

ORTHODONTIC AND PROSTHETIC REHABILITATION IN AMELOGENSIS IMPERFECTA - CASE REPORT

Paula Jiman¹, Andreea Simona Pop^{2*}, Dana Gabriela Festila³, Ada Gabriela Delean⁴
, Alexandrina Muntean¹

¹Iuliu Hațieganul University of Medicine and Pharmacy Cluj-Napoca, Romania, Faculty of Dental Medicine, Department of Paediatric Dentistry;

²Iuliu Hațieganul University of Medicine and Pharmacy Cluj-Napoca, Romania, Faculty of Dental Medicine, Department of Oral Rehabilitation, Oral Health and Dental Office Management;

³Iuliu Hațieganul University of Medicine and Pharmacy Cluj-Napoca, Romania, Faculty of Dental Medicine, Department of Orthodontics.

⁴Iuliu Hațieganul University of Medicine and Pharmacy Cluj-Napoca, Romania, Faculty of Dental Medicine, Department of Conservative Dentistry.

*Corresponding author; *e-mail*: spopandreea@yahoo.com

All authors contributed equally to the present study and should be regarded as main authors.

ABSTRACT

Aim of the study Amelogenesis imperfecta (AI) is a term that describes a group of inherited conditions that affect the structure and the appearance of dental enamel in both primary and permanent dentition. **Material and methods** The oral rehabilitation of patients with AI remains a challenge, for the clinician and for the patient, because it requires long-term collaboration. To illustrate the difficulties in treatment of such pathology, we present a clinical case. **Results** AI associated with crowding, canine inclusion, and severe enamel impairment with associated hyperesthesia, imposed complex orthodontic and prosthetic treatment. **Conclusions** Long lasting treatment that involves an interdisciplinary team of specialized dentists is required in order to properly manage a condition such as AI.

Key words: AI, orthodontics, prosthetics

INTRODUCTION

Amelogenesis imperfecta (AI) is a term that describes a group of inherited conditions that affect the structure and the appearance of dental enamel, in both primary and permanent dentition [1,2]. An average reported prevalence of AI is estimated to be <0.5% (less than one case in 200), varying on the country and region [1-7]. This condition also appears in conjunction with alterations in other intra-oral and/or extra-oral tissues, the term AI being additionally used to describe enamel phenotypes in other syndromes [1,2]. However, the presence of defective enamel without any underlying systemic defects is the characteristic trait of AI [2].

The formation of enamel, amelogenesis, has been described in three functional stages: presecretory, secretory and maturation stages.

In the presecretory stage, the organic matrix of the enamel is developed, while in the secretory phase the ameloblasts secrete the entire thickness of the enamel. Last of all, in the maturation stage, there is an increase in the inorganic content and the length and width of the prisms through an exchange of organic contents, water and ions [1].

The diverse clinical enamel malformations are believed to reflect the timing during amelogenesis in which disruptions occur. Hence, secretory stage defects result in a thin, hypoplastic enamel layer, while maturation stage defects result in an enamel that is of normal thickness, but pathologically soft [1]. Therefore, AI is presented to have various phenotypes, being caused by mutations or altered expression in five genes: *AMEL* (amelogenin), *ENAM* (enamelin), *MMP20*

(matrix metalloproteinase-20), *KLK4* (kallikrein-4) and *FAM83H* [1, 8-10]. There are more proteins with roles in cell-cell and cell-matrix adhesion, in transport and master controllers of amelogenesis. Moreover, there is also an emerging group of proteins that have functions in enamel development and that require further investigation for the time being [7,9,11].

Depending on the mutation involved, the inheritance pattern, the expression of matrix proteins and the associated biochemical changes, there are various clinical appearances of AI [1]. At the moment, there are phenotypes of AI described: hypoplastic, hypocalcified, hypo maturation and hypo maturation-hypoplasia with taurodontism [1,12-14]. This clinical classification can be, however, confounded by mixed phenotypes [9,12]. Different types of AI vary from defective enamel formation to complete deficiencies in mineral and protein content, with the mildest form of AI presenting discoloration and the most drastic form presenting pitting [1,2,15]

AI is characterized by hypo mineralisation and/or hypoplasia of the enamel, with reduced aesthetic appearance and discoloration of the teeth, increased tooth sensitivity and fragility [1,15]. AI causes patients' problems such as: wear of enamel with exposed dentin areas, masticatory difficulties, pain, early tooth loss and loss of vertical dimension [9,11,12,14-16].

Additionally, there are other reported clinical features of AI such as: microdontia, deviant crown and root morphology, root resorption, enlarged pulp chamber, pulp stones, gingival enlargement, gingivitis, periodontitis, tooth agenesis, dens in dente. Moreover, skeletal and orthodontic abnormalities, such as overbite, over jet, anterior and/or posterior open bite, cross bite and crowding have been reported to be

associated with AI [8,15,16].

MATERIAL AND METHODS

The oral rehabilitation of patients with AI remains a challenge, for the medical team, patient and family, because it requires long-term collaboration and involvement. To illustrate the treatment difficulties of such pathology, we present a clinical case.

12 years old girl, presented for physiognomic impairment during speaking and smiling.

We notice oval face form, avoiding the exposure of the teeth when smiling (Fig.1)



Figure 1. Face/profile photos before treatment

Patient profile appear convex through the anterior position relatively to nasal-frontal plane of the upper and lower lip and slightly retracted chin.

The endo-oral examination reveals: unsatisfactory oral hygiene, mixed dentition, inconsistent with age, changes in colour and shape of all teeth (fig.2).



Figure 2. The appearance of the dental

arches before treatment

The radiological investigation reveals: high position of the maxillary canines (the roots are completely formed, with the modified axis; the crowns overlap the roots of the lateral incisors); 5.3 and 6.3 persistent on the arch; 3.3 intramaxillary, rotated 90° (fig. 3).



Figure 3. Orthopantomography before treatment

Lateral cephalogram reveal skeletal class I (Wits=-1, ANB=2), normodivergent type (Y axis=65°, SN-MP=33°) (fig.4).



Figure 4. Lateral cephalogram before treatment

The subsequent diagnosis can be formulated: Amelogenesis imperfecta, Dental Maxillary Anomaly with crowding, persistence 53,63,73, inclusion 13, 23, 33, chronic marginal gingivitis. All the functions of the dento-maxillary apparatus are severely

affected. The main aetiology is hereditary. The therapeutic objectives comprise: control of teeth sensitivity, create space for permanent canine alignment, surgical exposure of all included canines and traction on the arches, functional and stable occlusion, prosthetic restoration in accordance with growth and development.

DISCUSSIONS AND EVOLUTION OF THE CASE:

In order to create space: 14, 24, 34, 44 were extracted. Orthodontic treatment was performed with fixed appliances using .022 metallic braces, Roth prescription. The alignment and leveling were accomplished in 8 months using Ni-Ti arches from .014 to .016X.022. The included canines (13, 23) were surgically discovered and buttons were bonded intraoperatively, and with the help of a padlock they were pulled with light forces onto a steel spring. The collage of orthodontic attachments was a challenge, given the alteration of the dental enamel.

At the end of the orthodontic treatment (patient age 15 years), patient face and profile are harmonious and balanced (fig.5).



Figure 5. Face/profile photos before treatment

Teeth hypersensitivity and dental aesthetic impairment required prosthetic rehabilitation, using provisional physiognomic crowns. The crowns were cemented with glass ionomer cements, to control and reduce tooth sensitivity and allow patient to maintain an

adequate oral hygiene(fig.6).



Figure 6. The appearance of prosthetically rehabilitated arches

The crowns have a long-lasting character, until the end of the growth processes, when complete rehabilitation, in accordance with periodontal criteria and aesthetic requirement can be made, by means of ceramic crowns.



Figure 7. Occlusion after orthodontic and prosthetic treatment

3rd molars eruption with odontal pathology increased painful clinical symptomatology, due to enamel mineralization disorders and, in this particular situation, the decision was made to extract all the wisdom molars.

During complex treatment patient compliance was adequate as a result of dentist permanent motivation and family support.

CONCLUSIONS

1. In order to minimize the dental pathology caused by the hypo mineralization, AI is a disorder that should be diagnosed in an early stage.

2. The oral rehabilitation of children and adolescents with AI is complex due to the

presence of dental and periodontal issues and a possibly altered eruption sequence.

3. Frequent dental appointments that involve an interdisciplinary team of specialized dentists are required in order to properly manage a condition such as AI.

REFERENCES

- 1 Gadhia K, McDonald S, Arkutu N, Malik K. Amelogenesis imperfecta: an introduction. *Br Dent J.* 2012 Apr 27;212(8):377-9.
- 2 Roma M, Hegde P, Durga Nandhini M, Hegde S. Management guidelines for amelogenesis imperfecta: a case report and review of the literature. *J Med Case Rep.* 2021 Feb 9;15(1):67.
- 3 Altug-Atac AT, Erdem D. Prevalence and distribution of dental anomalies in orthodontic patients. *Am J Orthod Dentofacial Orthop.* 2007 Apr;131(4):510-4.
- 4 Bäckman B, Holm AK. Amelogenesis imperfecta: prevalence and incidence in a northern Swedish county. *Community Dent Oral Epidemiol.* 1986 Feb;14(1):43-7.
- 5 Sedano HO. Congenital oral anomalies in argentinian children. *Community Dent Oral Epidemiol.* 1975 Mar;3(2):61-3.
- 6 Chosack A, Eidelman E, Wisotski I, Cohen T. Amelogenesis imperfecta among Israeli Jews and the description of a new type of local hypoplastic autosomal recessive amelogenesis imperfecta. *Oral Surg Oral Med Oral Pathol.* 1979 Feb;47(2):148-56.
- 7 Ortiz L, Pereira AM, Jahangiri L, Choi M. Management of Amelogenesis Imperfecta in Adolescent Patients: Clinical Report. *J Prosthodont.* 2019 Jul;28(6):607-612.
- 8 Poulsen S, Gjørup H, Haubek D, Haukali G, Hintze H, Løvschall H, Errboe M. Amelogenesis imperfecta - a systematic literature review of associated dental and oro-facial abnormalities and their impact on patients. *Acta Odontol Scand.* 2008 Aug;66(4):193-9.

- 9 Smith CEL, Poulter JA, Antanaviciute A, Kirkham J, Brookes SJ, Inglehearn CF, Mighell AJ. Amelogenesis Imperfecta; Genes, Proteins, and Pathways. *Front Physiol.* 2017 Jun 26;8:435.
- 10 Möhn M, Bulski JC, Krämer N, Rahman A, Schulz-Weidner N. Management of Amelogenesis Imperfecta in Childhood: Two Case Reports. *Int J Environ Res Public Health.* 2021 Jul 5;18(13):7204.
- 11 Dashash M, Yeung CA, Jamous I, Blinkhorn A. Interventions for the restorative care of amelogenesis imperfecta in children and adolescents. *Cochrane Database Syst Rev.* 2013 Jun 6;2013(6):CD007157.
- 12 Abd Alraheem I, Donovan T. Management of amelogenesis imperfecta in an adult patient: a short review and clinical report. *Br Dent J.* 2020 Aug;229(4):239-243.
- 13 Toupenay S, Fournier BP, Manière MC, Ifi-Naulin C, Berdal A, de La Dure-Molla M. Amelogenesis imperfecta: therapeutic strategy from primary to permanent dentition across case reports. *BMC Oral Health.* 2018 Jun 15;18(1):108.
- 14 Strauch S, Hahnel S. Restorative Treatment in Patients with Amelogenesis Imperfecta: A Review. *J Prosthodont.* 2018 Aug;27(7):618-623.
- 15 Sabandal MM, Schäfer E. Amelogenesis imperfecta: review of diagnostic findings and treatment concepts. *Odontology.* 2016 Sep;104(3):245-56.
- 16 Leevailoj C, Lawanrattanakul S, Mahatumarat K. Amelogenesis Imperfecta: Case Study. *Oper Dent.* 2017 Sep/Oct;42(5):457-469.
- 17 Kumar S, Gupta S. The restoration of function and esthetics of a patient with amelogenesis imperfecta using a combination of orthodontic and prosthodontic treatment: a case report. *J Contemp Dent Pract.* 2009 Nov 1;10(6):E079-85.