EAS Journal of Dentistry and Oral Medicine

Abbreviated Key Title: EAS J Dent Oral Med ISSN: 2663-1849 (Print) & ISSN: 2663-7324 (Online) Published By East African Scholars Publisher, Kenya

Volume-6 | Issue-3 | May-Jun-2024 |

Case Report

DOI:10.36349/easjdom.2024.v06i03.004

OPEN ACCESS

Rehabilitation of a Patient with Amelogenesis Imperfecta: A Multidisciplinary Approach

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Journal homepage: https://www.easpublisher.com



Abstract: Amelogenesis imperfecta (AI) is a genetic disorder affecting enamel development affecting its structure and clinical appearance. It can affect both primary and/or permanent teeth and may be linked with nephrocalcinosis. Autosomal dominant inheritance is the main transmission mode of AI, but autosomal recessive, X-linked can also occur in some cases. AI is a serious condition that can result in reduced oral health-related quality of life and causes some psychological disturbance This article presents a case report of a partially edentulous patient with amelogenesis imperfecta along with its features. **Keywords:** Amelogenesis imperfecta – prosthesis - esthetics.

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INTRODUCTION

Amelogenesis imperfecta (AI) is a genetic disorder affecting enamel formation resulting in defective enamel structure and composition. It may affect the primary and /or permanent dentition [1]. This condition may be associated with nephrocalcinosis [2, 3].

Inheritance is mostly autosomal dominant, however autosomal recessive, X-linked and sporadic cases can also occur spontaneously in members of the same family [4, 5].

The clinical manifestations of AI vary widely, ranging from discoloration and enamel hypoplasia to severe structural abnormalities [6].

AI can have significant impacts on dental health, aesthetics and quality of life.

This article describes rehabilitation of a partially edentulous patient with AI and its features.

CASE PRESENTATION

A 26-year-old female presented to the dental clinic of Monastir for esthetic reasons than for functional reasons.

Her two brothers had dental anomalies. Examination revealed no relevant medical history, apart from her dentition, and general development was normal.

Intraoral examination revealed an oligodontia in both arches, yellow discoloration and a decay in the left lower second molar (Fig. 1).

Periodontal examination revealed no signs of gingival inflammation neither teeth mobility except for the 37 around which a periodontal pocket was record.

Panoramic radiograph revealed the impaction of several teeth: the upper and lower anterior teeth (except the 23), the right upper and lower second molars. The root formation of the lower third molars was not complete. Also, a reduced enamel thickness was noted (Fig. 2). An extensive bone loss and a furcation involvement were noted around the left lower second molar (Fig. 3).

The clinical and radiographic features led to the diagnosis of amelogenesis imperfecta, hypoplastic type.

In order to preserve dental structures and their vitality, a comprehensive prosthetic restoration was being considered. However, the therapeutic decision must take into account several factors: aesthetic,

mechanical, economic. Unfortunately, orthodontic traction of the impacted teeth was not feasible due to non-sufficient posterior anchorage, therefore it was decided to retain them for bone preservation.

After discussion with the patient and due to her limited financial means, it was decided to proceed with an upper ceramic-metal bridge and a lower acrylic resin removable partial denture.

The patient's consent was taken.



Fig. 1: a. Maxillary arch, b. Mandibular arch



Fig. 2: Panoramic radiograph



Fig. 3: Retro-alveolar radiograph of the 37

Management:

A multidisciplinary approach involving prosthodontics and periodontics was adopted to address the patient's dental concerns comprehensively.

The objectives of the treatment plan were to:

- Reduce of dental sensitivities.
- Prevent tooth decay and maintain periodontal health.
- Ensure aesthetic and functional rehabilitation.
- Improve the self-esteem of the patient.

Oral Hygiene Instructions were given to the patient including regular brushing with fluoride toothpaste and the use of fluoride mouth rinses to minimize the risk of dental caries and maintain oral health.

Surgical phase: includes

- Extraction of the 37 because of the advanced periodontitis and the important bone destruction.
- Alveolectomy around the 17 to release the tooth and use it as posterior anchorage for the upper bridge (Fig. 4. a-b-c).
- Crown lengthening: Gingivectomy of the upper teeth to improve the alignment of gingival margins and to ensure a best retentive value.

Prosthodontic phase: includes a first step of studying the case for that

- First impressions of both arches were taken
- The occlusion was registered, the casts were mounted on the articulator and the mock-up was realized (Fig. 5. a-b).

This mock up allows to visualize the prosthetic project and to study its feasibility in terms of available vertical prosthetic space as well as to communicate with the patient. In our case the interocclusal space is sufficient to ensure the prosthetic rehabilitation so the vertical dimension occlusion of the patient will remain unchanged, whereas in other cases where vertical prosthetic space is insufficient, an OVD (occlusion vertical dimension) increase should be considered on the the patient condition that does not have temporomandibular disorders.

- Preparation of the upper teeth and fabrication of a temporary prosthesis which were made by the laboratory technician taking reference from the wax-up. In the practice, the teeth were prepared and the provisional ones are adjusted and rebased before being temporarily sealed (Fig. 6)
- At this stage, it was possible to perform gingival remodeling (coronary elongations). After periodontal surgery, the temporary prosthesis will remain in place for healing time (Fig. 7. a-b-c-d).
- A clinical observation phase is required for a few time to validate:
 - The preservation of pulp vitality,
 - Static and dynamic occlusion,
 - Neuromuscular adaptation (in case of an increased vertical dimension): Good masticatory function and the absence of neuro-musculo-articular pathologies related to the new vertical dimension
 - The marginal adaptation of restorations,
 - o Periodontal health and aesthetics



Fig. 4: a-b. Surgical release of the upper second molar, c. mucosal healing after two weeks



Fig. 5: a-b. Diagnostic wax-up



Fig. 6: Temporary prosthesis



Fig. 7: a-b. Coronary elongation, c-d. Relining of the temporary prosthesis

Once these conditions have been validated, second impressions were made with a metallic tray and silicone material of low and heavy viscosity (Fig. 8).

- Second registration of the occlusion and mounting casts on the articulator. Occlusal

reference was the centric relation as it was the only reproducible position

- Realization of the metal framework of the upper bridge and the metal copings were examined and the marginal limits were verified. An adequate interocclusal distance allowed ceramic application. The unglazed ceramic was clinically tried and returned to the definitive cast. the mounting of teeth in wax was also verified intraorally.

- Sealing of the fixed bridge and Insertion of the removable partial denture after occlusal adjustment (Fig. 9).
- Regular clinical and radiographic follow-ups are necessary to monitor the eruption progress of the impacted lower teeth and the patient's hygiene.



Fig. 8: Upper second impression



Fig. 9: Final result

DISCUSSION

Amelogenesis imperfecta has numerous classification systems and the most widely accepted is proposed by Witkop and Sank in 1976, which considers the inheritance pattern of the disorder, as well as its specific clinical characteristics [6].

AI may be associated with some other dental and skeletal developmental defects or abnormalities that can complicate the treatment, such as root resorption, attrition, delayed eruption, tooth impaction and agenesis of teeth [7, 8]. In our case, tooth impaction was present.

Management of AI patients should aim to establish health, function and aesthetics of the patient's own teeth and prevent or delay the need for extraction and prosthetic replacement. Hence, it requires a multidisciplinary team approach to rehabilitation, which may include a pediatric dentist, an oral surgeon, a periodontist, an orthodontist, and finally a prosthodontist.

This is essential to ensure good longevity to the restorations provided.

The therapeutic strategy depends on the following factors:

The patient's age and cooperation, the type of the dentition, the type and intensity of enamel involvement, the association or not with a skeletal, alveolar or functional anomaly, periodontal conditions and the socio-economic level [9, 10].

Thus, AI is supported through three phases:

A Temporary phase in temporary or mixed teeth immediately after diagnosis it's based on Information, motivation and cooperation of the patient and on improving periodontal health and preventive measures such as dietary advice and teaching a correct brushing method, sealants, caries treatment and limitation of future orthodontic needs.

The second phase in the transient phase in mixed dentition as soon as the objectives of the previous phase have been achieved. It aims to provide the child with an aesthetic and functional result until the end of growth and definitive treatment in adulthood.

During this phase a periodontal preparation may be done: coronary elongations [11], remodeling of soft tissue contours through gingivectomy, osteotomy or osteoplasty to prepare tissues for final prosthetic treatment. Polycarboxylate resin crowns in severe damage or resin crowns made in the laboratory may be used.

An orthodontic preparation is also recommended during this phase to minimize the duration of treatment using temporary prosthetic restorations as an anchor [12].

Regular follow up is necessary during this phase to ensure harmonious growth and balanced psycho-social development of the patient; and to control occlusal integration of the provisional restauration.

The Final phase, which is illustrated by our case, is performed in adult age with permanent teeth, it leads to improved aesthetics, optimization of occlusal functions and a good long-term prognosis of the prosthesis.

Actually, we assist to an advent in cosmetic dentistry thus, micro-abrasion can be used for moderate cases [13, 14]. Conservative care is based on reconstitution with glass ionomer cement or composites, but there are doubts about the quality of bonding, retention and durability of composites [15]. In fact, adhesion quality is reduced due to more porous enamel, with less mineral content and more protein content. For that, self-adhesive systems for a less aggressive bonding on hypomineralized enamel are recommended. A technique of deproteinization of enamel with sodium hypochlorite (NaOCI) may be used. NaOCI could improve the adhesion quality of the composite.

Despite these bonding difficulties, composites are considered a good material for transient aesthetic reconstructions [16, 17].

In some cases of AI where the teeth are unrestorable and the patient is seeking a fixed option, dental implants can be considered [18, 19]. Careful planning is essential and timing of extractions with respect to implant placement is very important in order to preserve bone. In cases where there is insufficient bone width, it may be possible to graft the bone with guided bone regeneration, A maxillary sinus grafting procedure may be carried out which can give extra height of bone for implant placement in the posterior maxilla.

Therefore, the dentist has to diagnose the condition as early as possible to offer early intervention and to avoid or minimize its effects on oral health and the person's psychology. Especially if renal involvement is here, an early diagnosis may limit complications because untreated nephrocalcinosis is associated with significant morbidity [20].

CONCLUSION

AI is a serious condition that can result in reduced oral health-related quality of life and causes some psychological disturbance. Hence, accurate diagnosis and appreciation of associated clinical anomalies in each case enable the dentist of early preventive measures and management techniques using a multidisciplinary approach. An early diagnosis leads to an easier prosthetic management allowing them to be accepted by their peers and society in general.

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Cite This Article: Sameh Aguir, Sameh Rezigui, Wafa Nasri, Abdellatif Chokri, Hiba Triki, Lamia Mansour (2024). Rehabilitation of a Patient with Amelogenesis Imperfecta: A Multidisciplinary Approach. *EAS J Dent Oral Med*, 6(3), 35-41.