

• Dentinogenesis imperfecta

Dentinogenesis imperfecta (DI) is an autosomal dominant trait, its frequency of occurrence is about 1 in 8000. This inherited dentin defect originates during the **histodifferentiation** stage of tooth development. The predentin matrix is defective resulting in amorphous, disorganized, and atubular circumpulpal dentin. Teeth are **blue-gray or brown** and **abrade rapidly**. Occasionally, these teeth become abscessed as a result of exposure of pulp horns caused by wear. Full coverage is the treatment of choice. Both the **primary** and **permanent** dentitions are affected in dentinogenesis imperfecta. **Important:** Radiographs of a preschool child with dentinogenesis imperfecta will show **obliteration of the pulp chambers** with secondary dentin, a characteristic finding. Roots of teeth usually are **narrower and appear more fragile**. Crowns generally appear more bulbous than usual due to the smaller roots. Dentinogenesis imperfecta can be subdivided into three basic types:

- **Shields Type I:** occurs with **osteogenesis imperfecta**. As a result of an inherited defect in collagen formation, there is brittle bones, bowing of the limbs, and blue sclera. Teeth have **amber translucent** color. Primary teeth affected more than permanent teeth.
- **Shields Type II:** also known as **hereditary opalescent dentin**, patients have only dentin abnormalities and no bone disease. Both primary and permanent teeth affected equally.
- **Shields Type III:** quite rare, only dental defects occur, similar to type II. Features of type III that are not seen in type I and type II include multiple pulp exposures, periapical radiolucencies, and a variable radiographic appearance. Seen exclusively in a triracial isolated group in Maryland known as the **Brandywine population**.

Amelogenesis imperfecta is one of the major defects of enamel. It is a hereditary disease characterized by faulty development of the enamel. There is normal pulpal and root morphology. There are four major categories according to the stages of tooth development in which each is thought to occur.

- **Hypoplastic Type:** occur in the **histodifferentiation** stage of tooth development. There is an insufficient quantity of enamel formed due to areas of the enamel organ that are devoid of inner enamel epithelium, causing a lack of cell differentiation into ameloblasts. Affects both primary and permanent dentitions. The affected teeth appear small with open contacts; clinical crowns contain very thin or nonexistent enamel.
- **Hypomaturation Type:** defect in enamel matrix **apposition** and is characterized by teeth having normal enamel thickness but a low value of radiodensity and mineral content.
- **Hypoplastic or Hypomaturation Type with Taurodontism:** is an example of inherited defects in both **apposition** and **histodifferentiation** stages in enamel formation. The enamel appears mottled with a yellow-brown color and is pitted on the facial surfaces. Molar teeth demonstrate taurodontism.
- **Hypocalcification Type:** is an example of inherited defect in the **calcification** stage of enamel formation. Quantitatively, the enamel is normal, but qualitatively, the matrix is poorly calcified. The enamel is soft and fragile and is easily fractured, exposing the underlying dentin, which produces an unesthetic appearance.