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

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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

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X-linked pattern

• Females
  • Alternating zones of normal and abnormal enamel

• Males
  • Similar to smooth type

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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

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• **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

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• 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

- Hypomaturation 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

• Hutchinson's teeth (incisors)

• Mulbery molars (Moon’s molar, Fournier’s molar)
• Hypocalcemia
  • Ca^{++} less than 6-8 mg / 100 ml

• Birth injuries
  • Turner’s teeth / turner’s hypoplasia

• Fluoride

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DENTINOGENESIS IMPERFECTA
• Also called as
  • *Hereditary opalascent dentin*
  • *Capdepont’s teeth*

• Hereditary developmental disturbance of dentin in absence of any systemic disorder

• In presence of systemic disorder ⇒ *Osteogenesis imperfecta with opalascent dentin*

• Autosomal dominant ⇒
  • chromosome 4
  • Dentin sialophosphoprotein (DSPP)
## CLASSIFICATION

- **Old classification**

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
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</thead>
<tbody>
<tr>
<td>Type I</td>
<td>DI associated with OI</td>
</tr>
<tr>
<td>Type II</td>
<td>DI without OI</td>
</tr>
<tr>
<td>Type III</td>
<td>Brandywine type</td>
</tr>
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</table>
New classification

<table>
<thead>
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</tr>
</tbody>
</table>
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

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RADIOGRAPHIC FEATURES

• DI type I
• Brandywine type

Shell teeth

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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

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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 FEATURES

- Coronal dentin normal

- Obliteration of pulp by calcified tubular dentin, osteodentin, fused denticles

- “Lava flowing around boulders” appearance

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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)

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CLINICAL FEATURES

• Maxillary anterior region > mandible

• Delay or lack of eruption

• Irregular shape

• Defective mineralization

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RADIOGRAPHIC FEATURES

• Enamel and dentin → very thin

• Pulp chamber → exceedingly large
HISTOLOGICAL FEATURES

• Marked reduction in amount of dentin

• Widening of predentin layer

• Large areas of interglobular dentin

• Irregular tubular pattern