

## CASE REPORT

# Functional and Esthetic Rehabilitation of a Young Patient with Amelogenesis Imperfecta

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## ABSTRACT

Amelogenesis imperfecta (AI) is a hereditary disorder that expresses a group of conditions that cause developmental alterations in the structure of enamel. Rehabilitation of patients with dental anomalies like AI is a challenge in terms of both function and esthetics for the patient. This article describes the sequenced treatment for a young male patient with mutilated natural dentition caused by AI of the hypoplastic type using Hobo's twin-stage technique of full mouth rehabilitation. The aim of the treatment was to restore esthetics and optimal masticatory function using an interdisciplinary approach. Electromyography was used as an advanced means of diagnosis and an aid to verify the clinical outcome.

**Keywords:** Amelogenesis imperfecta, Enamel, Full mouth rehabilitation, Hypoplastic.

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## INTRODUCTION

Amelogenesis imperfecta (AI) is a hereditary disorder involving both primary and permanent dentitions. It comprises a group of conditions causing alterations in the development and structure of enamel. The affected teeth exhibit normal dentin and root form and are found to be more resistant to decaying.<sup>1-3</sup>

The developmental alterations in enamel structure have been found to be unrelated to systemic disease.

Depending on the population studied, a prevalence of AI from 1:700 to 1:8,000 has been reported.<sup>4</sup> The clinical picture of AI presents with a thin enamel layer (hypoplastic), rough texture (hypomature), opaque white to yellow-brown color (hypocalcified), a mottled appearance, or association with one or two characteristics. To determine the presence of AI, an accurate diagnosis with other enamel defects and verification of alteration in symmetric pattern linked to genetic inheritance are mandatory.<sup>5</sup> Congenitally missing teeth, abnormalities in dental eruption, pulpal calcifications, anterior open bite, root and crown resorption, root malformations, hypercementosis, taurodontism, etc., are other features associated with AI.<sup>6</sup>

This article describes the sequenced treatment for a young male patient with mutilated natural dentition caused by AI of the hypoplastic type. The aim of the treatment was to restore esthetics and restore optimal masticatory function using an interdisciplinary approach.

## CASE REPORT

A 20-year-old patient reported to the Department of Prosthodontics with the chief complaint of sensitivity and difficulty in chewing food due to worn-out teeth. He gave a history of stained teeth since birth, which were worn out over the years. Patient was highly concerned with the unesthetic appearance of his teeth. He also reported to have a younger brother with hypoplastic spots on the teeth. Extraoral examination revealed pain on opening associated with the left temporomandibular joint (TMJ), deviation of the mandible to the left, tenderness on palpation with masseter muscle, positive clench test, and negative load test. Intraoral examination revealed missing maxillary and mandibular left first molars, generalized attrition, generalized brownish yellow staining of teeth, loss of enamel involving all surfaces, decreased dimension of teeth, mild gingivitis, anterior open bite, edge-to-edge incisal relation (Fig. 1A), group function occlusion, thick and ropy saliva, and a freeway space of 2 mm, root canal treated 47, 48, and a high smile line. Orthopantomogram (Fig. 1B), intraoral periapical radiographs, and extraoral TMJ radiographs were used for further investigation. Radiological findings were thin radiopaque layers of enamel with normal pulp chambers and root

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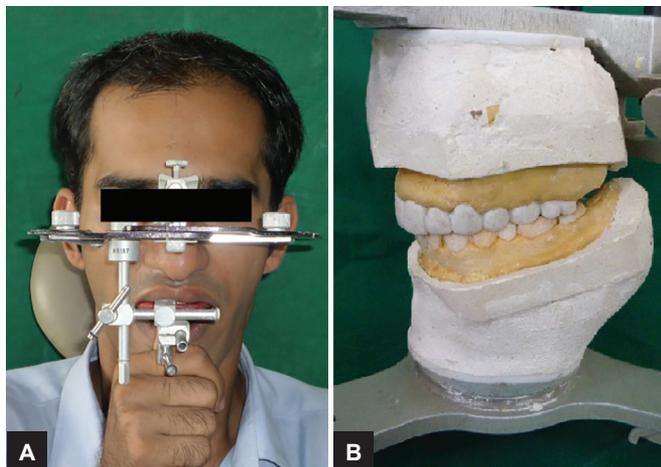
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**Figs 3A and B:** (A) Facebow record; and (B) wax mock-up on study models

**Phase III**

- Teeth preparation (segmental approach) and impression making using addition silicone double mix, double step (Ad-Sil putty and light body, Prime Dental Pvt. Ltd., Mumbai, Maharashtra, India).
- Deprogramming using Lucia jig (Pattern resin, GC Corporation, Tokyo, Japan) (Fig. 4A) followed by

bite registration using bite registration wax (Alu wax, Maarc, Shiva products, Thane, Maharashtra, India).

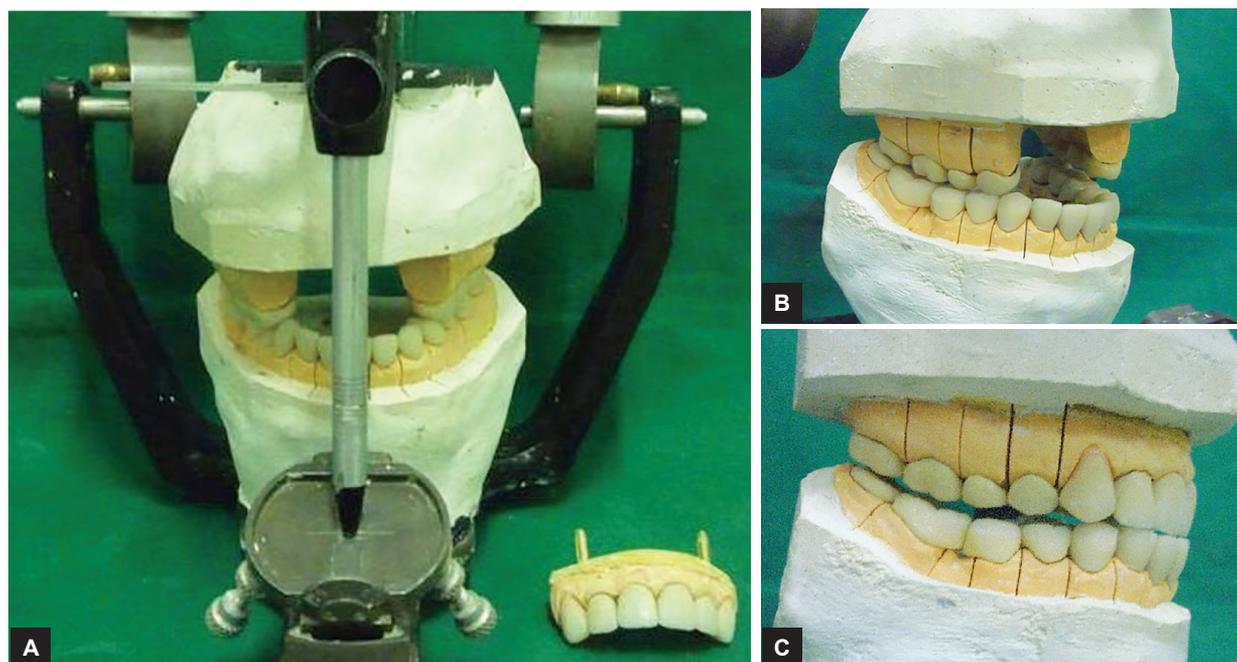
- Provisional acrylic resin crowns (heat-polymerized tooth-colored acrylic resin, DPI, Mumbai, India) (Fig. 4B), fabricated using wax mock-up as a reference at the planned vertical dimension, were cemented. (Patient was kept under review for 3 months to evaluate response).
- After evaluation and necessary adjustments, coping trials were taken (Figs 4C and D).
- Ceramic build-up was done using Hobo’s technique (Table 1 and Figs 5A to C) as a guidance to establish canine-guided occlusion.
- Posterior crowns were cemented first using glass ionomer cement (GIC luting cement, GC Corporation, Tokyo, Japan) followed by anteriors.
- Porcelain fused to metal restorations with 14 to 17, 24 to 27, and 34 to 37, 44 to 47; fabricated using Ni-Cr alloy coping (Bellabond plus, BEGO Co, Bremen, Germany) and layered with ceramic (VMK 95, VITA Zahnfabrik, Bad Säckingen, Germany).
- All-ceramic crowns (Lava, 3M ESPE, Seefeld, Germany) with 13 to 23, 33 to 43 (Fig. 6A).



**Figs 4A to D:** (A) Deprogramming using Lucia Jig; (B) provisionalization; (C) maxillary coping trial; and (D) mandibular coping trial

**Table 1:** Articulator adjustment values for twin-stage procedure

Conditions	Condylar path		Anterior guide table	
	Sagittal condylar path inclination	Bennett angle	Sagittal inclination	Lateral wing angle
Condition 1	25	15	25	10
Condition 2	40	15	45	20

**Figs 5A to C:** (A) Removable maxillary anterior segment; (B) Condition 1; and (C) Condition 2

#### Phase IV

- Oral hygiene reinforcement and dietary advice given.
- Postoperative evaluation (Figs 6B and C).
- Postoperative EMG records to evaluate effect of treatment (Fig. 6D).
- Recall and maintenance.

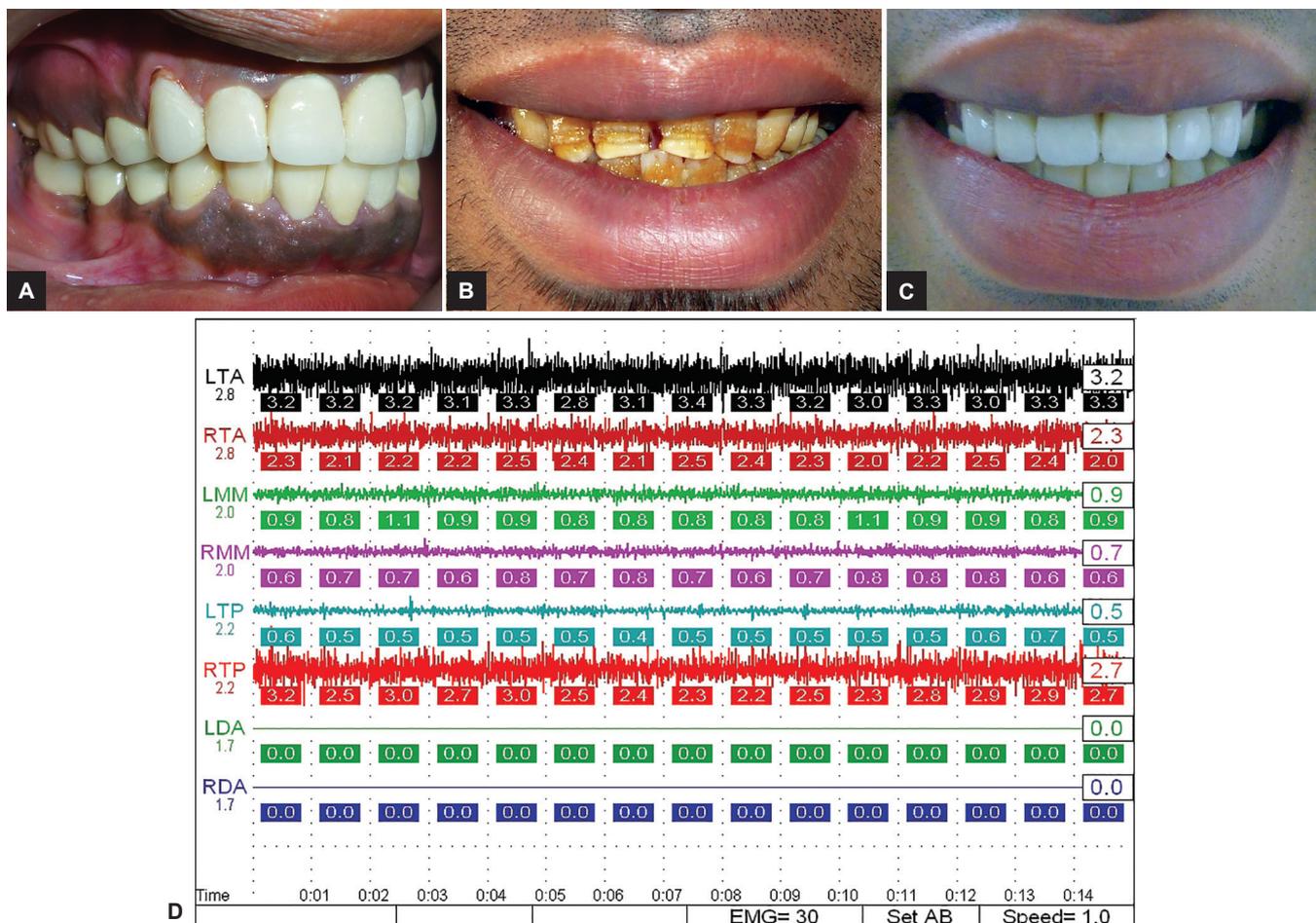
#### DISCUSSION

Various restorative procedures can be used to treat AI. The treatment for patients with AI is related to many factors including the age of patients, the type and severity of the disorder, its intraoral manifestation, esthetic and functional demands, and the socioeconomic status. Restoration of these defects is important from a functional as well as esthetic point of view, as it manifests a deep psychological impact on the patient (Figs 6B and C).

Classifications of AI are primarily based on mode of inheritance and phenotype. The most commonly used classification was proposed in 1988 by Witkop, and revised by Nusier in 2004. Depending on appearance of enamel and the hypothesized developmental defects, AI is classified into four patterns: Hypoplastic, hypomaturational, hypocalcified, and hypomaturational-hypoplastic.<sup>1-3,7</sup> The hypoplastic type is characterized by well-mineralized

enamel, but its amount is reduced as seen in the radiograph<sup>8</sup> (Fig. 1B).

Full mouth rehabilitation is a treatment modality that not only focuses on the esthetics and functional aspect of the dentition, but also extensively improves upon the health of the whole stomatognathic system. It ranges over an extensive period, so patient cooperation plays an important role. The segmental approach of teeth preparation was followed since it decreases chair-side time, requires segmental anesthesia, improves patient comfort, and helps maintain vertical dimension. In this case, progressive wear of teeth coupled with passive eruption maintained the vertical dimension, thus eliminating the need to raise the vertical dimension. Sufficient crown height was obtained by planned surgical crown lengthening. Putty (addition silicone) indices of the wax mock-up were used as a guide for teeth preparation and temporization. The occlusion was developed at the centric relation position of the mandible to provide an even and stable occlusion without pathogenic deflective contacts. Lucia jig was used for deprogramming the mandible from the learned neuromuscular pattern; to change it from tooth-guided intercuspal position to joint-guided centric relation contact position movement and facilitate easier maneuver into centric relation. Heat-cured acrylic resin provisionals were preferred over



**Figs 6A to D:** (A) Posttreatment occlusion; (B) pretreatment view; (C) posttreatment view; and (D) posttreatment EMG record

autopolymerizing acrylic resin due to its strength, durability, resistance to discoloration, and closer resemblance to final restorations. All-ceramic crowns for anterior teeth and porcelain fused to metal crowns for posterior teeth were used to provide satisfactory esthetic results. As the palatal and lingual surfaces of the teeth exhibited pitting and malformation, more conservative treatment options like composite laminates or veneers were not used.

In the Hobo’s technique used, a cast with a removable anterior segment was fabricated (Fig. 5A) and occlusal morphology of the posterior teeth was reproduced without anterior segment. This resulted in a cusp angle coincident with the standard values of the effective cusp angle (Condition 1) (Fig. 5B). This was followed by reproduction of anterior morphology using the anterior segment, thus providing anterior guidance, which produces a standard amount of disclusion (Condition 2)<sup>9</sup> (Fig. 5C). This method uses the standard values proposed in the twin-stage technique, and relies on the factor of cuspal angle to achieve a centric occlusion and an excursive disclusion. Condylar path has always been considered as one of the key determinants of occlusion. As per the work of Hobo and Takayama,<sup>9</sup> condylar path exhibits deviation and has a minor influence on disclusion.

Studies showed that with every degree of rise in horizontal condylar guidance, disclusion is increased by only 0.020 mm during protrusion, by 0.015 mm on nonworking side, and by 0.002 mm on the working side. Anterior guidance and the cusp angle are important determinants for achieving disocclusion and thus, the average angulations of the horizontal condylar guidance can be successfully used as a guide to achieve mutually protected occlusion.<sup>10</sup> The choice of the method for rehabilitation selected depends upon the clinical case, operator skills, and convenience.

The posttreatment EMG record showed definite improvement in muscle activity, indicating a positive treatment outcome (Fig. 6D). A combination of new generation materials, along with improved clinical procedures helps produce an esthetic, long-lasting, and functional outcome, which satisfies both the clinician and the patient.

**CONCLUSION**

The AI should be diagnosed at the earliest possible age and intervened with an interdisciplinary approach to provide a long-term predictable survival of restorations. The clinical outcome depends on the complexity of the case and execution of the proposed treatment plan.

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