

Case Reports

Comprehensive Prosthodontic Management of a Patient with Amelogenesis Imperfecta

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Amelogenesis imperfecta (AI) is a referred to a group of rare, inherited disorders characterized by abnormal enamel formation^{1,2}. It occurs as a result of mutations in five genes including; AMEL, ENAM; MMP20; KLK4 and FAM83H). It follows an autosomal dominant, autosomal recessive or x-linked pattern of inheritance. Usually both the primary and secondary dentition is affected. Phenotypically, AI is classified by the clinical researchers into 4 main types including; hypoplastic, hypomaturational, hypocalcified, and hypomaturational-hypoplastic^{2,3}.

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Clinically AI presents differently according to the type involved. Hypoplastic form of AI is characterized by reduced enamel thickness with yellowish-brown discoloration, rough or smooth and glossy attrited tooth surface, and with/without grooves and/pitting. Hypocalcified type is characterized by discolored-dark brown and softer enamel which wears down rapidly, though the size and shape of crown remains adequate. Whereas in hypomaturational type, enamel of adequate thickness is present but it is still softer than normal enamel with mottled-colored cloudy white/yellow/brown discoloration^{4,5}.

The most common complaints in the patient with AI revolve around compromised esthetics, dental sensitivity and functional limitation due to worn down teeth. These problems can eventually affect the psychological well

ABSTRACT

Each case of amelogenesis imperfect should be properly diagnosed and planned in order to achieve the best esthetic and functional outcome. The age, severity and type of amelogenesis imperfect along with the financial status of the patient should be considered while formulating a treatment plan for such patient. This case report outlines the stepwise management of a young female patient with Amelogenesis Imperfecta- Hypocalcified type.

Keywords: Full coverage restorations, Hypocalcified form, Inherited disorders, treatment

being of the patient, therefore it must be addressed in a timely manner⁶.

The aims of a treatment plan for a patient with AI should be timely and accurate diagnosis, reduction of pain/sensitivity, prevention and restoration followed by long term maintenance. A multidisciplinary approach is therefore needed to diagnose and carefully address the functional and esthetic complaints of these patients⁷. In this case report the aim of the author is to describe the possible prosthodontic management of a patient with amelogenesis imperfecta of the hypocalcified type, in order to improve both function and esthetics.

CASE REPORT:

A 26-year-old female reported to the prosthodontics

department, AFID with a chief complaint of compromised esthetics due to small sized discolored teeth and difficulty in chewing. The patient belonged to lower middle class. Medical history was noncontributory, although family history revealed that two of her cousins suffered from the same condition.

The general physical examination of the patient revealed a young patient of lean built, well oriented in time, space and person sitting comfortably in the dental chair. Her vitals were within normal limits. The extra-oral examination of the patient revealed a symmetrical face without any facial swelling, adequate mouth opening with no deviation or deflection upon opening. The patient had decreased lower facial height with no detectable pathology in temporomandibular joint. The intra oral examination revealed generalized discoloration, chipped off enamel and rough tooth surface. The teeth were small in size with crowding and plaque and calculus deposition. Soft tissue examination showed mild generalized gingivitis. (Figure 1)



Figure 1: Intra-oral view

Study casts, photographs and radiographs were acquired to aid in diagnosis of the condition. On the basis of these investigations and clinical findings, patient was diagnosed as Amelogenesis Imperfecta- Hypocalcified type.

Different treatment options were discussed in detail with the patient, which included veneers, full coverage restorations, overlay denture and over dentures. She opted for full coverage restorations. Before beginning the treatment, the patient was informed about the details of the procedure including the amount of tooth structure removal, the need for endodontic therapy, the expected clinical longevity and the duration of treatment. Treatment was started with the some objectives in mind including, conservation of maximum tooth structure, establishment/restoration of the lost vertical dimension of occlusion (OVD) to a physiological neuromuscular stable position, improved masticatory function and enhancement of the esthetics.

Patient was referred to the Periodontology department for Oral prophylaxis with thorough scaling and root planning. Moreover, the gingivitis was also addressed by the expertise of a periodontist.

All the completely erupted teeth (17, 16, 15, 14, 13, 12, 11,

21, 22, 23, 24, 25, 26, 27, 36, 35, 34, 33, 32, 31, 41, 42, 43, 44, 45, 46, 47) underwent Root Canal Therapy. Figure 2 shows the panoramic radiograph after completion of root canal therapy of all erupted permanent teeth.



Figure 2: Panoramic radiograph after completion of RCT's

Further maxillary and mandibular impressions were recorded for diagnostic casts. Diagnostic wax up was carried out at a re-established OVD and an occlusal splint was fabricated at the same OVD to evaluate the patient's adaptability to altered OVD. She was advised to wear the splint 24 hours, except during eating and teeth cleaning for the next 3 months. The increased OVD was well tolerated by her. Cranio-maxillary relationship was recorded using a facebow which was then transferred to the articulator (Figure 3 and 4)



Figure 3: Facebow transfer

Later on, a centric relation record was made at an increased OVD to mount the lower cast in relation with the upper one. PAP approach was followed for full mouth rehabilitation. Crown preparation of the lower right posteriors #44,45,46,47 was initiated followed by impression recording using Polyvinyl Siloxane material. Temporary crowns were made and cemented chair side, occlusal splint was adjusted according to the crowns.



Figure 4: Facebow record transferred to the articulator using a transfer jig

In the similar manner, the lower left posteriors were prepared followed by impression recording, chair side temporary crowns fabrication and cementation. The dental casts with the prepared teeth were articulated with the help of the CR record. Metal framework trial of the permanent restorations was carried out to check the marginal fit. (Figure 5)

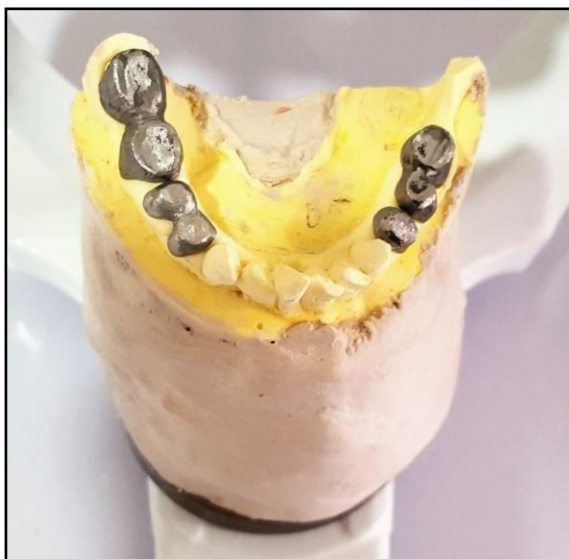


Figure 5: Metal framework trial of mandibular posteriors

The same procedure was repeated for maxillary right and left posterior teeth. In the same appointment, permanent crowns of the lower posteriors were cemented temporarily to assure any changes were further to be made. (Figure 6). The same protocol was repeated for the maxillary arch.

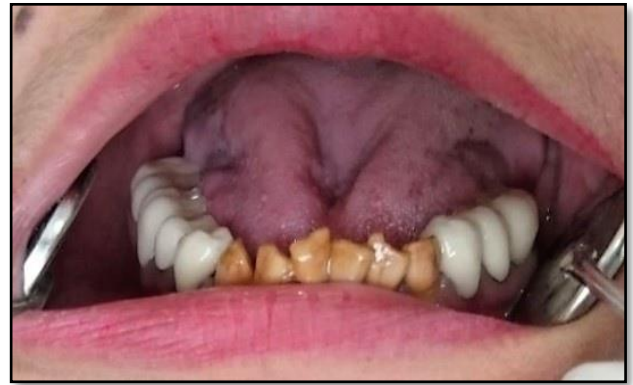


Figure 6: Mandibular posterior permanent teeth temporarily cemented

Later on, maxillary anterior teeth were prepared. Trial of the fabricated anterior restorations was successfully carried out and it was then cemented temporarily. Due to endodontic complications, the treatment plan had to be modified and lower central incisors were extracted. Crown preparations of lower anterior were carried out after 3 months of extractions. Once the temporaries were cemented in the patient's mouth, adjustments were done intra-orally to develop anterior guidance. After which impressions were record and the casts were mounted in the articulator using switch cast mounting technique and then a custom acrylic anterior guidance was developed on the articulator and the definitive prosthesis was being fabricated based on that guidance. Metal framework trial was carried out followed by preglaze trial. The restorations were found to be adequate at both the stages. (Figure 7)



Figure 7: Maxillary anterior prosthesis

The patient was kept under observation for 1 month and was preceded by permanent cementation of all the restorations. She was called for follow up of the prosthesis after a period of 3 months. She was happy and satisfied with both function and esthetics. A further satisfactory follow up was done at 1 and 3 year interval.



Figure 8: Pre and post treatment intraoral pictures

DISCUSSION:

AI presents as a clinical challenge for the dental practitioner as management of this condition is important not only from functional or esthetics standpoint, but from psychological aspect as well. Traditionally in the past, these patients were rehabilitated by removable complete dentures after serial extractions, which adversely affected the psychological well-being of the patient. Newer studies advocate following a multidisciplinary approach with considerations of certain factors including the age, type of AI, severity of the condition and financial aspects^{6,7}

Both direct and indirect treatment options are suggested for full mouth rehabilitation of patients with AI⁸⁻¹³. Few studies recommended direct composite restorations especially in young AI patients. These restorations offer certain benefits including, excellent esthetics, conservative approach, and improved wear^{8,9}. Conversely other clinical studies revealed poor prognosis of such restorations especially in AI - hypocalcified and hypomaturational type, where the enamel qualitative alterations produce poor etching pattern and weakened bond between enamel and resin^{10,11}. Moreover the 5 year survival rate of direct restoration is significantly low, 50% in AI patients as compared to healthy control group 80%.⁵. As of these limitations, full coverage restorations are usually advised as an alternative to direct restorations especially for severe AI cases, because there is no correlation between restoration longevity and severity and type of AI. Both all ceramic and porcelain fused to metal restorations provides excellent esthetic and function with clinical predictability and long survival rates¹¹⁻¹³. However, they involve a significant sacrifice of tooth structure and a high risk of pulp exposure in young patients with prominent pulp horns.

Novelli et al recommended using prefabricated composite veneers as an alternative treatment option for young AI patients where use of full coverage crowns can lead to endodontic complications. It is a simple clinical procedure that produces a restoration with proper shape, accurate anatomy, and glossy surface with minimally invasive tooth preparation¹⁴. However, due to lack of clinical studies, the clinical performance and longevity such restorations are not predictable.

Limitations: The shade of the restorations was selected as per the patient's choice, as she wanted pearl white teeth. Moreover, at the preglaze trial stage, there was disharmony in gingival zeniths, but the patient was satisfied as the cervical area was not visible during smile because of low smile line. She did not give us the consent for the customization.

CONCLUSION:

Each case of AI is unique and requires careful planning and management keeping certain considerations in mind. The final result depends on complexity of the case, attention to detail, use of recommended materials /armamentarium and execution of the properly designed treatment plan.

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