

INTRODUCTION

Amelogenesis imperfecta is a rare genetic disorder that affects tooth enamel. It can be seen as part of a syndrome or on its own, and clinical expression varies significantly. Prevalence in the United States is approximately 1 in 14,000. There are four main types: Type 1 (Hypoplastic), Type 2 (Hypomaturation), Type 3 (Hypocalcified), and Type 4 (Hypomaturation-hypoplastic with taurodontism). Each type is associated with its own physiologic cause and clinical appearance. Generalized dental findings associated with amelogenesis imperfecta include abnormal quality and/or quantity of enamel, delayed eruption, impacted teeth, pulp stones, enlarged pulp chambers, missing teeth, anterior open bite, and loss of VDO. Patients with amelogenesis imperfecta generally suffer from tooth sensitivity, functional concerns, poor esthetics, and an impacted quality of life.

CASE REPORT

This case presentation discusses a 14-year-old male patient who presented to UH Rainbow Babies and Children's Hospital for dental treatment due to amelogenesis imperfecta. This condition is also seen in the family history, as the patient's mother has the same diagnosis. The patient does not have any other significant medical history. His medications include fluticasone and a daily multivitamin. There are no known allergies.

The patient was first seen with UH on 8/18/2025. His chief concern was addressing sensitivity across the entire dentition and esthetics. Initial clinical exam revealed generalized malformed enamel, no carious lesions, unerupted #1, 15, 16, 17, 18, 31, 32, retained #H and R, and no missing or supernumerary teeth.

Various treatment plans were discussed: 1. Extraction of maxillary dentition for placement of a complete denture with expectation of future implants while restoring the majority of the mandibular dentition with zirconia crowns. 2. Addressing all maxillary and mandibular dentition with placement of stainless steel crowns on posterior dentition and resin crowns on anterior dentition with extraction of any/all non-restorable teeth. More definitive restorations will be decided upon by a general dentist when the patient is of the appropriate age. Option 2 was decided upon by providers, family, and the patient.

The patient was seen for dental treatment under general anesthesia on 9/04/2025. Stainless steel crowns were placed on #3, 4, 5, 28, 29, and 30. Extractions were completed on #H and R. The plan includes additional appointments for dental treatment under general anesthesia, and the patient was scheduled for 3/20/2026. This appointment was cancelled, and the patient has yet to reschedule.

CLINICAL AND RADIOGRAPHIC PRESENTATION

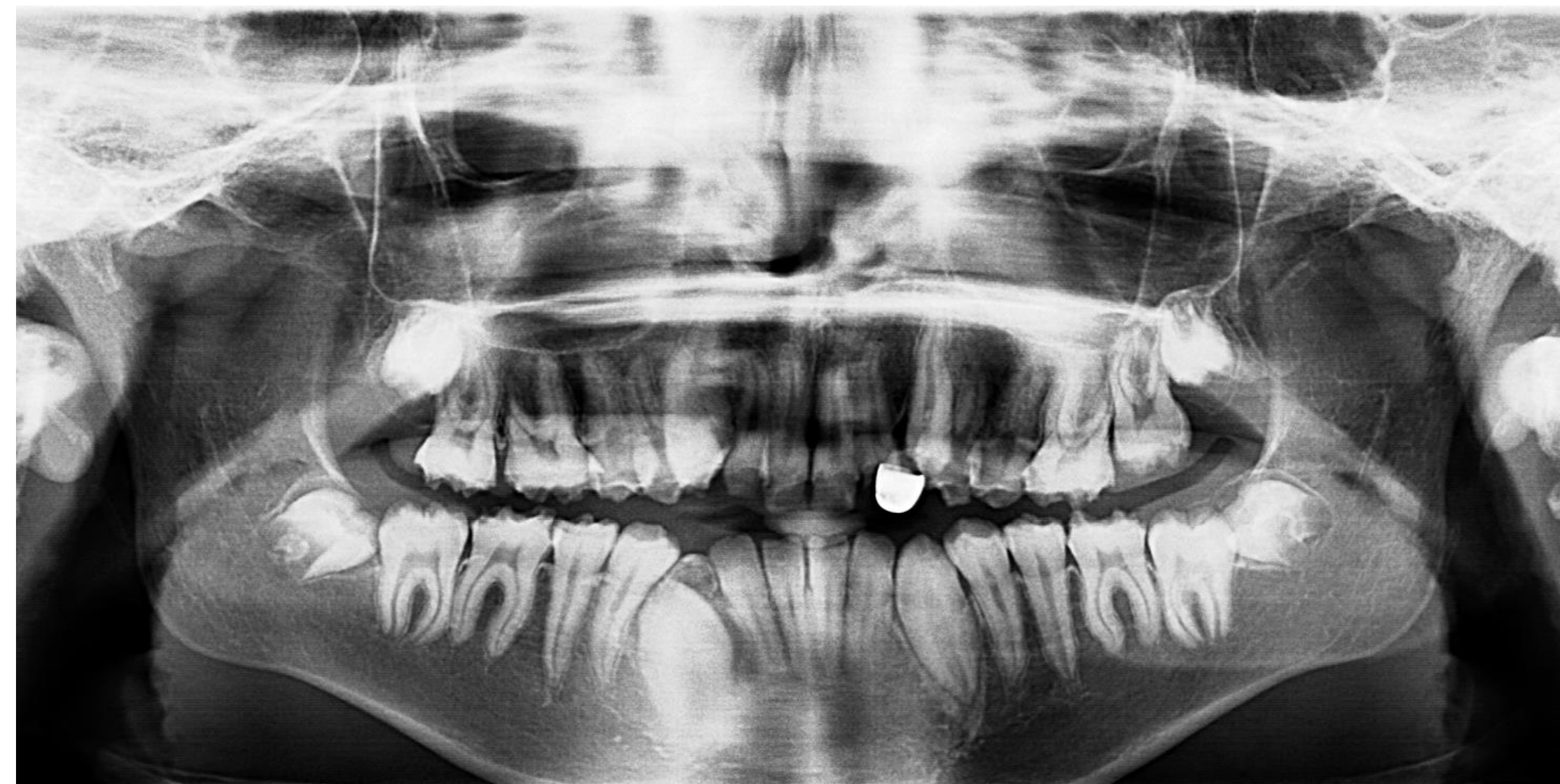


Figure 1. Panoramic radiograph was obtained on 8/18/2025. Findings include abnormally formed enamel and delayed eruption of permanent dentition.

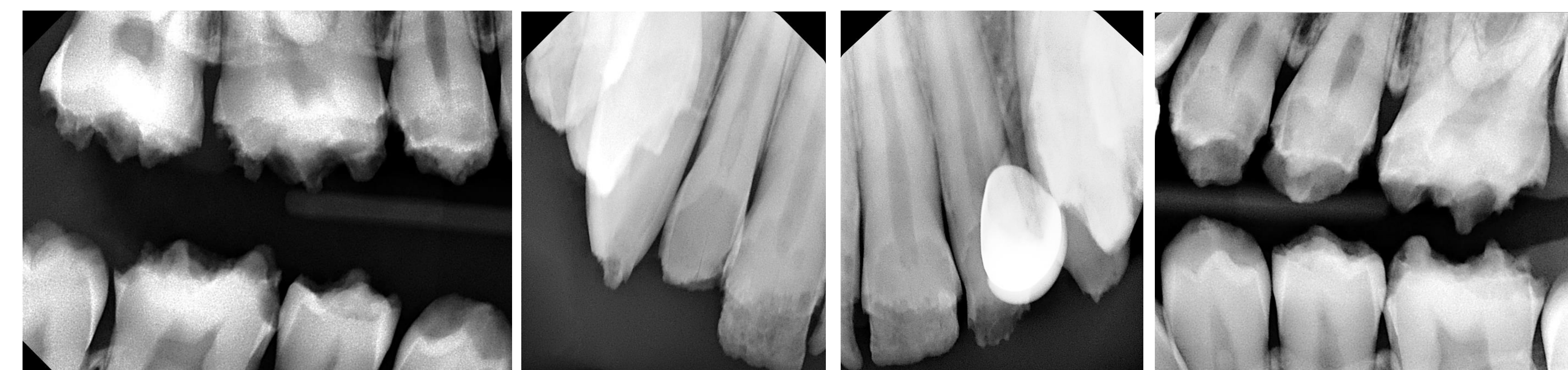


Figure 2. Intraoral radiographs were obtained on 8/18/2025. Malformed enamel is evident in all images with significant loss of tooth structure. A pulp stone can be noted in #21 in the left bitewing.



Figure 3. Clinical photos were obtained on 8/18/2025. Malformed enamel is evident in all images with discoloration and significant loss of tooth structure.

DISCUSSION

Amelogenesis imperfecta is a genetically inherited disorder that occurs due to defective ameloblasts affecting enamel formation and mineralization. The enamel that forms is generally soft and prone to decay, wear, and fractures.

Patients tend to have significant sensitivity when eating, drinking, and completing homecare routines. Homecare is often subpar due to discomfort, and the roughened enamel is more susceptible to calculus formation. Many patients are also negatively affected socially and mentally due to esthetics.

Extractions and removable appliances were considered optimal treatment in the past, but with advances in dentistry, the standard of care has shifted. Full coverage crowns are recommended to seal the entire tooth and establish VDO. This can reduce sensitivity which helps patient compliance with homecare, thereby improving oral health.

It has been shown that when treating affected teeth, there is difficulty obtaining profound anesthesia. The combination of this, dental anxiety, and generalized intraoral sensitivity, often leads to a recommendation for treatment under general anesthesia.

Treatment is best provided in stages: Initial phase (primary and mixed dentition), transitional phase (permanent dentition), and definitive phase (adulthood). Treatment is provided prior to adulthood to prevent symptoms, help avoid further complications, and improve esthetics, with the main goal of preserving as much tooth structure as possible.

A multidisciplinary approach should be utilized to provide the best outcome. This may include pediatric dentists, orthodontists, endodontists, prosthodontists, and periodontists. Early recognition and proper treatment of amelogenesis imperfecta can significantly change the lives of affected patients, which often results in positive behavior changes and improved quality of life.

REFERENCES

1. Wang CI, Sinada N, Schoenbaum TR. The dental management and prosthodontic reconstruction of patients with amelogenesis imperfecta: A narrative review. *Dentistry Review*. 2024;4(1). doi:10.1016/j.dentre.2024.100080
2. Herath CK, Jayasooriya P, Hewage P. Dental Management of a Child Patient with Amelogenesis Imperfecta. *Journal of South Asian Association of Pediatric Dentistry*. 2021;4(2):134-137. doi:10.5005/jp-journals-10077-3061
3. Ortiz, L., DDS, Pereira, A. M., DDS, Jahangiri, L., BDS, DMD, MMSc, & Choi, M., DDS, MS, FACP, MBA. (2019). Management of Amelogenesis Imperfecta in Adolescent Patients: Clinical Report. *Journal of Prosthodontics*, 28(6), 607-612. Retrieved from <https://rave.ohiolink.edu/ejournals/article/369930918>