Oral pathology 29/6/2015

Sheet # 4

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→All extra notes are underlined by dotted line.

Generalized enamel disturbances due to hereditary disturbances (genetic):

1. Affecting only Teeth: Amelogenesis Imperfecta
2. Generalized defects including teeth: Ectodermal dysplasia & Down syndrome

❋ Amelogenesis Imperfecta

- Inheritance: Autosomal dominant (most of the cases), recessive, X-linked
- Most of enamel on all teeth in both dentitions
- Other components of teeth (dentine, pulp, cement, …) are normal
- Family history
- Mutations in some genes that are related to enamel formation, could lead to amelogenesis imperfect: ENAM, AMELX, MMP20, KLK 4.
- No systemic disease (distinguished by their specific dental abnormalities).
- There are more than 16 forms of amelogenesis imperfecta that differ in the clinical presentation and the pattern of inheritance.
- The incidence varies from 1:1000 [the dr. said 1: 700 (in Sweden)] to 1:15000 (in USA) of the population.
- How to differentiate between Amelogenesis Imperfecta & chronological enamel hypoplasia or turner?
  AI affects only enamel, on most of the teeth in both dentitions.

  ➔ Q: Are there any reported cases of AI with no family history of the disorder?

Note: The enamel formed in two steps: formation of the matrix then calcification.

So AI types differ by what its affect:

- Affecting the matrix formation (the organic structure) …. Hypoplastic type → abnormal enamel thickness but with normal calcification.
- Affecting the calcification ….. Hypomineralized type → normal enamel thickness but abnormal calcification.

- Types:
  1. **Hypoplastic type**:
     - Thin enamel but normally mineralized …. normal hardness
     - E > dentine in radiodensity "as normal" …. you can distinguish enamel from dentine in radiographs.
     - Abnormal color and shape with spaces between teeth.
     - **Generalized (affect all enamel)**…. there will be smooth teeth with needle like cusps (pointed cusps) and sharp incisal edges (the structures that are mainly composed from enamel are mainly affected).
     - **Not all enamel** …. generalized roughness with pitting and vertical grooves.
  2. **Hypomineralized/hypomaturation type**:
     - Most common form
     - Enamel of normal thickness
     - Newly erupted: normal size & shape of teeth
     - Newly erupted teeth will have white chalky appearance then it became opaque, brown-yellow because it will be stained due to its high porosity.
     - Enamel soft chalky and easily removed (because of less minerals) → gross attrition
     - E = D in radiodensity…. you can't distinguish enamel from dentine in radiographs.
     - Teeth get darker in time because of the reduced mineral content

✓ To distinguish between Amelogenesis Imperfecta & other abnormalities: AI=defect affecting all teeth + in both dentitions + family history

Dentine

Now we will discuss disturbances affecting dentine, same as enamel due to:

- **Local causes** that may cause disturbance of the structure of dentine such as: local trauma or infection (turner teeth) or radiotherapy.

- **General causes**: due to Systemic (1) disturbances during dentin development or it could be due to genetic disease (2) such as Dentinogenesis Imperfecta.

a) Systemic causes include:

1. Rickets, will affect mineralization of dentine. Increase predentine width, hypocalcified dentine with increase in interglobular D.

2. Hypophosphatemia, increase in interglobular D, large pulp chamber & long pulp horns with cracked E (all these factors will lead to early pulpitis and loss of vitality).

3. Hypophosphatasia, deficiency of alkaline phosphatase enzyme, will lead to defect in the mineralization of bone and calcified structures, increase predentine and increase in interglobular D and large pulp chamber.

4. Juvenile hypoparathyroidism:
    - Small teeth w hypoplastic E and short roots
    - Prominent incremental lines in D
    - Disturbances in teeth eruption

5. Cytotoxic agents during dentin development(as taking chemotherapy): Prominent incremental lines in D

All of these systemic diseases produce characteristics changes in dentin at histological level; most of them result in increased predentin formation & interglobular dentin.

b) Dentinogenesis Imperfecta

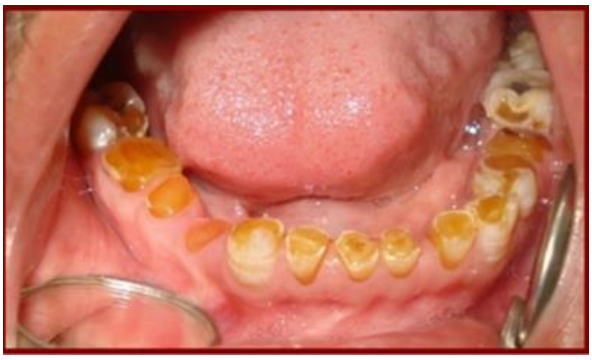
- Hereditary generalized disturbance affecting formation of dentine.
- It's a genetic disease inherited in an autosomal dominant manner/family history.
- affect dentitions, all teeth and surfaces as in Amelogenesis imperfect.
- Mutation in DSPP gene (Dentine Sialophosphoprotein)

- It has 3 types:
  - ꕥ **Type I**:
    - Dental abnormality associated with Osteogenesis Imperfecta (OI a bone disease).
    - Autosomal dominant (family history)
    - The presence of blue sclera which can help in diagnosis.
  - ꕥ **Type III -Brandywine isolate-** : [the dr. said that it's not important]
    - it's actually very rare & it happens in certain racial isolate in USA but otherwise it's rare & it's characterized by then dentin & large pulp chambers
  - ꕥ **Type II (Hereditary opalescent dentine أسنان براقة):**
    - The most common dental genetic disease, involving approximately 1in 6000 to 1in 8000 of the population. The type that is imp. & we must know about.
    - Teeth only affected no OI / Isolated defect in dentine.
    - Autosomal dominant
      - *Clinically* (spot diagnosis)
        - > all teeth & both deciduous and permanent dentitions are affected.
        - > Bluish-gray, brown/yellowish
        - > Soft dentine → attrition
        - > On eruption teeth have a normal contour but opalescent amber-like appearance كهرمان. Subsequently, they may have an almost normal color, following which they become translucent, and finally gray or brownish with bluish reflections from the normal, it is rapidly lost and the teeth then show marked attrition.
        - > ↓ caries
      - *Radiographs*
        - > appear as **thin, short, blunt roots** (this will affect the function of these teeth) & obliteration of pulp chamber with abnormal dentin, in addition the **crown appears bulbous** due

to constriction in the cervical region & because of this morphology "large crown & thin root" there is susceptibility of **root fracture**.

- *Histologically*
  - characterized by **normal enamel** & **normal mantle dentin (**the dentine immediately adjacent to the enamel or cementum**)** but the rest of dentin is **hypomineralized** so softer than normal **with irregular** & **wide** dentinal tubules, these tubules often **devoid of odontoblastic process and there are less number** of dentinal tubules.

- ✓ Originally it was though that a defective dentinoenamel junction (DEJ) was present and was resulting in chipping of enamel; SEM studies have discovered normal junction (no loss of scalloping at DEJ) and the reason for enamel loss is that the enamel is hard (normal enamel) and the dentine is softer than normal so when there is some pressure on the teeth, there will be fracture of enamel then chipping.
- ✓ **Attrition** in these teeth is very common, very **severe** and rapid but the teeth are **not particularly sensitive** because of absence of odontoblastic process & the obliteration of pulp chamber with abnormal dentin, the cause of attrition here is that enamel is normal but it is supported by weak-soft dentin the **enamel is easily lost & fractured** resulting in exposure of dentin (but **no pulp exposure**) consequently developing rapid attrition, in these cases **there is loss of crowns**, so **it's advisable & indicated to place full crown "full coverage** "in order to protect tooth structure , maintain vertical dimension & avoid tooth attrition.



➔ Questions:

1- Which is more common DI or AI?

2- What about dental caries?

3- Tooth sensitivity (more than or equal to normal people)?

4- Can we do Crowning (when we should make crowns 'time')?

c) **Dentinal Dysplasia**

- Autosomal dominant (its rare condition)
- affect dentin formation.
- 2 types:

ᘓ **Type I … is Radicular Dentine Dysplasia (Rootless dentine):**

- Most common type
- as the name implies (Radicular) the crown is normal but the defect affect mainly the root.
- After eruption of teeth it's of normal size, shape & color *clinically* but on X-ray (*radiographs)* you can see short conical blunt or even absent roots, and sometime the condition is described as rootless teeth because the teeth are very short or there is no root at all.
- similar to Dentinogenesis Imperfecta there is **obliteration of pulp chamber & root canals**, but here in some teeth the pulp chamber appears as thin line or **crescent** in shape. In addition it's very common in these cases to find **Periapical radiolucencies** without obvious case such as **caries.** The difference between Dentinogenesis Imperfecta and radicular dentin dysplasia is clinically that the color of teeth of DI is Bluish-gray but in case of radicular dentin dysplasia the crown is normal.
- *Histologically:* the radicular dentin is irregular and surrounded by numerous calcified spherical bodies (the odontoblast processes are winding between these calcifications) sometime described as water **streaming around boulders**.

➔ Q: Expected complications?

You can expect "because of the short root" the initial clinical sign of this disease is extreme mobility without evidence of periodontal disease. And some time there is continuous exfoliation of teeth either spontaneously or secondary to minor trauma.

-Less frequently delayed eruption is the presenting symptom.

- Fracture of roots during extraction

****In other word in radicular dentin dysplasia there is dysplasia of dentin of the root only so clinically the teeth might appear normal but in radiograph the roots appear abnormal.**

- ꕥ **Type II (Coronal Dentine Dysplasia):** the roots here are normal but the defect affect dentin in the crown only.
  - ❖ Primary teeth; clinically they are identical to Dentinogenesis Imperfecta clinically and with obliteration of pulp chambers & root canals. So in primary teeth we can't differentiate between Dentinogenesis Imperfecta & Coronal dentin dysplasia.
  - ❖ Permanent teeth; appear of normal color (clinically). Thistle-tube pulp chambers "wide pulp chambers" or the pulp chamber appear like flame & multiple pulp stone can be seen (in radiographs)
    - ✓ So if you have a patient with abnormal primary teeth and you are confused if its Dentiogenesis Imperfecta or Coronal dentine dysplasia, you have to wait until permanent teeth erupt.

d) Regional Odontodysplasia

- Affect all teeth components (enamel, dentine, pulp, cementum, alveolar bone).
- Regional: disturbance affect group of teeth (mostly in anterior maxilla "incisors & canines")
- Very rare & of unknown etiology, not hereditary.
- Characterized by delay or failure of eruption.

- Irregular & hypoplastic enamel.
- Thin dentine
- The dentin is also hypoplastic (hypocalcified) with increased in the interglobular dentin.
- Pulp stones and widely open apices.
- Large & wide pulp chambers
- Focal calcifications in the dental follicle
- Enamel and dentin have the same density so we can't differentiate between them because both are hypocalcified & due to large pulp chamber surrounded by radiopaque rim so **they described as** Ghost teeth. أشباح