

# Amelogenesis Imperfecta : An Adolescent Case Report

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## ABSTRACT

Amelogenesis Imperfecta is a genetic disease characterized by the abnormal formation of enamel.<sup>1</sup> Affected patients have unique restorative needs throughout development and maturation of the permanent dentition. This case report highlights the dental treatment over the course of over three years in an adolescent female with hypocalcified AI and her dental treatment through adolescence. From age 10 years through age 14 years, full mouth rehabilitation was completed under general anesthesia on two occasions and several in-office restorative treatments were completed including endodontic treatment and anterior and posterior full coverage restorations. This case report summarizes her care and factors related to success and re-treatment.

## BACKGROUND

Amelogenesis imperfecta refers to a group of rare inherited disorders characterized by abnormal enamel formation without any other abnormalities of the body.<sup>1</sup> AI is classified into four main types and includes 14 recognized subtypes.<sup>2</sup> The main types are assessed via clinical appearance, radiographic features, and enamel thickness, while subtypes are determined via mode of inheritance and genetic mutation. Type 1 Hypoplastic AI, the most common type, features enamel that fails to reach normal thickness and shows varied presentation such as rough, smooth, grooves, pitted, or discolored. Type 2 Hypomaturation AI is discolored with a rough, pitted texture of normal enamel thickness but of poor quality that is brittle. Type 3 Hypocalcified AI is similar to type 2 however, the enamel is chalky, tartar builds up quickly, and enamel can fracture with an explorer or scaler. Type 4 Hypomaturation-Hypoplastic with Taurodontism shows thin enamel, small teeth, wide spacing and enlarged pulp chambers.<sup>2</sup> Genetic inheritance may be X-linked (AMELX), autosomal dominant (FAM83H), and autosomal recessive (ENAM, MMP20).<sup>3</sup>

### CLINICAL FINDINGS in AI include: <sup>1,4</sup>

- Poor esthetics
- Tooth sensitivity
- Poor gingival condition and oral hygiene
- Decreased OVD through loss of tooth structure
- Increased tendency for impacted permanent teeth and associated anomalies
- Increased open bite tendency
- Decreased masticatory function
- Lower self-esteem
- Eruption disturbances

**MODALITIES OF TREATMENT** involve dental and psychosocial management.<sup>1,4</sup> During primary dentition, treatment may include placement of SSCs on primary molars to prevent development of caries, attrition of defective enamel, and to maintain space and OVD. In mixed dentition, goals are to preserve tooth structure, maintain vitality, decrease sensitivity and improve esthetics. Once adulthood is reached, to attain function, esthetics, and restored VDO, a multidisciplinary approach including prosthodontics, periodontics, orthodontics, and orthognathic surgery may be indicated.<sup>4,5</sup>

## CASE REPORT

### Patient AS:

- 13-year, 9-month old female presented to the University of Michigan School of Dentistry Clinic for a limited oral examination on 11/12/2021.
- Initially presented as a new patient 10/08/2018
- Medical History: Amelogenesis Imperfecta Type III (Hypocalcified), mandibular cyst, maxillary cyst
- Surgical History: dental rehabilitation (2015, 2017, 2018, 2019, 2021), enucleation of anterior maxillary and mandibular cysts (s/p 2018, 2019), tonsillectomy and adenoidectomy.
- Medications: None
- Allergies: azithromycin (GI distress)
- Family history: mother reports AI runs in the family and mother reports that she has no remaining teeth.
- Social history: No exposure to alcohol or smoking.



### Dental History

#### 01/09/2018: C.S. Mott Children's Hospital

- Enucleation of 2 mandibular cyst and 1 maxillary cyst

#### 10/08/2018 University of Michigan

- Referred by a private pediatric dental office, "Anxious - hx of full mouth rehab at Mott/Hurley. Presents with multiple failing restorations, abscess, PARL, oral path referred to UMHS OMFS. Unable to coordinate tx orally due to patient's insurance. She would benefit from more aesthetic tx."
- Exam reveals sensitivity to probing/explorer touch on #4, #8, #9 (recurrent decay), #10, #13, #20, #23, #25. #10, #23, and #25 show missing strip crowns. #24 shows pulp necrosis and symptomatic apical periodontitis. #K-over-retained
- Behavior: anxious, concerned about needles
- OR Referral sent for treatment of enamel hypoplasia and recurrent decay impacting eight teeth.



#### General anesthesia 03/13/2019

- Diagnosis: #24 grade 2 mobile & PARL, AI, recurrent caries Pediatric OMFS
- Tx Completed: #24 -EXT, enucleation, and curettage of cyst Pediatric Dentistry
- Tx Completed: #4 SSC, #6 composite resin crown, #10 composite resin crown, #13 SSC, #20 SSC, #23 composite resin crown, #25 composite resin crown, #30 SSC

#### Clinical Exam 09/20/2020 University of Michigan

- Periodic oral exam reveals a missing SSC, and recurrent decay on several teeth. A second rehabilitation under general anesthesia was planned.

#### General anesthesia 01/06/2021

##### Pediatric Dentistry

- Tx Completed: #2 SSC and removal of distal operculum using Bovie instrument, #4 SSC, #5 SSC, #6 composite resin crown, #8 facial composite, #10 composite resin crown, #11 composite resin crown, #12 SSC, #19 SSC, #21 SSC, #29 SSC

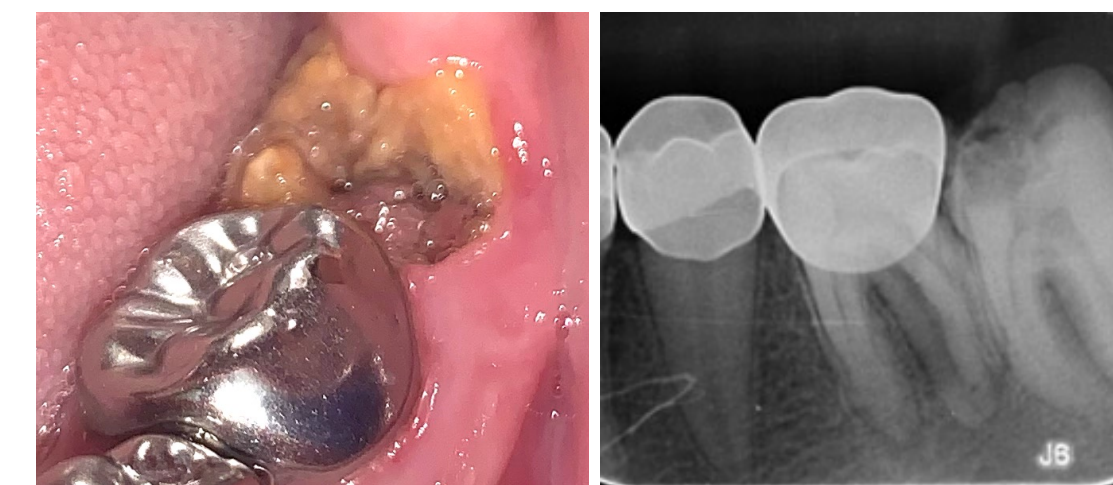
### Problem Focused Exam 02/09/2021

- CC: "One week ago she bit into an apple slice and her upper front tooth cracked"
- Findings reveal endo-treated #9 resin fracture with recurrent caries near facial cervical area, subgingival about 1mm. Slight PARL noted with PDL widening on distal aspect. Negative to palpation & percussion testing.
- Tx completed: #9 strip crown replaced



### Clinical Exam 10/05/2021

- Periodic exam reveals #2 distal overhang, #10 and #11 missing crowns with cold sensitivity, #18 pre-eruptive caries, #26 missing resin crown, and #19 indicated for RCT. #Treatment under general anesthesia was planned
- Tx plan: #2-SSC, #10-resin crown, #11-resin crown, #18-IPT/MO/SSC, #19-RCT/SSC, #26-resin crown



### Problem Focused Exam 11/12/2021

- CC: "my lower tooth crown came off & I am here to get it fixed so I can go back to school"
- Findings reveal endo-treated #25 has missing crown
- Tx completed: #25 strip crown replaced



### Problem Focused Exam 3/9/2022

- CC: "I have pain on lower front and lower left"
- Findings reveal facial surface of strip crown missing on endo-treated #26 and grossly carious non-restorable #18
- Tx completed: #26 strip crown replaced



## CONCLUSIONS

- Treatment of AI in the transitional and early permanent dentition is challenging. If unable to delay care until eruption of all permanent teeth, multiple visits to protect and treat permanent teeth as they erupt may be indicated.
- Pulp vitality is a common concern with affected young permanent teeth and endodontic treatment may be necessary.<sup>4</sup>
- Additional re-treatment in young adulthood is necessary for an optimally esthetic outcome.

## REFERENCES

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