CASE REPORT

Interdisciplinary Treatment of a Patient with Amelogenesis Imperfecta

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Amelogenesis imperfecta (AI) is a clinically and genetically heterogeneous group of hereditary disorders primarily affecting enamel formation by ameloblasts. Different manifestations of AI—hypoplastic, hypomineralized, or hypomature enamel—can coexist in the same patient and even in the same tooth.1 Although most of the 50% of AI patients who have malocclusions also show skeletal anterior open bites, the significance of this association has not been elucidated.2 Other cases involve deep bites secondary to crumbling dental tissue. The unesthetic appearance of AI patients’ teeth often erodes their self-esteem and disrupts their social lives.3

Successful management of AI requires early recognition, preventive counseling, emotional support, and individualized restorative care adapted to the phases of tooth development. Although some dentists prefer to delay definitive rehabilitation until eruption of the permanent dentition is complete, the potential psychological impact of this disorder during childhood and adolescence requires that esthetic treatment be started as early as possible, at least for the anterior teeth.

Diagnosis and Treatment Planning

A 14-year-old male with severe hypoplastic AI was referred to our orthodontic clinic (Fig. 1). Over the previous 10 years, he had received preventive and restorative treatment in both the primary and permanent dentition. His chief concerns were dental sensitivity...
Fig. 1 14-year-old male patient with amelogenesis imperfecta, bimaxillary protrusion, mandibular crowding, and mild open bite.
Fig. 2 A. Patient after 24 months of treatment, following restoration of upper incisors and canines with porcelain veneers. B. Superimposition of cephalometric tracings before and after orthodontic treatment.
and extreme dissatisfaction with the color of his teeth.

The clinical examination showed a convex facial profile with competent lips and frontal symmetry. The patient had a Class I molar relationship with minor mandibular anterior crowding, a blocked-out upper right canine, and a mild open bite. Tooth shades varied from light to dark yellow. Hypoplastic caries and multiple composite restorations were present, and the cusps of some teeth had crumbled. Metal crowns had been placed on all first molars. Oral hygiene was good. The panoramic x-ray showed a full complement of permanent teeth, including the developing third molars, and normal root anatomy. The lateral cephalogram revealed a severe skeletal Class II malocclusion with a high mandibular plane angle, a hypoplastic mandible, and dentoalveolar bimaxillary protrusion.

The aims of treatment were to restore a normal smile, eliminate dental hypersensitivity, and improve masticatory function using orthodontic, cosmetic, and restorative procedures. Considering the bimaxillary protrusion and arch-space discrepancy, the orthodontic treatment plan involved the extraction of two maxillary and two mandibular premolars, which would allow us to align the dental arches, retract the anterior teeth, and improve the interincisal angle while maintaining the Class I molar and canine relationships. Esthetic and restorative objectives included the placement of porcelain laminate veneers on the anterior teeth and porcelain bridges in the four posterior quadrants.

Treatment Progress

We decided to extract the second premolars in both arches because they were heavily restored. To reduce the risk of bond failures, we considered banding the first premolars after treating the caries in the upper incisors, but the final decision was to bond a conventional multibracketed appliance by means of a light-cured composite. Treatment began with sectional springs to retract the first premolars and then the canines, using a palatal bar as anchorage. The incisors were bonded only after the canines had been completely retracted. Residual spaces were then closed with continuous closing arches.

Although the severe dental discoloration raised some concern that the abnormal enamel might impede proper adhesion, no brackets failed during treatment. Some enamel did fracture off the upper and lower left first premolars, and a restoration on the lower right first premolar partially detached, during debonding and removal of residual adhesive. Throughout orthodontic treatment, the patient’s dentist carefully monitored his progress and administered prophylaxis periodically to prevent caries formation. The patient’s hygiene and cooperation were good.

Treatment Results

Active orthodontic treatment lasted 24 months, followed by retention with upper and lower Essix® retainers. Restorations were placed in two phases, for financial reasons: first on the upper incisors and canines (Fig. 2A) and, one year later, on the remaining affected teeth (Fig. 3). The posterior regions were restored with four porcelain-fused-to-metal bridges from the premolars to the second molars, according to the ideal occlusal plane.

Post-treatment photographs demonstrated a substantial change in the overall health of the dentition and a dramatic improvement in the smile. Cephalometric superimpositions showed a satisfactory occlusion and correction of the bimaxillary protrusion (Fig. 2B). Two years after debonding, the occlusion and the restorations have remained stable. The patient expressed great satisfaction with the interdisciplinary treatment, stating that his dental hypersensitivity had disappeared and, more significant, that he had gained a much better self-image and level of integration with his peers.

Discussion

The process of AI involves interactions of proteins and enzymes that contribute to the synthesis of hydroxyapatite. Mutations in four genes responsible for normal enamel formation have been linked to different forms of AI, which can display autosomal dominant or recessive, sex-linked, and sporadic inheritance patterns. The reported prevalence of AI...
The four major types of AI, based primarily on phenotype—hypoplastic, hypomaturation, hypocalcified, and hypomaturation-hypoplastic with taurodontism—are subdivided into 15 subtypes according to phenotype and mode of inheritance. Hypoplastic forms of AI are characterized by inadequate enamel, which can be very thin and appear pitted and ridged, even though translucency and hardness are normal. Interdental spacing may be present, and the patient may experience thermal or osmotic hypersensitivity. In hypomineralized forms of AI, the enamel is soft, rough, discolored, and highly prone to attrition. In hypomature forms, enamel thickness and hardness are normal, but the surface shows a whitish discoloration. Hypomineralized and hypomature
forms of AI often overlap. The term dysmineralization has been proposed to describe the spectrum of AI enamel defects.\textsuperscript{1}

Diagnosis of AI is based on clinical observation, family history, and pedigree plotting. Panoramic radiographs can reveal unerupted or spontaneously resorbing teeth, and periapical radiography can show the contrast between enamel and dentin in cases of dysmineralization. Differential diagnosis depends on the exclusion of extrinsic environmental factors and developmental disturbances—primarily dental fluorosis, other forms of chronological enamel dysplasia,\textsuperscript{1} molar-incisor hypomineralization,\textsuperscript{9} and regional odontodysplasia.\textsuperscript{10}

Early, individualized care and support are critical to the successful management of AI. The primary dentition must be protected by preformed metal crowns on the posterior teeth and either polycarbonate crowns or composite restorations on the anterior teeth. Upon eruption of the first permanent teeth, the pediatric dentist, restorative dentist, and orthodontist must design an interdisciplinary plan that includes hygiene motivation, restorative procedures to reduce dental hypersensitivity, and placement of preformed crowns when hypoplasia and crumbling predominate. Genetic counseling for affected individuals and their families is mandatory. Severe AI causes great physical and emotional suffering. Beginning in childhood, the patient experiences dental hypersensitivity and the need for numerous restorative procedures to counter progressive deterioration. The esthetic consequences of AI are also emotionally upsetting, especially during childhood and adolescence.\textsuperscript{3,11,12}

Although our patient had undergone numerous restorations since early childhood, treatment of his malocclusion had been delayed. This is a common approach among dentists treating children and adolescents with AI, reflecting concerns about the risk of bonding failures, caries progression, and enamel fractures during orthodontic treatment. Hypoplastic enamel is often well mineralized, however, and generally allows normal bonding with resin-based composites.\textsuperscript{13} Adhesion to hypocalcified and hypomature teeth with little residual normal enamel is often problematic, but no relevant evidence-based recommendations have been made.\textsuperscript{14} Sapir suggests that the tooth surfaces to be bonded should be clinically assessed for the severity of hypomineralization as well as the smoothness, hardness, and color of the surface enamel.\textsuperscript{14} Yellow or brown discolorations tend to be deeper and more porous than those with a creamy-white appearance.\textsuperscript{15,16} Teeth showing milder defects can be bonded with self-etching primers (SEPs) and conventional composite resins. SEPs have been recommended as an alternative to phosphoric acid to reduce the risk of enamel fracture while minimizing etching depth and conditioning-related enamel loss.\textsuperscript{17} They tend to make the bonding procedure more complicated,\textsuperscript{6,18-21} however, and can interfere with self-curing resins.\textsuperscript{22} In cases of excessive enamel loss and where dentin is exposed, the dentin becomes hypermineralized, which can affect adhesion. Longer etching time does not increase the microtensile bond strength of AI-affected primary dentin and is harmful to normal dentin.\textsuperscript{23}

The most serious discolorations can be treated by composite restoration before bonding. To enhance the shear bond strength of composite to hypocalcified enamel, several authors recommend the application of 5% sodium hypochlorite, which eliminates some of the protein from the hydroxyapatite after acid conditioning of the enamel and dentin.\textsuperscript{24,25} Fluoride-releasing, resin-modified glass ionomer cements may be an alternative to composites for use before bonding with SEPs,\textsuperscript{26} but their efficacy with AI enamel has not been evaluated.\textsuperscript{14} Banding can be an option for severely affected teeth, especially canines, although band removal increases the risk of fracture in fragile teeth that are prone to crumbling.

Intensive follow-ups, prophylactic treatment, and dietary consultation are recommended to counteract the possibility of caries progression due to poor oral hygiene during orthodontic treatment.\textsuperscript{27} Enamel fractures are another potential risk, especially in debonding. Our patient experienced enamel fractures and partial detachment of a restoration during appliance removal.

Esthetic treatment of the incisors was particularly challenging in this case because the intense
discoloration of the teeth prevented the use of more transparent veneers. For financial reasons, esthetic and prosthetic treatment had to be performed in two phases. In the first stage, we placed veneers on the upper incisors and canines so the patient could enjoy his new smile and its psychological benefits as soon as possible. Esthetic treatment was completed with the remaining veneers and crowns in the second phase.

Conclusion

Early interdisciplinary treatment of amelogenesis imperfecta at the same time as the malocclusion not only improves the overall health and appearance of the dentition, but greatly enhances the patient’s emotional well-being and quality of life. These advantages make a strong case against unjustified delays in care.

REFERENCES