

# MANAGEMENT ENIGMA OF AMELOGENESIS IMPERFECTA IN YOUNG AGE....A REPORT OF TWO CASES

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## ABSTRACT

Amelogenesis imperfecta (AI) is a hereditary condition that affects tooth enamel without systemic involvement. The main clinical characteristics are extensive loss of tooth tissue, poor esthetics and tooth sensitivity. Early recognition followed by appropriate preventive care and oral rehabilitation is essential in the successful management of AI. This report describes the different inheritance and clinical presentation along with management modalities in two patients. Difference in clinical presentation between boys and girl is also highlighted. Both the patients reported with the carious exposure, with general dentinal sensitivity and poor esthetic appearance. The first phase of the treatment was preventive measures to improve dental and periodontal health. On the second phase the molars were endodontically treated and covered with stainless steel crowns. A direct veneering composite resin restoration and heat cure full veneer crown restoration for esthetic rehabilitation were done in third phase. The performed treatment achieved satisfactory esthetics and function.

## KEY WORDS

**Amelogenesis imperfecta, stainless steel crown, acrylic crown**

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## INTRODUCTION

Amelogenesis imperfecta (AI) is a group of inherited disorders characterized by abnormal enamel formation. These non-syndromic disorders are not associated with defects of other parts of the body or any other health problem<sup>1</sup>. These enamel defects are as a result of gene mutations associated with amelogenin protein and enamelin protein. AI follows an autosomal dominant, autosomal recessive or X-linked pattern of inheritance. The reported prevalence of AI is highly variable, it is reported to be 1:14,000 in the USA and 1:700 in Europe.<sup>1</sup> There are 3 types of amelogenesis imperfecta: hypoplastic, hypocalcification, and hypomaturation types. Amelogenesis imperfecta can cause white flecks, narrow horizontal bands, lines of pits, grooves, and discoloration of the teeth varying from yellow to dark brown.<sup>1,2</sup> Enamel hypocalcification is a defect in the mineralization process. In this form, enamel is soft and friable.<sup>3</sup> Hypomaturation is an abnormal occurrence in the final stages of the mineralization process. The clinical features distinguish the hypoplastic and hypocalcified types. In the hypoplastic forms, the enamel does not develop to its normal thickness. The hypomaturation forms differ from hypocalcification in that the enamel is harder, with a mottled opaque white to yellow-brown or red-brown color, and tends to chip from the underlying dentin rather than wear away.<sup>2,4</sup> Restoration of these defects is important not only because of esthetic and functional concerns, but also because there maybe a positive psychological impact for the patient. Treatment planning for patients with amelogenesis imperfecta is related to many factors: the age and socioeconomic status of the patient, the type and severity of the disorder, and the dentition at the time the treatment is planned.<sup>4</sup> Pitted enamel surfaces may predispose AI teeth to plaque accumulation. Meticulous oral hygiene has to be maintained at a high level if a favorable long-term prognosis for restorative procedures is to be achieved.<sup>5</sup>

## CASE REPORT 1

A 14-year-old girl reported to the Dept of Pediatric & Preventive Dentistry of Dr. R. Ahmed

Dental College & Hospital with the chief complaint of pigmented, fragile teeth. After obtaining past dental history it was found that the deciduous teeth had also been affected. A complete dental and medical examination of the index case was performed and 12 family members were also examined. After prophylaxis, all teeth were dried and the defects were recorded according to the Developmental Defects of Enamel Simplified Index-DDE (FDI, 1992)<sup>6</sup>. Based on the genetic and clinical examination of proband and family members, a diagnosis of autosomal dominant hypomaturation AI (ADAI) was determined (Figure 1). Several family members had similar features. Medical history revealed well complied childhood immunization and others were noncontributory.

### Radiographic and Clinical Examination

An intra- and extra-oral radiographic examination was performed, and no significant difference in hard tissue contrast was noted between

enamel and dentin (Figure 2). Oral hygiene was poor and the gingival tissue was boggy and swollen (Figure 3). The clinical examination revealed the presence of vertical grooves, linear defects, pigmented pits and areas with thin enamel which was very pathognomonic in female patients having AI. Other presentation included diffuse opacities, teeth with shape and color alterations and areas with fractured enamel (Figures 3 and 4). The posterior teeth presented with carious exposures in all the lower first molars with history of occasional pain (Figures 5). It appeared that cariously involved molars did not show any sign of loss of the vertical dimension (Figure 6). Also noticed in this case is the cross bite in the left canine region which made the treatment more complex.

A three-phase treatment plan was proposed:

- 1) Preventive care
- 2) Restorative procedures
- 3) Case follow-up

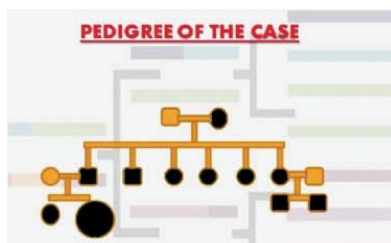


Figure 1: Autosomal dominant presentation



Figure 2: No hard tissue contrast between enamel & dentin



Figure 3: Swollen boggy gingival with left canine in cross bite Relation



Figure 4: Vertical grooves

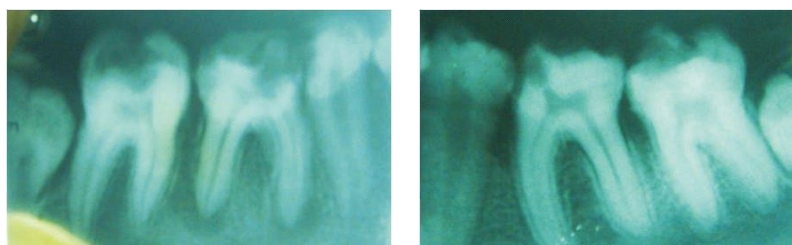
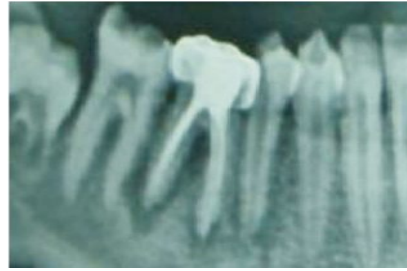


Figure 5: Carious exposures in 36, 46



Figure 6: Sharp cuspal height with no loss of vertical dimension



**Figure 7: Endodontically treated lower molars**



**Figure 8: Stainless steel crown on molars**



**Figure 9: Preparation of mandibular anteriors for full veneer heat cure acrylic crown**

**Figure 10: Post Operative smile with heatcure acrylic crown**

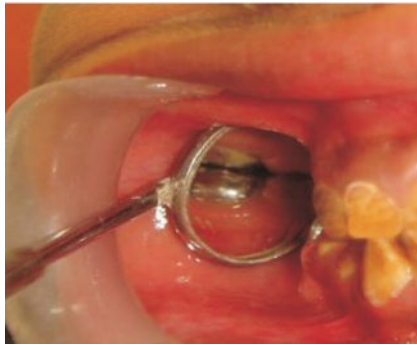
**Figure 11: Patchy Discolouration and gross deformity**

## MANAGEMENT

Since the patient presented poor oral hygiene, preventive care of oral hygiene instructions and dietary advice, were given along with fluoride mouthwash. Regarding restorative procedures, direct and indirect composite restorations along with heat-cure acrylic crown were considered. Firstly the posterior teeth (36 and 46) (Figures 7) were endodontically treated and filled. During a subsequent visit stainless steel crowns (3M ESPE) were selected for all the first permanent molars to maintain the existing vertical dimension and prevent fracture of crown. Initially one sided both upper and lower crowns were fabricated taking guidance of the occlusion of the opposite side since vertical dimension was to be maintained in that existing height (Figure 8). The upper and lower anteriors whose labial aspects were affected (Figures 2,3) veneering with composite, were planned initially. The degree of enamel defect, intensity and deepness of colour alteration and/or extrinsic pigmentation was

considered as criteria of the stripping thickness. But due to the problems associated with mineralization, initial bonding failure had been noticed in this patient. Considering this fact and age of the patient, full veneer heat cure acrylic crown has been planned for all the anteriors as an intermediate restoration since patient was having severe sensitivity and difficulty in managing proper oral hygiene. Accordingly the teeth were prepared with diamond burs (SHOFU Inc.) under local anesthesia. Maxillary anteriors were prepared first followed by mandibular anteriors (figure 9). Impression was taken with rubber base impression material and sent to lab for fabrication of the crown. The crowns were constructed as a separate unit and cemented with luting GIC. After 3 month follow up a dramatic improvement in esthetic; gingival health were noticed in this patient (Figure10).





**Figure 12. Maintenance Of Vertical Dimension By Stainless Steel Crown**



**Figure 13: composite vennering**



**Figure 14: 3 months follow up**

## CASE REPORT 2

A 10 year-old boy was referred to the pediatric dentistry dept of, Dr. R Ahmed Dental College & Hospital for evaluation and treatment for his unaesthetic appearing yellowish teeth. Prior to treatment, detailed medical and dental history was obtained from the parents. The dental, family history revealed that none of parents, brothers or relatives suffered from similar dental conditions. Intra-oral examination showed loss of enamel from incisors and molars, but no marginal gingivitis was seen (Figure10)

All the teeth were covered by hypoplastic enamel. The molars were severely affected with loss of occlusal enamel layer, resulting in a reduction of the occlusal vertical dimension and carious exposure were found in left sided upper & lower permanent first molars (26 & 36). The exposed dentin seemed normal. The dental family history, clinical and radiographic features were consistent with a possible diagnosis of idiopathic hypocalcified-hypoplastic amelogenesis imperfecta.

In order to restore the function, esthetics and prevent further loss of tooth structure, the treatment was planned to be conducted in three phases. The first phase of the treatment started with preventive measures to improve dental and periodontal health. In the second phase pulpally involved 26 & 36 were endodontically treated and filled with GIC. The patient's vertical dimension was reduced a little due to wearing out of cuspal enamel. But permanent increase of vertical dimension was not feasible as permanent second molar has not erupted and occlusion is in transitional phase. To maintain existing vertical dimension preformed stainless steel

crowns were constructed for all the permanent first molars. (Figure 12). To restore the esthetics a direct composite veneering was advocated in this patient. In labial veneer preparation, a minimum chamfer was obtained with diamond burs (KG Sorensen). The proximal contact area was maintained. Then, 37% phosphoric acid etching was performed for 15 seconds and a bonding agent was applied (Single Bond, 3M ESPE). A thin layer of an opaque shade of composite (Charisma OA20, Heraeus Kulzer, Germany) was initially applied to mask the remaining spot (Figure 13), followed by a hybrid resin composite (Charisma color A20 and I, Heraeus Kulzer, Germany) and a microfilled resin composite (Filtek All 0 color A2, 3M ESPE, USA) (Figure 9). The lower canines were restored with opaque and microfilled composites. Finishing and polishing were achieved with abrasive discs (Sof-lex/3M ESPE). The patient has been put on periodic checkup. After 3 months follow up existing restoration were seen in good health with improvement in patient's outlook and confidence. (Figure 14)

## DISCUSSION

Treatment of patients with AI requires special care. These patients are emotionally upset due to the poor appearance of their teeth. So esthetic improvement can positively increase their self-confidence and improve their behavior. However, the restoration of esthetics and function of teeth in child patients suffering from amelogenesis imperfecta often poses a great challenge to the dentist<sup>7</sup>. Poor oral hygiene is a recognized problem in patients with AI, because of the rough enamel surface which causes

plaque retention and due to the sensitivity experienced when brushing which was evidently present in the first case. Therefore, oral hygiene instructions are essential and corresponded to the first phase of the treatment<sup>1,5</sup>. Nowadays, there is a range of materials used to restore the teeth that includes the use of composite resin, polycarbonate crowns; stainless steel crowns (SSCs), full ceramic crown, metal ceramic crown, glass ionomer cement and functional maintenance dispositive to restore a mutilated dentition<sup>8</sup>. In most cases, full coverage is desirable for posterior teeth due to the extensive loss of enamel and also to prevent further loss of tooth structure and contact point<sup>9</sup>. In mixed dentition and early permanent dentition, SSCs are the most effective type of restoration<sup>8,9</sup>. The SSCs are extremely durable, relatively cheap, subject to minimal technique sensitivity during placement, and offers the advantage of full coronal coverage<sup>10</sup>. For anterior esthetics in young children composite veneering is supposed to be a good option. But due to cost and difficulty in achieving isolation, heat cure acrylic crown can be a good alternative<sup>11</sup>. It provides great esthetic along with full coronal protection. It has been seen that in some patients of AI due to constant irritation in teeth due to thin layered enamel, there is gradual receding of the pulp chamber due to formation of the reparative dentin<sup>12</sup>. So amount of reduction for full coverage restoration will not lead to the exposure of pulp. This same thing can be noticed in the first case. But for the boy this amount of reduction will lead to exposure of pulp so the change of modalities to composite veneering was considered for that patient. A better understanding of AI is required in order to make a correct diagnosis. Therefore, early prevention measures and an appropriate restorative treatment may avoid further damage. During post treatment follow up, after the restorative procedures, the patient's dental hypersensitivity disappeared completely and the chewing function increased leading to good physical wellbeing. It became evident that this was a strong motivation to improve her behavior and the level of cooperation during treatment. However a long term follow up is most desirable.

## CONCLUSION

AI can significantly affect the children psychologically and socially specially during the growing phase of life'. So the treatment during the mixed dentition and early permanent dentition must be performed with special consideration, in order to restore both the function and the child's confidence to prevent serious problems in the future occlusion and social adaptation of the patient.

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