

Dental Rehabilitation for Pediatric Patient with Amelogenesis Imperfecta: A Case Report

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Abstract

Amelogenesis imperfecta (AI) is a developmental disturbance that interferes with normal enamel formation in the absence of a systemic disorder. A seven-year-old Saudi healthy boy was presented with dissatisfaction of his teeth appearance, mild lower posterior teeth sensitivity, poor masticatory efficiency and low self-esteem. The clinical and radiographic results indicated a hypomaturation and hypoplasia with taurodontism according to Witkop's classification of AI. Because of uncooperative behavior, treatment under general anesthesia was planned to manage the child behavior. The primary goal of treatment was to restore and preserve of remaining tooth structure against further sensitivity and wear. Many factors should be considered in treatment planning of AI cases such as the patient's age, socioeconomic status, type of AI, and severity of the condition. The early diagnosis is very important in such enamel defects, because it not only determines the appropriate clinical intervention but also minimize damages to the tooth structure.

Keywords: Amelogenesis Imperfecta; Composite Restoration; Stainless Steel Crowns; Restorative Treatment

Introduction

Amelogenesis imperfecta (AI) is a developmental disturbance that interferes with normal enamel formation in the absence of a systemic disorder and affects all the primary and permanent dentitions [1]. Specific gene mutations proven to cause AI which include enamel proteins, either structural (amelogenin, enamelin, ameloblastin, c4orf26) or enzymatic (kallikrein 4, MMP20); some others mutations in transcription factors (MSX2, DLX3), cellular proteins (WDR72, FAM83H, COL17A1), cellular receptor (ITGB6) and calcium carrier (SL-C24A4) are causal factors [1]. According to the population studied, the prevalence of AI is estimated to be lower than 0.5% [2]. Many classifications of AI have emerged long time ago based on clinical features (phenotype) and mode of inheritance pattern (genotype). AI can be inherited by autosomal dominant, autosomal recessive, x-linked or sporadic inheritance [3]. One of the most common classification of AI was introduced by Witkop in 1988. Based on clinical features and mode of inheritance, AI was classified into four major types: hypoplastic, hypomaturation, hypocalcified, and hypomaturation-hypoplastic with taurodontism and with 15 subtypes [3]. The oral manifestations of AI include quantitative and qualitative enamel deficiencies, low dental caries susceptibility, rapid attrition, loss of vertical dimension, excessive calculus deposition, gingival hyperplasia as well as impacted permanent teeth and ectopic eruption [4]. In addition, the incidence of anterior open bite in hypomaturation AI, hypoplastic AI and hypocalcified AI are 31 percent, 50 percent and 60 percent, respectively [5]. This report presents the clinical dental management of pediatric patient with AI in early mixed dentition.

Case Report

The patient, a seven-year-old Saudi healthy boy, was referred to King Saud Medical City - Riyadh, dental clinics from a primary health care center presented with dissatisfaction of the appearance of his teeth, mild lower posterior teeth sensitivity, poor masticatory efficiency and low self-esteem.

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The patient was examined dentally; photographs (Figure 1) and dental radiographs (Figure 2 and 3) were obtained. The family history revealed that the parents are first cousins and his father is having full dental rehabilitation. Intra-oral clinical examination revealed an early mixed dentition and poor oral hygiene with generalized mild plaque-induced gingivitis. The primary teeth showed short clinical crowns with white/yellow to brown color, hypoplastic, chipped enamel and lack proximal contact as well as exposed dentin in posterior teeth and dental caries. The enamel layer of all permanent teeth was very rough and thin. Symmetrical "U-shaped" maxillary and mandibular dental arches. Both vertical dimension and facial height were reduced because of attrition of primary molars.



Figure 1: Intra-oral preoperative photographs



Figure 2: Pre-operative panoramic radiograph.

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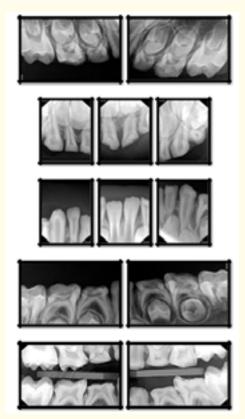


Figure 3: Pre-operative periapical and bitewing radiographs.

The panoramic (Figure 2), bitewing and periapical (Figure 3) radiographic examination, revealed an early mixed dentition period with all permanent teeth present except the third molars and covered by thin layer of enamel. The maxillary first permanent molars showed hypo- taurodontism. Enamel contrast almost similar to dentin. The dental age is late compared to his chronologic age.

Discussion

The clinical and radiographic results indicated a hypomaturation and hypoplasia with taurodontism according to Witkop's classification [3]. Patients with AI are consider as high risk according to caries-risk assessment as they need high levels of oral health care [6,7].

The early diagnosis is very important in such enamel defects to determine the appropriate clinical intervention and minimize further damages to the tooth structure. However, the cooperation and motivation of both the patient and parents are important for the successful management of AI. Teeth discoloration and crown appearance have significant psychological and social effects in patients with AI [8]. Moreover, AI is a condition that reduces oral health-related quality of life [9].

Current guidelines for restorative treatment in young children and adolescents suggest covering the surface with direct composite resin until adulthood and recommend stainless steel crowns (SSCs) for primary and permanent molars with developmental defects [10,11]. Patients with AI often ask for a more permanent therapy at an earlier age [12].

The primary goal of treating the present case was to restore and preserve the remaining tooth structure against further sensitivity and wear while avoiding the negative psychological effects. After diagnosis, the treatment planning was proposed with emphasis on behavior guidance and preventive program to stop the tooth destruction and improve the gingival health. The preventive program consisted of

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plaque control program, fluoride program, desensitizing agents may diminish tooth sensitivity, dietary analysis and advice, recall visits for examination and reinforcement, and providing the parent and child with simple educational materials.

Prior to treatment, because of uncooperative behavior, treatment under general anesthesia (GA) was accomplished to manage the child behavior. Other behavior management techniques were also considered in the follow up visits including: tell, show, and do, distraction and positive reinforcement. A written informed consent was signed by the father. Direct composite restorations were done on the anterior teeth to enhance the patient's esthetic. Five percent sodium hypochlorite (NaOCl) solution was applied for one minute on the tooth structure to enhance bonding between composite restoration and tooth structure [13]. However, the long-term adhesive bond strength of AI enamel and dentin surfaces were not known [14]. In addition, SSCs were done on posterior primary and permanent molars with minimal preparation to preserve as much tooth tissue as possible for later restorative preparations. Unrestorable primary incisor teeth were extracted. Regular periodic assessment can identify permanent teeth needing care as they erupt.

In six months recall visit for the present case, all the restorations are intact. The patient showed much improvement in his oral care habits (reduction of sugary snack, brushing regularly), and the gingival tissue healed with no inflammation (Figure 4 and 5).



Figure 4: Six months' post-operative photographs.



Figure 5: Six months' post-operative radiographs.

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Conclusion

Many factors should be considered in treatment planning of AI cases such as the patient's age, socioeconomic status, type of AI, and severity of the condition, and association with other medical conditions. The early diagnosis is very important in such enamel defects, because it not only determines the appropriate clinical intervention but also minimize damages to the tooth structure and maximize the treatment options available for the permanent dentition. The dental treatment for patients with AI are challenging and requiring a multidisciplinary approach on the long term.

Conflict of Interest

Authors declare no conflict of interest associated with this publication.

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