelogenesis imperfecta (AI) has been described as a complex group of hereditary conditions that disturbs the developing enamel and exists independent of any related systemic disorder. This clinical case report describes the diagnosis and management of hypoplastic amelogenesis imperfecta in a 5-year-old child. The treatment objectives were to improve aesthetics, improve periodontal health, prevent further loss of tooth structure, and improve the child's confidence. The treatment plan was to restore the affected teeth with full coverage restorations. Treatment involved placement of composite strip crowns on maxillary anterior teeth and stainless steel crowns on the posterior teeth followed by fluoride varnish application in the upper and lower arches. A 6-month follow-up showed great aesthetic and psychological improvements in the patient.

Keywords: Amelogenesis imperfecta, Deciduous dentition, Composite strip crowns, Stainless steel crowns

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Introduction

Hereditary defects of enamel are the most common anomalies of dental hard tissues. Amelogenesis imperfecta (AI) has been described as a complex group of hereditary conditions that disturbs the developing enamel and exists independent of any related systemic disorder[1,2,3]. This inherited condition occurs due to disturbance in the enamel proteins during amelogenesis which results in abnormal enamel formation. This disturbance can lead to anomalies in enamel quantity, structure or composition[4,5,6,7]. The mode of inheritance of AI can be either as autosomal dominant or autosomal recessive or X-linked[8]. AI is the oldest known hereditary disorder affecting enamel observed in humans[9]. This genetic disorder can affect any number of teeth and can occur in both primary and permanent dentition which results in the teeth being small, pitted, grooved and fragile with discolouration[10]. The incidence of AI ranges from 1:14,000 to as high as 1:700 children depending on the population under study and the diagnostic criteria employed[11]. In children, hypoplastic type of AI is more prevalent[12,13,14].

AI is classified into 4 major types and 14 subtypes depending on the clinical manifestation and genetic factors. The 4 main types are hypoplastic, hypocalcification, hypomaturation and hypomaturation-hypoplastic with taurodontism[15]. Hypoplastic type occurs due to defect in the enamel matrix formation which is characterised by reduced thickness or amount of enamel[16,17]. It can be localised or generalised but the calcification of the enamel is normal[5]. Hypocalcified type occurs due to defect in the calcification of enamel wherein the enamel develops to normal thickness but is soft, friable and wears away easily from the underlying dentin[16]. In hypomaturative type the enamel has normal thickness but compared to hypocalcified type it is harder shows mottling with opaque white to yellow-brown or red-brown discolouration and tends to chip away easily than wearing away[2]. In hypomaturation-hypoplastic with taurodontism type, there is mixture of hypomaturation and hypoplastic defects with taurodontism[17].

The common complications of AI regardless of the types are tooth sensitivity, poor aesthetics, decreased occlusal vertical dimension, difficulty in maintaining oral hygiene ultimately leading to decreased self confidence in cases involving anterior teeth[12,18]. Other rare complications include multiple impacted teeth, congenitally missing teeth and anterior open bite[19]. Treatment of AI becomes imperative not only for restorative and aesthetic reasons but also because of the psychological impact on the children it causes. We hereby report a case of Amelogenesis Imperfecta in primary dentition with full mouth rehabilitation.

Case Report

A 5-year-old healthy female child reported to Department of Paediatric Dentistry with chief complaint of discoloured teeth present since eruption. Medical history was not contributory. Family history revealed that no family member or relative had such a presentation. The parent confirmed that the patient's social life was affected, and she was teased by her friends due to the poor appearance of her teeth. A comprehensive extraoral and intraoral clinical examination was done, and radiographs and photographs were taken. Clinical examination revealed patient had complete set of primary dentition. The enamel was yellowish brown in colour,**(Figure 1, 2)** soft in consistency with attrited cuspal contour. Dental caries was seen in primary right upper second molar.**(Figure 3)** The radiographic examination revealed loss of enamel and discontinuity in the anterior teeth. Based on clinical and radiographic findings, the case was diagnosed as hypoplastic amelogenesis imperfecta.

The treatment objectives were to improve aesthetics, improve periodontal health, prevent further loss of tooth structure, and improve the child's confidence. The treatment plan was to restore the affected teeth with full coverage restorations.

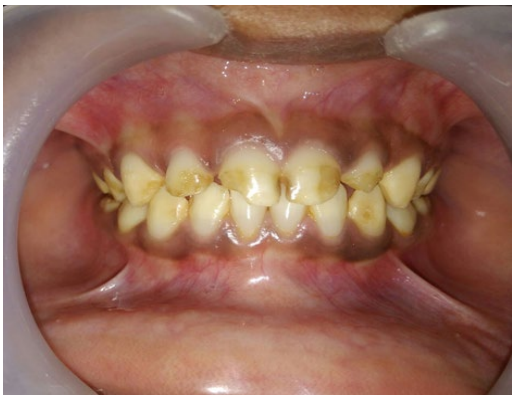

Figure 1: Pre-operative picture of occlusion

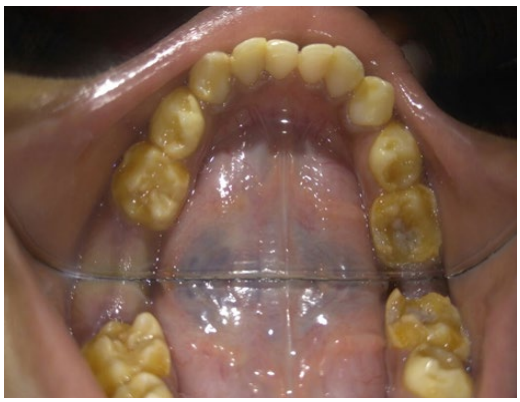

Figure 2: Pre-operative picture of mandible

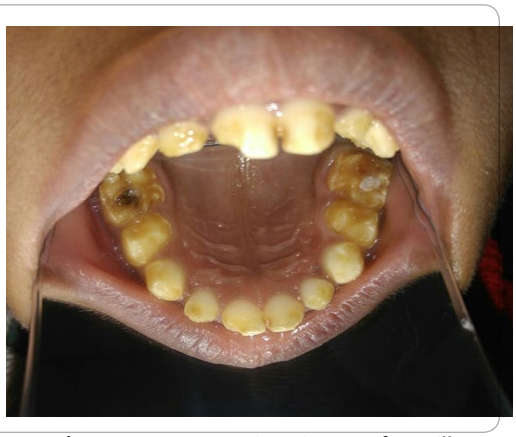

Figure 3: Pre-operative picture of maxilla

Anterior teeth were prepared to receive composite strip crowns. **(Figure 4)** After the tooth preparation try-in was done with anterior strip crowns (3M ESPE) and size 3 was selected for all anterior teeth. Etching was done with 37% phosphoric acid on all the anterior teeth. Bonding agent (Te-Econom Bond, Ivoclar Vivadent) was then applied according to the manufacturer's instructions. Composite resin(Filtek Z350 by 3M ESPE) was packed into the strip crowns and was bonded to the prepared anterior teeth and was light cured. The composite strips were removed using a BP blade. Finishing and Polishing was done.**(Figure 5)**

Dental caries was excavated from primary right upper second molar and vital pulp therapy was carried out. The Posterior teeth were prepared to receive stainless steel crowns. Stainless steels crowns (3M ESPE) were adapted on all primary molars and luted with type II Glass ionomer cement(GC).**(Figure 6,7)** Fluoride varnish application (Fluor Protector, Ivoclar Vivadent) was done in the upper and lower arches. Aesthetics and psychology of the patient had been greatly improved when the patient was followed up after 6 months.

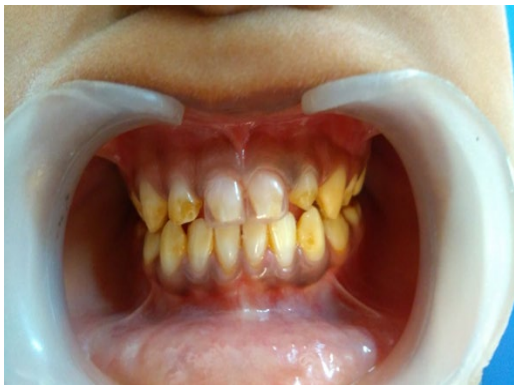

Figure 4: Preparation for placement of strip crowns

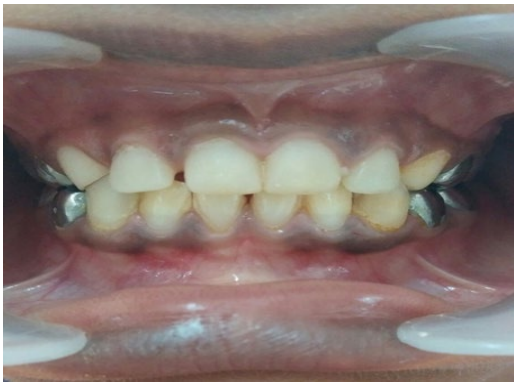

Figure 5: Placement of strip crowns

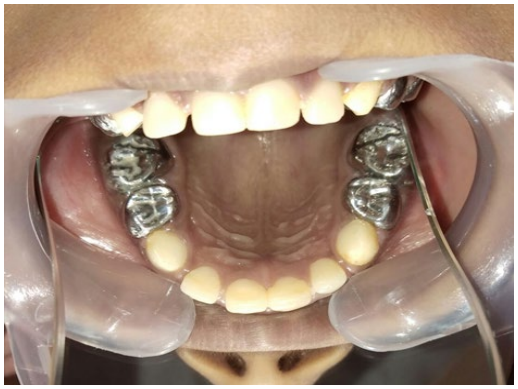

Figure 6: Placement of Stainless steel crowns on Maxillary molars

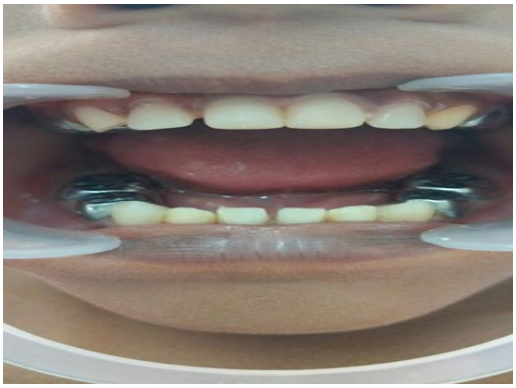

Figure 7: Placement of Stainless steel crowns on mandibular molars

Discussion

Amelogenesis imperfecta has many forms and the children are affected at a very early age. There are 3 stages in enamel formation: 1) the translation and secretion of an extracellular matrix, 2) mineralisation of the matrix, 3) matrix removal and crystalline growth or maturation of enamel[5]. Any disruption in these stages results in Amelogenesis imperfecta[20] with different subtypes of the condition arising from disruptions in the different phases of the stages[21].

Hypoplastic AI occurs due to the disruption in the secretion of enamel matrix which is the first stage in enamel formation. It is characterised by local or generalised decrease in the thickness of the enamel with teeth being yellowish brown in colour, have a rough texture and wide spaces between them. Hypocalcified type occurs due to defect in the mineralisation of the enamel matrix which is the second stage in amelogenesis. The thickness of the enamel is almost normal but due to the deficient mineral content, the enamel is soft, friable, and subject to rapid wear. Hypomaturation type is due to defect in matrix removal which is the third stage in amelogenesis resulting in decreased mineral deposition and increased matrix retention. The enamel is of normal thickness but compared to the hypocalcified type, the enamel is harder, mottled, brown-yellow and tends to chip away rather than wearing away. These variants of AI may overlap clinically. The degree of mineralisation determines the various treatment options as thin or poorly mineralised enamel is difficult to bond to. Hypoplastic type has variable mineral content while the hypocalcified and hypomaturation type has low mineral content[22].AI is reported to be caused due to the mutations in any of the following genes/locus AMELX, ENAM, MMP20, KLK4, FAM83H, WDR72, C4orf26 and FAM20A[9].

The complications of AI may result in problems in mastication and aesthetics. The management of a patient with AI is dependent on many factors like age, socioeconomic status, the type and the severity of the condition. Preventive methods for AI in primary dentition include dietary advice, oral hygiene instructions and topical fluoride application[23]. Four weeks of fluoride usage may result in reduction in dental sensitivity. With the possibility of the disorder presenting in permanent dentition too, it is the responsibility of the dentist to counsel the patient's parents about its effects in permanent dentition. If needed the parents can be encouraged to seek genetic counselling[24].The treatment of AI is multidisciplinary and it includes paedodontists, orthodontists, oral surgeons, and prosthodontists. For primary teeth the treatment options include use of glass ionomer cements, composite resins, full coverage restorations like stainless steel crowns, lab fabricated crowns, strip crowns, porcelain veneers. If anterior teeth cannot be restored and extractions are needed, an aesthetic bridge can be used. In case of multiple extractions, overdentures can be used too. Treatment of AI is a long process and cooperation from the child and motivation from the parents go a long way in success of the treatment.

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