

MANAGEMENT OF AMELOGENESIS IMPERFECTA
- AN ESTHETIC REHABILITATION

ABSTRACT

Amelogenesis imperfecta (AI) is a diverse collection of inherited diseases that exhibit quantitative or qualitative tooth enamel defects in the absence of systemic manifestations. Also known by varied names such as Hereditary enamel dysplasia, Hereditary brown enamel, Hereditary brown opalescent teeth, this defect is entirely ectodermal, since mesodermal components of the teeth are basically normal. The AI trait can be transmitted by either autosomal dominant, autosomal recessive, or X-linked modes of inheritance. Genes implicated in autosomal forms are genes encoding enamel matrix proteins, namely: enamelin and ameloblastin, tuftelin, MMP-20 and kallikrein – 4. This case report deals with the treatment of Amelogenesis Imperfecta in 20 year old patient.

Key words: Full Mouth rehabilitation, enamel, dental, genetic, inherited Amelogenesis Imperfecta (AI)

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Date of Submission : 2/1/16

Date of Acceptance : 20/1/16

INTRODUCTION

Amelogenesis Imperfecta represents a group of conditions, genomic in origin affecting the structure and clinical appearance of the enamel of all or nearly all the teeth which may be associated with morphologic or biochemical changes elsewhere in the body¹. The term, AI was introduced by Weinmann and co-workers. It is a hereditary defect of enamel affecting both the primary and permanent

dentition² with mutations in five genes—AMEL, ENAM, MMP20, KLK4, and FAM83H³. Transmission of the gene is autosomal dominant, recessive or X linked with an prevalence of 1:700 to 1:14,000⁴.

It is an exclusive ectodermal disturbance, related to alterations in the organic enamel matrix which causes white flecks, narrow horizontal bands, lines of pits, grooves, and discoloration of the teeth varying from yellow to dark brown

prone to rapid wear and breakage^{5,6}. With generalized compromise of enamel, there is loss of vertical dimension as well as lack of interproximal contacts, resulting in food lodgement and problems associated with it. In most cases the esthetic disability is striking³. AI has been reported to be associated with other dental conditions such as Taurodontism, pulpal calcifications, multiple tooth impactions, oligodontia, hyper-cementosis, distorted roots, aberrant dentin formation and marked follicular hyperplasia and malocclusion^{2,7,8,9}.

According to Witkop, AI can be classified into four main types: (1) hypoplastic; (2) hypomaturational; (3) hypocalcified; and (4) hypomaturational-hypoplastic with taurodontism². Each main type of AI may be further divided into several subtypes, depending on clinical features and inheritance patterns. Other forms of enamel dysmineralization will exhibit a pattern based upon the time of insult, thus affecting the enamel forming at the time. In contrast, AI will affect all teeth similarly and can have a familial history. Fluorosis can mimic AI, but usually the teeth are not affected uniformly, often sparing the premolars and second permanent molars¹⁰.

The condition is a social hindrance to the patient and treating such individuals brings drastic transformation in their psychosocial behaviour and approach towards life¹⁰. The treatment of patients with AI should be planned taking both the clinical and the emotional demands into consideration. Therefore, the planning of esthetic restorations on the anterior teeth may not only preserve the tooth structure but also eliminate the adverse social impact of their teeth¹¹.

CASE REPORT

A 20-year-old female patient was referred to the Department of Conservative Dentistry and Endodontics, Genesis Institute

Of Dental Sciences and Research, Ferozpur with a chief complaint of unesthetic smile due to discoloured & rough front teeth. The family history revealed that her parents had a consanguineous marriage (1st cousins) and there was no history of any other members of the family having a similar discoloration of the teeth. No significant medical history was recorded. Clinical examination of the patient revealed that there was thin enamel with dark brown pigmentation on the facial surface, enamel pit defects and light brown bands on the incisal third of the maxillary anterior teeth (FIG 1).

A treatment plan was developed with the following aims: to reduce the reported sensitivity of the teeth and to improve the esthetics. Esthetic rehabilitation with All Ceramic Crowns was planned but due to financial issues PFM crowns were desired by the patient. Root canal treatment was performed w.r.t 11,12,21,22 as there was insufficient tooth structure and further reduction of tooth would lead to pulpal exposure. The tooth preparation was done for porcelain fused to metal crowns on left and right maxillary incisors. The prepared teeth were then restored with temporary heat cure acrylic crowns for 3 months for evaluation of esthetic satisfaction of the patient. After 3 months, the temporary crowns were replaced by PFM crowns in the anterior teeth (FIG 2).

DISCUSSION

The term "Amelogenesis Imperfecta" describes a diverse group of hereditary conditions primarily affecting the quality and/or quantity of dental enamel. The affected teeth show a soft enamel of normal thickness that chips and wears easily and has a radiodensity similar to that of dentin^{12,13}.

The results of clinical and radiographic evaluations indicated that the patient in the present case had hypomaturational form of AI. All the teeth are misshapen, and spotted. The



FIG 1

The exposed dentin was hypersensitive. From history & clinical examination it was diagnosed as – Amelogenesis imperfecta of the Hypomature type. Patient had not undergone any restorative treatment for esthetics before and now desired to improve the appearance of her anterior teeth.



FIG 2

Oral hygiene instructions were given and the patient was recalled after 1 month for follow up. A follow up of 24 months showed that esthetic expectations of the patient were completely fulfilled. Thus, with completion of the treatment, we could not only conserve the remaining tooth structure but could also improve esthetics of the patient.

insufficiency of the enamel makes the teeth extremely sensitive to contact and thermal stimuli. These problems combine to make early diagnosis essential and immediate treatment a necessity, even for the youngest patients^{14,15}. Currently, the cases of AI are restored with adhesive restorative techniques, overdentures, fixed partial dentures, full porcelain crowns, porcelain fused-to metal crowns and inlay/onlay restorations constitute the contemporary treatment modalities. However, full porcelain restorations are becoming increasingly popular, because of their inheriting esthetics, excellent biocompatibility and improved physical properties. Moreover, the advances in the field of esthetic dentistry, especially in bonding to dentin, help practitioners to restore function and esthetics to an acceptable level^{16,17,18}. But, marginal adaptation and bonding problems have been pointed out as disadvantages of laminate veneers¹⁹.

In the present case, metal-reinforced porcelain restorations were preferred considering patient's socioeconomic status and to redouble the mechanical durability, recover esthetics and protect the residual dentin.

It is very important that the treatment plan is such formulated that it prevents the development of psychological problems arising from the appearance of teeth affected by Amelogenesis Imperfecta²⁰. Following the treatment, the psychological transformation of this 20 year old girl was spectacular as was evident from her confident smile during the follow up visits.

CONCLUSION

This clinical report describes the oral rehabilitation of a 20 year old girl patient affected by Hypomature Amelogenesis Imperfecta. The treatment plan for cases of AI is related to many factors : the age of the patient, the socioeconomic status of the patient, the type and severity of the disorder, and the intraoral situation at the time the treatment is planned. Thus, rehabilitation in this case included anterior PFM crowns to eliminate tooth sensitivity, improve the esthetics and restore function.

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