

Review Article

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Amelogenesis Imperfecta: Longevity of Clinical Treatment in Pediatric Dentistry

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Abstract

Amelogenesis Imperfecta (AI) is a rare, hereditary disorder that causes a defect in enamel mineralization in the primary and permanent dentitions. Amelogenesis imperfecta is generally categorized as hypoplastic, hypocalcified, or hypomaturation form based on the primary enamel defect. However, distinctive clinical features may be seen in each variant. Moreover, compromised esthetic appearances, tooth sensitivity and loss of occlusal vertical dimension are the common clinical problems observed in these variants. The objective of this review was to discuss the management strategies for patients with amelogenesis imperfecta. Definite diagnosis and adequate treatment planning are ensured for patients clinically presented with different forms of AI. Clinicians should provide preventive care and establish an early permanent therapy plan for these patients. Nevertheless, an interdisciplinary approach is required to improve both esthetic and functions. Moreover, there is a need for long-lasting restorative solutions for AI patients.

Keywords: Amelogenesis imperfect, Crowns therapy, Composite restorations, Stainless crowns, Porcelain therapy

Introduction

Amelogenesis Imperfecta (AI) is a rare, hereditary disorder that causes a defect in enamel mineralization in the primary and permanent dentitions. Several problems might be associated with the Amelogenesis imperfecta: hypersensitivity issues, wear, restorations that might require replacement, gingivitis, aesthetic problems that ultimately result in social avoidance. Patients with Amelogenesis imperfecta can be clinically presented with rapid tooth loss or enamel fractures as well as alterations in the enamel morphology that may compromise the aesthetic appearance and masticatory function [1]. Clinical treatment plays a significant role to address the esthetic appearance of affected teeth, reduce hypersensitivity, and optimize the masticatory function and the preservation of tooth structure thereby improving the quality of life for these patients [2].

Amelogenesis Imperfecta is generally categorized as hypoplastic, hypocalcified, or hypomaturation form based on the primary enamel defect. However, distinctive clinical features may be seen in each variant. Moreover, compromised esthetic appearances, tooth sensitivity and loss of occlusal vertical dimension are the common clinical problems observed in these variants. The mildest clinical problems were visible in the pitted hypoplastic type AI while the most severe form was observed in the hypocalcified type of AI [3].

Proper diagnosis and management are required that might improve the clinician's dental care. Comprehensive management sometimes necessitates interdisciplinary treatment planning. The comprehensive management of AI should include active follow-up involving oral hygiene care instructions and dietary modifications. Preventive care should be addressed individually based on the patient's risk assessment. A close follow-up is crucial to achieving long-term success and outcomes [4]. The objective of this review was to discuss the management strategies for patients with amelogenesis imperfecta.

Overview

Numerous studies have discussed the strategic management required for treating Amelogenesis imperfect to improve the quality of life. A Randomized controlled trial by Lundgren GP et al discussed the importance of crown/prosthetic therapy in young individuals suffering from AI revealed excellent results clinically. The RCT included 27 patients ranging between 11-22 years suffering from severe AI. A total of 119 Procera crowns and 108 IPS e.max Press crowns were placed on the patient's mouth. Longevity, clinical success rates, and survival rates were assessed of 227 crowns for 4.3-7.4 years. Overall, 79% (193) crowns were followed for around 5 years. However, results stated the survival rate was 99.6% and the success rate 94.7% while 95% crowns had the acceptable quality. Moreover, 4% of the crowns required adjustment due to suboptimal marginal integrity. Hence, sensitivity issues were resolved or decreased after crown therapy (p<0.001). Therefore, Ceramic crown therapy in adolescents and young adults with a severe form of AI showed promising results of excellent survival and the long-term success rates and longevity with fewer adverse events reported [5,6].

On the other hand, seven patients with a severe form of AI aged 16 to 23 years who were treated with Porcelain crowns participated in an interview study. The interviews were analyzed using thematic analysis. The analysis process stated three main features: disturbances

in daily lifestyle, management of disturbances and normalization of daily routine. These are the main features of patients presented with AI and they were experiencing the enamel disturbances primarily caused by AI. Severe pain condition and sensitivity issues were the common problems reported by these patients. Some of them were experiencing feelings of embarrassment and social avoidance. After the placement of the Porcelain crowns, they described their feeling like a normal patient. This study supported the use of porcelain crowns can have a positive impact on a patient's lifestyle [1]. Likewise, a study evaluated the oral health-related quality of life (OHRQoL), dental fear and dental beliefs before and after early prosthetic crown therapy for AI during the adolescence stage. A total of 69 patients were included with AI aged ranging 6-25 years (33 males and 36 females). Patients were instructed to complete three questionnaires measuring OHRQoL (OHIP-14), dental fear and dental beliefs respectively. 26 patients suffering from AI between ages 9 and 22 yr. received crown therapy and responded to the questionnaires before and after the crown placements. OHIP-14 scores were significantly higher among the patients than the healthy controls. Quality of life problems in the 26 patients with severe AI decreased significantly (p<0.001). However, early prosthetic therapy didn't increase dental fear or negative attitudes toward the dental treatment [7].

Restorative dental treatment of patients with a generalized form of AI is a challenging task. The treatment approach requires interdisciplinary management of several dental disciplines such as restorative, orthodontic, and prosthetic dental specialties. Several clinical reports stated the definitive prosthetic rehabilitation might restore both esthetics and function in patients with a severe form of AI [8,9].

A 5-year-old patient diagnosed with AI in the primary dentition. The treatment comprised of oral rehabilitation of the primary molars with stainless steel crowns and resin-filled celluloid crowns. The main perspectives of this treatment in a patient with AI were to enhance the esthetic appearance, restore masticatory function and to eliminate the teeth sensitivity. Treatment ultimately ended up with a positive psychological impact and thereby aided in improved quality of life. This clinical report supported the use of cost-effective crowns such as stainless crowns, and resin-filled celluloid forms to re-establish the oral functions and improve the child's psychological development [10].

Another case report was documented with an 8-year-old girl suffering from AI. A hypoplastic form of AI was diagnosed in the permanent dentitions. A multidisciplinary treatment approached was performed and categorized into preventive, orthopedic and rehabilitation stages. Initially, the preventive treatment was implemented using fluoride varnish applications to protect the fragile enamel and thereby aided in reducing the tooth sensitivity issues. The patient received an interceptive orthopedic treatment to establish the cross-relationship of the arches during the six months. Lastly, indirect composite resin crowns were performed in the rehabilitation stage to establish the vertical dimension in the posterior teeth while anterior teeth were restored with direct composite resin restorations. After 18 months of long term follow up, dental hypersensitivity and improvements in aesthetic appearances and functions as well as enhance the quality

of life was observed [11].

Another report discussed the full mouth rehabilitation of deciduous teeth affected by AI. The treatment included full mouth reconstruction with stainless steel crowns on posterior teeth and indirect composite veneers on anterior teeth was found effective in restoring esthetics, masticatory function, and improved self-confidence [12].

A 4(1/2) year old patient was diagnosed with AI. The treatment included the oral rehabilitation of the primary molars with stainless steel crowns and resin-filled celluloid forms that were placed on both the maxillary and mandibular primary incisors and canines. Normal occlusion was established that resulted in improved masticatory function and thereby played a significant role in the patient's psychological behavior and systemic health [13].

Sometimes the amelogenesis imperfect is co-existed with a special clinical condition anterior open bite. Diagnosis and treatment planning of AI patients presented clinically with open bite require a comprehensive and multidisciplinary approach to address all dental, occlusal, development, skeletal and soft tissues problems associated with these clinical conditions [14].

A clinical report discussed the treatment plan for a patient affected by AI with anterior open bite supported the placement of resin composite laminate veneers on maxillary anterior teeth and stainless steel crowns for posterior teeth that can reduce sensitivity issues thereby resulted in improved aesthetics and masticatory function [15].

Therefore, amelogenesis imperfecta affected with an anterior open bite requires a multidisciplinary team of specialists. It usually starts with conservative steps taken during the adolescence stage and the final prosthetic rehabilitation with all-ceramic crowns after reaching the adulthood [16].

A study was conducted to determine the effect of deproteinization on the clinical success of composite crowns in hypo calcified amelogenesis imperfect affected permanent dentitions. A total of 32 permanent teeth in 4 healthy children were restored with strip crowns and composite resin. Hence, the deproteinization procedure did not affect the clinical success of the adhesive restorations, however, composite restorations were clinically predictable in children with hypocalcified amelogenesis imperfect in long-term follow-up [17]. Another study also supported the use of composite prostheses helped in restoring esthetic, function and improved the quality of life [18].

Nevertheless, the longevity of dental restorations in patients with AI is limited. Therefore, a retrospective study included 82 patients with AI revealed that Patients with hypo mineralized/hypomaturized AI have restorations of shorter longevity than those with hypoplastic AI (p<0.01). Moreover, Porcelain crowns had significantly longer survival than composite resin materials in the AI group (p<0.001). Therefore, a study supported the importance of establishing an early permanent therapy plan for these patients to avoid frequent dental visits. There is a need for long-lasting restorative solutions for patients with AI [19].

Classification of AI

Based on various criteria	Hypoplastic	Hypocalcified	Hypomature
Clinical appearance	enamel is thin and stained, but normally calcified	soft enamel that can be removed without difficulty	enamel is of normal thickness, but of reduced hardness
Color	varies from yellow/brown to red/brown	varies from yellow/brown to red/brown	varies from yellow/brown to red/brown

Classification is mainly based on various criteria (pattern of inheritance, phenotypical abnormalities and molecular disorders, biochemical analysis of the enamel)

Conclusion

Definite diagnosis and adequate treatment planning are ensured for patients clinically presented with different forms of AI. Clinicians should provide preventive care and establish an early permanent therapy plan for these patients. Nevertheless, an interdisciplinary approach is required to improve both esthetic and functions. Moreover, there is a need for long-lasting restorative solutions for AI patients.

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