

## Developmental Alterations of Teeth II

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## Amelogenesis Imperfecta pp: 99-106

### General Information

- Classification can be impractical for clinicians
- Problems arise in one or more of the three stages of enamel formation
  - Elaboration of enamel matrix; hypoplastic
  - Mineralization; hypocalcified
  - Maturation; hypomaturation

### General Information

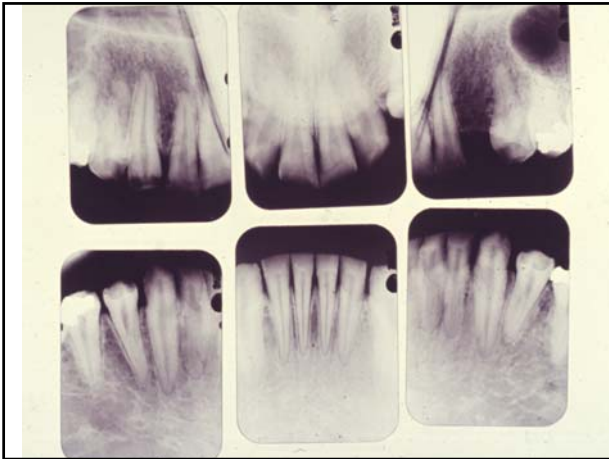
- Absence of systemic disorder
- Can be part of a syndrome
- Many types
- Different modes of inheritance
- Different phenotypes in a single family
- Same phenotype with different gene mutations
- Homozygotes differ from heterozygotes
- 1:800 – 1:15,000 (clustering)
- Both dentitions

### Genes and Phenotypes

- AMELX (amelogenin)
  - X-linked
  - Diffuse smooth hypoplastic and hypomaturation
- ENAM (enamelin)
  - AD, AR
  - Generalized pitting and thin enamel
- MMP-20 (enamelysin)
  - AR pigmented hypomaturation
- KLK4 (kallikrein-4)
  - hypomaturation
- DLX3 (genes of craniofacial development)
  - Hypoplastic/hypomaturation and taurodontism
- AMBN (ameloblastin)
  - Indication for strong association

### Hypoplastic type

- Inadequate deposition of organic matrix
- Normal mineralization
- Radiographic contrast
- Seven types



## Hypoplastic type Generalized Pitted

- A.D.
- Pinpoint/head pits in rows or columns
- In-between enamel normal
- Across the surface
- Buccal surface more severely affected
- Does not correlate with pattern of environmental damage

## Hypoplastic type Generalized Pitted



## Hypoplastic type Generalized Pitted



## Hypoplastic type Generalized Pitted



## Hypoplastic type Generalized Pitted



### Hypoplastic type Localized Pitted

- A.D. or A.R. (more severe)
- Pinpoint/head pits in rows
- Linear depression or area of hypoplasia
- Middle third of buccal surface
- Incisal or occlusal unaffected
- All teeth or some teeth
- Primary or both dentitions

### Hypoplastic type Localized Pitted



### Hypoplastic type Diffuse Smooth A.D.

- Thin, hard, glossy
- Like crown preparations, open bite
- Opaque white to brown
- X-ray: peripheral thin enamel outline
- Unerupted exhibit resorption

### Hypoplastic type Diffuse Smooth A.D.



### Hypoplastic type Diffuse Smooth A.D.



### Hypoplastic type Diffuse Smooth X-linked dominant

- Males
  - Thin, hard, glossy
  - Like crown preparations, open bite
  - Opaque white to brown
  - X-ray: peripheral thin enamel outline
- Females
  - Alternating vertical bands of normal and abnormal enamel
  - Lyonization

### Hypoplastic type Diffuse Smooth X-linked



### Hypoplastic type Diffuse Rough

- A.D.
- Thin, hard, rough
- Tapering of occlusal and incisal surfaces
- Open contact
- Open bite

### Hypoplastic type Diffuse Rough



### Hypoplastic type Diffuse Rough



### Hypoplastic type Enamel Agenesis

- No enamel
  - Some enamel is however present in some cases
- Yellow teeth
- Tapering
- There is also a type of enamel agenesis associated with nephrocalcinosis

### Hypomaturation Type

- Defect in the maturation of enamel crystals
- Normal shape
- Mottled appearance
- White, yellow or brown
- Enamel is soft
- Radiodensity similar to dentin or reduced

### Hypomaturation Type



### Hypomaturation Type Diffuse Pigmented A.D.

- Mottled brown
- Chipping from dentin with an explorer
- Very uncommon anterior open bite
- Soft similar to hypocalcified
- Calculus

### Hypomaturation Type Diffuse Pigmented A.D.



### Hypomaturation Type Diffuse Pigmented A.D.



### Hypomaturation Type Diffuse X-linked

- Deciduous are opaque white with mottling
- Permanent are yellow-white; darken with age
- Fast enamel loss
- Reduced contrast
- Females exhibit lyonization
  - Random asymmetric bands
  - Transillumination highlights bands

### Hypomaturation Type Diffuse X-linked





Hypomaturation Type  
Diffuse X-linked (female)



Hypomaturation Type  
Diffuse X-linked (female)



Hypomaturation Type  
Snow-capped Teeth

- X-linked, A.D.?
- Zone of white opaque enamel on incisal and occlusal surface (1/4 to 1/3 of the surface)
- Looks like fluorosis
- Anteriors, anteriors/bicuspid, premolars/molars
- Both dentitions

Hypomaturation Type  
Snow-capped Teeth



Hypomaturation Type  
Snow-capped Teeth



Hypocalcified Type

- A.D. or A.R. (more severe)
- No significant mineralization
- Normally shaped teeth at eruption
- Enamel very thin and easily lost
- Yellow or brown color
- Calculus
- Open bite

### Hypocalcified Type



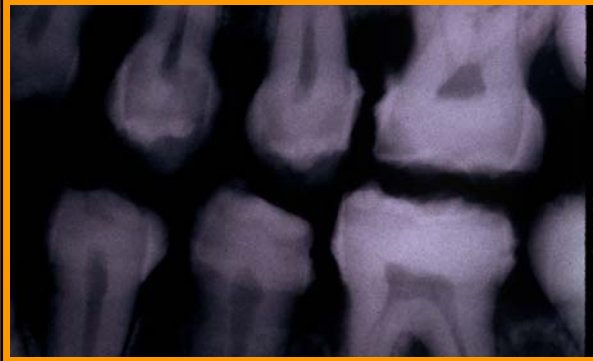
### Hypocalcified Type



### Hypocalcified Type



### Hypocalcified Type



### Hypomaturation/Hypoplastic

- Enamel hypoplasia and hypomaturation
- One kindred
- Present in tricho-dento-osseous syndrome
  - Kinky hair
  - Osteosclerosis
  - Brittle nails
  - Taurodontism

### Tricho-dento-osseous syndrome



## Tricho-dento-osseous syndrome



## Tricho-dento-osseous syndrome



## Tricho-dento-osseous syndrome



## Amelogenesis Imperfecta Treatment

- Restorations as soon as possible
- Dentures (overdentures)
- Veneers in mild cases
- Glassionomers for better adhesion to dentin

## Dentinogenesis Imperfecta pp: 106-108

## General Information

- Two types
  - Dentinogenesis imperfecta with osteogenesis imperfecta
    - COL1A1 and COL1A2
  - Hereditary opalescent dentin
    - DSPP (dentin sialophosphoprotein)
      - Also in DDI
    - (Brandywine isolate)
- 1:8,000
- Both dentitions
- Deciduous more severely
- Permanent incisors, first molars
- Least 2<sup>nd</sup> and 3<sup>rd</sup> molars



## General Information

- Blue/brown discoloration
- Bulbous crowns with cervical constrictions
- ~100% penetrance; variable expressivity
- Enamel defects (hypoplasia)
- Generally obliterated pulp chambers and canals except shell teeth (Brandywine isolate)
- Periapical pathology
- Treatment very difficult

## Osteogenesis/Dentinogenesis Imperfecta



## Osteogenesis/Dentinogenesis Imperfecta



## Osteogenesis/Dentinogenesis Imperfecta



## Osteogenesis/Dentinogenesis Imperfecta



## Osteogenesis/Dentinogenesis Imperfecta



## Hereditary Opalescent Dentin



## Hereditary Opalescent Dentin



## Shell Teeth



## Dentin Dysplasia pp: 108-112

## General Information

- A.D.
- Two types
  - Radicular type (Type I)
  - Coronal type (Type II)
- No correlation with systemic disease
  - DD-like changes in teeth of patients with
    - Calcinosis universalis
    - Rheumatoid arthritis
    - Tumoral calcinosis
- DDII and HOD

## Dentin Dysplasia Type I

- Rootless teeth
- Severity varies even in the same patient
- A.D. 1:100,000
- Normal color and anatomy of crown in both dentitions
- Dentinal disorganization
- Tooth mobility and premature exfoliation
- Teeth fracture easily

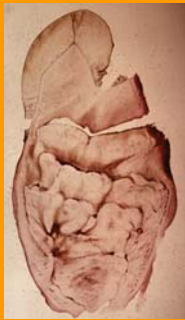
## Dentin Dysplasia Type I

- Little or no detectable pulp
- Crescent or chevron-like pulp chamber
- Pulp stones in less severe cases
- Multiple periapical lesions (caries, exposure)

## Dentin Dysplasia Type I



## Dentin Dysplasia Type I



## Dentin Dysplasia Type II

- Form of hereditary opalescent dentin
- Normal root length
- Multiple periapical lesions uncommon
- Deciduous teeth like HOD
  - Amber to brown color
  - Bulbous crowns, cervical constrictions, thin roots and obliterated pulp chambers (not before eruption)
- Permanent teeth
  - Normal color
  - Enlarged pulp chambers (flame-like)

## Dentin Dysplasia Type II



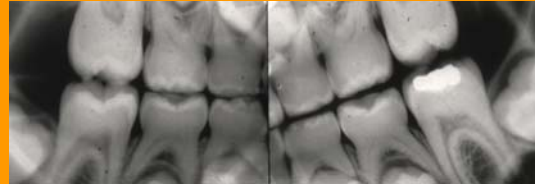
## Dentin Dysplasia Type II



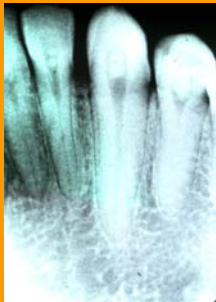
## Dentin Dysplasia Type II



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## Dentin Dysplasia Type II

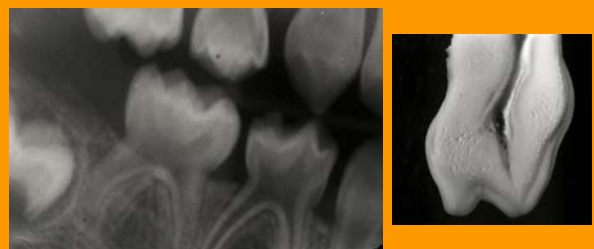


## Vitamin D-resistant Rickets pp: 847-848

## General Information

- X-linked dominant
- Lyonization in females
- Hypophosphatemia
- Decreased intestinal absorption of calcium
- Rachitic changes
- Teeth have large pulp chambers and pulp horns extending to the DE junction
- Microexposures
- Multiple periapical lesions

## Vitamin D-resistant Rickets



## Hypophosphatasia pp:845-847

### General Information

- Alkaline phosphatase mutation
- A.D. or A.R.
- Decreased levels of alkaline phosphatase
- Increased levels of phosphoethanolamine
- 4 types
  - Perinatal: lethal
  - Infantile: problems after 6 month, failure to grow, nephrocalcinosis, lithiasis
  - Childhood: later age, open fontanelles with premature fusion, “beaten copper” skull radiolucencies
  - Adult: mild
- Rickets
- Premature loss of teeth
- Teeth lack cementum

### Hypophosphatasia



### Hypophosphatasia



## Regional Odontodysplasia

### General Information

- **Non-hereditary**
- Can be associated with other pathoses
  - Ectodermal dysplasia, hypophosphatasia, vascular abnormalities
- Various theories of pathogenesis
  - Local circulatory deficiency, trauma, infection, mutation?, meds
- Maxilla > mandible
- Usually one quadrant
  - Occasionally normal tooth between malformed
- Both dentitions



## General Information

- Affected teeth fail to erupt frequently
- Yellow/brown color
- Ghost teeth

## Regional Odontodysplasia



## Regional Odontodysplasia



## Regional Odontodysplasia



## Regional Odontodysplasia

