



Preventive Strategy of Primary Teeth Affected by Amelogenesis Imperfecta: Case Report

Ariri El Mehdi*, Lferd Meriem and Chhoul Hakima

Department of Pediatric Dentistry, University of Mohammed V Rabat, Morocco

Abstract

Amelogenesis Imperfecta (AI) is defined as a result of specific genetic defects affecting the deposition, mineralization and maturation of enamel in both the primary and the permanent dentitions. The clinical issues are sensitivity (particularly to cold), rapid wear and poor aesthetics, all of which can vary depending on the type of AI. Other anomalies with AI include disturbances in eruption, anterior open bite, pulpal calcifications, pathological root or crown resorption and taurodontism, all of which can present in the pediatric patient. This case report describes the preventive strategy of a case with amelogenesis imperfecta in a young patient who has 4-year-old with 18 months follow-up.

Keywords: Amelogenesis imperfecta; Preventive strategy; Primary teeth

Introduction

Amelogenesis imperfecta has been defined as a complex group of hereditary enamel defects not associated with evidence of systemic disease. It can affect all or some teeth in deciduous and/or permanent dentition. The prevalence of AI varies according to different studies ranging from 1/700 to 1/14.000 [1,2]. These differences are mainly due to diagnostic or demographic criteria or to mutant genes in the studied population. Different classifications of AI have been reported in literature, usually related to phenotype characteristics (clinical and radiological appearance) or to the description of the mode of inheritance of the disease (autosomal dominant/recessive). Clinically four types of AI have been recognized according to enamel types: Type 1 (hypoplastic with a deficiency in the quantity of enamel); Type 2 (hypomaturation, with an opaque and chalky enamel), Type 3 (hypocalcified, characterized by a normal amount of poorly mineralized enamel) and Type 4 (hypoplastic-hypomaturation enamel associated with taurodontism). In some classifications, the type 4 is also related to other dental anomalies such as multiple impacted teeth and congenitally missing teeth [3-5]. The Problems associated with amelogenesis imperfecta include an increased incidence of plaque deposits and consequent gingivitis, dental hypersensitivity, loss of vertical dimension due to a rapid wear of the dentition, poor esthetic appearance and the potential need for lifelong, extensive restorative care. These in addition, psychological problems need to be considered, mainly related to the difficulties that these patients report in their social interaction due to the unesthetic appearance of their smile. The challenges faced in managing a young patient with Amelogenesis Imperfecta (AI) are numerous and prevention is the most important tool in any clinician's armamentarium [6,7]. It is particularly important when teeth are known to be vulnerable. This report describes the preventive strategy of a case with hypoplastic amelogenesis imperfecta in a young patient who has 4 years old, with 18 months of follow-up.

Case Presentation

A 4-year-old female patient presented at to the Department of Pediatric Dentistry and Prevention of Rabat University. She was referred with an unsightly appearance affecting all the primary teeth and dental sensitivity (Figure 1). The family history was negative for the disease. Past medical history was reviewed and there was no remarkable report. The initial clinical examination did not reveal any extra-oral pathology signs. Dental examination revealed poor esthetics due to a yellowish aspect of all maxillary and mandibular primary teeth with no dental caries. Radiographic examination showed generalized loss of enamel with high pulp horns and no periodontal defects were present. The occlusal function showed several characteristics typical of a first dental class. On evaluation of the vertical dimension of occlusion using phonetics, interocclusal measurements and facial appearance as guides, it was determined that the vertical dimension of occlusion was maintained. After doing a professional Tooth brushing with fluoride toothpaste, dental discoloration still existed in all maxillary and mandibular primary teeth. Based on the clinical and radiographic findings,

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*Correspondence:

Ariri El Mehdi, Department of Pediatric Dentistry, University of Mohamed V Rabat, Mohammed V Military Hospital of Rabat, Morocco, Tel: +212-668607363;

E-mail: haririelmehdi@gmail.com

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Figure 1: A 4-year-old patient affected by hypo mineralized AI. Clinical examination revealed pain during brushing and hot and cold sensitivity. Enamel was yellow, easily chipping, with loss of dental structure.

a diagnosis of hypoplastic amelogenesis imperfecta was made. After explaining to the patient and her mother this enamel defects, we opted for a preventive strategy to reduce sensitivity, to protect dental structure, to prevent caries, occlusal disorder and periodontal disease. The first aim of our preventive strategy was to confirm diagnosis of this enamel defects. After accomplishing a professional Tooth brushing with fluoride toothpaste to avoid the confusion with an extrinsic-tint, we studied clinical and radiographic signs and we concluded a diagnosis of hypoplastic amelogenesis imperfecta. In order to reduce dental sensitivity, to prevent caries and periodontal disease and to protect dental structure, we used topical fluoride in the form of a varnish to foster the protection of the smooth surfaces of primary teeth and the remineralization of the first carious lesions. It was applied twice every month during our preventive strategy. The enamel alterations, which are characteristic of the disease and observed at maxillary and mandibular anterior teeth (51-52-61-62-71-72-81-82) were protected using a direct composite resin. The main goals of this stage were represented by the need to protect the occlusal dental surfaces from the abrasion and provide a satisfactory esthetic result. Dental abrasions particularly in posterior teeth, when if not protected may determine the loss of the vertical dimension as well as a diffuse and marked dentinal hypersensitivity. To prevent the occlusal disorder, we used Performed Metal Crowns (PMCs) restorations on the occlusal surfaces of maxillary and mandibular primary molars. After assessing the tooth shape, contact points/areas and the occlusion, orthodontic separators were used to create space for fitting a performed metal crown at the mesial and distal contacts. The patient was seen 3 days later for removal of the separator and the inter-proximal area of the gingiva was inspected. Different sizes of crowns was selected until we find one which covers all the cusps with no tooth preparation and approached the contact points with a slight feeling of “spring back”. Then, we loaded the crown generously with a glass ionomer luting cement from the base upwards, fitting the crown and we did a first stage seating. After wiping the excess cement away, we checked fit and we did second stage seating. Then we removed excess cement, flossed between the contacts and checked occlusion and discharge. Anecdotally, preformed metal crown placed high in occlusion do not cause problems for children and avoidance of interferences of >1.5 mm are recommended. It has been suggested that interferences <1 mm are well tolerated with dento-alveolar compensation occurring within a few weeks [8,9]. In summary, young children appear to have an adaptable masticatory system in which changes occur quickly. After finishing and polishing, the crowns restorations were checked radiographically. At last, in order

to maintain all results and a high oral health status, the patient was instructed to use Tooth brushing with fluoride toothpaste three times a day (250 to 550 ppmF) and we used topical fluoride in the form of a varnish (10%) twice yearly during our post-preventive strategy [10], and the mother was informed about the importance of maintaining meticulous oral hygiene and regularly returning for clinical and radiograph monitoring. The patient was seen every 3 months and she was asymptomatic. After 18 months, clinical and radiographic signs were still stable with a good aesthetics and periodontal health.

Discussion

The term Amelogenesis Imperfecta (AI) refers to a heterogeneous group of genetic disorders characterized by defects in enamel formation of the teeth in the absence of any generalized or systemic diseases. It can affect all or some teeth in deciduous and/or permanent dentition. Recent studies have identified five genes as being responsible for AI by mutation or altered expression: AMEL (amelogenin), ENAM (enamelin), MMP20 (matrix metalloproteinase-20), KLK4 (Kallikrein-4), and FAM83H4 [5]. A diagnosis of amelogenesis imperfecta can present challenges for patient and clinician alike. This can be further complicated in a child due to cooperation and the ever-changing dentition. As with all children, initial experience of the dental profession plays a part in their cooperation for the future. Children with AI often require extensive dental treatment throughout their lives, therefore it is essential to ensure the initial experiences are positive [7]. However, this can be hard to achieve when a child may be experiencing sensitivity when brushing their teeth and eating/drinking cold things. Seeing a child as early as possible will enable the clinician to build up a rapport with the child and parents/carers.

In our case, the patient was diagnosed with hypoplastic amelogenesis imperfecta and an early preventive strategy was realized to reduce sensitivity, to protect dental structure, to prevent caries,

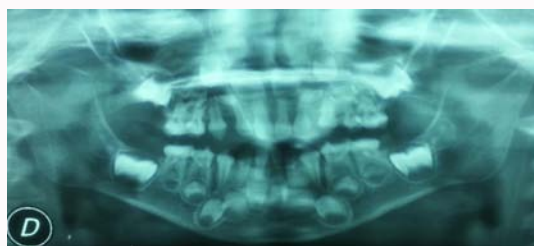


Figure 2: Panoramic X-ray showing a thin layer of enamel on all temporary teeth.



Figure 3: Oral management was realized under local anesthesia through four visits. Stainless steel pediatric crowns were realized on primary molars and direct composite restorations were done in anterior teeth with 18 months follow up.

occlusal disorder and periodontal disease. Prevention is the most important tool in any clinician's armamentarium. It is particularly important when teeth are known to be vulnerable, as is the case in AI. Oral hygiene can be particularly difficult for this group of patients due to sensitivity when brushing. Advising the use of warm water for tooth brushing will go some way to relieving symptoms when rinsing. Regular use of fluoride mouthwashes can also help to reduce sensitivity and prevent caries in children old enough to be trusted not to swallow it. However, the discomfort associated with brushing cannot always be overcome and cases where there are significant plaque and calculus deposits are not unusual. In addition, psychological outcomes need to be considered, mainly related to the difficulties that these patients report in their social interaction due to the unesthetic appearance of their smile.

Clinical management may vary according to the type and severity of the disease, the age of the patients, as well as their socioeconomic status. For these reasons, a wide range of possible and different treatments for teeth restoration of patients affected by AI has been reported in literature. With regard to the possible treatments related to the clinical case here reported, the therapies vary from a conservative to a prosthetic approach [11]. In particular, the conservative treatment of the affected teeth may be used during childhood, as transitional treatment, but cannot be used for a longer period because it does not satisfy esthetic needs and it would not prevent the abrasion of posterior occlusal surfaces over time with consequent loss of the vertical dimension. The preventive strategy described in the present case report is reasonably simple. In the anterior teeth, direct composites have the advantage that they can be started as soon as there is sufficient tooth erupted and added to as further eruption occurs. However, the child and parent should be warned that with continuing eruption and gingival maturation, the margins of any restoration will become visible and additional treatment will be required, at intervals, to maintain good aesthetics [12]. In the posterior regions, Preformed Metal Crowns (PMCs) restorations were placed on the occlusal surfaces of primary molars. Orthodontic separators between molars will assist placement of crowns [8]. No tooth preparation is required to undertake this treatment, assuming the dentition is caries free. In an anxious or young child compromise may be necessary, in which case; second deciduous molars are considered to be higher priority to maintain the occlusion and placing aesthetic veneers on anterior teeth may be used as acclimatization and help to motivate the child and family [13-21]. However, there is always a risk that the child may not return as their primary complaint may be addressing the aesthetics.

As with all dental procedure, following-up is of critical importance. The patient should be seen at 3, 6 and 12 months and yearly at mixed and permanent dentition. Esthetics and periodontal status should be confirmed both clinically and radiographically at these control visits. In this case after 18 months, clinical and X-ray examinations showed satisfactory esthetic and functional outcomes

and a good state of periodontal health (Figure 2 and 3).

Conclusion

Developmental disturbances such as amelogenesis imperfecta not only impair the function and aesthetics but also have a profound psychological effect on the patient's self-esteem and confidence especially in childhood. An early preventive strategy is essential to reduce sensitivity, to protect dental structure, to prevent caries, occlusal disorder and periodontal disease.

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