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Oral Rehabilitation of Primary Dentition Affected by Amelogenesis Imperfecta: A Case Report

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Abstract

Aim: The purpose of the case report was to describe the treatment of a 4½-year-old boy with amelogenesis imperfect (AI) in the primary dentition.

Background: AI is a hereditary condition that affects the development of enamel, causing quantity, structural, and compositional anomalies involving all dentitions. Consequently, the effects can extend to both the primary and secondary dentitions.

Report: A 4½-year-old boy was brought to the dental clinic complaining of tooth hypersensitivity during meals. The medical history and clinical examination were used to arrive at the diagnosis of amelogenesis imperfecta. The treatment was oral rehabilitation of the primary molars with stainless steel crowns and resin-filled celluloid forms of both maxillary and mandibular primary incisors and canines.

Improvements in the patient's psychological behavior and the elimination of tooth sensitiveness were observed, and the reestablishment of a normal occlusion resulted in improved eating habits. The child was monitored in the Pediatric Dentistry Clinic at four-month intervals until the mixed dentition stage.

Summary: The oral rehabilitation of young children with AI is necessary to reestablish the stomatognathic system function, so important for a child's systemic health. An adequate medical history and a careful clinical examination were essential for a correct diagnosis. Treatment was rendered that was appropriate for the child's age and clinical/psychological characteristics.

Clinical Significance: Cost-effective restorative techniques involving stainless steel and composite-resin crowns are shown for the restoration of a young patient with amelogensis imperfecta.

Keywords: Amelogenesis imperfecta, primary dentition, crowns, pediatric dentistry

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Introduction

Amelogenesis imperfecta (AI) actually is a group of hereditary conditions that affect the development of dental enamel, causing anomalies in the enamel quantity, structure, and composition.[1-3] Usually, both the primary and secondary dentitions are affected. This rare dental disease was first described by Spokes[4] in 1890 as "hereditary brown teeth;" however, until 1938 it was not classified as a separate entity from dentinogenesis imperfecta.[5] The estimated prevalence of AI ranges from 1:14,000 to 1:700, depending on the diagnostic criteria and the population studied.[6] This clinical condition has previously been reported by Ayers et al.,[1] who described a case report of a seven-year-old girl and her family with a history of hipoplastic amelogenesis imperfecta. Additionally, in 2006 Vitkov et al.[3] described a case report of five patients between 3, 5, and 10 years of age with AI and the treatment options included were composite-resin crowns and veneers in all teeth. Moreover, Türkün[7] reported a case of hypomaturated type AI in a 16-year-old young man that was successfully treated with different types of resin composites.

Patients with amelogenesis imperfecta are often esthetically affected, due to tooth discoloration and extensive loss of tooth tissue leading to hypersensitivity.[8] Usually, the pulp and the dentin are normal[9] and the histologic alterations include a hypoplastic enamel layer, positive birefringence, generalized pitting, roughness with irregular generally cracked borders, and a porous surface.[10]

The original division of AI into hypoplastic and hypocalcified types by Weinmann et al.[11] in 1945 has since been further classified. It has been reported that the primary structure for the classification of AI is based on the mode of inheritance, with the phenotype being the secondary discriminator.[2,12] In addition, molecular defects and biochemical analysis also contribute to the classification of AI.[2] Normally, genomic and biochemical information are not present and, therefore, the diagnosis is based on clinical manifestation, as well as an inheritance pattern. The patterns of inheritance of AI may be X-linked, autosomal dominant, or recessive.

Witkop[13] recommended classifying AI into three groups: hypoplastic AI, hypocalcified AI, and hypomaturated AI. The hypoplastic type results from an error that affects the dental tissue protein matrix; the calcification is normal but characterized by a local or generalized decrease in the amount of enamel thickness. In contrast, the hypocalcified AI is caused by a defect in the initial nucleation of enamel crystallites. The enamel is completely formed on a normal matrix; however, it is not fully mineralized. In hypomature AI, there is a defect in the removal of extracellular protein, which results in decreased mineral deposition and increased matrix retention.[14] Moreover, the major forms of AI can be divided into 14 subtypes, regarding the predominant clinical manifestation and inheritance mode.[13]

The present case report describes the management of a child with severe amelogenesis imperfecta in the primary dentition.

Case Report

A 4½-year-old Caucasian male was brought by his mother to the Department of Pediatric Dentistry at Piracicaba Dental School, University of Campinas, SP, Brazil. The main reason for the dental visit was a complaint of hypersensitivity of the teeth during meals. The family history revealed that this disease had already affected his mother and maternal relatives (grandfather, cousin, aunt, and two uncles) (Figure 1). Clinical examination was carefully undertaken and generalized tooth wear was observed, as well as enamel loss affecting the entire primary dentition, with the exception of some cervical areas (Figure 2). An interproximal intraoral radiographic examination was performed, and no difference in the hard tissue contrast was noted. A detailed medical history associated with clinical evidence and family history provided information that led to the diagnosis of autosomal dominant hypocalcified amelogenesis imperfecta.

Due to the loss of vertical dimension and teeth characteristics, the treatment of choice was oral rehabilitation with stainless steel crowns on the maxillary and mandibular primary molars and resin-filled celluloid forms for both the maxillary and mandibular primary incisors and canines. The stainless steel crown treatment was undertaken

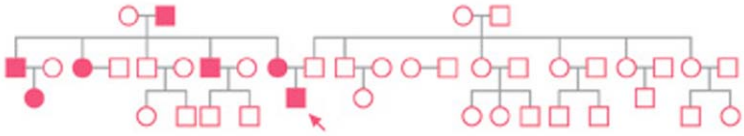

Figure 1. Family tree showing the hereditary pattern of amelogenesis imperfecta.
Note: Square: male; circle: female. The solid symbols represent the maternal family members affected by AI. The arrow points to the patient described in the present case report.

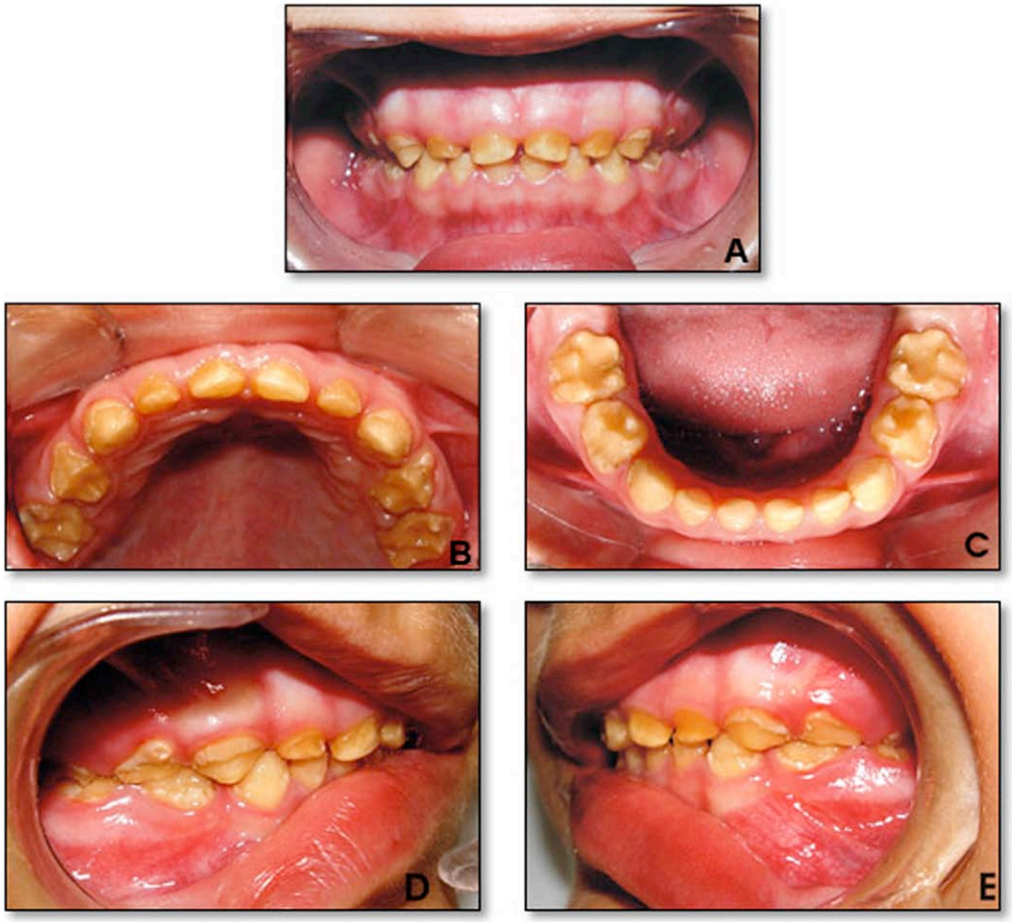

Figure 2. A. Frontal view of amelogenesis imperfecta in a 4 1/2 year-old child; **B.** Maxillary occlusal view; **C.** Mandibular occlusal view; **D.** Right lateral view; **E.** Left lateral view.

as follows: The stainless steel crown sizes were selected and early adjustments made based on diagnostic casts. The interproximal and occlusal surfaces of the teeth were prepared, and a feather edge gingival finishing line was prepared with no ledges or steps detectable. The preparations were made with short, tapered diamond burs (KG Soresen, São Paulo, SP, Brazil). Final adjustments on the stainless steel crowns were made with a carburundum disc (Saint-Gobain Abrasivos Ltda, Carapicuíba, SP, Brazil). The walls of the crowns were adjusted with #114 pliers to achieve a satisfactory fit, and the crowns were polished with rubber abrasive points (KG Soresen, São Paulo, SP, Brazil). The tooth surfaces were cleaned with a slurry of pumice and water and rinsed with the aid of high-volume suction before being isolated with cotton rolls. Fuji IX glass ionomer cement (GC America Inc., Alsip, IL, USA) was mixed according to the manufacturer's instructions and placed inside the crowns. Firm seating pressure was exerted on each crown using finger pressure alone. The patient's vertical dimension of occlusion was reestablished with the aid of the stainless steel crowns.

As for the restoration of the maxillary and mandibular primary incisors and canines, composite resin–filled celluloid forms (TDV Dental, Pomerode, SC, Brazil) were used in accordance with the manufacturer's instructions. The procedure used was as follows: Cut the forms with scissors so they fit directly over the teeth; perforate the lingual surface to allow composite resin to be removed on seating; clean the teeth with a slurry of pumice and water; rinse and evacuate; isolate the teeth with cotton rolls; acid-etch for 30 seconds with 37 percent phosphoric acid (3M/ESPE, St. Paul, MN, USA); rinse and evacuate the water; change the cotton rolls; gently air dry; apply two layers of Single Bond adhesive® (3M/ESPE, St. Paul, MN, USA) and light-cure following the manufacturer's instructions; apply a light-cured composite resin Z250® (3M/ESPE, St. Paul, MN, USA) inside the celluloid forms; seat the forms firmly with finger pressure; remove material excess with a probe (or other instrument); light cure the restorations; remove the outer forms; check the occlusion; and then finish and polish (Figure 3).

Instructions were given to the child's mother regarding oral hygiene and eating habits with the recommendation to avoid excessive force on the anterior teeth by cutting the child's food into small, bitesize pieces. These recommendations were essential and of great importance for treatment longevity. After four months, the treatment was completed, and the patient was recalled weekly during the first month to emphasize these measures. The patient has since been attended at the Pediatric Clinic every four months for other adjustments and preventive maintenance.

After 16 months, the maxillary and mandibular permanent incisors and first molars erupted showing the same clinical characteristics as the primary teeth. As a result, temporary restorations

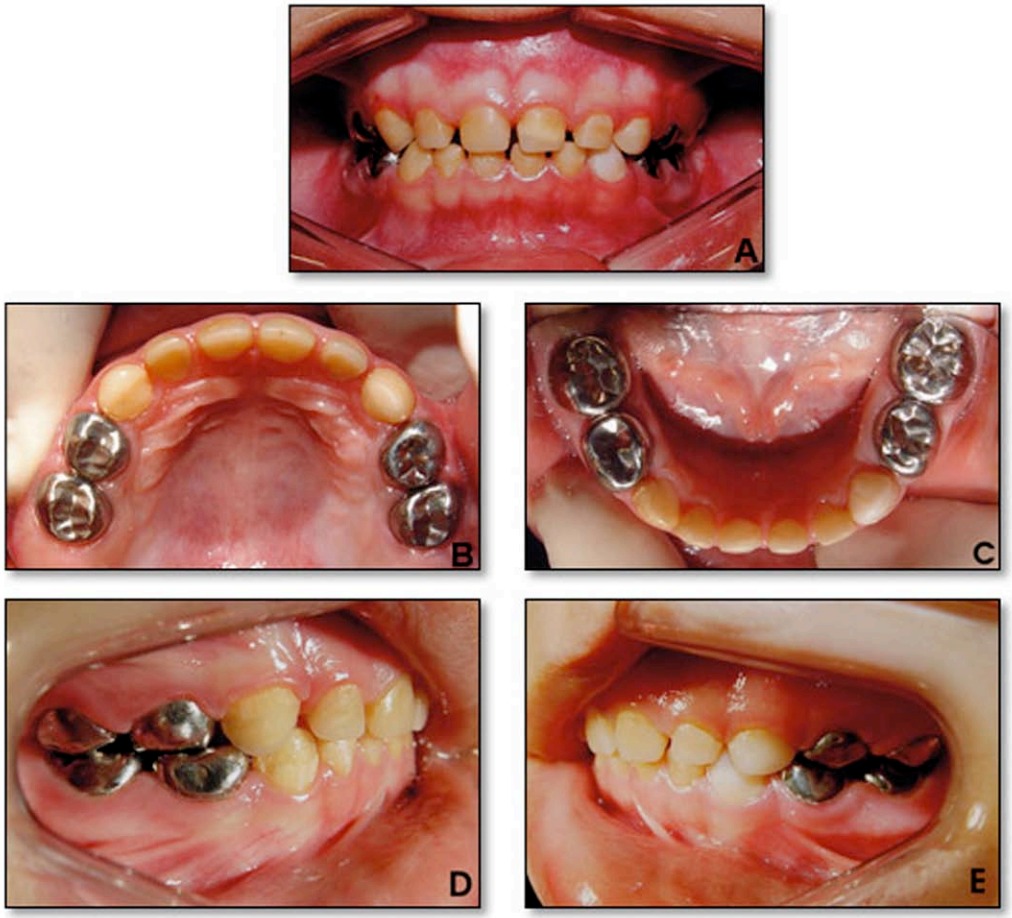

Figure 3. A. Frontal view of the completed treatment with stainless steel crowns and composite resin filled celluloid forms in a 4 1/2 year-old child; **B.** Maxillary occlusal view; **C.** Mandibular occlusal view; **D.** Right lateral view; **E.** Left lateral view.

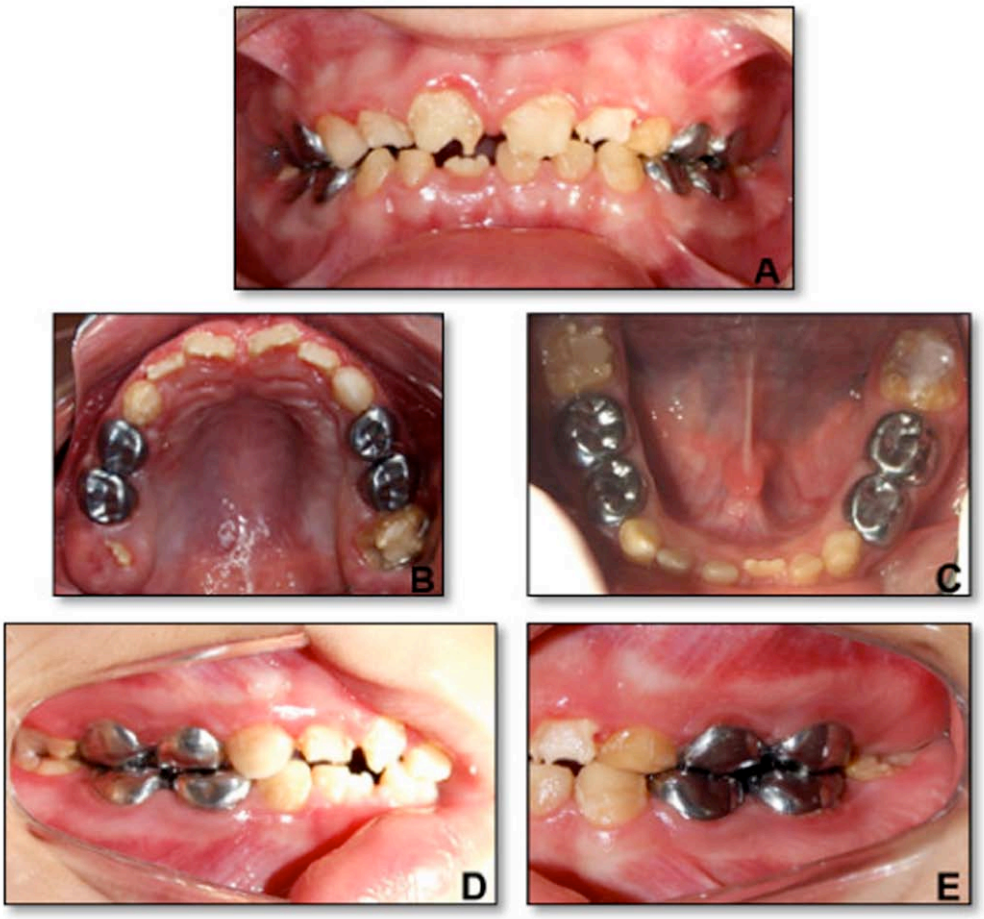

Figure 4. A. Sixteen months after completion of the treatment in the primary dentition – **A.** Frontal view; **B.** Maxillary occlusal view; **C.** Mandibular occlusal view; **D.** Right lateral view; **E.** Left lateral view.

were placed on the occlusal surface of the permanent molars with conventional Fuji IX GP glass ionomer restorative material (GC America Inc., Alsip, IL, USA). The permanent incisors were not restored because they had not erupted completely (Figure 4). The child's mother was strongly advised to seek prosthetic treatment for the permanent teeth at the Department of Oral Rehabilitation.

Discussion

Amelogenesis imperfecta is a rare disease that can compromise the entire primary and permanent dentition and becomes increasingly more severe with age.[7] In the present case report, the reestablishment of function and esthetics in a young patient with AI could be achieved thanks to advances in the field of restorative dentistry, coupled with a dedicated team approach.

The type of amelogenesis imperfecta in this case report was classified according to the hereditary pattern (genotypes) and clinical features (phenotypes).[1,15,16] The chalky dull color of the enamel represents low mineralization, clinically expressed by pigmented, softened, and easily detachable enamel structure. The family history, as revealed in the medical history, indicated the same condition existing in other family members on the mother's side of the family and this aided in the diagnosis of hypocalcified amelogenesis imperfecta.

Several treatments are available for pediatric patients with AI, such as multiple extractions, use of light-cured composite, polycarbonate crowns, space maintainers, and stainless steel crowns to restore a mutilated dentition.[17,18] The stainless steel crowns are generally used for primary molar teeth with developmental defects such as dentinogenesis imperfecta, hypocalcifield teeth, or huge tooth destruction due to caries.[6,19]

Additionally, stainless steel crowns are useful to reduce tooth hypersensitivity and to restore occlusion, allowing the patient to eat adequately and to maintain effective oral hygiene.[20,21] Moreover, due to the precocious age of the patient and considering the longevity of the maxillary and mandibular primary molars in the arches (approximately six years), stainless steel crowns were placed.[22] These crowns are able to duplicate the anatomy of primary molars with accuracy, considering the contour, occlusal surface, and life-like height. The crowns are pre-crimped at the cervical margin to provide good retention and a snap fit. When marginal adaptation is achieved and the oral hygiene is satisfactory, the crown smoothness contributes to the reduction of bacterial biofilm accumulation.[23] Considering all these factors, stainless steel crowns were the treatment of choice for the rehabilitation of the maxillary and mandibular primary molars in this particular case report.

With respect to the primary anterior teeth restorations, advances in esthetic dentistry, particularly in bonding to dentin, allowed for the reestablishment of function and acceptable esthetics, especially with respect to the short primary teeth cycle.[3,7] Moreover, thanks to new polishing systems, esthetics, and color stability, longevity can be achieved successfully with resin-based restoration.[7] Despite our esthetics-driven society, with high expectations for oral health and appearance, it is critical to recognize that good teeth conditions are important in the development of optimal psychosocial health.[24] In this same context, the use of resin-filled celluloid forms in the maxillary and mandibular primary anterior teeth was the most suitable treatment for this 4½-year-old boy.

This dental treatment played an important role in improving the patient's psychosocial development. When he first arrived in the Pediatric Dentistry Clinic, he was very shy, tooth hypersensitivity during meals made it difficult for him to eat well, and these factors resulted in inappropriate psychological behavior. After the completed treatment of the maxillary and mandibular primary molars, a visible and substantial improvement was soon observed in his eating habits, according to his mother. The child's behavior changed in a positive manner and upon return to the clinic, he appeared to be happier and more outgoing. He talked freely and played with everyone in the treatment room.

Additionally, the early interventions were essential for the reestablishment of the child's masticatory function, as well as the entire primary dentition esthetics. However, it should be emphasized that regular follow-up is extremely important for the long-term maintenance of this complex treatment. Moreover, additional intervention should be planned for the permanent dentition using appropriate restorative and prosthetic techniques and materials. This will help the patient to avoid the fate of his other family members who did not receive adequate therapy for their condition. They had all their teeth removed prematurely and now need maxillary and mandibular complete dentures. It is also important to highlight that oral rehabilitation during early childhood entails many challenges. Patients and parents may not have an adequate understanding of amelogenesis imperfecta or appreciate the extensive nature of the treatment required. A successful outcome requires collaboration by all concerned due to the complexity of care and the need to attend multiple, lengthy appointments over a longer period of time as opposed to short appointments for the placement of simple, direct restorations.

Conclusion

The oral rehabilitation of a young child with amelogensis imperfecta is shown to reestablish the stomatognathic system function and to contribute to the improvement of the patient's systemic health. A detailed medical history as well as a thorough and comprehensive clinical examination are essential for a correct diagnosis. Appropriate treatment should be provided for the patient's age and clinical/psychological characteristics. The treatment performed in this case report was considered successful. The patient no longer experienced thermal sensitivity, had his occlusion reestablished at an appropriate vertical dimension, and demonstrated improvements in his eating habits, in his oral hygiene, and, finally, in his psychological behavior.

Clinical Significance

Cost-effective restorative techniques involving stainless steel and composite resin crowns are shown for the restoration of a young patient with amelogenesis imperfecta.

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