

# **AMELOGENESIS IMPERFECTA**

- Autosomal dominant
- Autosomal recessive
- X – linked
  
- Types
  - Hypoplastic ( 60-73%)
  - Hypocalcified ( 7%)
  - Hypomature (20-40%)

# ETIOLOGY

- Genes involved
  - *Amelogenin* (*AMELX* and *AMELY*) on chromosome X
- Other genes involved
  - AMBN → ameloblastin
  - *ENAM* gene → Enamelin
  - Enamelysin
  - Kalikryn 4
  - Tuftelin

# CLINICAL FEATURES

- Hypoplastic type
  - Autosomal or X-linked
  - Generalized or Localized
  - Smooth, Rough or Pitted



## Generalized pitted variety



- Buccal surface more severely involved

- Arranged in rows or columns

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# Smooth type



- Enamel is thin, hard and glossy
- Opaque white to translucent brown in colour
- Teeth shaped like crown preparations
- Open contact points
- Anterior open bite

## X-linked pattern



- Females
  - Alternating zones of normal and abnormal enamel
- Males
  - Similar to smooth type



# Rough pattern



- Enamel is thin, hard and rough surfaced
- White to yellow white
- Crown preparation appearance
- Open contact points
- Anterior open bite

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# Enamel agenesis



- Total lack of enamel formation
- Yellow brown hue
- Rapid attrition

- Hypomaturation type

- Defect in maturation of enamel crystal structure
- Shape of tooth is normal
- Enamel is soft
  - Tends to chip away
  - Can be punctured by a dental explorer
- Mottled in appearance
- Agar brown colour





- Hypocalcified type

- Enamel matrix is laid down normally but no significant calcification

- Teeth normal in shape at time of eruption

- Enamel is very soft and easily lost

- Yellow, brown or orange staining





# RADIOGRAPHIC FEATURES

- Hypoplastic type
  - Thin peripheral rim of enamel
  - Enamel can be distinguished from the underlying dentin
- Hypomaturational and hypocalcification type
  - Contrast between enamel and dentin is lost

# ENVIRONMENTAL CAUSES OF ENAMEL HYPOPLASIA

- Nutritional deficiency and exanthematous diseases
  - Vitamin A and C deficiency
  - Measles, chickenpox, scarlet fever

- Congenital syphilis

- Hutchinsons teeth (incisors)

- Mulberry molars (Moon's molar, Fournier's molar)





- Hypocalcemia
  - $\text{Ca}^{++}$  less than 6-8 mg / 100 ml
- Birth injuries
  - Turner's teeth / turner's hypoplasia

- Fluoride



# **DENTINOGENESIS IMPERFECTA**

- Also called as
  - *Hereditary opalascant dentin*
  - *Capdepont's teeth*
- Hereditary developmental disturbance of dentin in absence of any systemic disorder
- In presence of systemic disorder →  
*Osteogenesis imperfecta with opalascant dentin*
- Autosomal dominant →
  - chromosome 4
  - Dentin sialophosphoprotein (DSPP)



# CLASSIFICATION

- Old classification

Type I	DI associated with OI
Type II	DI without OI
Type III	Brandywine type

# CLASSIFICATION

- New classification

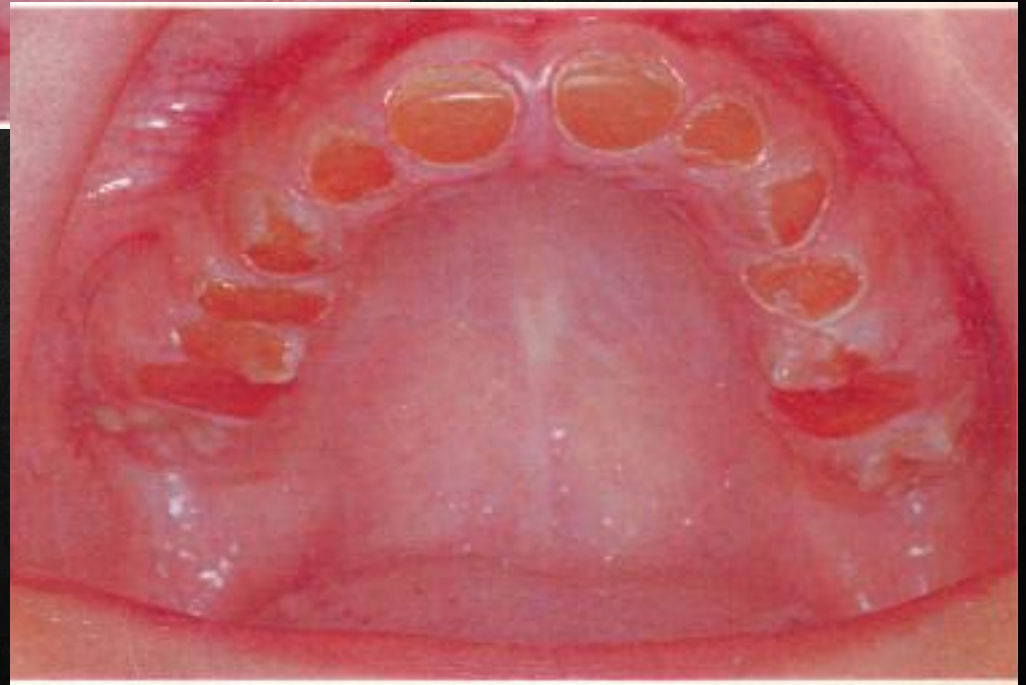
Type I	DI without OI
Type II	Brandywine type

# CLINICAL FEATURES

- DI type I
  - Blue gray or amber brown opalescent hue
  - Bulbous crowns
  - Narrow roots
  - Obliterated pulp chambers and root canals







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- Brandywine type
  - Dentin is amber colored and smooth
  - Crowns wear rapidly after eruption
  - Multiple pulp exposures

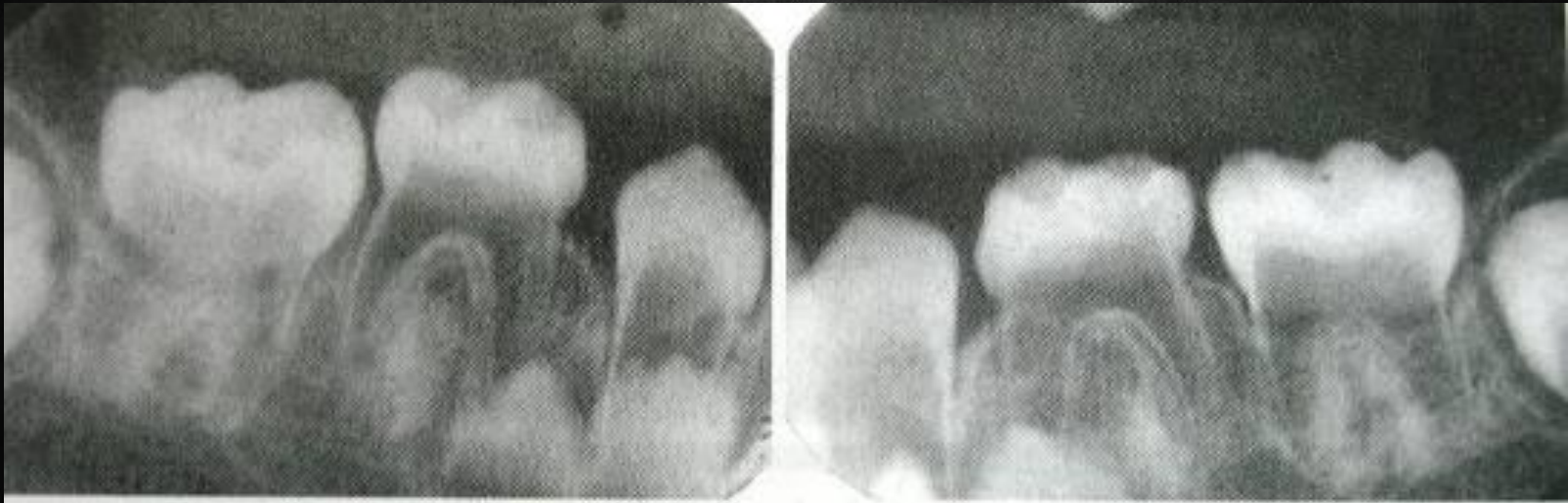
# RADIOGRAPHIC FEATURES

- DI type I





- Brandywine type



## ***Shell teeth***

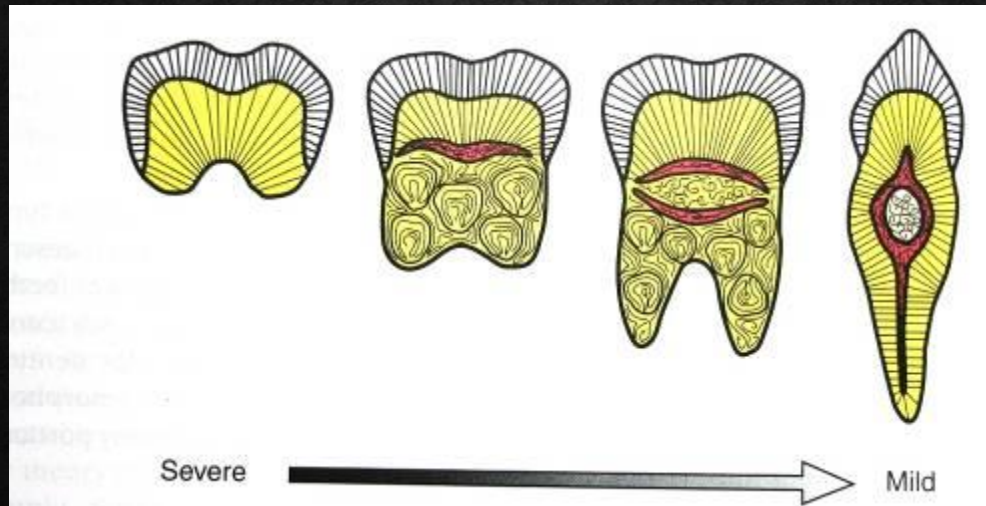
# **DENTIN DYSPLASIA**

- Rare disturbance in dentin formation characterized by
  - Normal enamel
  - Atypical dentin + abnormal pulp morphology
- Autosomal dominant trait



# CLASSIFICATION

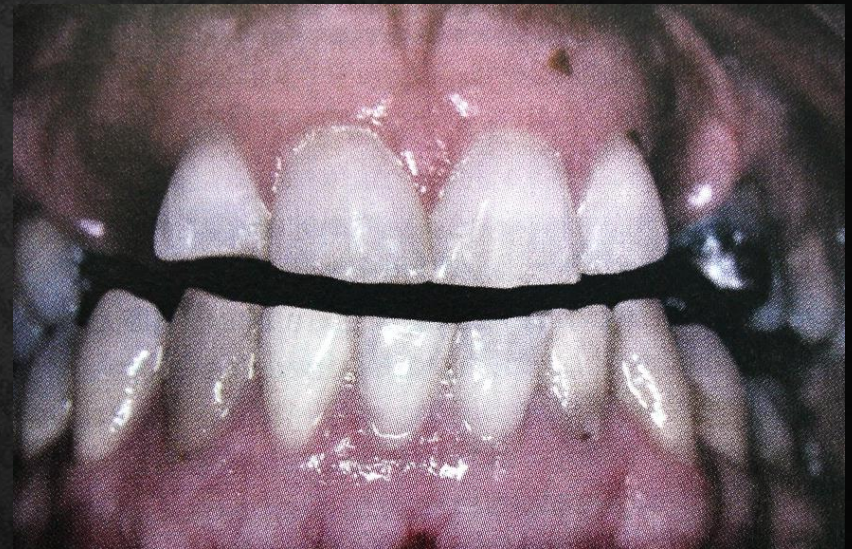
- Type 1: Radicular dentin dysplasia (rootless teeth)



- Type 2 : Coronal dentin dysplasia

# RADICULAR DENTIN DYSPLASIA

- Autosomal dominant
- Both dentition affected
- Clinically → Appears normal
- Root is stunted





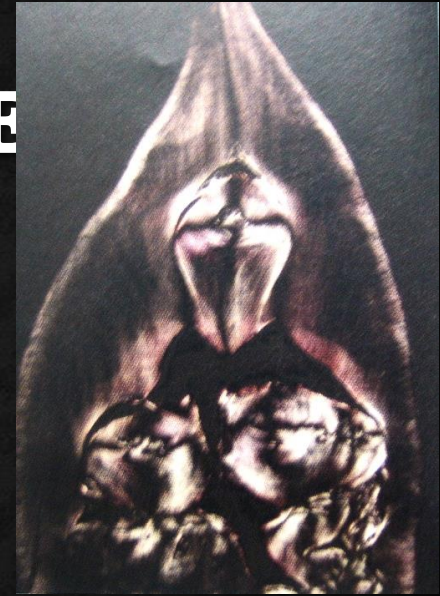
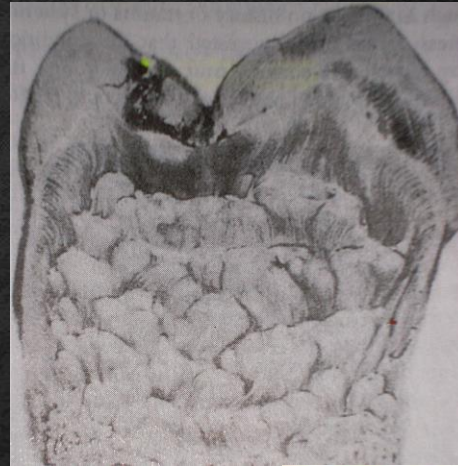
- Radiographically
  - Roots are short and blunt or conical
  - Obliteration of pulp chamber and root canal
  - PA granuloma / cyst without obvious reason





# HISTOLOGIC FEATURE

- Coronal dentin normal
- Obliteration of pulp by calcified tubular dentin, osteodentin, fused denticles
- *“Lava flowing around boulders”* appearance





# CORONAL DENTIN DYSPLASIA

- Autosomal dominant
- Both dentition affected
- Deciduous teeth
  - Appear yellow brown to blue
  - Complete obliteration
- Permanent normal
  - Thistle tube
  - Pulp stone most characteristic



# RADIOGRAPHIC FEATURES

- Deciduous teeth
  - Complete obliteration
  
- Permanent normal
  - Abnormally large pulp chambers  
→ *Thistle tube appearance*
  - Pulp stones



# HISTOLOGIC FEATURES

- Deciduous teeth
  - Amorphous and atubular dentin
- Permanent teeth
  - Multiple pulp stones

# REGIONAL ODONTOGENIC DYSPLASIA

- Odontodysplasia
- Odontogenic dysplasia
- Odontogenesis imperfecta
- Ghost teeth
  
- Etiology → unknown
  - Somatic mutation
  - Latent viral infection
  - Vascular malformation (associated vascular nevi)

# CLINICAL FEATURES

- Maxillary anterior region  
> mandible
- Delay or lack of eruption
- Irregular shape
- Defective mineralization





# RADIOGRAPHIC FEATURES

- Enamel and dentin → very thin
- Pulp chamber → exceedingly large



**Ghost teeth**

# HISTOLOGICAL FEATURES

- Marked reduction in amount of dentin
- Widening of predentin layer
- Large areas of interglobular dentin
- Irregular tubular pattern























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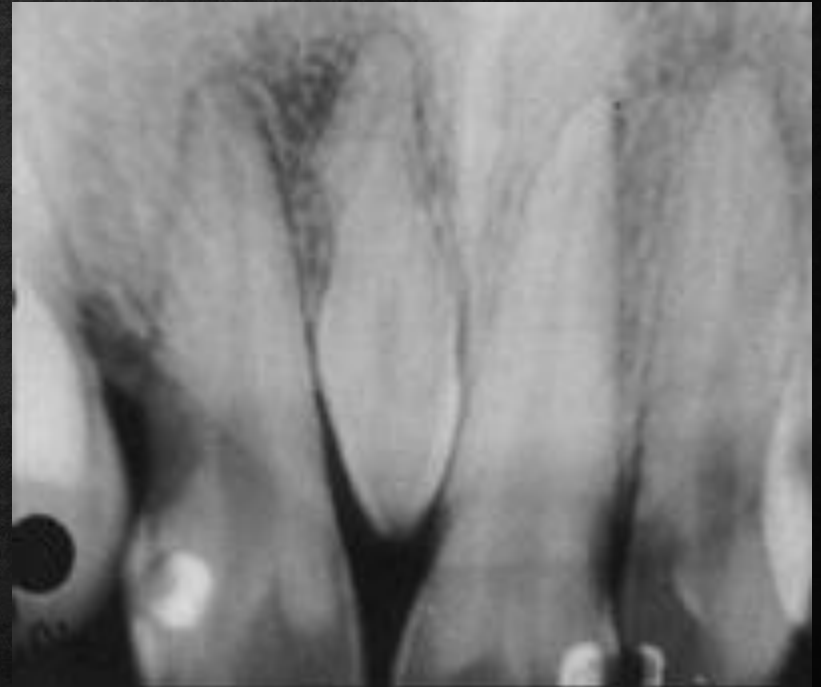


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