



In The Name Of GOD

Amelogenesis imperfecta

*H.yousefi*

# ● Amelogenesis imperfecta

- presents with abnormal formation of the enamel or external layer of teeth. Enamel is composed mostly of mineral, that is formed and regulated by the proteins in it.
- Amelogenesis imperfecta is due to the malfunction of the proteins in the enamel: ameloblastin, enamelin, tuftelin and amelogenin.
- People afflicted with amelogenesis imperfecta have teeth with abnormal color: yellow, brown or grey. The teeth have a higher risk for dental cavities and are hypersensitive to temperature changes. This disorder can afflict any number of teeth.



- Amelogenesis imperfecta is a disorder of tooth development.
- This condition causes teeth to be unusually small, discolored, pitted or grooved, and prone to rapid wear and breakage. Other dental abnormalities are also possible.
- These defects, which vary among affected individuals, can affect both primary (baby) teeth and permanent teeth.
- Researchers have described at least 14 forms of amelogenesis imperfecta. These types are distinguished by their specific dental abnormalities and by their pattern of inheritance.

- Symptoms of Amelogenesis Imperfecta

- Brown discoloration of teeth
- Yellow discoloration of teeth
- Thin tooth enamel
- Soft tooth enamel
- Easily damaged teeth





## ● Examinations

- No unusual or abnormal findings were identified during the extraoral examination.
- Intraoral examination revealed a generalized yellow-colored dentition Examination of the oral soft tissues revealed no unusual findings and no bony abnormalities.
- Radiographic examination revealed teeth with thin amounts of enamel and areas where the enamel had fractured away .

# ● Disorder Subdivisions:

- Hypoplastic AI (Type I)
- Hypomaturation AI (Type II)
- Hypocalcified AI (Type III)
- hypomaturation/hypoplastic(type IV)

- The most common form, the hypoplastic type, is deficient in normal enamel.
- 
- The crown of the teeth appear blanched, "snowcapped," yellow-brown, pitted, or grooved.
- Radiographic examination usually shows a full complement of teeth, but the crowns of the teeth either have very thin enamel or lack enamel completely.
- The teeth actually resemble crown preparations with characteristic excessive interdental spacing.





# ● Clinical and radiographic features

- The hypoplastic is a deficient amount of enamel. Depending on the subtype classification, the crowns of the teeth may appear as one of the following:
  - opaque white, translucent brown, yellow-brown, pitted, or grooved.
  - Radiographically, the teeth exhibit abnormally thin amounts of enamel, or completely lack enamel.
  - The radiodensity of the thin enamel present is greater than that of dentin. Because of the lack of enamel, the teeth clinically exhibit excessive interdental spacing.
  - The teeth often are described as resembling crown preparations.

- In the hypocalcified form of amelogenesis imperfecta, the teeth are formed with a very soft enamel due to the incomplete mineralization of the enamel structure.
- A sufficient amount of enamel is present, but there is no significant calcification. In this type, the enamel is quickly lost shortly after the teeth erupt into the oral cavity.
- The teeth affected by hypocalcified amelogenesis imperfecta appear honey-brown in color with a roughened-surface texture. Radiographically, the teeth appear to lack enamel, or the enamel is less radiodense than dentin.

- In the hypomaturational form of amelogenesis imperfecta, the normal amount of enamel is present and some mineralization does occur. However, there is a decreased amount of secondary mineralization (maturation) that occurs in the enamel. As a result, the enamel is softer than normal and tends to chip away from the underlying dentin. Fracturing of the enamel is common.
- The use of a dental explorer with pressure may pit the surface. In this type of amelogenesis imperfecta, the crowns contact interproximally.
- The affected teeth may appear chalky white and rough or grooved.
- Radiographically, the affected enamel exhibits a radiodensity that is similar to dentin

- In the hypomaturation/hypoplastic form of amelogenesis imperfecta there is both the lack of enamel and the lack of enamel maturation.
- 
- The most significant defect is that of the lack of enamel. The thin enamel that is present exhibits a lack of secondary mineralization.
- The teeth appear yellowish with opaque mottling, pitting, and attrition.
- Radiographically, the enamel and dentin appear to have a similar density. Large pulp chambers also may be seen.

- How common is amelogenesis imperfecta?
- The exact incidence of amelogenesis imperfecta is uncertain. Estimates vary widely, from 1 in 700 people in northern Sweden to 1 in 14,000 people in the United States.

- How do people inherit amelogenesis imperfecta?
- Amelogenesis imperfecta can have different inheritance patterns depending on the gene that is altered. Most cases are caused by mutations in the ENAM gene and are inherited in an autosomal dominant pattern. This type of inheritance means one copy of the altered gene in each cell is sufficient to cause the disorder.
- Amelogenesis imperfecta is also inherited in an autosomal recessive pattern; this form of the disorder can result from mutations in the ENAM or MMP20 gene. Autosomal recessive inheritance means two copies of the gene in each cell are altered.
- About 5 percent of amelogenesis imperfecta cases are caused by mutations in the AMELX gene and are inherited in an X-linked pattern. A condition is considered X-linked if the mutated gene that causes the disorder is located on the X chromosome, one of the two sex chromosomes. In most cases, males with X-linked amelogenesis imperfecta experience more severe dental abnormalities than females with this form of this condition.
- Other cases of amelogenesis imperfecta result from new mutations in these genes and occur in people with no history of the disorder in their family.





- Treatment and prognosis:
- The treatment and prognosis for teeth with amelogenesis imperfecta vary with the severity of the enamel involvement. Variants that include rapid enamel loss and attrition require full-coverage crowns as soon as it is practical. When excessive crown loss occurs, restoration is not possible.
- In the types of amelogenesis imperfecta that demonstrate less enamel loss, the aesthetic appearance is often the primary concern. Full-coverage crowns can be used to improve the clinical appearance of the crowns.

- Crowns are sometimes being used to compensate for the soft enamel.
- Usually stainless steel crowns are used in children which may be replaced by porcelain once they reach adulthood. In the worst case scenario, the teeth may have to be extracted and implants or dentures are required.