

Amelogenesis Imperfecta - Functional and Esthetic Rehabilitation: A Case Series

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Abstract:

The term amelogenesis imperfecta (AI) refers to a group of rare autosomal disorders expressed as defects in the enamel formation of teeth.^{1,2} The disorder is known to affect both primary and secondary teeth and may manifest with symptoms ranging from mild discoloration leading to compromised esthetics to complete mutilation of dentition leading to a loss of vertical dimension and compromised states of function.³ The restoration of vertical dimension loss in patients with enamel defects associated with AI presents a challenge for prosthodontists. Restorative treatment modalities vary from basic oral hygiene maintenance instructions to complex partial and full coverage restorations.³⁻⁵ The following case series details rehabilitation of two individuals affected by AI with completely different treatment modalities.

Key Words: Amelogenesis imperfecta, bonded restorations, esthetic rehabilitation, functional rehabilitation, hereditary disorders, oral rehabilitation

Introduction

Amelogenesis imperfecta (AI) is a term used to describe a set of hereditary disorders that express as a set of conditions, which result in deviations from the normal quality and/or quantity of enamel.¹ AI may express as an autosomal dominant or recessive genetic trait. In rare instances, it is known to demonstrate a gender-linked inheritance pattern.

Prior research indicates the prevalence to range from 1:718 to 1:140,000 with no racial predilections.⁶

The disorder is characterized by diffuse involvement of both primary and secondary dentitions. Although the enamel itself may be defective, the underlying root morphology along with quantity and quality of dentin appears to be largely unaffected.

They are classified primarily based on the phenotype, radiographic appearance, and mode of transmission:

1. Hypo-plastic: Well mineralized enamel with a reduction in quantity. The thin enamel allows dentin to show through

resulting in yellowish discoloration. The enamel present may be pitted and rough or smooth and glossy.

2. Hypomaturation: Results from a defect in the final stages of maturation of enamel matrix. The enamel typically chips off the underlying dentin. The enamel present is softer than normal and appears more opaque and yellow-brown in color.
3. Hypo-calcified: Enamel is normal in quantity but poorly mineralized. The crowns of teeth are anatomically sound on eruption but become mutilated when in function due to the poor mineralization of enamel. It is easily scraped off resulting in characteristic defects on the tooth surface.
4. Hypomaturation hypo-plastic: Described typically as a combination of 1 and 2 and is seen in the systemic disorder, tricho-dento-osseous syndrome.^{3,7-9}

The wide plethora of presentations of this disorder does not permit a single course of the treatment for every patient. The treatment protocol has to be tailored to meet the needs of individual patients and the severity of the affliction. Compromised esthetics, sensitivity to mechanical and/or chemical irritants and loss of vertical dimension leading to a breakdown of the masticatory apparatus are the most common problems associated with this disorder.³

The treatment of AI must be multidisciplinary with initial extractions and periodontal therapy ranging from oral prophylaxis to surgical interventions such as crown lengthening procedures and root coverage procedures. The prosthodontic rehabilitation of these patients may require as little as few dental restorations to full mouth reconstruction to regain lost vertical dimension with occlusal and esthetic makeovers.¹⁰ The degree of mineralization and quantity of the enamel present is a critical deciding factor in the selection of bonded partial or full coverage restorations.

This case series documents two cases of AI with different multidisciplinary treatment protocols that were customized to the individual needs of each patient.

Case Report

Case 1

A 22-year-old male presented to the Department of Prosthodontics, Saveetha Dental College and Hospital, Chennai, India with a primary complaint of severe esthetic and functional problems. He complained of generalized sensitivity on consumption of hot and cold beverages.

On thorough evaluation of the patients' history, no evidence of tetracycline use was elicited in the patient or his mother during pregnancy. The generalized enamel hypoplasia of both the primary and permanent dentition; and absence of systemic diseases that could have lead to generalized enamel hypoplasia resembling AI confirmed the diagnosis.

A palpatory and auscultatory examination of the temporomandibular joint (TMJ) was conducted along with a Krogh-Poulsen test. No temporomandibular disorders were evident. There was a marked reduction in the occlusal vertical dimension of the patient with an increase in freeway space and deterioration of the masticatory mechanism in general.

The radiographic assessment revealed no impacted permanent teeth, cysts or any other anomalies.

In the second appointment, we prepared diagnostic casts with impressions obtained in Irreversible hydrocolloid (Neocolloid-Zhermack SpA, Badia Polesine (RO), Italy) material and poured in Type III Gypsum (Orthokal-Kalabhai Karson Ltd. Vikhroli (W), Mumbai 400 079, India). An arbitrary facebow record was obtained with a spring-bow (Hanau Spring-bow - Whip MixCorp. 361 Farmington Ave, Louisville, KY 40209, USA). The centric relation and protrusive records were then made using Aluwax (5260 Edgewater Dr, Allendale, MI 49401, USA). Using the principles of phonetics and esthetics, the freeway space and desired vertical dimension was estimated at this point. The study models were mounted on a semi-adjustable articulator (Whip MixCorp. 361 Farmington Ave, Louisville, KY 40209, USA) using the facebow, centric and protrusive intra-oral records. A mock up and diagnostic wax-up were done at the planned vertical dimension followed to establish idealized anterior tooth morphology, esthetics and lateral, and protrusive guidance. An occlusal splint replicating the desired vertical dimension was fabricated in clear acrylic and worn by the patient over a period of 3-month for at least 22 h a day. It was well-tolerated.

Provisional crowns were then fabricated using the diagnostic mock-up as a template prior to the preparation of teeth along with a stent replicating the desired positions of the gingival zenith lines (Figure 1). This served as a guide in the crown lengthening procedure. Tooth preparations to receive Porcelain fused to metal crowns were carried out immediately after the crown lengthening procedure, and the temporary crowns were relined and cemented in place in the same appointment (Figure 2). The centric contacts, lateral and protrusive guidance was verified and refined intra-orally.

The temporary restorations were kept in place for a month during which time the patient was asked to report to the department on a weekly basis for further evaluation and adjustments of occlusion. The restorations were well-tolerated and required minimal adjustments during this time. The

anterior restorations were removed twice for adjustment and polishing of the gingival segment to aid in papilla fill. Porcelain fused to metal restorations were fabricated in accordance with the provisional restoration and cemented with glass ionomer cement (GC-Fuji-I, GC India Inc.) (Figures 3 and 4).

The patient was recalled to the department at 1 week, 3 months, 6 months, and 1-year intervals. During the follow-up, he was



Figure 1: Stents for crown lengthening procedure of anterior teeth.



Figure 2: Tooth preparation for porcelain fused to metal restorations and prefabricated temporary restorations.



Figure 3: Post rehabilitation intra oral view.

comfortable with the rehabilitation. He was well satisfied with the esthetics and function.

Case 2

A 19-year-old girl reported to the department with a primary complaint of unaesthetic front teeth. She had an otherwise healthy and functional dentition and no complaints of loss of function or tooth sensitivity.

A thorough evaluation of the patients past medical history was carried out to rule out long-term consumption of drugs, which could lead to similar symptoms of yellowish discoloration of teeth with the pitted appearance of the enamel. No positive history was elicited in this regard. She also had a history of similar defects being present in the primary dentition. Fluorosis was also ruled out due to distinct differences in the clinical presentation. The patient's sibling also demonstrated similar defective enamel. These observations were consistent with a diagnosis of AI. Radiographic findings of decreased radio-opacity of the enamel corroborated the diagnosis. TMJ evaluation yielded no signs or symptoms of dysfunction.

Intraoral examination confirmed no loss of vertical dimension and the tooth anatomy was intact. Generalized discoloration of teeth was present. A single tooth crossbite was recorded in relation to the 23 and 34. The patient was presented with a comprehensive treatment plan including orthodontic correction of the crossbite. As an alternative, she was recommended corrective therapy with endodontics and fixed dental prosthesis in the 23 and 34. As her primary concern was the esthetics in her anterior dentition, she requested that treatment is limited to a conservative esthetic correction involving only the maxillary and mandibular anteriors. She was informed about the esthetic and possible functional compromise. A final treatment plan of rehabilitation of esthetics with ceramic laminate veneers on all anterior teeth with the exception of 23 was planned and received the patient's approval.

Diagnostic impressions were made in irreversible hydrocolloid impression material (Neocolloid - Zhermack SpA, Badia Polesine [RO], Italy) and pore in Type III Gypsum (Orthokal- Kalabhai Karson Ltd. Vikhroli (W), Mumbai 400 079, India). An arbitrary facebow record was obtained using a spring-bow (Hanau Spring-bow- Whip MixCorp. 361 Farmington Ave, Louisville, KY 40209, USA) and mounted on a Hanau Wide-View semi-adjustable articulator (Whip MixCorp. 361 Farmington Ave, Louisville, KY 40209, USA) using a centric and protrusive wax bite record.

An anterior esthetic mockup was done. Since it was decided to forego the 23 for esthetic rehabilitation, a part masking would be achieved by only partly correcting the proclination of the 22. Tooth preparation was completed retaining the contacts

of natural dentition with a lingual wrap preparation and long chamfer finish lines (Figure 5). Direct composite provisional restorations were fabricated and cemented in place. Laminate veneers were prepared with e-max alumina and cemented in place (Figure 6) with Panavia A2 (Kuraray Medical Inc. Japan) resin cement. A debonding of the ceramic veneer in 21 was reported after 3 months and required re-cementation. At 1 year follow-up patient reported, no further problems with the rehabilitation.



Figure 4: Post rehabilitation extra oral view.



Figure 5: Tooth preparation for laminate veneers with gingival retraction.



Figure 6: Post-operative view with patient in mandibular protrusion.

Discussion

Active dental intervention in a complex AI case may span several decades, involving the participation of various dental specialties. The age-related treatment approach involves:

- Temporary or Initial phase (primary dentition 2-6 years)
- Transitional phase (early mixed dentition 6-11 years)
- Final permanent phase (mixed to permanent dentition).¹¹

Both cases presented here reported for treatment at a later stage in life where we undertook the final permanent stage of treatment. Lindunger and Stenberg reported in their study that half of a sample of patients with AI expressed a desire to have started their rehabilitation before the age of 16.¹² Lower self-esteem, social avoidance, negative psychological outcomes, and the high cost of extensive restorative care are other problems that patients with AI face. An early intervention with a multi-disciplinary approach improves the psychological state of patients making them more tolerant of further treatment in the transitional and final phase of treatment. Bouvier *et al.*, reported a case of a 10-year-old girl in whom they performed initial treatment for anteriors. This resulted in a spectacular psychological transformation and motivated the patient for further treatment.¹³ Had the patient in the second case report received an early intervention it may have convinced her for a more comprehensive treatment plan that was presented to her at the onset.

It is well-documented that patients with AI often present with anomalies such as impacted permanent teeth and follicular cysts. The tendencies of such anomalies may be up to six-fold that of a normal unaffected individual. Bearing this in mind, it is of paramount importance that a thorough radiographic evaluation must be performed prior to development of a treatment plan.^{5,14,15} Both cases reported in this series presented no such anomalies.

The hypoplastic variant of AI typically presents with a sufficient quantity of enamel to function well with bonded restorations such as direct or indirect composite or ceramic to meet the esthetic challenge presented by the discolored teeth.² This was evident in the second case in the report where a sufficient quantity of tooth material permitted the use of more conservative treatment to meet the esthetic need of the patient. The teeth despite being discolored exhibited enough tooth structure to permit a sufficient bonding surface for adhesive restorations. A noteworthy study while studying micro-tensile bond strength of enamel from the hypoplastic variant reported a lowering and bond strength as compared to normal enamel¹⁶ and this may be the cause of debonding of the veneer in the 21 regions.

The most severe problems occur with the variants that demonstrate the lowest amount of enamel such as the hypocalcified and the thin smooth hypoplastic. The patients present with loss of vertical occlusal dimension and masticatory

dysfunction. The treatment plan must be developed with a view to prevent further breakdown while bearing in mind the increased susceptibility of these teeth to dental caries. The lost dimension in the first patient was attributed to the deficits in enamel structure and the erosion of the exposed dentin. Full coverage restorations with predetermined increased dimension was hence the treatment of choice for the first patient. The coverage will prevent further compromise of dentition while maintaining masticatory efficiency.¹⁷ The increase in vertical dimension must be gradually obtained with sufficient time provided to a patient to acclimatize. In this case, an increase of 3 mm was sufficient to obtain desired esthetics and function. The splint must be worn for a sufficient period of time usually a minimum of 3 months with about 22 h of wear per day.

The clinical crown height of the natural dentition had been limited by the breakdown of the natural dentition in the posterior region. This is a common finding in patients of AI. Full mouth flaps with crown lengthening procedure are usually required to provide an optimum clinical crown height to allow retention of full coverage restorations.¹⁸ To aid in maximizing retention, the final restorations were fabricated in splinted units in the posterior region.

Patients after an extensive rehabilitation must be motivated to practice good oral hygiene maintenance with adjuncts to dentifrices such as mouthwashes, dental floss, and interdental brushes. The patients, in this case, series maintained meticulous oral hygiene in the year that they were followed up after treatment and required minimal prophylactic intervention on the recall visits.

Conclusion

The importance of early intervention is profound in improving the psychological well-being and quality of life of patients afflicted with AI.

The predictable, stable and long-term outcome can be achieved as a result of thorough investigation and treatment planning. The cohesive intervention of multiple disciplines is required to achieve this end. Treatment must be directed at the achievement of a comfortable, pain-free, esthetically acceptable and functionally sound masticatory apparatus. With the achievement of these objectives, we as health care providers will be able to enable these patients to lead a normal life without social, emotional and physical disturbances.

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