DENTINOGENESIS IMPERFECTA

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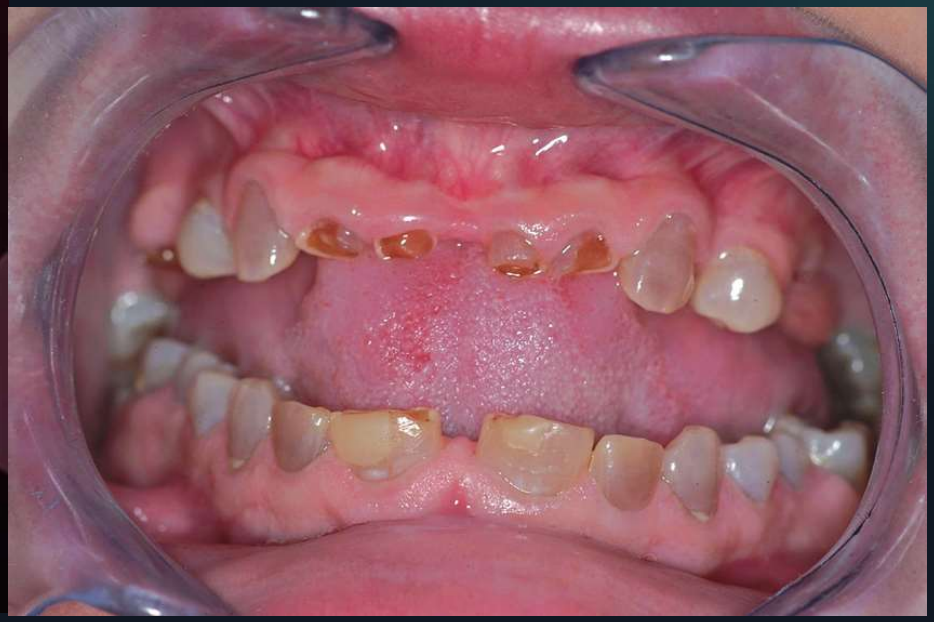
What is Dentinogenesis imperfecta?

- Also known as Capdepont teeth
- A tooth development disorder
- A type of dentin dysplasia
- Affects both primary and permanent teeth
- A condition which causes teeth to be discolored, resulting in blue-grey or yellow-brown color
- Enamel appears normal, but is prone to rapid wear and breakage due to lack of support by abnormal underlying dentin

Etiology

- It is hereditary, in an autosomal dominant manner
- Caused by mutations in the Dentine Sialophosphoprotein (DSPP) gene
- DSPP gene alters proteins involved in formation of dentin, leading to production of abnormally soft and less mineralized dentin



Different Types of Dentinogenesis Imperfecta

Type I

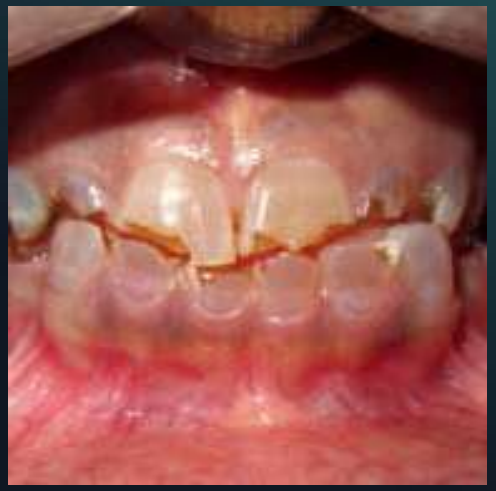
- Occurs in people with osteogenesis imperfecta
- Have mutation in COL1A1 or COL1A2

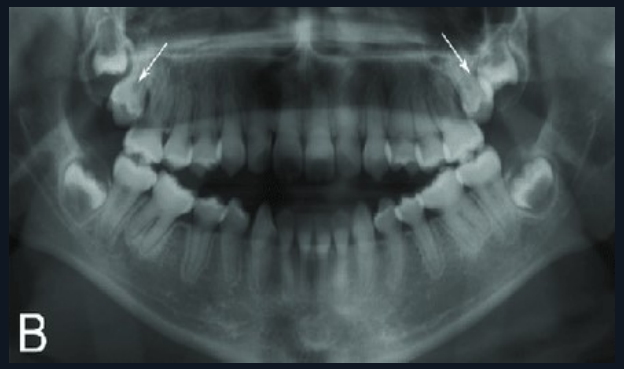
Type II

- Most common
- Occurs in people without inherited disorders
- Teeth have short roots and missing pulp chambers
- Progressive hearing loss during old age

Type III

- Very rare
- Occurs in people without inherited disorders
- Teeth have shell-like appearance and multiple pulp exposures





Treatment

- Remove sources of infection or pain
- Restore aesthetics, which includes amalgam, veneers, crowns, caps, bridges
- If majority of teeth are lost, then dentures or dental implants may be necessary
- Protect teeth from wear
- Some dentists recommend resin restorations or whitening teeth

Role of Dental Team

- Ask for thorough family medical history
- Suggest genetic testing
- Look for specific signs and symptoms and do clinical examination
- Get x-rays of teeth

References

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