The Restorative Management of Amelogenesis Imperfecta in the Mixed Dentition

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Amelogenesis impefecta is a hereditary condition affecting the formation of enamel in which the rough enamel surface can compromise periodontal health and the esthetics. Affected posterior teeth usually exhibit interproximal space loss which makes restoration of the primary molars difficult. This article describes a technique, using separators to regain interproximal space prior to the placement of stainless steel crowns on the molars of a girl with amelogenesis imperfecta.

Key words: *amelogenesis imperfecta, enamel hypoplasia, stainless steel crown, composite, space loss, separators.*
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INTRODUCTION

Amelogenesis imperfecta is a hereditary condition which results in abnormal development of enamel on the teeth. It presents as a series of manifestations of enamel malformation predominately affecting all the teeth in both the primary and permanent dentitions. It has been recognized as an isolated anomaly, or as an integral feature of certain syndromes, such as amelo-onchohypohidrotic, Morquio, trichodento-osseus, Kohlschütter and dystrophic epidermolysis bullosa, oculo-dento-osseous dysplasia, pseudohypo-parathyrodism, tuberosis sclerosis and vitamin D-dependent rickets.[1] However, Witkop[2] proposed that the term amelogenesis imperfecta should be limited to those inherited congenital defects that primarily affects enamel formation which are not accompanied by morphological or metabolic defects in any other body system.

The diagnosis of amelogenesis imperfecta is usually based on generalized enamel hypoplasia in both the primary and permanent dentitions; a family history of the condition; and the absence of systemic diseases that may cause generalized enamel hypoplasia resembling amelogenesis imperfecta.[3]

The prevalence reportedly ranges from 1:14,000 in the USA[2] to 1:4000 in Sweden.[4] Two genes are know to be responsible for the complex process of enamel formation and maturation, namely the amelogenin and enamelin genes. Alteration in the amelogenin, the ectoderm derived protein involved in the control of enamel mineralization, can result in improper differentiation of the enamel during matrix formation, maturation and organization.[5-7]

In 1971, Rao and Witkop[8] listed 11 varieties of amelogenesis imperfecta and they suggested that additional types might be present. The classification of Winter and Brook[7] was based on the genotypes, radiographic appearance and morphological pattern of enamel. Nevertheless, the classification suggested by Witkop,[2] which is based on the pattern of inheritance and the main morphological features in clinical and radiological terms, is commonly used. Essentially, there are three main phenotypes of amelogenesis imperfecta, depending on the stage of enamel formation that is primarily affected; they are recognized as hypoplastic, hypomaturation and hypocalcified types.

The hypoplastic type which accounts for 60-73% of cases mainly affects females. The enamel is usually thin but of normal hardness. The surface is usually smooth but with localized areas of thicker enamel resulting in an abnormal crown contour. Alternatively, the enamel may be pitted or have horizontal or vertical ridges. Female carriers of the X-linked type may have alternating vertical ridges of normal and pitted enamel, known as the Lyonization effect.[10] In addition, some teeth may exhibit delayed or failed eruption, or even undergo resorption.

The hypomaturation type is less common and affects 20-40% of cases and usually occurs in males. The thickness of the enamel is initially normal ; however, it is lost soon after eruption because of its reduced hardness. Therefore, rough and highly sensitive dentin is exposed. The enamel is dark yellow to brown in color. Because of the X-linked mode of inheritance, female carriers may have vertical stripes of opaque white enamel alternating

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with normal enamel (Lyonization effect).[10]

Approximately 7% of individuals affected by amelogenesis imperfecta have the hypocalcified variant. The enamel is insufficiently mineralized and extremely soft. Therefore, the discolored enamel is easily removed. Interestingly, the enamel at the cervical portion is frequently better calcified than that on the rest of the crown. The hypoplastic and hypocalcified variants may occur together. Overlapping features have been identified at the micromorphological[11,12] and microradiographical levels.[13] The existence of the various sub-classifications has recently been questioned, because the clinical and histological variability connected with the same pattern of inheritance suggests that the genetic defect, together with biological variations, could explain the differences in clinical expression. Therefore, a new classification of amelogenesis imperfecta based on the molecular genetics and mode of inheritance, has been postulated.[14]

The rough enamel surface, which is susceptible to extrinsic staining and the abnormal shape of the crown, cause a major esthetic problem. In addition, the rough enamel[1] surface, which usually extends subgingivally, predisposes to plaque retention and calculus formation. Therefore, the gingival health of many of these teeth is often compromised. Thermal sensitivity of the affected teeth which is relatively common in the more severe variants of the hypocalcified and the smooth and thin hypoplastic types. Several investigators have also reported delayed eruption and impaction of teeth[4,9,15] and even accelerated dental development.[16]

An accurate diagnosis is important to exclude the presence of a systemic condition that exhibits generalized enamel hypoplasia as an accompanying feature; thus the appropriate genetic counseling can be scheduled. Additionally, by establishing the type of amelogenesis imperfecta, the most appropriate form of restorations can be prescribed for the affected teeth.

The psychological impact on an individual who has teeth affected by amelogenesis imperfecta cannot be underestimated. The individual may be too embarrassed to smile because they are unhappy with the appearance of their teeth.[17] Therefore, the planning of esthetic restorations on the anterior teeth may not only preserve the tooth structure and reduce sensitivity, but also eliminate the adverse social impact of their teeth.

This report describes the clinical management of a case of amelogenesis imperfecta in the early mixed dentition, with special emphasis on the clinical procedures used to regain space prior to restoration of the molars.

CASE PRESENTATION

A 7 years 2 months old Chinese girl attended the clinic because of discolored primary and permanent teeth which manifested some thermal sensitivity. The family history revealed that her paternal grandparents, father and uncles suffered from similar dental conditions (Figure 1). Clinically, the child's oral hygiene was unsatisfactory and she exhibited moderate chronic marginal gingivitis. All of her erupted teeth were covered by discolored hypoplastic enamel.

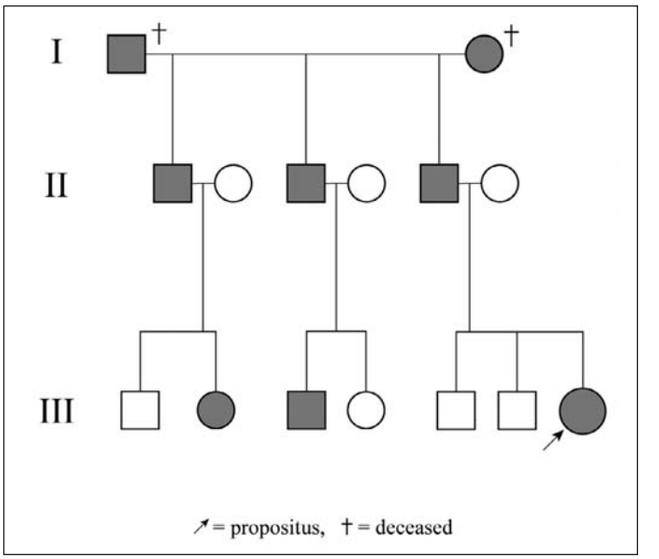

Figure 1: Pedigree of the family with amelogenesis imperfecta.

Her primary teeth and permanent incisors were darkish-yellow in color. Although the enamel of the incisors was smooth the surface contours were irregular. Attrition of the enamel was evident on her primary teeth resulting in a reduction in the occlusal vertical dimension, and the interproximal spaces had been lost between the primary molars and canines (Figure 2).

Two supernumerary teeth were located in the anterior of the maxilla; one erupted between the maxillary permanent central incisors while the other was inverted and impacted palatal to the maxillary left permanent central incisor (Figure 3). The bitewing radiographs showed that the teeth were covered by only a thin layer of enamel on the occlusal and proximal surfaces. The pulp morphology was normal (Figure 4). Also, there were varying degrees of interproximal wear and there were open contacts between the primary molars. The clinical and radiographic features and the family history were consistent with a diagnosis of x-linked hypoplastic amelogenesis imperfecta.

The treatment objectives were to improve the esthetics, eliminate the tooth sensitivity, prevent further loss of tooth structure, modify the child's attitude and behavior towards dental treatment and improve her periodontal health. As part of the treatment plan, the treatment alternatives were explained to the child and her parents. This included the amount of tooth structure that would need to be removed, the expected clinical longevity of the restorations and the length of the treatment period. After considering all of the treatment options, it was decided to place direct composite veneers on the permanent incisors and stainless steel crowns on the primary and permanent molars; the parents understood that this could temporarily compromise the esthetics in the molar regions.

The erupted supernumerary tooth was extracted prior to the restorative treatment. However, so as not to compromising the growth of the adjacent incisors, surgical removal of the impacted supernumerary tooth was postponed until the root development was completed.

A hybrid composite was chosen to restore the defective tooth structure of the permanent incisors. No preparations were preformed on the incisors, other than cleaning with a rotary bristle brush with

Figure 2: Pre-operative clinical pictures showing the rough yellowish brown colored enamel on all of the teeth.

Figure 3: The pre-operative upper anterior occlusal radiograph indicating the presence of impacted and erupted supernumerary teeth.

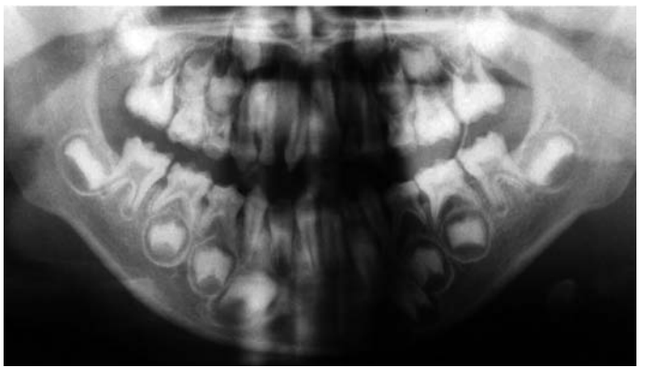

Figure 4: The radiographic appearance of the dentition (a) panoramic radiograph showing diminished enamel on the erupted teeth and all of the developing teeth consistent with the patient's chronological age.

Figure 4: (b) The bite-wing radiographs showing the lack of radiographic differentiation between the enamel and dentin and spacing between the molars with evidence of associated space loss.

Figure 5: Rubber ring separators inserted between (a) the primary molars prior to preparation of the mandibular second primary molars for stainless steel crowns.

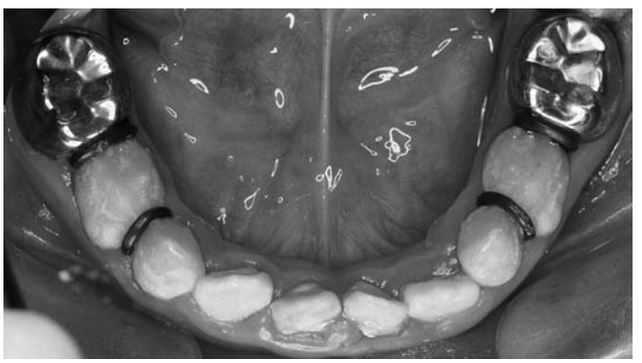

Figure 5: (b) mesially and distally on the primary molars immediately after placement of stainless steel crowns on the second primary molars.

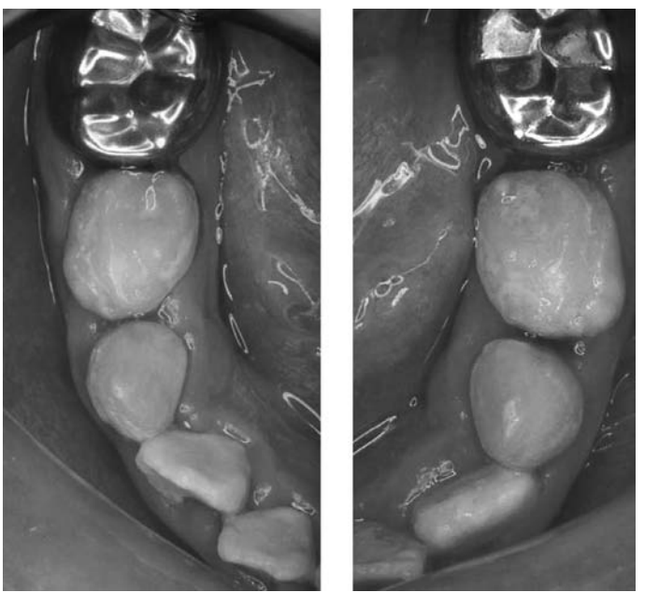

Figure 6: The amount of space that was regained mesial and distal to the mandibular primary molars is evident after removal of the separators.

Figure 7: Post-operative pictures showing the composite veneers on the incisors and stainless steel crowns on the primary and permanent molars.

pumice prior to acid-etching with 35% phosphoric acid for 30 seconds followed by rinsing with water spray. The labial surfaces of the maxillary and mandibular incisors were then directly restored with resin composite (TPH Spectrum, Dentsply). Conservative tooth preparation was performed on the primary canines prior to placement of the direct composite restorations.

Due to attrition of the interproximal hypoplastic enamel, space had been lost between the molars. In order to regain this lost space, orthodontic rubber ring separators (Ormco) were placed two days prior to the placement of stainless steel crowns (Figure 5a). Since the first permanent molars were not fully erupted, the sequence of the restorative treatment began with the primary second molars, followed by the first primary molars and then the first permanent molars. Furthermore, preformed stainless steel crowns were placed bilaterally at each visit so as to balance the occlusion and alter the vertical dimension.

By using rubber ring separators, the interproximal spaces were opened by 0.8 to 1.0mm, which was adequate for the placement of a stainless steel crown. The preformed stainless steel crowns (3M)

Figure 8: The post-operative panoramic radiograph after restorations of all the erupted teeth and extraction of the erupted supernumerary tooth.

were placed on the mandibular primary second molars following minimal slice preparations of the teeth. At the same visit separators were then placed on the mesial and distal surfaces of the mandibular first primary molars (Figure 5b). At the subsequent visit, the interproximal spaces adjacent to the mandibular first primary molars had been regained and the teeth were then restored with preformed stainless steel crowns (Figure 6). Using the same procedures, all of the erupted primary and permanent molars were restored according to the pre-determined sequence of treatment (Figure 7). The adaptation and quality of the margins of the preformed stainless steel crowns were evaluated using a panoramic radiograph (Figure 8). The stainless steel crowns on the first permanent molars are considered to be only temporary restorations. Once the second permanent molars and the premolars have established the level of the occlusal plane, the stainless steel crowns on the first permanent molars will be replaced by cast full-coverage restorations.

DISCUSSION

The successful management of amelogenesis imperfecta during childhood requires the cooperation and motivation of both the patient and parents need to be fully assessed before a definitive treatment plan is formulated.[18] Usually, the treatment will extend over many years and long term success will depend on regular attendances for restorative procedures and the maintenance of a high level of oral care.

Frequent topical fluoride applications and dietary control are strongly recommended to prevent caries. Plaque retention and calculus formation resulting from the rough enamel surfaces necessitate high levels of oral health care.[3] The exposed dentin can be sensitive to such stimuli as sweet, hot and cold; topical fluoride applications can control this until definitive restorations can be placed. The newly available Tooth Mousse (GC®) which contains Recaldent® CPP-ACP may prove useful in this regard.

The range of materials used to restore the teeth may include glass ionomer cement, composite resin, and stainless steel crown.[3] Unfortunately, in most cases full-coverage is desirable for posterior teeth due to the extensive loss of enamel also to prevent further loss of tooth structure.[19] In the primary and early mixed dentitions stainless steel crowns are the most effective type of restorations. The insertion of rubber separator rings prior to the placement of stainless steel crowns obviates the need for proximal reduction of tooth structures to allow the stainless steel crowns to be inserted with minimal tooth reduction.[20] However, the enamel can be stripped away if a tight fitting stainless steel crown is forced into place on a tooth. Also, good margins on the preparation may be difficult to achieve because the enamel can easily be made to flake off; this means that care should be executed when placing and removing the stainless steel crown during trial fitting. Glass ionomer cements are currently the luting agent of choice.

It is important to provide an esthetically pleasing and functional dentition. In the primary dentition, the anterior teeth can be restored with composite resin strip crowns; while glass ionomer cement is an effective intermediate restorative material. In the mixed dentition, if there is enamel available for bonding, then composite resin veneers may be used to mask the discoloration and improve the crown morphology.[3,21] However, the etching pattern of amelogenesis imperfecta is atypical due to abnormal prism structures,[22] so pretreatment of the affected enamel by sodium hypochlorite solution to enhance bonding has been suggested.[23] Additionally, because the thickness of enamel is also reduced, conservative tooth preparation, or even no preparation for direct composite veneer restoration is recommended.[24] In adolescents, porcelain veneers are also likely to be useful; however, their use in cases with amelogenesis imperfecta has not been extensively reported. Full-coverage adhesive composite crowns or polycarbonate crowns have also been advocated,[25] but strict oral hygiene measures need to be implemented because of the plaque retentive nature of the margins of the restorative materials and passive eruption, which will inevitably expose more defective enamel. Porcelain jacket crowns, which provide esthetic permanent restorations, have reportedly been successful in affected adults, but their use in young patients is contraindicated because of the presence of large pulp chamber and the likely need for frequent replacement due to passive eruption.

The psychosocial effects of amelogenesis imperfecta on affected individuals are significant.[17] Although there are technical difficulties associated with performing extensive restorative care in the mixed dentition, they are outweighed by the psychosocial benefits to the affected child. Nevertheless, preserving as much tooth structure as possible is highly desirable because the restorations will need to be replaced several times during adulthood.

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