Amelogenesis Imperfecta - A Case Report

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BACKGROUND

- Amelogenesis Imperfecta (AI) is a group of hereditary developmental disorders that affects enamel formation
- The prevalence in the general population is estimated to be around 1/14000
- There are no other syndromic, metabolic, or systemic conditions associated with AI
- Patient's chief complaints are often regarding poor esthetics, sensitivity, chewing difficulties, and loss of tooth structure
- Associated clinical characteristics include short clinical crowns, malformed roots, taurodontism, congenitally missing teeth, supernumerary teeth, anterior open bite, and abnormal growth of the maxilla and mandible

GENETICS

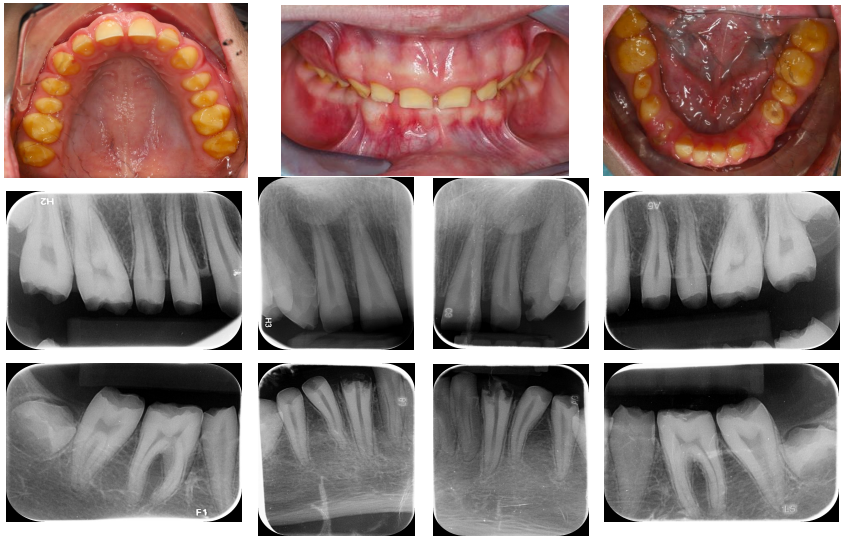
- There are several key genes involved in the enamel formation process
- There are multiple known mutations that are known to cause AI with different inheritance patterns depending on the affected genes
- Some genes that are known to involved in AI include AMELX, ENAM, MMP20, KLK4, DLX3
- Inheritance patterns include autosomal dominant, autosomal recessive, and X-linked recessive

TYPES

- Type I: Hypoplastic - quantitative deficiency with lack of enamel
- Type II: Hypomaturation - qualitative deficiency in maturation of crystalline structure
- Type III: Hypocalcification - qualitative deficiency in mineralization
- Type IV: Hypoplastic, Hypomaturation, and Taurodontism - combination of type I and type II features, with taurodontism seen as well

MANAGEMENT

- In less severe cases: regular supervision and fluoride to reduce sensitivity
- In more severe cases: SSC on posterior teeth to preserve vertical dimension and reduce sensitivity, and composite or veneered crowns in anterior teeth that may need to get replaced
- Patient should be referred in early permanent dentition to be evaluated for proper dental age to receive permanent restorations



CASE REPORT

- A 13y11m M presents to pediatric dental clinic for a new patient exam
- Patient has non-contributory medical history, is currently taking no medications, and has no known drug allergies
- Chief complaint is sensitivity to hot and cold
- History of AI on his maternal side
- Extraoral exam: No pathology detected
- Moderate generalized gingivitis
- Generalized spacing on maxilla and mandible
- Class I R and L molar classification
- Undeterminable canine classification
- OB: 10mm, OJ: 5mm
- Caries #11-ml
- Fracture to the root evident on #22 and #27
- Oral hygiene instructions: recommended to use soft-bristled toothbrush due to sensitivity, interproximal brush for flossing due to generalized spacing
- #11-ml filled with glass ionomer
- Referral placed to oral surgery for extractions of #22 and #27
- Referral placed to graduate restorative department in order to evaluate dentition for timing of permanent crown placement.

REFERENCES

1. Guideline on Dental Management of Heritable Dental Developmental Anomalies. (2016). *PubMed*, *38*(6), 302–307. https://pubmed.ncbi.nlm.nih.gov/27931469
2. Koch, G., Poulsen, S., Espelid, I., & Haubek, D. (2017). *Pediatric dentistry: A Clinical Approach*. John Wiley & Sons.
3. Ortiz, L., Pereira, A. M., Jahangiri, L., & Choi, M. (2019). Management of amelogenesis imperfecta in adolescent patients: clinical report. *Journal of Prosthodontics*, *28*(6), 607–612.
4. Strauch, S., & Hahnel, S. (2018). Restorative Treatment in Patients with Amelogenesis Imperfecta: A Review. *Journal of Prosthodontics (Print)*, *27*(7), 618–623. https://doi.org/10.1111/jopr.12736