ABSTRACT
Management of a patient with amelogenesis imperfecta is a challenge for the clinician. The treatment options vary considerably depending on several factors, such as the age of the patient, socioeconomic status, periodontal condition, loss of tooth structure, severity of the disorder, and most importantly the patient cooperation. Complete oral rehabilitation may involve adhesive porcelain veneers, dentine bonded crowns and eventually full coverage gold, porcelain bonded to metal or all ceramic crowns.

Keywords: Amelogenesis imperfecta, Amelogenin, Enamelin, Enamelysis, Kallikrein.


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INTRODUCTION
Amelogenesis imperfecta is a developmental disturbance that interferes with normal enamel formation in the absence of a systemic disorder. In general, it affects all or nearly all of the teeth in both the primary and permanent dentitions.1 Amelogenesis imperfecta may be inherited by X-linked or sporadic inheritance. The different clinical manifestations of amelogenesis imperfecta have a specific gene anomaly associated with each phenotype. Specific mutations proven to cause amelogenesis imperfecta include: amelogenin (AMELX), enamelin (ENAM), kallikrein 4 (KLK4), enamelysis (MMP-20) and FAM83H.1 Amelogenesis is a two-staged process where a protein rich matrix is initially laid down during the secretary phase, followed by the mineralization phase where the proteins are replaced by hydroxyapatite crystals. This results in the highly mineralized enamel structure.2 Amelogenesis imperfecta has been classified into four types: hypoplastic, hypomaturation, hypocalcified, and hypomaturation-hypoplastic with taurodontism. Each type has subtypes differentiated by mode of inheritance.4-9

Enamel hypoplasia results in a decreased quantitative enamel formation. The enamel in hypocalcification appears normal but poorly mineralized while hypomaturation results in an abnormal mineralization in the final stages of tooth formation. The most common form, the hypoplastic type, is deficient in normal enamel. The crowns of the teeth appear blanched, snow-capped, yellow-brown, pitted or grooved. Radiographic examination usually shows a full complement of teeth, but the crowns of the teeth either have very thin enamel or lack enamel completely. Dental radiography in form of orthopantomogram (OPG) and full mouth intraoral radiographs plays a vital role in diagnosing the difference in density of enamel in amelogenesis imperfecta patients and normal patients along with dentin thickness, pulp canal and root length.3 This case report represents the complete oral rehabilitation of a patient with amelogenesis imperfecta.

CASE REPORT
A 16-year-old male patient (Fig. 1) reported to the Department of Pedodontics and Preventive Dentistry, Dasmesh Institute of Research and Dental Sciences, Faridkot, with a chief complaint of discolored teeth since childhood (Figs 2A to C) and associated intermittent pain in right and left lower back teeth since 3 to 4 months. Pain was dull and nonradiating. Patient had undergone a dental
treatment from local dentist wrt same teeth few months back. Patient resided in a nonfluoridated area since birth. His natal, postnatal and medical histories were not significant. His mother and maternal aunt also suffered from the same condition. Patient had similar appearance of teeth in primary dentition.

On intraoral examination, it was found that whole permanent dentition exhibited yellowish brown discoloration without superficial chipping away of enamel. 36, 46 and 47 were restored by previous dentist. Radiographic examination revealed inappropriate root canal treatment with overhanging margins of restorations wrt same teeth (Fig. 3). Mesioangular impaction wrt 48 was present. Dental caries wrt 35, 37 and 45 were present. A provisional diagnosis of hypoplastic amelogenesis imperfecta was proposed.

The complete oral rehabilitation included oral prophylaxis, topical fluoride application restorations wrt 35, 37, 45, composite veneering of all maxillary and mandibular anterior teeth (Figs 4 and 5). Retreatment of 36, 46, 47 was done as patient had pain in previously root canal treated teeth (Fig. 6). Patient again reported with
sensitivity and pain wrt 35, 37 and 47, so root canal treatment was also carried out in the respected teeth. Extraction of impacted mesioangular 48 was done. Esthetic rehabilitation of 35, 36, 45 and 46 was done with porcelain fused to metal crowns (Fig. 5). Follow-up revealed a healthy and a satisfied patient after the treatment (Fig. 7).

**DISCUSSION**

Reversible and noninvasive treatment with composite resin should be considered before the more destructive treatment options. The use of composite resins allows restoration of esthetics, which is most important to the patient while preserving tooth tissue. Clinicians often avoid using composite resins, as they are susceptible to staining and technique sensitive. Staining can be effectively managed by regular polishing of the restorations. If necessary the surface layer can be removed and the restoration with a new surface layer without causing further damage to the underlying tooth structure. If the composites fracture or chip they can also be repaired easily without the need for removing the whole restoration. Composite resins should, therefore, be considered as the initial restorative material of choice for all patients, especially when the patient is in their late teens and early twenties, as a medium term option. When they start to repeatedly fail or the maintenance burden becomes too great the treatment could progress to more invasive techniques. This may involve adhesive porcelain veneers, dentin bonded crowns and eventually full coverage gold, porcelain bonded to metal or all ceramic crowns.

**CONCLUSION**

Rehabilitation of patients with amelogenesis imperfecta requires careful planning with the most important factor to consider being the age and cooperation of the patient. Management of these patients through childhood and the early teens is mainly focused around counseling, prevention and preservation of the deciduous, mixed and adult dentition.

**REFERENCES**