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Case Report

Amelogenesis imperfecta – A case series

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ABSTRACT:
Amelogenesis Imperfecta (AI) represents a group of developmental conditions, genomic in origin, affecting deposition (hypo-plastic), calcification (hypo-calcification), or maturation (hypomaturation) of dental enamel. In general, it affects all or nearly all of the teeth in both the primary and permanent dentitions leading to functional as well as aesthetic inadequacies. This case report discusses the management of hypo-plastic AI through direct composite veneering in three sisters aged 20yrs, 17yrs, 15yrs. This approach enhanced aesthetics as well as boosted their self- esteem.
Keywords: Amelogenesis Imperfecta, enamel hypoplasia, hypoplastic teeth, dental care

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INTRODUCTION

Amelogenesis imperfecta (AI) is defined as a group of hereditary developmental defects of the dental enamel affecting both primary and permanent dentition.[(1)]It exhibits quantitative or qualitative enamel abnormalities in the absence of any systemic manifestations. Depending on the population studied and the criteria used for the diagnosis, the prevalence of this disorder varies from1:700 to 1:16,000.(2)

On the basis of clinical, radiographic and histopathologic findings, it can be generalised into threetypes- (3)

- Hypo-plastic (deficiency in organic matrix deposition)
- Hypo-mineralisation (deficiency in organic matrix mineralisation)
- Hypo-maturation (defect in enamel crystal maturation)

Another variant of AI is the hypo-plastic, hypo-maturation type associated with taurodontism. Witkop and Sauk further classified the four types of AI according to the specific features seen clinically **(Table no I**).(3)

Clinically, hypo-plastic form of AI is represented by:(4)

- Thinner enamel, having adequate hardness and mineralisation.
- Smooth and glossy crown or rough having pits and ridges
- opaque white to yellowish-brown discolouration
- Loss of proximal teeth contact with flattening of the occlusal surfaces due to attrition

Hypo-calcified AI is represented by:(4)

- Normal enamel thickness
- Softer consistency which is easily worn off
- Appears chalky to dark brown in colour.

Hypo-maturation variant is represented by:(4)

- Normal enamel thickness
- Softer enamel consistency
- Yellow/ brown mottled coloured with snow caps
- Enamel usually harder than the hypo calcified variant.

Radiographically, a thin radiopaque layer of enamel can be seen in hypo-plastic AI. Unlike the hypo-plastic type where the radiodensity of the enamel appears normal, both hypo-calcified AI and hypo-maturation AI show lower radiodensity. The radiodensity of enamel is less than that of dentine in hypo-calcified form and similar to dentin in hypo-maturation form.(4)

In this case report, direct composite veneering technique was used for the management of AI keeping in mind the severity of the disease as well as patient

affordability. It was decided to place direct composite veneers on the facial/ buccal surfaces of the teeth which fall into the smile line, that is upto the 2nd molar of both upper and lower quadrants.

Type I hypoplastic
IA hypoplastic, pitted autosomal dominant
IB hypoplastic, local autosomal dominant
IC hypoplastic, local autosomal recessive
ID hypoplastic, smooth autosomal dominant
IE hypoplastic, smooth X-linked dominant
IF hypoplastic, rough autosomal dominant
IG enamel agenesis, autosomal recessive
Type II hypomaturation
IIA hypomaturation, pigmented autosomal recessive
IIB hypomaturation
IIC snow capped teeth, X-linked
IID autosomal dominant?
Type III hypocalcification
IIA autosomal dominant
IIB autosomal recessive
Type IV hypomaturation — hypoplastic with taurodontism
IVA hypomaturation — hypoplastic with taurodontism, autosomal dominant
IVB hypoplastic — hypomaturation with taurodontism, autosomal dominant

TABLE NO. I (Witkop and Sauk classification of AI)

CASE REPORT

This clinical report outlines the management of three sisters (Case 1- 20 years, Case 2 -17 years, Case 3 -15 years) who reported with chief complain of discoloured teeth. They also expressed extreme dissatisfaction with their appearance while smiling. History revealed that their deciduous teeth were also similarly discoloured. Family history revealed that their mother and maternal grand father were also suffering from the same condition. Other than this, their past medical history was non-contributory.

PROCEDURE

The facial/ buccal surfaces were prepared with chamfer finish line. After cleaning the prepared tooth with pumice and bristle brush, acid etching was done using 37% phosphoric acid (Ivoclar N- Etch) for 15 secs. The acid was rinsed off with water spray and the tooth surface was dried. Dentine bonding agent (Tetric N-Bond, Ivoclar) was applied and light cured. After this, an opaque layer of composite resin was applied to mask the underlying dark coloured structures which was followed by application of a translucent layer of restorative composite resin mimicking the enamel (Tetric N- Ceram, Ivoclar). And finally the direct composite veneers were polished using discs (3M ESPE Composite polishing discs). ***(Figure 1)***

FIGURE 1 (DIRECT COMPOSITE VENEERING TECHNIQUE)

CASE 1

Clinical examination showed that all anterior and posterior teeth presented with yellow-dark brownish discolouration with snow capped 11, 21. The surface of the crown was rough with hard consistency and no signs of chipping. Anterior open bite was another positive finding. Other intra- oral findings included grossly decayed 16 and disto-occlusal caries in relation to 46. There was no tender on percussion and patient was not willing to undergo any endodontic treatment.

Panoramic radiograph showed a thin radiopaque layer of enamel with greater radiodensity ascompared to the underlying dentine. Final diagnosis of diffuse rough hypo-plastic amelogenesis imperfecta was made.

The pre-op and post-op photos are shown in ***figure 2a*** and ***figure 2b*** and panoramic radiograph in ***figure 2c***.

FIGURE 2.1 (PRE-OP AND POST-OP)

FIGURE 2.2 (POST-OP)

FIGURE 2.3 (OPG)

CASE 1

CASE 2

All anterior and posterior teeth presented with yellow-brownish discolouration. Localised pitting was seen in relation to 12, 11, 21, 22. The crown surface was rough with hard consistency and no signs of chipping. There was anterior open bite along with reduction of height of the clinical crown in upper anteriors.

Panoramic radiograph showed a thin radiopaque layer of enamel with greater radiodensity ascompared to the underlying dentine. Final diagnosis of localised pitted hypo-plastic amelogenesis imperfect was made.

The pre-op and post-op photos are shown in ***figure 3a*** and ***figure 3b*** and panoramic radiograph in ***figure 3c***.

FIGURE 3.1 (PRE-OP AND POST-OP)

FIGURE 3.2 (POST-OP)

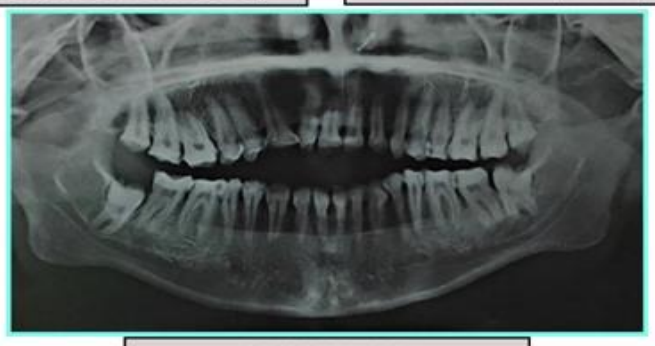

FIGURE 3.3 (OPG)

CASE 2

CASE 3

All anterior and posterior teeth presented with diffuse yellow- brownish discolouration. The surface of the crown was smooth with hard consistency and no signs of enamel chipping.

Panoramic radiograph showed a thin radiopaque layer of enamel with greater radiodensity ascompared to the underlying dentine. Final diagnosis of diffuse smooth hypoplastic amelogenesis imperfect was made.

The pre-op and post-op photos are shown in ***figure 4a*** and ***figure 4b*** and panoramic radiograph in ***figure 4c.***

FIGURE 4.1 (PRE-OP AND POST-OP)

FIGURE 4.2 (POST-OP)

FIGURE 4.3 (OPG)

CASE 3

DISCUSSION

Many treatment modalities have been proposed for management of AI depending upon the patient's age, severity of the condition, socio-economic status and other intra oral findings. This includes microabrasion, composite veneers, porcelain laminates, metal ceramic or all ceramic crown.

According to ***Robinson et al***,(5) and ***Turkun LS et al***,(6) composite resin veneers are considered to beone of the best ways to preserve tooth structure. However, due to poorly mineralised & friable enamel seen in amelogenesis imperpecta, the bond between the resin and enamel surface may not bevery strong.

A case report by ***Venezie RD et al***,*(7)* indicated that the bond is weaker when compared to normal enamel. But with newer technologies, it is now possible to provide treatment to an acceptable level to meet the aesthetic and functional demands of the patient with AI.(8)

All zirconia crowns or 3/4 zirconia cores can also be used as it requires minimal occlusal reduction of 0.5mm. These are relatively less prone to fracture when compared to all ceramic restorations but at the same time they are less economic.

Moreover, as per study done by ***Meijering AC et al(10)***, patient satisfaction success rate of 80-100% was found for veneer restorations. Crown restorations are relatively more invasive when compared to veneers and it is not justified to cut the lingual portions of the crown if the patient is not having any discomfort other than the aesthetics.

It is critical to identify at early stages and take further necessary precautions so as to prevent functional and social trauma in children and young adults. There has been mention of multiple extractions in AI case

followed by rehabilitation with complete denture prosthetics. Adolescents subjected to such harsh treatment have a negative psychological impact on their self esteem.

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