

Esthetic and Functional Rehabilitation of a Case of Amelogenesis Imperfecta With A Multidisciplinary Approach Using an Innovative Sequentially Staged Technique –A Case Report

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Introduction:

Amelogenesis imperfecta (AI) presents with a rare abnormal formation of the enamel^[1] or external layer of the crown of teeth. Enamel is composed mostly of mineral that is formed and regulated by the proteins in it. Amelogenesis imperfecta is due to the malfunction of the proteins in the enamel: Ameloblastin, Enamelin, Tuftelin and Amelogenin.

Amelogenesis imperfecta can have different inheritance patterns depending on the gene that is altered. Mutations in the ENAM gene are the most frequent known cause and are most commonly inherited in an autosomal dominant pattern. This type of inheritance means one copy of the altered gene in each cell is sufficient to cause the disorder.

About 5% of Amelogenesis imperfecta cases are caused by mutations in the AMELX gene and are inherited in an X-linked pattern. A condition is considered X-linked if the mutated gene that causes the disorder is located on the X chromosome, one of the two sex chromosomes. In most cases, males with an X-linked form of this condition experience more severe dental abnormalities than affected females. Amelogenesis imperfecta represents an inherited group of anomalies of enamel formation with an incidence of 1:718 to 1:14,000.²

The clinical features distinguish the three types.^{1, 3}

- Hypoplastic—The enamel does not form in normal thickness.
- Hypocalcified—Enamel thickness on newly erupted teeth closely approaches that of normal teeth, but the enamel is soft, friable and can be easily removed from the dentin.
- Hypomaturational type—Develop enamel of normal thickness. The Hypomaturational type differs from Hypocalcification in that enamel is harder, with a mottled opaque white to yellow brown to a red brown color and tends to chip from the underlying dentin rather than wear away. Treatment planning for patients with Amelogenesis imperfecta is multifactorial: the age and socioeconomic status of the patient, the type and severity of the disorder, the intraoral situation at the time of treatment planned.⁵ If teeth affected with Amelogenesis imperfecta are not detected and treated early; further deterioration of the existing condition will occur with damage to the periodontal tissues, thus further complicating the treatment plan and prognosis of the patient.

The common restorative problems associated with AI are sensitivity, loss of vertical height, dysfunction and esthetics. Restoration of these defects is important in meeting the esthetic, functional, and

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psychological concerns of the patient. Recent materials and improved clinical procedures have enabled esthetic and functional rehabilitation for patients with such severely worn out dentition.² The following clinical report demonstrates a multidisciplinary approach in treating a patient who presented with Hypomaturation type (Type III) of Amelogenesis Imperfecta along with malocclusion.

Case Report:

The patient reported to the clinic with the chief complaint of esthetic concern along with malpositioned teeth and expressed her desire for correction of the same. On examination it was seen that the patient required orthodontic correction before any concrete restorative procedures were planned. All radiographs were taken, and preliminary impressions made using alginate. Vertical dimension was evaluated clinically. Diagnostic wax up was planned at the existing vertical dimension and it was seen that there was no clinical loss of vertical dimension.

Face bow recording was done and wax up was achieved on a semi adjustable Arcon articulator. Patient's informed consent was taken before proceeding with the treatment. The treatment planning was done with a multidisciplinary approach with all concerned specialists coming together for a consensus on the desired approach.

Orthodontic treatment was initiated. The case was planned as a non-extraction case as it was desirable for an esthetic outcome. Orthodontic treatment was done with Beggs technique. The patient was scheduled on

regular monthly appointments till completion of treatment.

This was followed by esthetic periodontal procedure where recontouring of the gingival zenith was done to enhance the gingival contour.

Diagnostic wax up was completed with the vertical dimension maintained (Figure). The wax up helped in assessing the outcome of the final prosthesis and it also helped in fabricating the temporary restorations⁵. The maxillary and the mandibular first molars were prepared and temporized in one appointment. In the subsequent appointment, the permanent crowns were cemented with the same teeth and the rest of the posterior teeth were prepared. This method was adopted to maintain the vertical dimension of occlusion of the patient. The third appointment consisted of permanent cementation of the remaining posterior final crowns along with preparation and temporization of the anterior teeth. The final appointment consisted of permanent cementation of the final crowns on the mandibular and maxillary anteriors. A group function type of occlusal scheme was provided to the patient (Figure 11) and regular follow-up with good oral hygiene maintenance was advised (Figure 12). This approach which was sequentially staged helps in rehabilitating the patient quickly and predictably. The limitations of this technique of rehabilitation is that it is applicable only in cases that do not involve a raise in vertical dimension of occlusion.

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Discussion

The clinical presentation of AI varies with the type. The Hypoplastic type shows well mineralized enamel, but its amount is reduced as seen in the radiograph. Clinical management of AI can range from preventive interventions to complete rehabilitation depending on severity of the case⁴⁻⁷. In this case, the patient presented with a decreased vertical dimension and a freeway space of 5 mm, hence it was decided to increase the vertical dimension by 3 mm.^{8,9} The severe wear of anterior teeth facilitates the loss of anterior guidance, which protects the posterior teeth from wear during excursive movements. Collapse of posterior dentition results in loss of normal occlusal plane and decreased vertical dimension. The choice of restoration in this case was porcelain fused to metal as this would double the mechanical durability, recover esthetics, and protect the residual dentin.^{9,10} the rehabilitation was carried out using an innovative sequential approach where segmental preparation and temporization along with permanent rehabilitation was simultaneously carried out in different segments to enable speedy rehabilitation. This approach also helped in maintaining the vertical dimension of occlusion throughout the entire treatment as the keys of occlusion i.e. the first molars were rehabilitated first.

Conclusion

Amelogenesis imperfecta is multi-factorial and clinical symptoms variable. The treatment of such cases involves a thorough analysis of the relationship between natural teeth and the stomatognathic

system¹⁰. Rehabilitation of a patient presenting with this condition was carried out using an innovative sequential approach.

Clinical significance

The above case reflects the importance of the use of prosthodontic principles and strategic planning in addition to a multidisciplinary approach in managing a young female patient of Amelogenesis imperfecta. the clinical approach adopted in this case also helps in speedy rehabilitation along with maintenance of vertical dimension of occlusion.

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