PROSTHETIC REHABILITATION OF AMELOGENESIS IMPERFECTA – A CASE REPORT

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Abstract

This clinical report describes the oral rehabilitation of a young adult patient diagnosed with hypomature amelogenesis imperfecta. The specific objectives of this treatment were to eliminate tooth sensitivity while enhancing esthetics and restoring masticatory function. Porcelain fused to metal full veneer crowns were placed on anterior and posterior teeth to modify the occlusion and to improve the esthetics. Clinical examination 12 months after treatment revealed no evidence of disorders associated with the restored teeth or their supporting structures.

Key words: Amelogenesis Imperfecta, Hypoplastic full mouth rehabilitation

Introduction

Amelogenesis Imperfecta (AI) has been defined as a group of hereditary enamel defects not associated with evidence of systemic disease. There are at least 12 distinct types of amelogenesis imperfecta[1, 2] based on combination of clinical, radiographic, histological, and genetic criteria. On the clinical and radiographic basis alone, 3 broad groups can be distinguished: (1) hypoplasia, in which the enamel is reduced in quantity but is relatively well-mineralized; (2) hypocalcification, in which enamel is formed in relatively normal amounts but is poorly mineralized; and (3) hypomaturation, in which the final stages of the mineralization process are abnormal. Various authors have classified amelogenesis as:

Weinmann *et al.*, 1945 [3]	***Two types based solely on phenotype: hypoplastic and hypocalcified***
Darling, 1956 [4]	*Five phenotypes based on clinical, microradiographic and histopathological findings.* Hypoplastic Group 1 – generalised pitting Group2 – vertical grooves (now known to be X-linked AI) Group 3 – Generalised hypoplasia Hypocalcified Type 4A – chalky, yellow, brown enamel Type 4B – marked enamel discolouration and softness with post-eruptive loss of enamel Type 5 – generalised or localised discolouration and chipping of enamel
Witkop, 1957 [5]	*Classification based primarily on phenotype. 5 types:* 1. Hypoplastic 2. Hypocalcification

	3. Hypomaturation 4. Pigmented hypomaturation 5. Local hypoplasia Added mode of inheritance as further means of delineating cases.
Schulze, 1970 [6]	*Classification based on phenotype and mode of inheritance.*
Witkop and Rao, 1971 [7]	*Classification based on phenotype and mode of inheritance. Three broad categories: hypoplastic, hypocalcificied, hypomaturation.* **a.** Hypoplastic Autosomal dominant hypoplastic-hypomaturation with taurodontism (subdivded into a and b according to author) Autosomal dominant smooth hypoplastic with eruption defect and resorption of teeth Autosomal dominant rough hypoplastic Autosomal dominant pitted hypoplastic Autosomal dominant local hypoplastic X-linked dominant rough hypoplastic **b.** Hypocalcified Autosomal dominant hypocalcified **c.** Hypomaturation X-linked recessive hypomaturation Autosomal recessive pigmented hypomaturation Autosomal dominant snow-capped teeth White hypomature spots?
Winter and Brook, 1975 [8]	*Classification based primarily on phenotype. Four main categories: hypoplasia, hypocalcification, hypomaturation, hypomaturation-hypoplasia with taurodontism, with mode of inheritance as a secondary means of sub-classification.* **a.** Hypoplasia Type I. Autosomal dominant thin and smooth hypoplasia with eruption defect and resorption of teeth Type II. Autosomal dominant thin and rough hypoplasia Type III. Autosomal dominant randomly pitted hypoplasia Type IV. Autosomal dominant localised hypoplasia Type V. X-linked dominant rough hypoplasia **b.** Hypocalcification Autosomal dominant hypocalcification **c.** Hypomaturation Type I. X-linked recessive hypomaturation

**(Table adapted from article by Peter JM Crawford, Michael Aldred and Agnes Bloch-Zupan, Amelogenesis imperfecta; Orphanet Journal of Rare Diseases 2007, 2:17)*

The clinical features that distinguish the hypoplastic from hypocalcified are that in the hypoplastic forms, the enamel does not develop to its normal thickness whereas the hypocalcified forms, the enamel thickness on the newly erupted teeth closely approaches that of normal teeth, however the enamel is soft, friable, and can easily be removed from the dentin. In contrast to hypoplastic types, the hypomaturation types develop enamel of normal thickness. Notably the hypomaturation forms differ

from hypocalcification in that the enamel is harder, with a mottled opaque white to yellow-brown or red-brown color, and tends to chip from the underlying dentin rather than wear away.[9]
According to Seow[10], the primary clinical problems of AI are esthetics, dental sensitivity, and loss of occlusal vertical dimensions. However, the severity of dental problems experienced by patients varies with each type of AI. Historically, treatment of patients has included multiple extractions and the fabrication of complete dentures. These options are psychologically harsh particularly when the problem must be addressed in adolescent patients. The treatment plan for patients with AI is related to many factors including the age of the patient, the socioeconomic status of the patient, the type and severity of disorder, and the intraoral situation at the time the treatment plan is developed. This clinical report describes the treatment sequence of a 19-year-old female patient with hypomature amelogenesis imperfecta.

Clinical Report

A 19-year-old girl who was self-conscious about tooth appearance and suffered from considerable tooth sensitivity presented for treatment. The patient was referred to the Department of Prosthodontic Dentistry in PMS College of Dental Science and Research, Trivandrum for evaluation and treatment. Prior to treatment, a detailed dental, medical, and social history was obtained from the patient.

Clinical and radiographic examination of the patient revealed total loss of enamel from the tooth surface, a asymptomatic Temporomandibular Joint with functional Angle Class I dental relationships with deep bite and a deep carious lesion in relation to 46, root stump in relation to 16 and bucally placed 25 (Fig 1,2, 3& 4).

Biochemical tests were done such as blood investigation and saliva collection using oragene kits, the enamelin gene was mapped to chromosome 4q21, to check for gene mutations.
However, evaluation of the gene did not reveal any mutation(s) in the coding regions that were associated with the disorder.

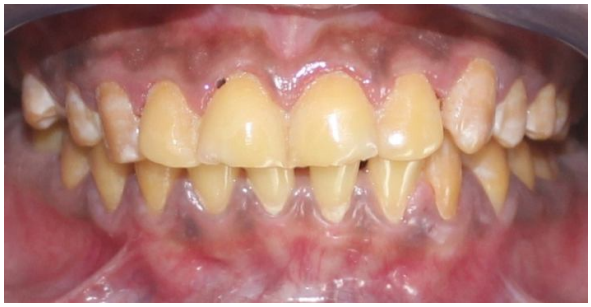
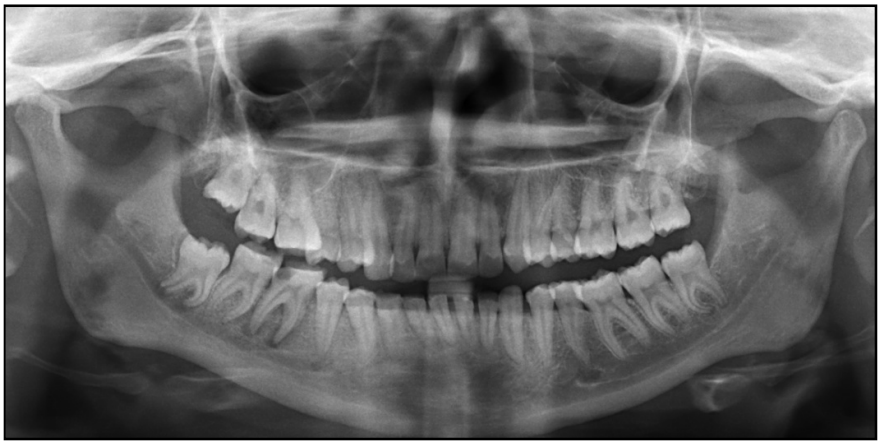


Figure: 1



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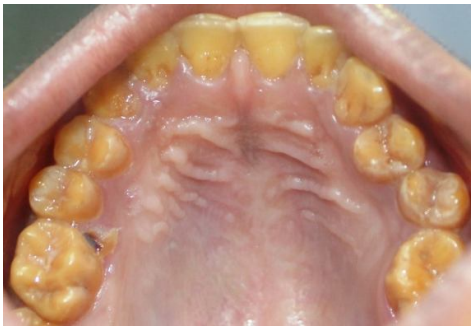

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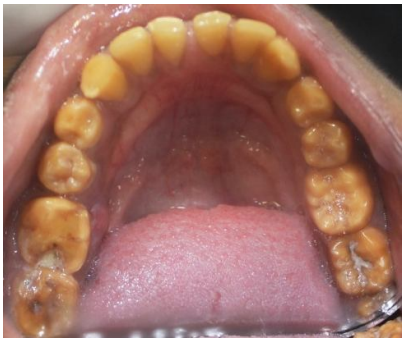


Figure: 4

Due to the complex needs of the patient, an interdisciplinary approach was followed. A treatment plan was developed with the aims of restoring the carious teeth, restoring masticatory function, reducing the reported tooth sensitivity, restoring lost vertical dimension and improving the patient's appearance with full veneer crowns.

After the extraction of root stump in relation to 16 and the prescribed healing period an accurate diagnostic casts were made with irreversible hydrocolloid (Neocolloid, high precision impression material, Zhermack), and was mounted in centric relation based on occlusal records (occlufast, Zhermack) on a semi-adjustable articulator (Hanau Wide- Vue Arcon articulators) using a face-bow (Hanau springbow) transfer. (Fig 5 & 6).

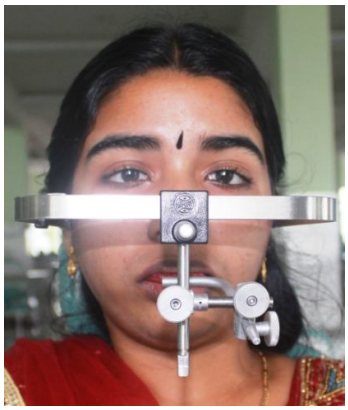
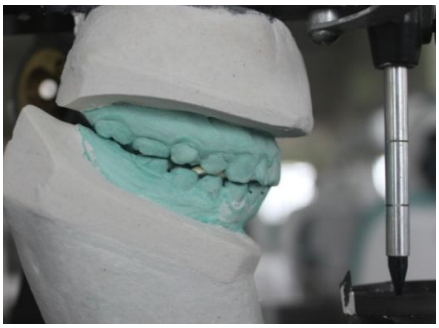


Figure: 5

Figure: 6

On the articulator, vertical dimension was increased by 2 mm as determined and confirmed by clinical evaluation (Fig 7). A diagnostic wax up was performed perfecting the occlusion with functional posterior and anterior guidance providing disocclusion on protrusive and lateral excursions in canine guided occlusion. Tooth preparation was done intraorally one arch at a time. Impressions were made in addition silicone polyvinyl siloxane (Aquasil Soft putty/ regular set, Dentsply & 3 M ESPE Express XT light body) and the provisional restorations were made using putty index of the mock wax up and was luted with zinc phosphate cement (De Trey Zinc, Zinc phosphate cement, Densply)(Fig 8). Occlusal relations were checked and then refined in patient's mouth.



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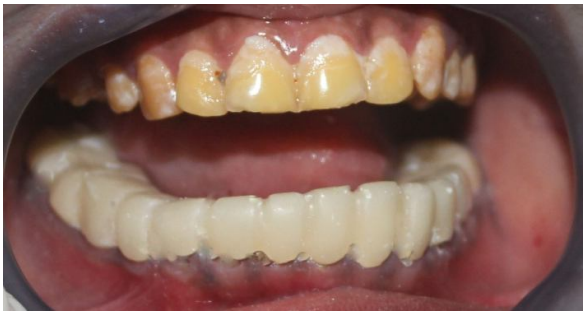
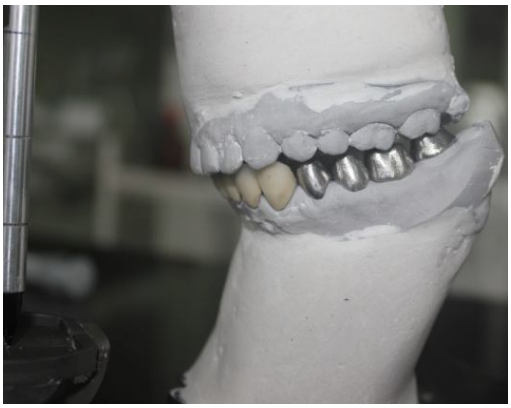


Figure: 8

Jaw relations were recorded with provisionals in the lower arch. Anterior bisque and posterior metal trial was done first in the semiadjustable articulator followed in the patient's mouth and checked for anterior guidance and clearance in excursive movements(Fig 9). After the cementation of the porcelain fused to metal full veneer crowns in relation to the lower arch using Glass ionomer cement (GC Fujicem paste pak). Upper anterior bisque trial and posterior metal done was done similarly to the lower jaw duplicating the contours of the provisionals. (Fig. 10,11,12,13,14,15,16).



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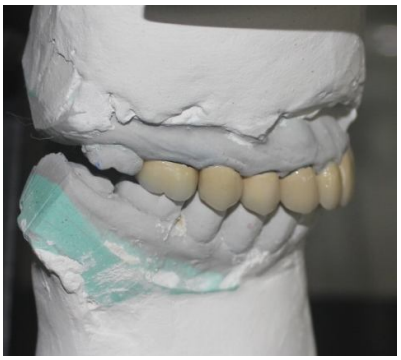

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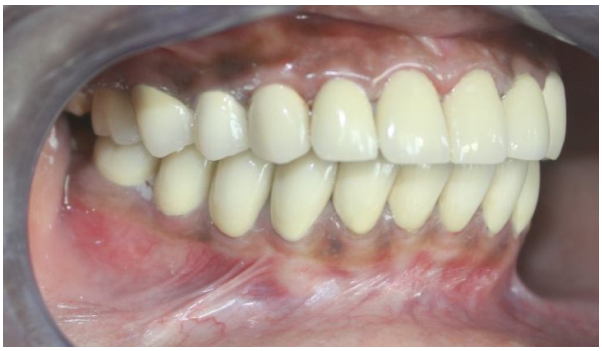

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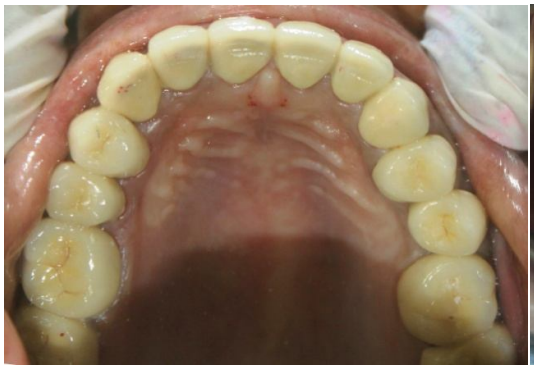

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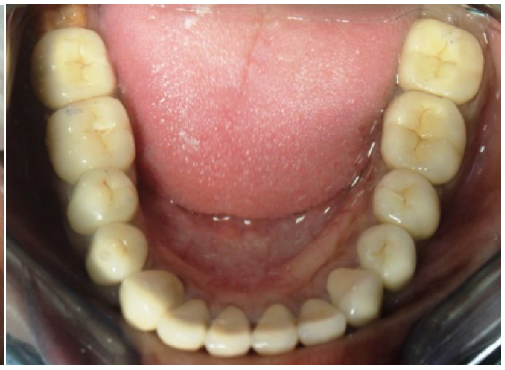

Figure: 14

Figure 15

Figure: 16

Summary

The supportive clinical care needed by these individuals is substantial both in terms of clinical and emotional demands. Adolescents in particular have been known to become reclusive and withdrawn, and even inclined to suicide because of their disfigured teeth and related to aesthetics.

Treatment is as ever based on the principles of prevention before intervention. However, in these patients' cases, intervention will likely be earlier and more radical than for others. The progression of treatment during childhood has been described as a temporary phase followed by a transitory phase [11].

In infancy, the primary dentition is protected by the use of preformed metal crowns on posterior teeth; either polycarbonate crowns or composite restorations are used on anterior teeth. The eruption of the permanent dentition, beginning at six years of age, presents a particularly difficult period. Some of the forms of AI present with hypersensitive teeth or with teeth that crumble, and both presentations provide a very real disincentive to good oral hygiene and are very difficult to restore. Those cases with enamel which is reasonably hard (*i.e.* less hypomineralised) and thin (*i.e.* more hypoplastic) lend themselves fairly readily to the use of preformed metal crowns on posterior teeth, as they erupt and composite restorations on anterior teeth. These latter may need to be added to as more of the cervical part of the tooth is revealed. Restorative treatment requires local analgesia at least.

Children with AI are not without malocclusions and it is important that a restorative dentist and an orthodontist are involved with the pediatric dentist in the care plan from the child's early age. It is the pediatric dentist's role to deliver the patient to the restorative dentist a patient who is motivated, with good oral care practices and with no treatment option compromised by previous activity. The anterior open bite seen in some cases of AI requires consideration of surgical as well as restorative management.

This clinical report describes the procedure for full mouth rehabilitation of a hypomature type of amelogenesis imperfecta using porcelain fused metal ceramic crowns. The report describes a method of rehabilitating one arch at a time for patient comfort and convenience and at the same time gives a perfect occlusion which is critical for longtime maintenance and success of the restorations.

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