Management of Amelogenesis Imperfecta in the Mixed Dentition: A Case Report

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Background and Clinical Manifestations

Background

Amelogenesis imperfecta (AI) is a group of inherited developmental enamel defects affecting all or nearly all teeth in the primary and permanent dentitions, without an associated systemic disorder. Prevalence ranges from approximately 1:700 to 1:14,000 depending on population. AI may be inherited in X-linked, autosomal, or sporadic patterns, and each phenotype correlates with specific gene mutations (e.g., AMELX, ENAM, KLK4, MMP-20, FAM83H). Unlike acquired enamel defects, AI presents with uniform involvement of teeth and often a positive family history.

Clinical Manifestations

The most common presenting complaints are tooth sensitivity and compromised esthetics. The four major AI types include hypocalcified, hypomaturation, hypoplastic, and hypomaturation-hypoplastic with taurodontism, each with multiple inherited subtypes.

Clinical findings may include altered eruption patterns, rapid attrition, excessive calculus accumulation, gingival hyperplasia, and low caries susceptibility. Pathologic associations include impacted or ectopically erupting permanent teeth, enlarged follicles, congenitally missing teeth (notably second molars), pulp calcifications, and crown or root resorption. Anterior open bite is frequently associated, particularly in hypocalcified AI.

Treatment Considerations

Management of AI in pediatric dentistry is age- and dentition-dependent, with special challenges in the mixed dentition. The primary goals are to preserve tooth structure and vitality, reduce sensitivity, maintain vertical dimension and arch integrity, and support normal eruption.

- Primary dentition: Stainless steel crowns are recommended for primary molars to prevent attrition and maintain occlusal vertical dimension. Alternative restorations include polycarbonate crowns, glass ionomer cement (GIC), or composite resin.
- Mixed dentition: Treatment is complex due to ongoing eruption. Stainless steel crowns are commonly placed on young permanent molars. Anterior teeth may be restored with GIC or composite, recognizing that retreatment is often necessary. Definitive rehabilitation is deferred until full eruption of the permanent dentition.
- Permanent dentition: Treatment focuses on restoring function, esthetics, sensitivity control, and vertical dimension, often requiring a multidisciplinary approach.

Restorative planning should be guided by AI subtype. Hypocalcified AI often requires full-coverage restorations due to poor enamel bonding, while hypoplastic AI allows for more conventional preparations. Across all stages, conservative treatment and clear communication with patients and families regarding expectations and retreatment are essential.

Case Report and Discussion

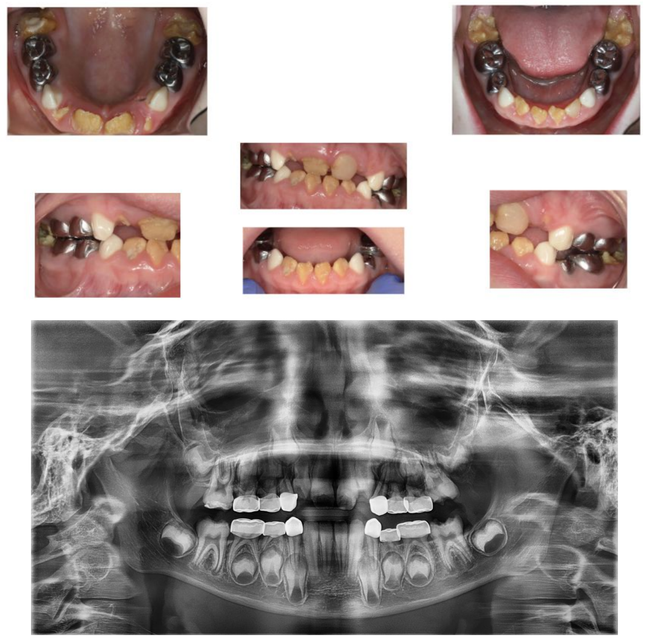

Fig 1. Clinal photographs and Panoramic radiograph of the patient at age 8y 11mo

An 8-year-11-month-old female with a history of amelogenesis imperfecta (subtype unspecified) was referred to the University of Michigan for evaluation and management options. The patient previously underwent treatment under general anesthesia with full coronal coverage of all primary teeth due to severe sensitivity and esthetic concerns. Clinical examination revealed a cooperative patient in the mixed dentition with good oral hygiene, no caries, and enamel defects consistent with AI. Extraoral and soft tissue examinations were within normal limits. Occlusal findings included Class I molar and canine relationships bilaterally, maxillary crowding, buccal eruption of teeth #7 and #10, zero overbite and overjet, and an anterior crossbite between teeth #8 and #25. Radiographic and clinical examination identified a lost restoration on tooth #9 but no active pathology. The patient was referred for orthodontic consultation, which recommended reevaluation in six months to allow further eruption of the maxillary lateral incisors before initiating treatment, with a likely future need for maxillary expansion and 2×4 orthodontic appliances. Restorative planning will be coordinated with orthodontics, with consideration for stainless steel crowns on permanent molars to manage sensitivity and maintain vertical dimension, interim anterior restorations with the understanding of frequent retreatment, and referral to prosthodontics to establish long-term care.

Discussion

1. Understanding what type of AI you are dealing with can help dictate how you prepare the tooth, with the hypocalcified type often requiiring full coverage and the hypoplastic type requiring adequate preparation
2. In primary dentition full coverage works best
3. In the mixed dentition frequent re-treatments will likely occur and transient restorations are indicated
4. Want to preserve as much tooth structure as possible
5. Managing concerns and expectation with the family is important

Subtypes of Amelogensis Imperfecta

AI Type	Primary Defect	Enamel Thickness	Hardness / Mineralization	Key Clinical & Radiographic Features
Hypocalcified	Defective mineralization	Normal	Poorly mineralized; soft enamel	Enamel fractures easily, often smooth surface, rapid wear, severe sensitivity
Hypomaturation	Defect in matrix protein degradation	Normal	Reduced hardness	Enamel chipping and breakdown, decreased radiographic opacity near EDJ, often mild and underdiagnosed
Hypoplastic	Defective enamel matrix formation	Reduced	Normal mineralization; hard enamel	Thin enamel, pitting or grooving, rough or smooth surface, normal radiographic contrast
Hypomaturation-Hypoplastic with Taurodontism	Combined matrix and maturation defects	Variable (often normal)	Reduced hardness	Chipped enamel, opaque white coloration, taurodont molars, associated with tricho-dento-osseous syndrome

Table 1. Subtypes of amelogenesis imperfecta

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