Amelogenesis Imperfecta - Multidisciplinary Approach: A Case Report*

Introduction

Inherited conditions of the enamel and dentin are determined by genes that regulate functions unique to the highly differentiated and specialized odontogenic cells. Some gene mutations that affect the structure or composition of enamel usually result in alterations that are detectable only in enamel, and the resultant inherited basic defects present at birth are described as amelogenesis imperfecta (AI).

Gene mutations in the enamel matrix produce 1 of the following results: insufficient enamel being formed (hypoplasia), a marked deficit of initial calcification of the organic matrix (hypo-calcification), or a defect in the formulation of crystalline apatite in the enamel rods (hypo-maturation). The term enamel maturation means progression of enamel mineralization. Water and protein are removed from the enamel extra-cellular matrix coincidently with increasing calcium hydroxyapatite crystal formation. Sometimes, mature enamel is formed that contains 96% inorganic substance. Structural and/or regulatory genes associated with enamel code for types I, IV, and VII collagen, amelogenins, enamelin, and enamel peptidases.

The human amelogenin (AMEL) gene is located on the distal portion of the short arm of the X chromosome in the p22.1 to p22.3 region. This locus is also near the centromere on the Y chromosome (possible at the proximal long arm Yq11 region). It is hypothesized that mutations of the human amelogenin gene are associated with the various X-linked types of AI. The Y chromosomal localization might be associated with genes that participate in regulating tooth size and tooth shape. The estimated prevalence for all types of AI combined is 1 in 14,000. An excellent classification of AI based on the clinical and radiographic appearance of the enamel defect and on the mode of inheritance of the trait has been recently proposed.

The treatment plan for patients with AI is related to many factors, including 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 plan is developed. According to Seow, important clinical problems of AI are aesthetics, dental sensitivity, and loss of occlusal vertical dimension. However, the severity of dental problems experienced by patients varies with each type of AI. Historically, treatment of patients has included multiple extractions and the fabrication of complete dentures. These options are psychologically harsh when the problem must be addressed in adolescent patients. This case report...
describes the treatment of a 24-year-old female patient with AI using metal-ceramic fixed partial dentures (FPDs) in the full mouth coverage.

A Case Report

A 24-year-old female patient was referred for treatment of attrition and aesthetic problems of her teeth. She was very self-conscious about the appearance of her teeth. A detailed medical, dental, and social history was obtained. Photographs and dental radiographs were made.

Physical examination revealed a person who appeared healthy, well developed, and well nourished, and who had normal skin and adnexa. No facial abnormality or asymmetries were noted. The patient’s oral hygiene was extremely good. The lips, tongue, palate, floor of the mouth, and cheeks exhibited no abnormalities. The enamel layer was absent in the full portion of the teeth. The gingival tissue and interdental papillae were good. Pocket depth was normal. The patient was caries-free, and did not exhibit any periodontal problems. The patient had bilateral-balanced occlusion. It was determined that the patient’s inter-occlusal distance was reduced because of an absence of teeth and enamel hypoplasia. Therefore the patient’s vertical dimension was altered (Fig. 1).

Laboratory studies were not remarkable. Blood chemistry included serum calcium, phosphorus, and alkaline phosphatase. Other tests used were urine-analysis, serology, fasting blood glucose, and haematology.

The patient’s diagnostic casts were made, as were face-bow and protrusive records. Casts were mounted in centric relation.

None of the unerupted teeth were removed in the maxilla and the mandible for preventing the clefts instead of bone loss and also patient did not accept the removal of unerupted teeth.

A treatment plan was developed with the following aims: to improve the aesthetics and to restore masticatory function. Fabrication of metal-ceramic FPDs for maxillary and mandibular teeth was planned. The patient was informed of the diagnosis and the treatment plan, which she accepted.

The vertical dimension of occlusion was determined by the usual clinical tests and was set on the articulator (Dentatus Type ARH, Sweden). No tooth reduction was made except minor preparations to remove undercuts. Impression for teeth were made with elastomeric impression material (One Time Perfect®, Type 3, Detax, Germany). Temporary restorations were made with self-curing acrylic resin (Temdent Classic®, Weil-Dental GmbH, Germany) and were luted with temporary luting agents (Cavex Temporary Cement®, Cavex Holland BV, Holland) for 3 weeks to tolerate new vertical dimension of occlusion (Fig. 3). The patient was then monitored systematically over a 2-month period to check for any functional or articulation problem following the change in vertical dimension of occlusion. After 2 months of monitoring, the patient had no problem.

Thereafter, a clinical crown lengthening surgery was made for maxillary anterior right and left central and lateral teeth. A periodontal pack was placed for the first postoperative week, and the patient was instructed to use 0.2% chlorhexidine mouthwash twice daily for 2 weeks. Postoperative healing was rapid, and the patient experienced no discomfort. The resulting gingival contour was an enormous improvement, and the patient was pleased with the result (Fig. 4).
2 weeks later, healing was sufficient to continue treatment. Impression for teeth were made with the same elastomeric impression material. Casts were fabricated. A trial evaluation of the metal substructure, prior to glazing of the ceramic material, enabled final occlusal refinement. Metal-ceramic FPDs for maxillary and mandibular teeth were cemented with zinc-policarboksilate cement (Adhesor® Carbofine, Spofa Dental, Frankfurt, Germany) using the manufacturer’s recommended power/liquid ratio (Fig. 5). The patient was observed at intervals of 3 months and then once a year (Fig. 6).

**Discussion**

The complexity of the management of patients with AI supports the suggestion that the prosthodontic profession should have a key position in the rehabilitation of rare disorder. Treatment of patients with AI should start with early diagnosis and intervention to prevent later restorative problems. However, some patients may not seek treatment until young adulthood, after advanced tooth wear and associated tooth sensitivity, functional, and aesthetic problems that have already occurred, leading to the need for complex restorative treatments. Fixed restorations also allow these patients to avoid social problems that are associated with partial or full dentures, particularly in young people.

Patients with AI need not to be faced with extraction of their teeth at an early age. A definite advantage is gained by combining the disciplines of periodontology and prosthodontics.

**References**


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