

Amelogenesis Imperfecta : A Case Report

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Abstract

Amelogenesis imperfecta is a hereditary condition where enamel formation is disturbed resulting in defects in mineralization or matrix formation. Restoration of the dentition poses great difficulties especially when all the teeth are severely affected. Treatment aims to relieve pain or tooth sensitivity, to preserve as much tooth tissue as possible while preventing further tooth loss, to maintain masticatory function and last but not least, to improve the appearance as this has great psychological impact on the patient's confidence. This article describes treatment of a 17 year old boy who presented with a severe form of amelogenesis imperfecta.

Key Words: Amelogenesis Imperfecta (AI), Enamel, Esthetics, Function, Genetic.

Introduction

Amelogenesis Imperfecta (AI) is a collective term for a number of conditions resulting from abnormal enamel formation. Many cases are inherited, either as autosomal dominant, autosomal recessive or as an X-linked trait. There have been a number of classifications, based primarily on the phenotype.¹⁻³ However, a recent classification based on the molecular basis for the observed enamel phenotype may help clarify the commonly seen overlap in phenotypes. According to this classification, AI is a group of conditions, genetic in origin, which affect the structure and clinical appearance of the enamel of all or nearly all the teeth, and which may be associated with morphologic or biochemical changes elsewhere in the body.⁴ This enamel anomaly affects both primary and permanent dentition.^{4,7} The incidence of AI has been reported as varying between 1:700 and 1:16 000, depending on the diagnostic criteria and the population studied.^{2,7,9} The appearance depends on the type of AI, varying from the mild hypomature 'snow-capped' enamel to the more severe hereditary hypoplasia with thin, hard enamel which has a yellow-brown appearance. Dental problems, which depend on the severity of the condition, include sensitive teeth and poor appearance due to tooth loss and staining. If tooth tissue loss is severe there is vertical loss resulting in reduced masticatory function and poor appearance. Pulpal involvement may occur in severe cases, and because the roughness makes cleaning more difficult, gingivitis and periodontitis may develop. As AI affects both primary and permanent dentition, prevention of future dental problems must start from the beginning, by educating the parents and reviewing the patient regularly. Attention must also be paid to the psychological health of the patient.¹⁰

Case Report

The patient was a healthy 17-year-old boy referred to the author's clinic by a general dental practitioner in May 2010. He was complaining of poor appearance, sensitive teeth, and was unable to eat properly. He had a severe form of AI, with absence of all the enamel when first seen (Fig. 1). His primary teeth had also been affected but he was not sure to what extent. Those teeth present were heavily stained, some had deep carious lesions with loss of tooth structure and the exposed dentine was relatively softer than the normal dentine. The teeth were vital, firm, and not tender to percussion. Only some teeth were sensitive to cold. These were vital and radiographs showed narrow pulps and root canals. The periodontal tissues were healthy. All second premolars except 25 and all wisdom teeth (18, 28, 38, 48) were missing. Deciduous tooth (1E) was retained.

His elder brother also suffered from the same type of problem. The elder brother's dental health was even worse than the patient's and he had been essentially treated with extraction and the provision of dentures. Treatment objectives for this patient were-

1. Improvement of esthetics.
2. Prevention of caries and gingivitis.
3. Prevention of further deterioration of the remaining dentition.
4. Management of sensitive teeth.
5. Patient education and motivation.

There were several treatment options: provisional crowns, composite restorations or occlusal splint at a predetermined vertical dimension and then definitive crowns when the patient adapted to the new vertical dimension. The option involving provisional crowns was selected because all the teeth urgently required protection and their appearance was poor.

As such treatment entails a very long time commitment, patient cooperation is essential if these objectives are to be achieved.

Treatment Plan

Treatment was divided into four phases-

Phase 1

1. Oral hygiene instruction and dietary advice.
2. Extraction of 1E and 46.
3. Root Canal Treatment (RCT) of 11, 12, 13, 14, 21, 22, 23, 24, 25, 31, 32, 33, 34, 41, 42, 43, 44, 47 (Fig. 3).

Phase 2

1. Mounting of study models and evaluation of occlusion.
2. Diagnostic waxing up of upper and lower posterior teeth.
3. Discussion of treatment options.
4. Re-organize jaw relationship raising the incisal pin by 4 mm.
5. Anterior occlusal wafer prepared for new

anterior raised occlusion.

6. Teeth 14, 16, 17, 24, 25, 26, 27, 34, 36, 37, 44, 47 prepared and impressions taken for provisional crowns.
7. Temporary bridge from 14 to 17, 24 to 27, 34 to 37 and 44 to 47 prepared and temporary crown cementation at pre-determined vertical dimension (Fig. 2).
8. Provisional acrylic crown cementation then regular patient review until new occlusal scheme problem free.

Phase 3

1. Three months after adaptation to the new occlusal vertical dimension (OVD), the new occlusal relationship was recorded using new study models and face-bow records.
2. Diagnostic waxing up of the upper and lower anterior teeth was done
3. Lower posterior teeth were replaced with metal crowns.
4. Upper posterior teeth were replaced with metal crowns.
5. Upper anterior teeth were prepared for metal ceramic crowns.
6. This was followed by provision of lower anterior metal ceramic crowns (Fig. 4, 5).

Phase 4

1. Oral hygiene reinforcement.
2. Dietary advice.
3. Regular dental checkups for caries and periodontal diseases.
4. Regular review of occlusion.

Discussion

Amelogenesis imperfecta is an inherited disorder, ranging from a defect in enamel formation to coexistence of other medical disorders.¹¹⁻¹³ As both the primary and permanent dentition are affected, preventive measures should be started, even before the teeth erupt.

In this case, the patient presented with severe tooth wear due to inherited abnormal enamel development. The poor appearance was due not only to the innate color of the teeth, but also to increased staining, chipping and attrition of the teeth. Discomfort was due to wide areas of exposed dentine and pulpal involvement of some teeth.

Treatment options for such a case include the following-

1. Extraction of retained deciduous tooth and a non restorable lower molar. Root Canal Treatment (RCT) of teeth which are pulpally involved. Provisional crowns made to the length and shape required for permanent posterior crowns. Definitive crowns made when the patient has adapted to the new vertical dimension.
2. Provisional crowns made to the length and shape required for permanent anterior crowns. Posterior teeth restored

with composite to the new vertical dimension. Definitive crowns made when the patient has adapted to the new vertical dimension. Extraction of the non-restorable tooth.

3. Composite restorations of all teeth to the new vertical dimension. Definitive crowns when the patient has adapted to the new dimension. Extraction of the non-restorable tooth.
4. Occlusal splint at the new vertical dimension, then definitive crowns made when the patient has adapted to the new occlusal relationship. Extraction of the non-restorable tooth.

The first option was selected because all his teeth urgently required protection and the appearance was poor. Provisional crowns also provided valuable information about future restorations. As treatment required a very long time commitment, cooperation was stressed right from the beginning.

Re-organization of the occlusion was undertaken because all the teeth required restoration and the OVD had to be altered.¹⁴ Evaluation of the patient's ability to adapt to the new OVD and establishment of a stable retruded contact position (RCP) must be acquired prior to the definitive treatment. Either an occlusal splint or provisional restorations like provisional crowns and intermediate composite can be used. The latter option was preferred as the need to convert the splint into temporary restorations could be omitted; at the same time the teeth were protected, masticatory ability improved. Stabilization of the jaw with posterior provisional crowns to the new vertical dimension was carried out initially. Excursive interferences were adjusted and definitive treatment could be started when a stable RCP was acquired with no change occurring between two successive appointments and the patient adapting to the new OVD. The time needed for this ranges from a couple of weeks to several months¹⁵. In order to analyze the appearance and the patient's ability to adapt to the new OVD, definitive treatment should not begin for at least one more month.^{16,17} In this case, adaptation took longer than 1 month. Owing to clinical examinations, definitive treatment was initiated 2

months later. After the clinical adaptation to new OVD, the posterior teeth were definitively restored. Shallow anterior guidance was planned to reduce stresses on the incisors, resulting in shallower cusps in the posterior crowns.

Removal of caries had to be done very carefully as the defective dentine was softer than normal dentine. Because the defective dentine involved a large area and in many areas was quite subgingival, there was a problem with obtaining all the preparations in one impression. Electrosurgery was considered initially, but later abandoned after minimal success and because most of the teeth were involved. It would be very traumatic and uncomfortable for the patient.

The morphology of the anterior crowns was constructed according to the racial characteristics of the patient. Owing to the highly discolored dentine a more opaque ceramic material had to be used.

Due to the poor financial condition of the patient only anterior teeth (14 to 24 and 34 to 44) were made of metal ceramic. Posterior teeth were made up of metal. Although the crowns could have been improved further, he was very happy to accept them; thus no further alteration was needed.

Construction of 28 crowns demanded very strong laboratory support, so good laboratory backup was important.

During the whole process oral hygiene and dietary advice were reinforced so that future periodontal and caries problems could be prevented. This periodic review of the patient's oral hygiene and periodontal health must be stressed and maintained in order to achieve long term success. Psychological health is also an important issue in AI patients.¹⁰ Although the appearance of the restorations could have been better, the patient was very happy with the outcome (Fig. 4) and mastication was satisfactory.

Besides achieving the objectives set out at the beginning of the treatment, psychologically he became a happier and more confident person.

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Legends

- Fig. 1: Pre-Operative- (A) Frontal view; (B) Maxillary arch; (C) Mandibular arch.
 Fig. 2: Posterior Temporary Bridge- (A) Maxillary arch; (B) Right Mandibular arch; (C) Left Mandibular arch.
 Fig. 3: (A) Pre-Operative OPG; (B) Post-Operative OPG.
 Fig. 4: Post-Operative Extra Oral view.
 Fig. 5: Post-Operative Intra oral view- (A) Frontal view; (B) Maxillary arch; (C) Mandibular arch; (D) Right occlusal; (E) Left occlusal.

