



Case Series

**A case series on amelogenesis imperfecta in
young patients**

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ABSTRACT

There are multiple challenges in treating patients with amelogenesis imperfecta (AI). When we think about pediatric patients, this becomes much more obvious. This case series report addresses different case reports of pediatric patients who present with imperfect amelogenesis. Patients with type I hypoplastic amelogenesis imperfecta, hypomaturational type amelogenesis imperfecta, and sporadic hypoplastic and hypomaturational type amelogenesis imperfecta were identified as cases I, II, and III, respectively. This case series has covered several therapies

Keywords: *Amelogenesis imperfecta, hypoplastic, hypomaturational, sporadic amelogenesis imperfecta*

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INTRODUCTION

Amelogenesis imperfecta has an impact on teeth, both primary and secondary. Depending on the kind of Amelogenesis imperfecta¹, clinical issues can include sensitivity (especially to cold), quick wear, and poor aesthetics. Addressing these issues, symptom relief, maintaining face height, and improving aesthetic attractiveness are treatment objectives. Open bites of anterior teeth, pulp calcification, pathological root or crown resorption, and taurodontia are other anomalies linked to amelogenesis imperfecta that might affect young patients.^{2,3}

Not only is it challenging for kids to react to amelogenesis imperfecta, but they also face additional challenges because of their inexperience with the dentist, self-awareness, fear of the dentist, and parental expectations. Like with any youngster, first impressions in the dentistry field have an impact on subsequent cooperation.

Not only is it challenging for kids to react to amelogenesis imperfecta, but they also face additional challenges because of their inexperience with the dentist, self-awareness, fear of the dentist, and parental expectations. Like with any youngster, first impressions in the dentistry field have an impact on subsequent cooperation. That's why it's critical to ensure that the initial encounter goes well. A doctor can establish a positive rapport with a child and parents by seeing them as soon as possible. The consequences of frequent visits on education and difficulties for working parents should be considered when scheduling appointments to support families.

CASE REPORT 1:

A 13-year-old girl complained of anterior upper teeth discoloration and forward eruption of her front teeth when she went to the pediatric department of Krishnadevaraya Dental College in Bengaluru. The main thing on the list was appearance. There was no evidence of a family history. Upon clinical inspection, there was a widespread yellowish discoloration, rough surface, pitted and grooved enamel, and decreased enamel thickness. To assess tooth morphology and developmental abnormalities, an orthopantomogram and intraoral periapical radiograph were obtained. Following cross-departmental consideration, type I hypoplastic amelogenesis imperfecta was determined. In this instance, composite veneers were the preferred treatment for the lower first permanent molars and the upper and lower front teeth, taking cost into account.



Figure 1: shows the pre-operative clinical photograph of generalised enamel involvement

CASE REPORT 2:

A five-year-old female patient reported at the dental department with her main complaint being discoloration of her freshly erupted permanent teeth. She was sensitive as well. There was no evidence of a family history. A generalized discoloration was found during the clinical examination. The enamel had a typical thickness and was pitted and rough. To assess tooth morphology and developmental abnormalities, an orthopantomogram and intraoral periapical radiograph were obtained. It was determined after cross-departmental debate that the amelogenesis imperfecta was of the hypomaturation kind. Oral hygiene instructions were given, and parents were encouraged to visit the clinic regularly until all permanent teeth had fully erupted.



Fig 2: shows the orthopantomogram image of the patient showing generalised enamel loss.

CASE REPORT 3:

The main complaint of a 13-year-old boy who came to the department was that he had sensitive and permanently discolored teeth. Clinical examination revealed an open bite, widespread discoloration, and a class III molar relation. Enamel that chipped easily, a decrease in enamel thickness, surface roughness, and varying degrees of structural loss were also seen. Enamel matrix defects resulted in grooved and pitted enamel, particularly in the anterior upper teeth and the first permanent molars. Consanguineous marriage history was mentioned. To assess dental morphology and developmental abnormalities, lateral cephalograms, orthopantomograms, and intraoral periapical radiographs were obtained. After extensive cross-departmental debate, it was determined to be a hypoplastic, hypo maturation, and sporadic amelogenesis imperfecta type since the permanent dentition was afflicted while the deciduous dentition was untouched. both the patient and the parents.



Figure 3: shows the clinical picture of the patient showing generalised hypoplastic enamel

DISCUSSION

Amelogenesis imperfecta refers to a genetic disorder-related systemic deficiency in enamel production in the primary and permanent teeth. Dental abnormalities that are inherited might be sporadic, autosomal recessive, X-linked, or autosomal dominant. Specifically, defects in the expression or mutation of certain genes, including amelogenin, kallikrein, and enamelin, are dysfunctional in the binding of proteins that make enamel⁴. Clinical presentations comprise four categories of amelogenesis imperfecta⁵. The most well-known type, Type I, has a hypoplastic structure and a lower enamel concentration. The teeth show varied degrees of defects⁶, roughened surfaces, and decreased enamel thickness, as shown in case 1.

Because of flaws in the maturation of proteins within the enamel matrix, type II, also known as hypomaturity, has fragile, mottled enamel. Moreover, the dentin and dental enamel may separate. Normal enamel thickness is observed in amelogenesis imperfecta type II. Ameloblasts secrete enamel matrix proteins normally, but during the maturation phase, this normal reabsorption of released proteins does not take place. Then, as seen in instance number 2, a significant amount of organic matter is still present in the tooth enamel⁷. Type III (hypocalcification) is associated with mineralization defects and occurs during an eruption of normal-thickness enamel. Due to insufficient mineralization, tooth enamel wears away rapidly and becomes less opaque on X-rays.

As seen in case number 3 type IV manifests as a mixed appearance of hypoplasia and maturation combined with taurodontia^{4,8}. Due to the structure of enamel hypersensitivity, plaque accumulation, and poor aesthetics have been reported⁹. Multidisciplinary patient care is recommended to prevent tooth decay, gingival inflammation, open bites, or loss of vertical dimension. Different treatment options have been reported depending on the patient's age, socio-economic condition, and severity of the malformation¹⁰. While stainless steel crowns, strip crowns, and compomer restorations are common in primary dentition, the challenge for mixed and permanent dentition in young adults is caring for the dentition as it grows. While ceramic crowns and veneers are preferred for adults, CAD/CAM composites offer the opportunity for high-quality restorations for growing children. Prevention is the most important tool in a clinician's arsenal. Oral hygiene can be particularly difficult for patients with AI because of sensitivity. While brushing, use warm water. The use of fluoride mouthwash can also help reduce tooth sensitivity and prevent tooth decay in children old enough to avoid swallowing. Local anesthesia (LA) is an essential tool to facilitate treatment in these children.

When dealing with AI in children, the goal should be to manage any symptoms or complaints the child may have. X-rays are a valuable tool for assessing tooth decay, but they also provide information about the thickness and quality of tooth enamel. Delayed eruption, taurodontia, and idiopathic resorption may also be seen. If there are no signs of wear, minimal intervention and monitoring is the preferred treatment plan. This makes treatment decisions much easier for adult patients. However, if the child experiences tenderness, occlusal wear, or is dissatisfied with aesthetics, intervention is necessary.

For children who require care of their deciduous dentition, Anterior teeth treatment is required to improve aesthetics with glass ionomer or composite direct veneer. steel crowns or restorations with glass ionomer cement on the occlusal surface of deciduous molars where SSC placement is not advised. Orthodontic separators between the molars assist in the placement of the SSC.¹⁴ There is no need to prepare the teeth for this treatment unless there is caries in the dentition. For apprehensive children, compromises may be needed. In this situation, the following is true: The second deciduous molars have a higher priority in maintaining occlusion. Placing aesthetic veneers boosts patient confidence but might not return as the primary complaint is addressed. Permanent dentition is similarly affected by amelogenesis imperfecta and should be protected as quickly as possible.

Treatment requirements in this phase include onlay/crown (Gold) or stainless steel crown¹⁵ on the fully erupted first molar. SSC also has the advantage of requiring less cooperation from the child and can be done in a single appointment. Nevertheless, restorations (Gold) are further reliable, and supragingival margins can be given for improving oral hygiene and the health of the gingiva.⁹ Because of takes a longer time for teeth to emerge, impairment can arise in the future. In these conditions, we might be required to think about restoration using glass ionomer up until full eruption.

In a study by Coffield et al., amelogenesis imperfecta has been shown to negatively impact well-being in adulthood,¹³ and this may also apply in childhood. Kids with amelogenesis imperfecta need higher dental requirements and can develop numerous problems. For anterior permanent centrals and laterals, composite veneers provide better aesthetics and decrease incisor attrition. Direct composite has the benefit that it could be given as sufficient teeth have erupted and can be topped up as more teeth emerge.

CONCLUSION

A multidisciplinary team including general dentists, pediatric specialists, orthodontic clinicians, and dental hygienists would be helpful for properly managing patients with amelogenesis imperfecta. As several amelogenesis imperfecta cases are being reported, it is very important to know the complications, clinical features, and management for better diagnosis and clinical management of the problem.

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CONFLICTS OF INTEREST

There are no conflicts of interest

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