Amelogenesis Imperfecta and Dentinogenesis Imperfecta

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Section D230
Amelogenesis Imperfecta

- Amelogenesis imperfecta is a congenital disorder with an abnormal formation of the enamel that fails to develop properly.

- It is caused by mutations in a variety of genes that are important for enamel formation.
Clinical Appearance

- People with Amelogenesis Imperfecta will have small, yellow, or brown teeth that are very prone to damage and breakage.
- Teeth can appear smooth or wrinkled.
- Variable status of enamel, possible exposure of dentin.
- Loss of inter-proximal contacts “Picket Fence Appearance.”
3 common types of Amelogenesis Imperfecta

- **Hypo-maturation**: Defect in the maturation of enamel crystals. With normal shape and mottled appearance. Vulnerable to attrition.

- **Hypo-plastic**: Enamel of reduced thickness due to defect in the formation of normal matrix. Pitting and grooves, enamel is hard and translucent.

- **Hypo-calcified**: Enamel matrix is formed in normal quality but poorly calcified. During eruption enamel is normal in thickness but weak with an opaque appearance.
Radiographic Appearance of Amelogenesis Imperfecta

- Hypo-maturation: Radiographically, enamel appears less radio-opaque than dentin.
- Hypo-plastic: Radiographically, the enamel contrast normally from dentin.
- Hypo-calcified: Radiographically, the enamel contrast is less than or equal to dentin.
Amelogenesis Imperfecta
Hypo-Plastic
Dentinogenesis Imperfecta

- A genetic disorder of tooth development which causes teeth to be discolored, translucent and weaker than normal.

- Dentinogenesis imperfecta type I occurs as part of osteogenesis imperfecta, which is caused by mutations in one of several other genes. Mutations in the DSPP gene have been identified in people with dentinogenesis imperfecta type II and type III.

- Dentinogenesis imperfecta affects an estimated 1 in 6,000 to 8,000 people.
Clinical Appearance of Dentinogenesis Imperfecta

- In dentiogenesis imperfecta the enamel is usually lost early due to loss of scalloping at the DEJ.
- People with dentinogenesis imperfecta will have teeth that may be gray or yellowish brown in color and exhibit translucent or opalescent hue.
Types of Dentinogenesis Imperfecta

• Type I- Occurs in patients affected by osteogenesis imperfecta, a genetic condition in which bones are brittle and easily broken.

• Type II- Occurs in people without hereditary disorders. It is an autosomal dominant trait.

• Type III- Known as “Brandywine type” it is a rare condition that exhibits pulp exposures and periapical lesions in deciduous dentition. It is found only in a population of southern Maryland, USA.
Radiographic appearance of Dentogenesis Imperfecta

- Teeth affected by dentogenesis imperfecta radiographically typically appear to have thin and spiked roots, bulb or bell shaped crowns with constricted cervical areas, obliteration of coronal and radicular pulp chamber which can vary depending on age.

- Radiographically Type I and Type II show total obliteration of the pulp chamber.

- Radiographically Type III shows thin dentin and extremely enormous pulp chamber.
Dentogenesis Imperfecta

Panoramic Radiograph showing obliterated pulp chambers
References

