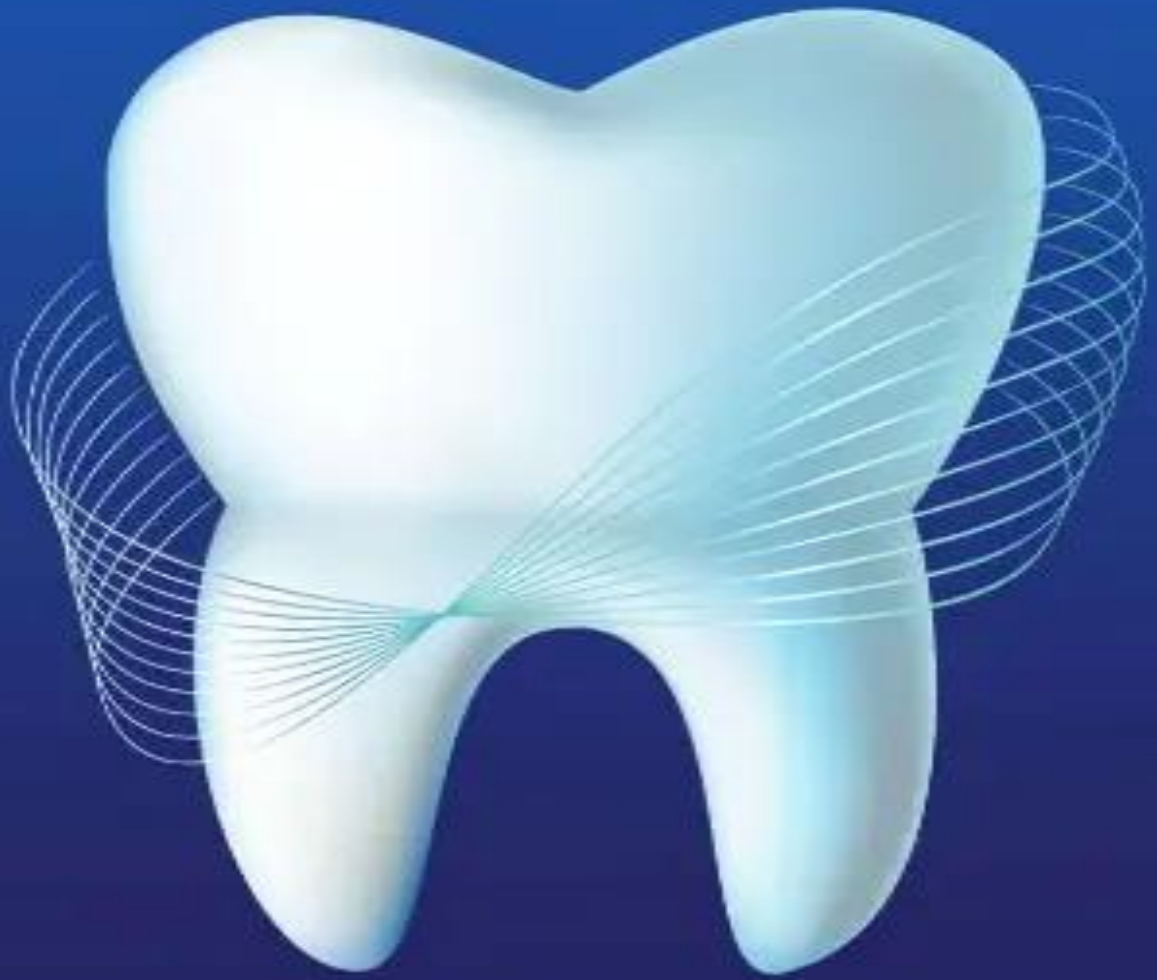
Amelogenesis Imperfecta
(imperfect enamel development)



Disturbances in the structure of Enamel

Disturbance in the structure of enamel occurs as a result of environmental factors or hereditary factors.

Environmental factors are:-

1-bacterial and viral infections (syphilis, measles, scarlet fever).

2-inflammation.

3-nutritional deficiencies (Vit. A,C. and D and Calcium).

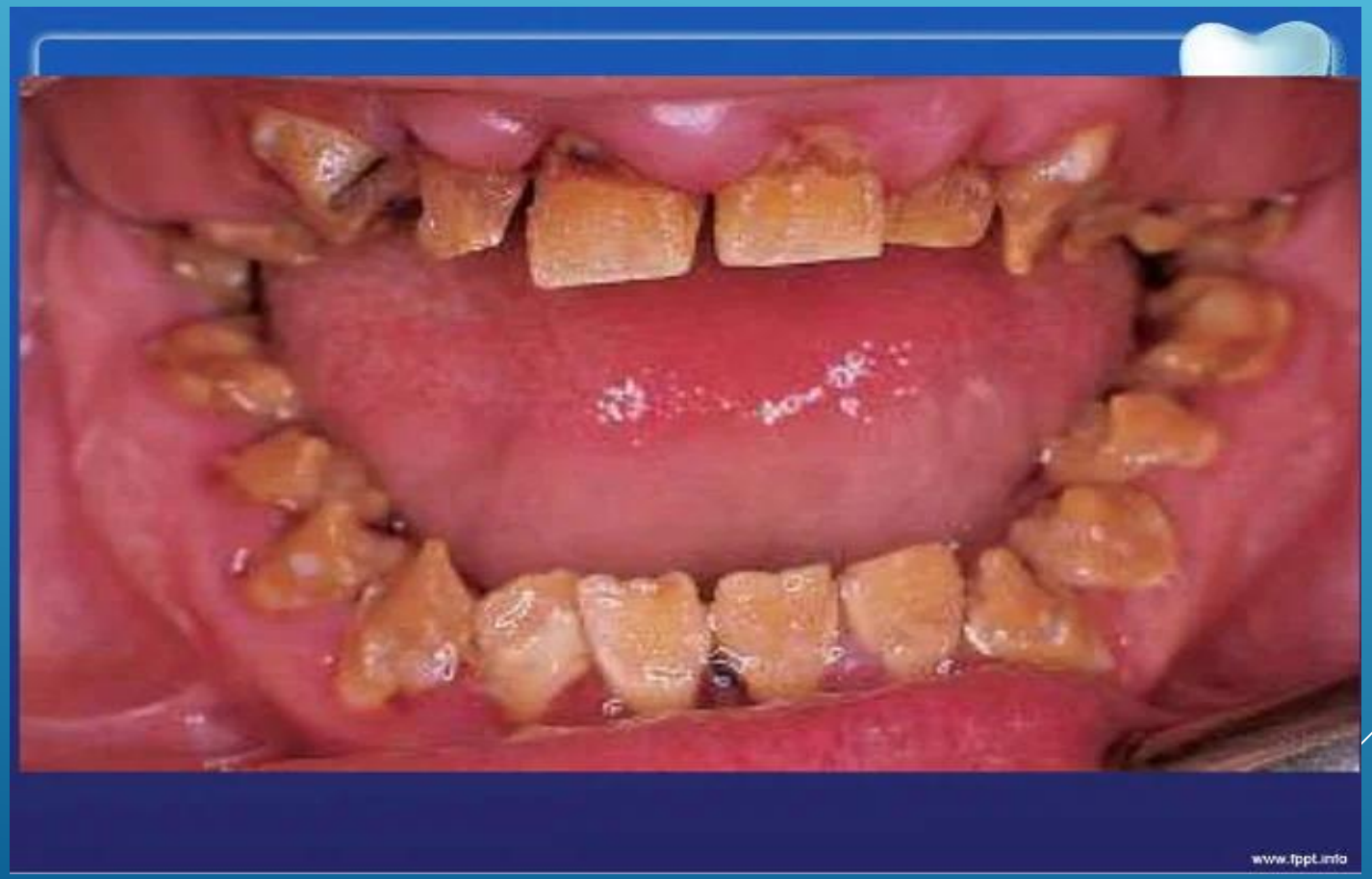
4-Trauma.

The enamel disturbances may be localized to one or two teeth (focal), or it may affect many teeth or all teeth (generalized) and that depends on the etiological factor.

Otherwise known as……

- A I
- Hereditary Enamel Dysplasia
- Hereditary Brown Enamel
- Hereditary Brown Opalescent Teeth

The structure and clinical appearance of dental enamel of nearly all teeth are affected in both primary and secondary dentition and is characterized by hypomineralisation and/or hypoplasia with discoloration, sensitivity and fragility. This condition causes teeth to be unusually small, discolored, pitted or grooved, and prone to rapid wear and breakage. Other dental abnormalities are also possible.



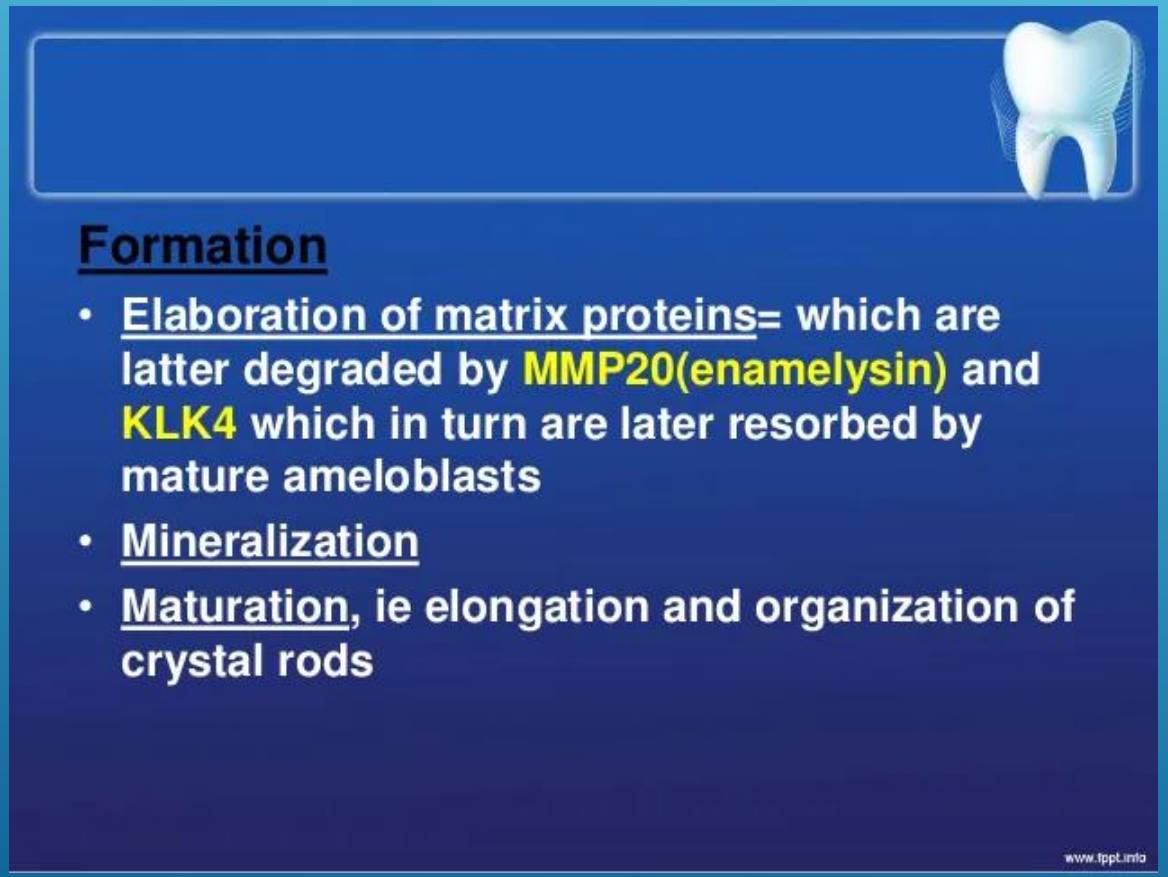
What is Amelogenesis Imperfecta ?

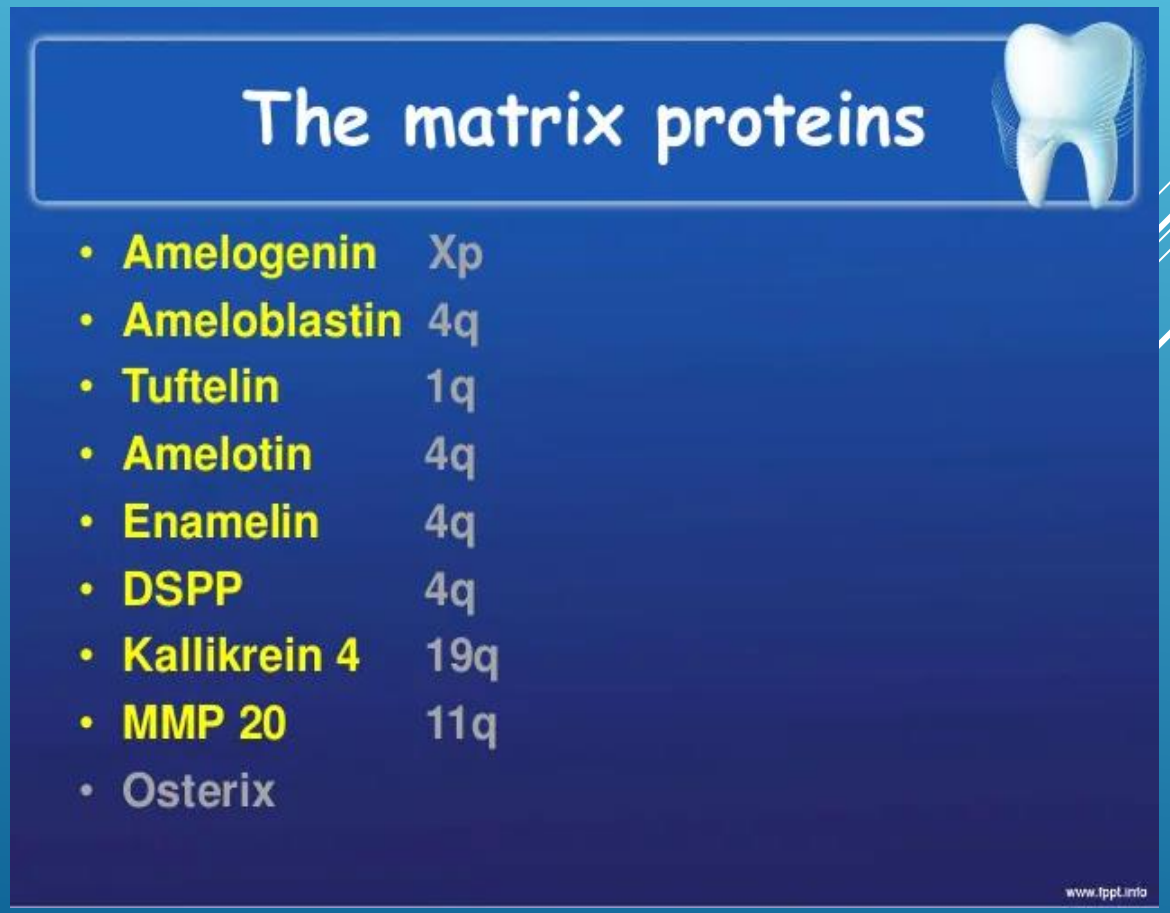
- Amelogenesis Imperfecta represents a group of **hereditary defects of enamel** unassociated with any other generalized defects. It is entirely an **ectodermal disturbance** , since the mesodermal components of the teeth are basically normal.

Development of enamel..

- 3 stages..
  - formative stage
    deposition of organic matrix.
  - Calcification stage
    matrix mineralization
  - Maturation stage
    crystallites enlarge and mature

Enamel is composed mostly of minerals (95% to 98% of it is calcium and phosphate ions that make up strong hydroxyapatite crystals), that are formed and regulated by the proteins in them. ***AI* is due to the malfunction of the proteins in the enamel (ameloblastin, enamelin, and amelogenin) as a result of abnormal enamel formation via amelogenesis**.

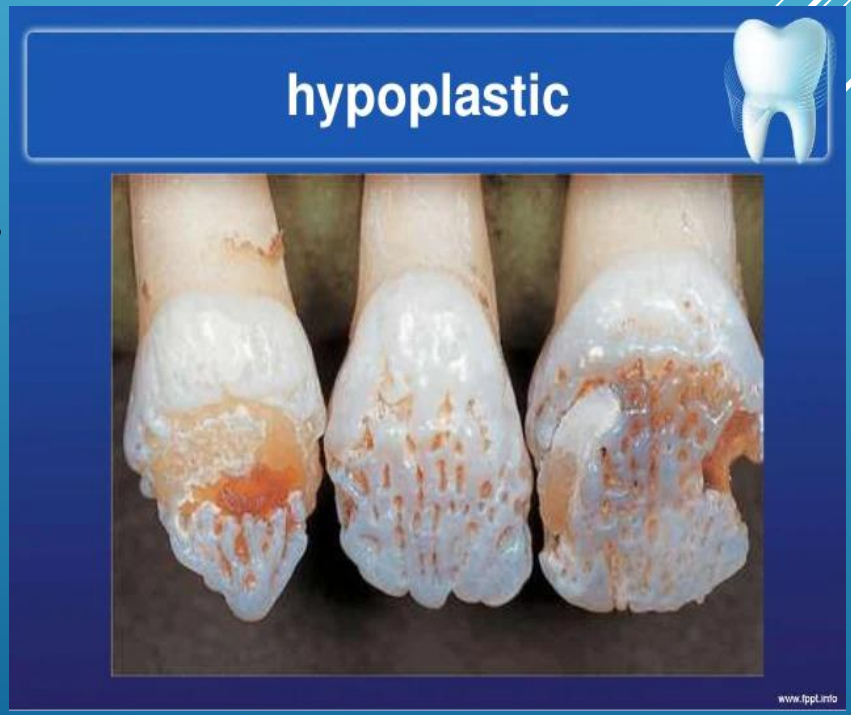




1- Hypoplastic *AI*

Which exhibits **decreased enamel matrix formation** caused by interference in the function of ameloblasts. Teeth erupt with **insufficient amounts of enamel**, ranging from pits and grooves in one patient to complete absence (aplasia) in another. Because of reduced enamel thickness in some cases, abnormal contour and absent interproximal contact points may be evident.

- Enamel of reduced thickness due to a defect in the formation of normal matrix.
- Pitting and grooves.
- Hard and translucent enamel.
- Radiographically, the enamel contrasts normally from dentine.



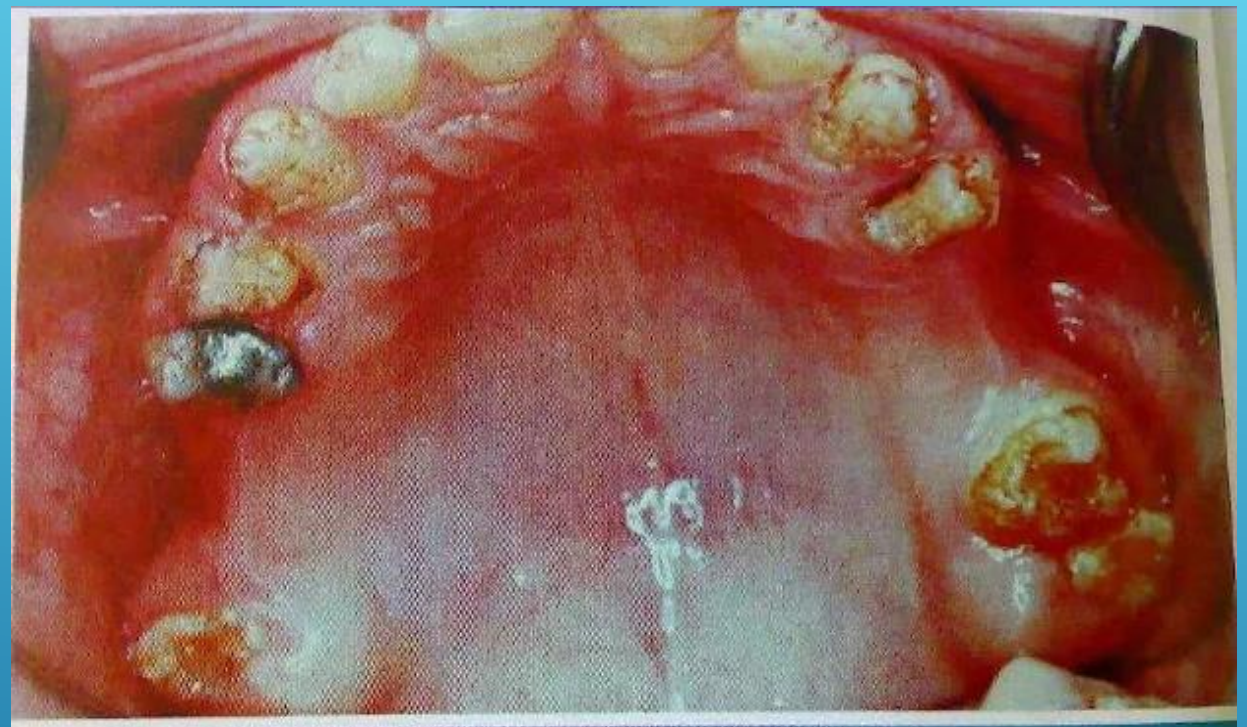


Figure 2-90 • **Hypoplastic amelogenesis imperfecta, generalized pitted pattern.** Same patient as depicted in Figure 2-89. Note diffuse involvement of all maxillary teeth, which is inconsistent with environmental damage. (Courtesy of Dr. Joseph S.

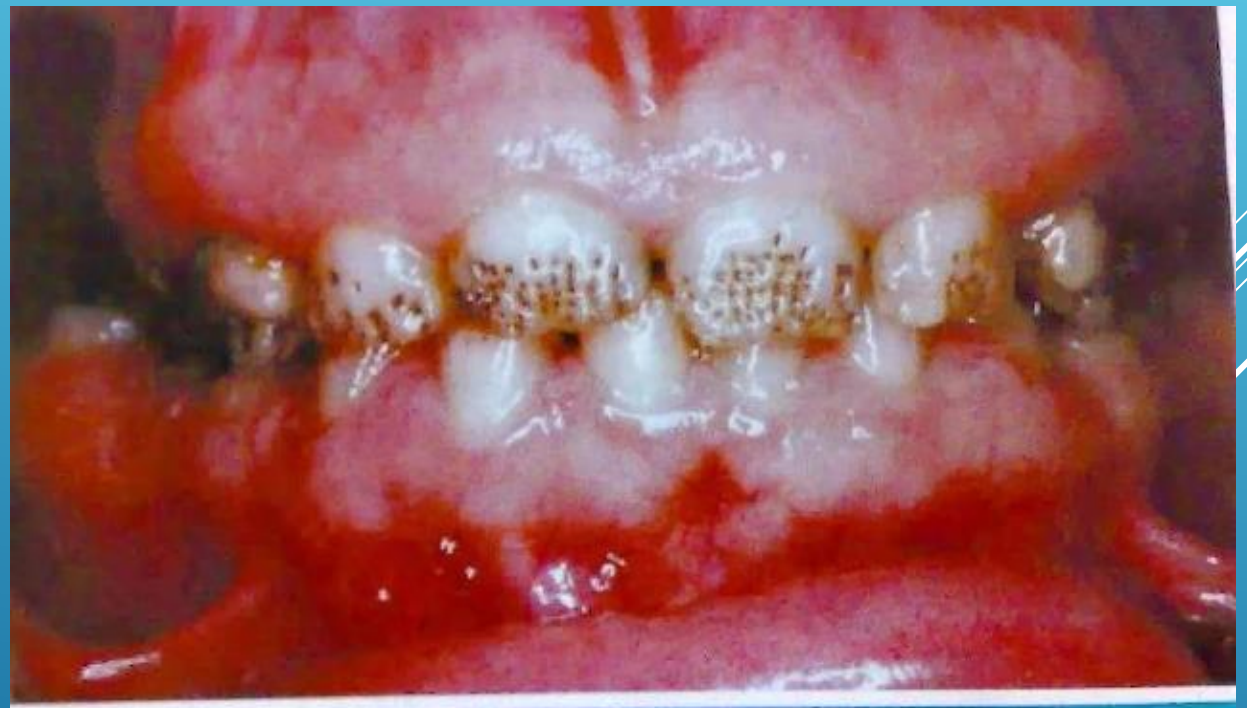
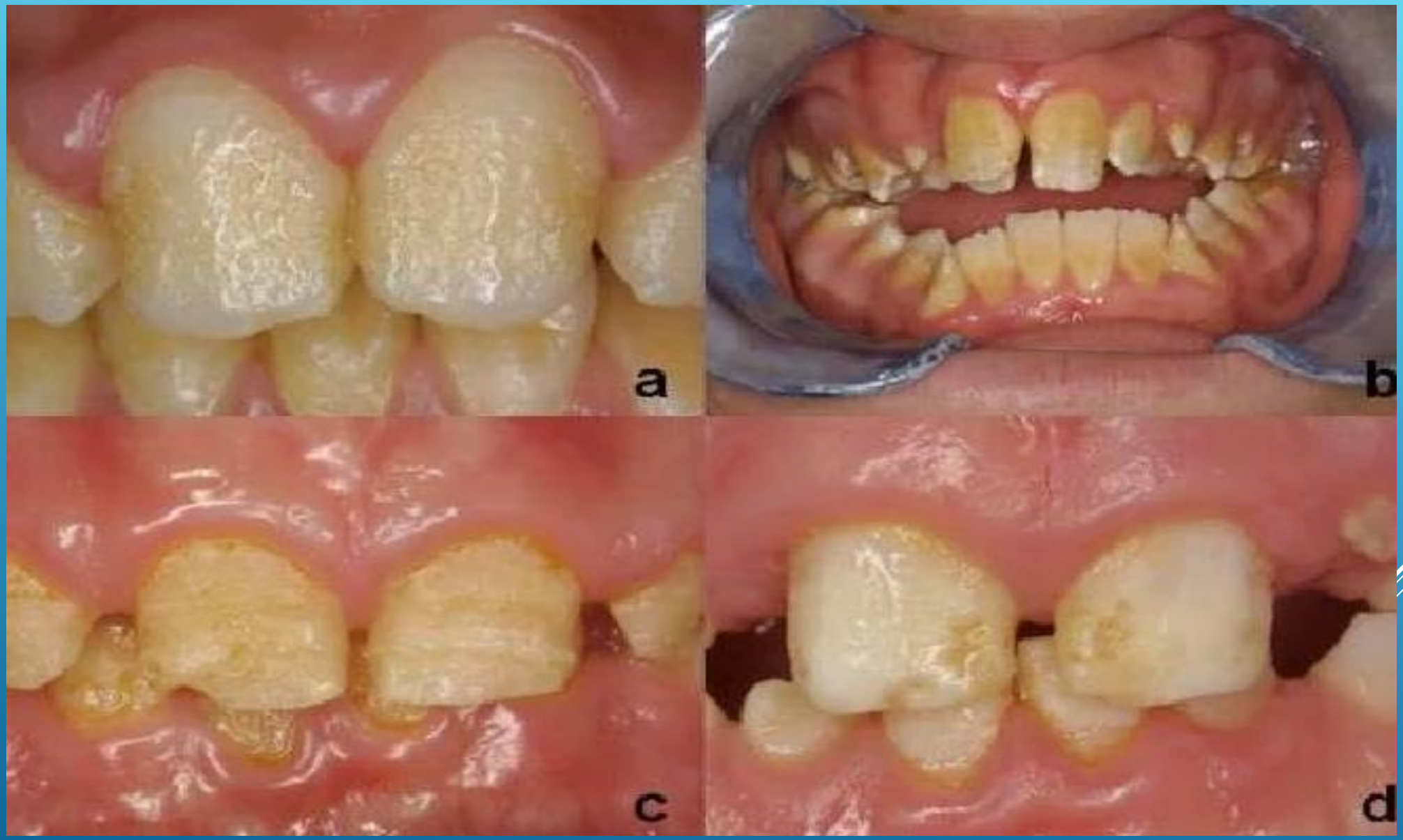


Figure 2-89 • **Hypoplastic amelogenesis imperfecta, generalized pitted pattern.** Note the numerous pinpoint pits scattered across the surface of the teeth. The enamel between the pits is o normal thickness, hardness, and coloration. (From Stewart RE,

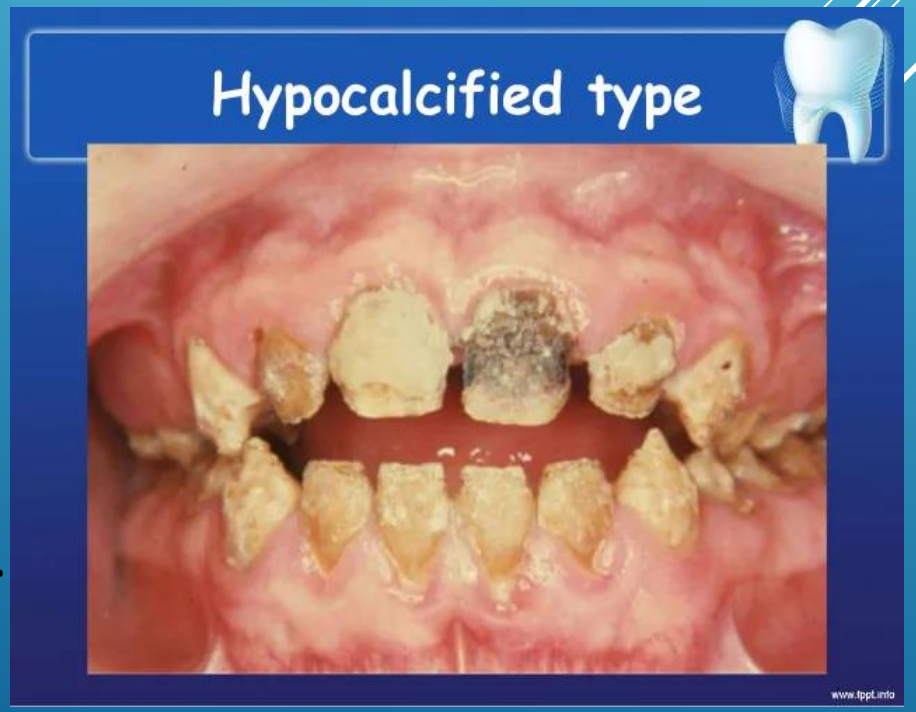

a
b
c
d

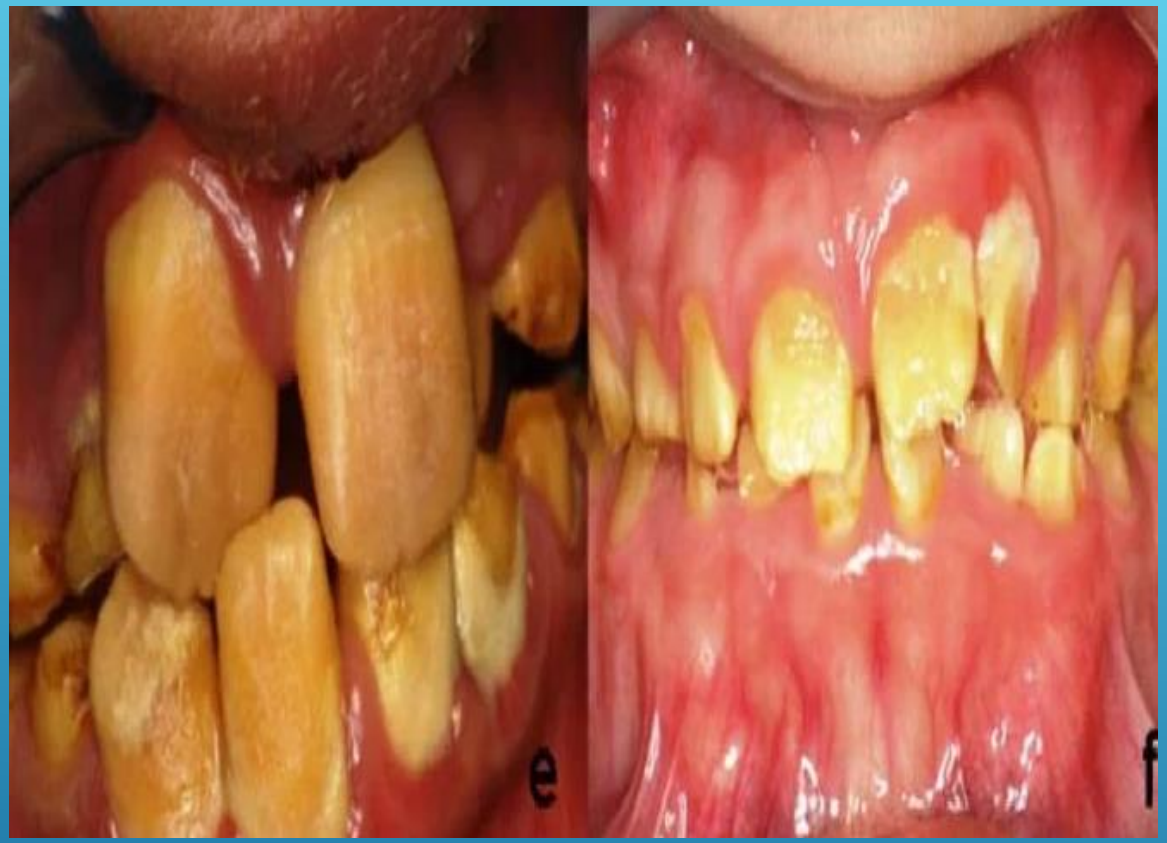
2- Hypocalcified *AI*

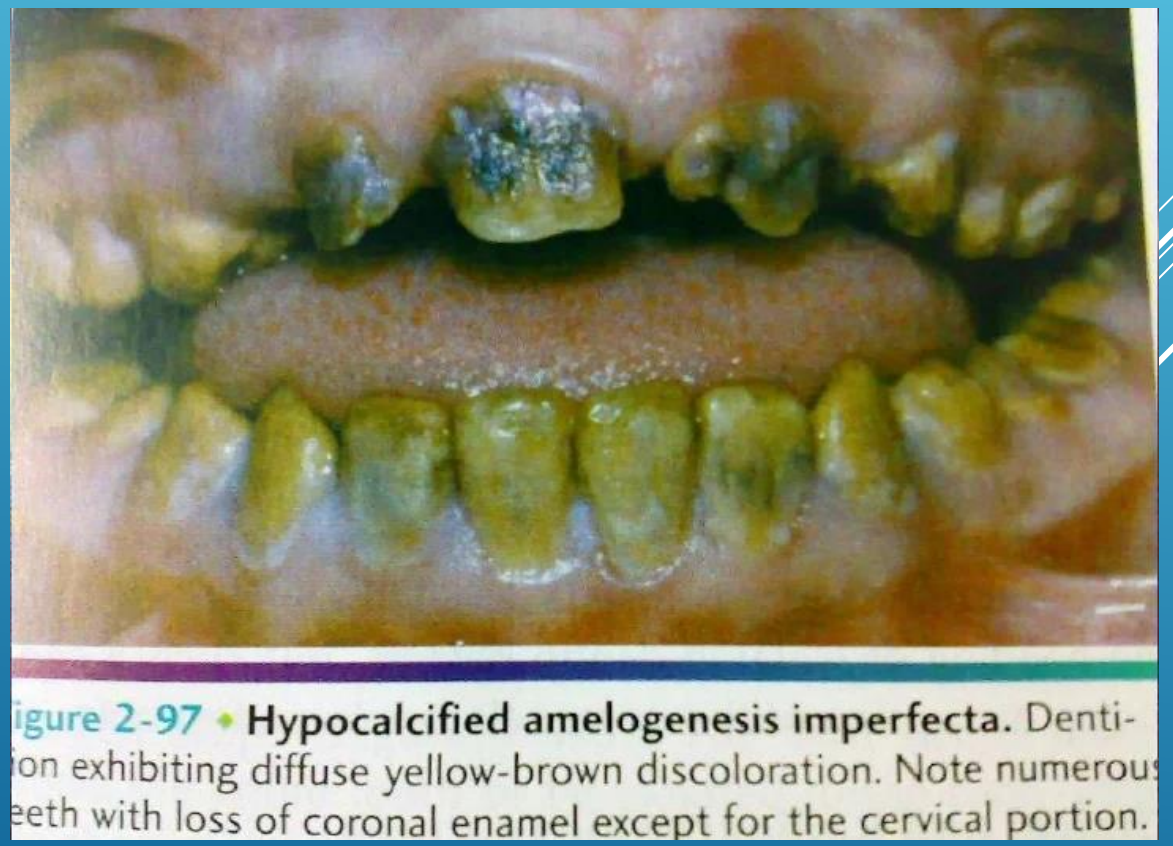
Which exhibits a severely defective form of **mineralization of the enamel matrix.**

The quantity of **enamel is normal**, but it is **soft and friable**, so that it fractures and wears readily. The color of the teeth varies from tooth to tooth and from patient to patient—from white opaque to yellow to brown. Teeth also tend to darken with age as a result of exogenous staining.

- Defect in enamel calcification
- Enamel of normal thickness
- Weak in structure
- Appears opaque or chalky
- Teeth become stained and rapidly wear down
- Radiographically, enamel is less radio-opaque than dentine.





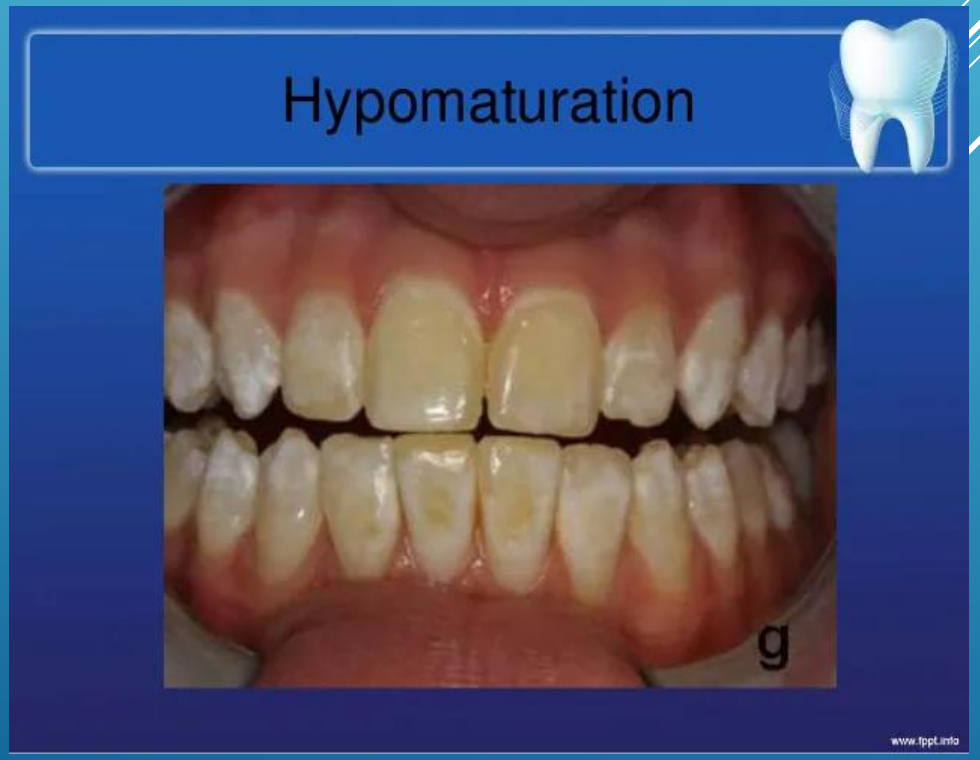


igure 2-97 • **Hypocalcified amelogenesis imperfecta.** Denti-
ion exhibiting diffuse yellow-brown discoloration. Note numerous
eeth with loss of coronal enamel except for the cervical portion.

3- Hypomaturation *AI*

Which exhibits **less** severe **mineralization** with focal or generalized areas of **immature enamel crystallites**.

- Enamel of normal thickness but mottled in appearance
- Slightly softer than normal and vulnerable to tooth wear, but not as severe as the hypocalcified type
- Radiographically, similar radiodensity as dentine.



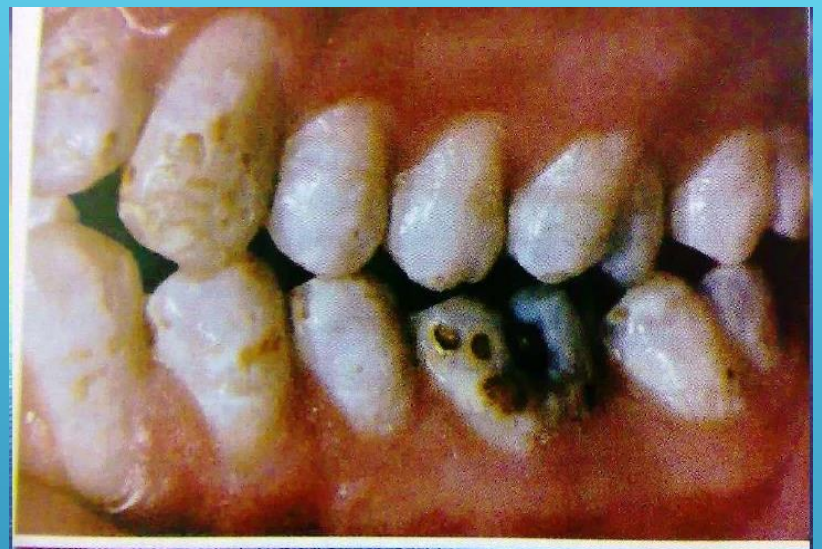


Figure 2-95 • **Hypomaturation amelogenesis imperfecta.** Dentition exhibiting mottled, opaque white enamel with scattered areas of brown discoloration.

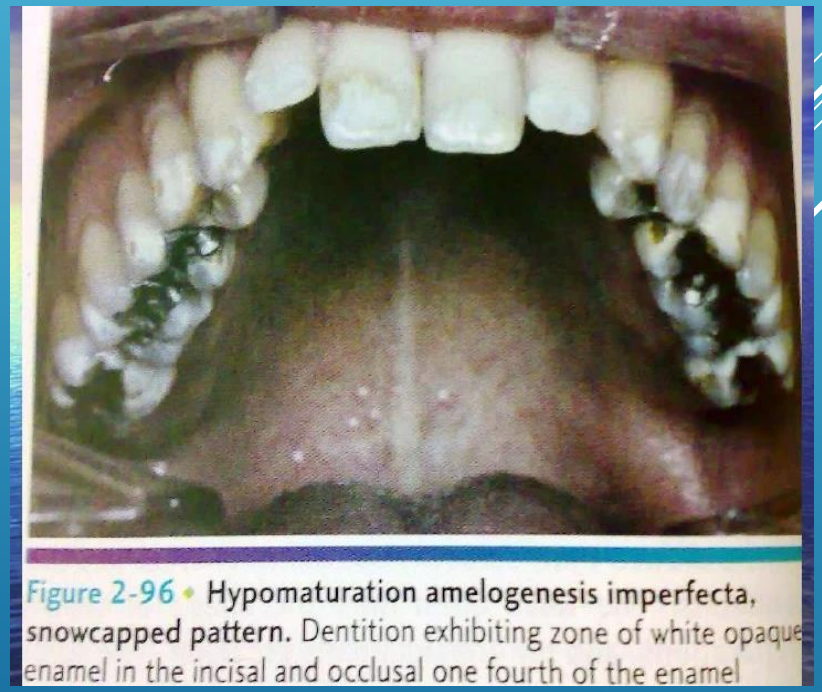


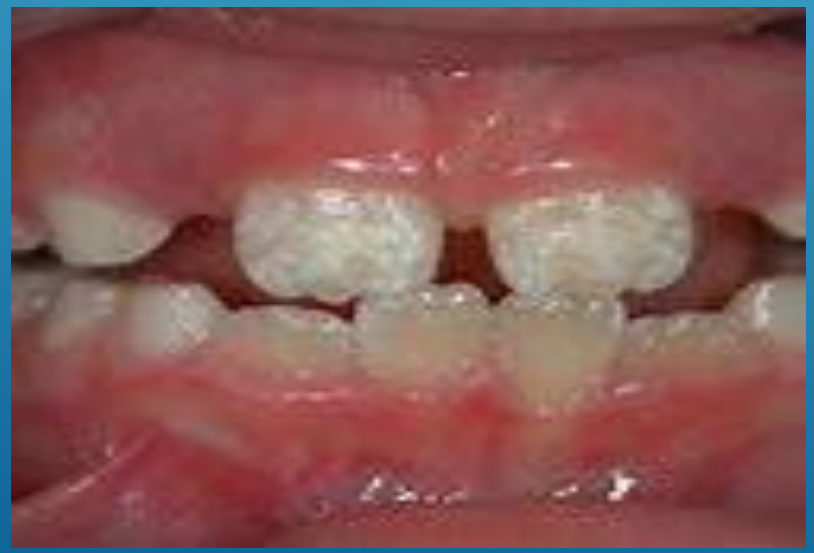
Figure 2-96 • **Hypomaturation amelogenesis imperfecta, snowcapped pattern.** Dentition exhibiting zone of white opaque enamel in the incisal and occlusal one fourth of the enamel

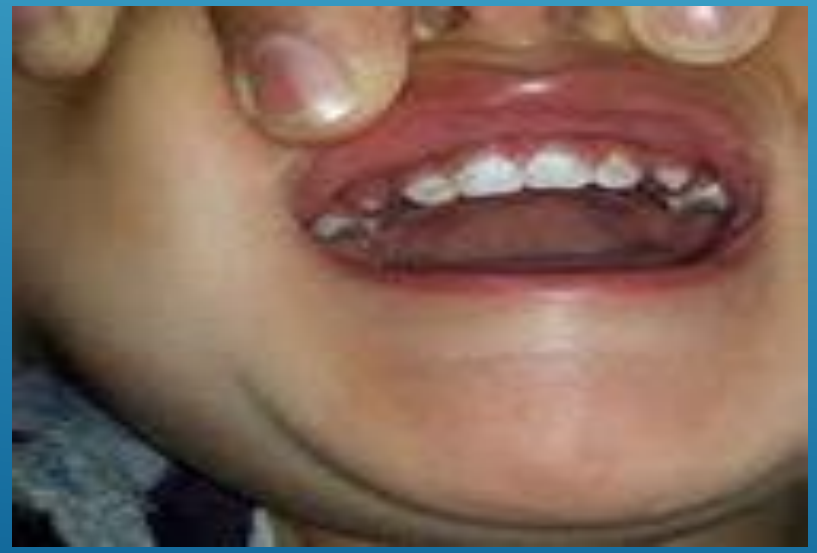
Enamel hypoplasia (EH)

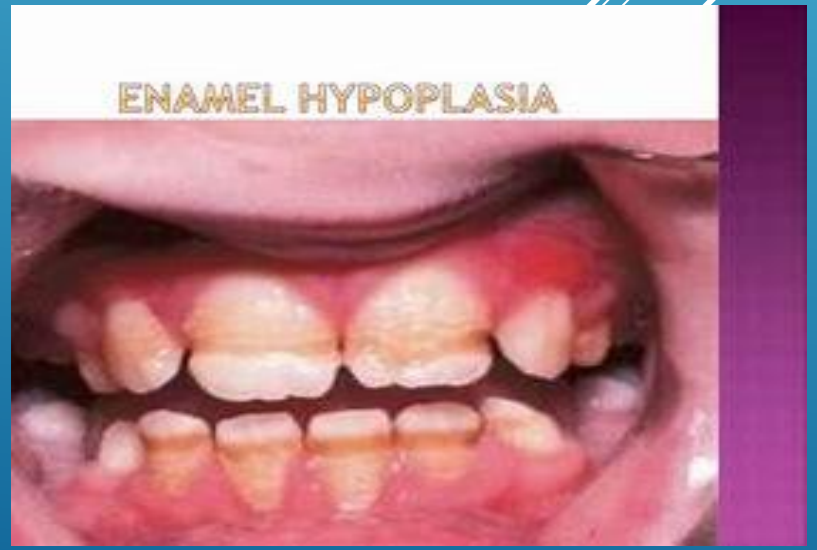
Enamel hypoplasia is a term that denotes incomplete or underdeveloped tooth enamel.

EH is a developmental defect that results in **inadequate enamel**.

It can affect both baby teeth and permanent teeth. In severe cases, no enamel forms on the teeth, and in standard cases, the tooth enamel is thin and weak. ***EH*** thus, is a surface defect of the tooth crown that is caused by a disturbance of enamel matrix secretion, defective calcification or defective maturation.

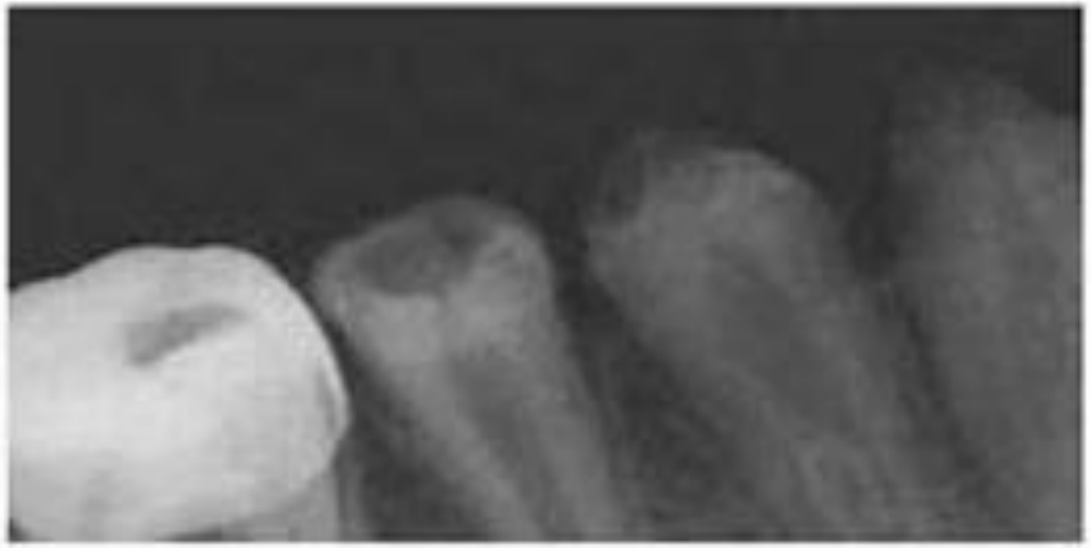




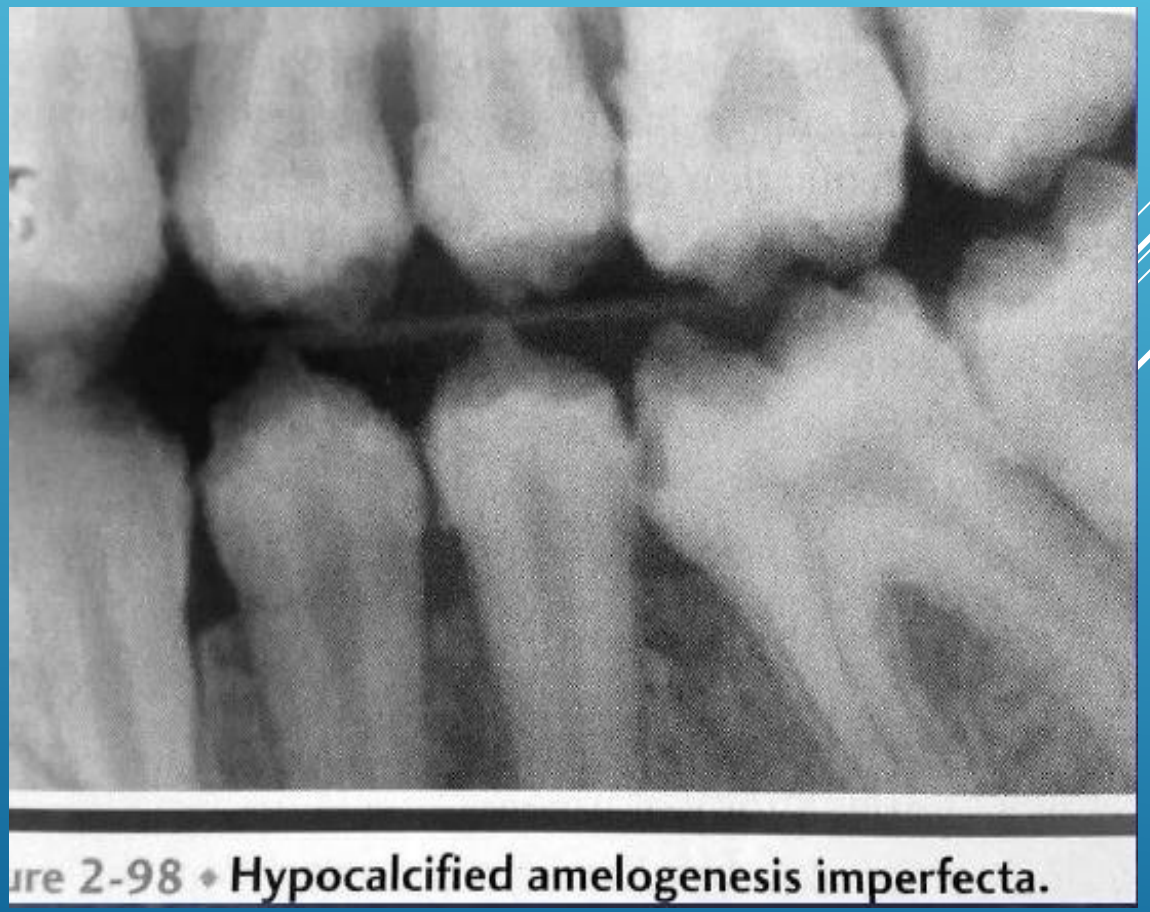


Radiographic Features

- The enamel may appear totally absent
- When present may appear as a thin layer , chiefly over the tips of the cusps & on the interproximal surfaces.
- In some cases calcification is so much affected that enamel & dentin seem to have the same radio density, making differentiation between the two difficult.



Amelogenesis imperfecta. This is a radiographic view of amelogenesis imperfecta showing the altered thickness and shape of the enamel crown that is characteristic of this disease. In this patient the condition was inherited as an autosomal dominant trait..

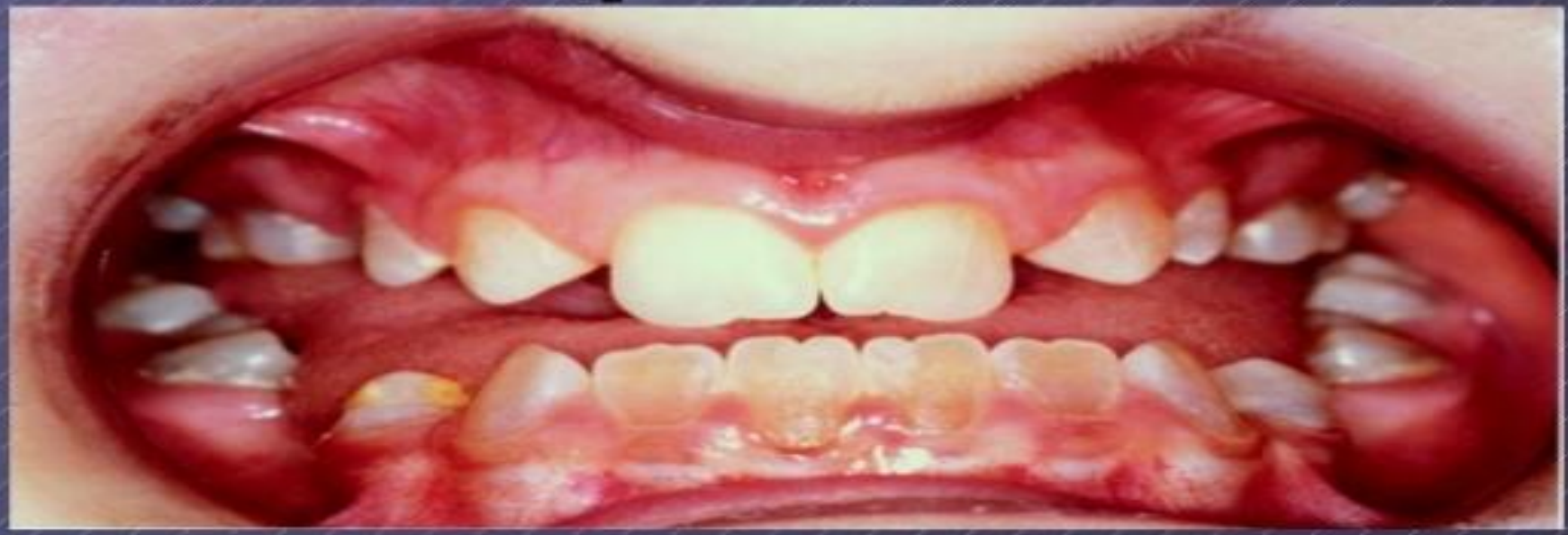


ure 2-98 • Hypocalcified amelogenesis imperfecta.

Histological Features

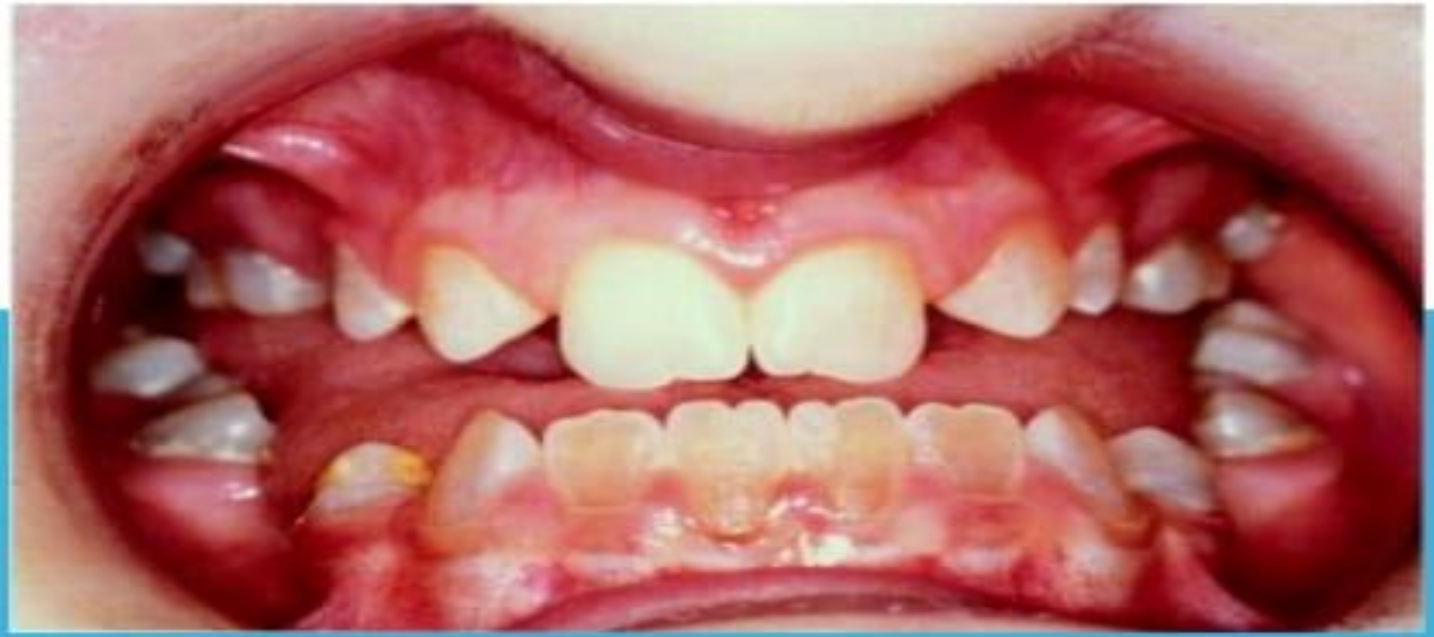
- Hypoplastic type—disturbance in the differentiation or viability of ameloblasts.
- Hypocalcification type– defects of matrix structure and of mineral deposition.
- Hypomaturation type– alteration in enamel rod & rod sheath structures.

Dentinogenesis imperfecta

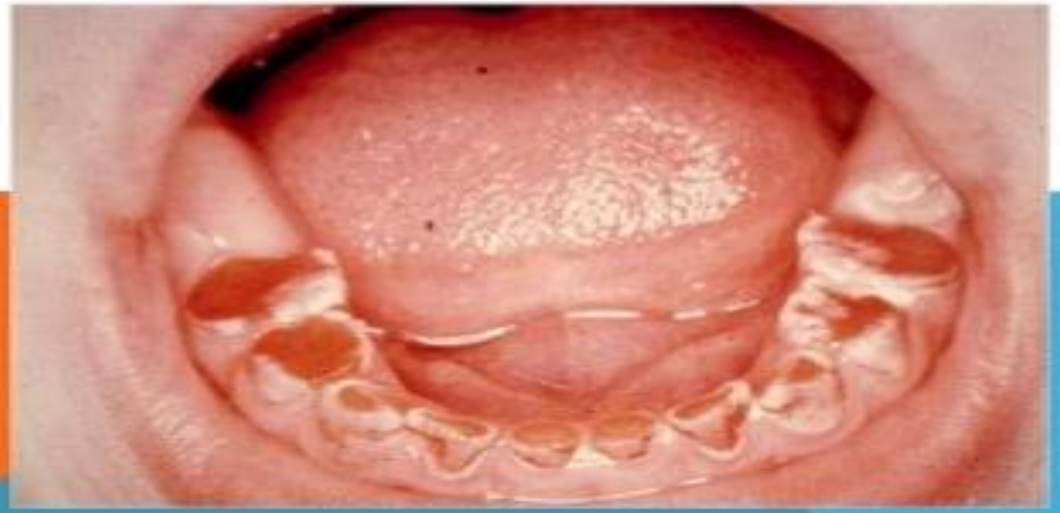


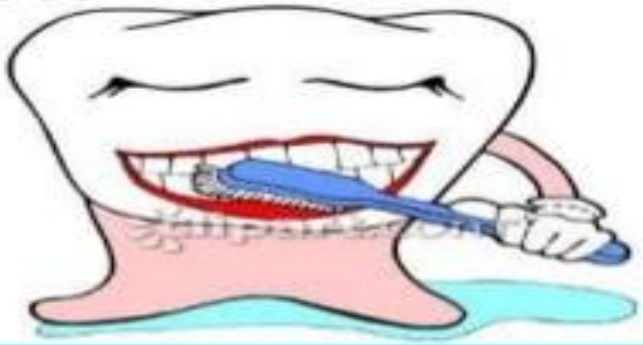
DENTINOGENESIS IMPERFECTA

- is a *genetic disorder of tooth development.*
- This condition causes teeth to be *discolored* (most often a blue-gray or yellow-brown color) and translucent.
- Teeth are also *weaker* than normal, making them prone to rapid wear, breakage, and loss.
- These problems can affect both **primary teeth** and **permanent teeth**.



- This condition is inherited in an *autosomal dominant pattern*, which means one copy of the altered gene in each cell is sufficient to cause the disorder.
- Dentinogenesis imperfecta affects an estimated **1 in 6,000 to 8,000 people.**
- Dentinogenesis imperfecta represents a group of hereditary conditions that are characterized by **abnormal dentin formation.**
- These conditions are genetically and clinically heterogenous and can affect only the teeth or can be associated with the condition osteogenesis imperfecta.





DENTINOGENESIS IMPERFECTA

DENTINOGENESIS IMPERFECTA

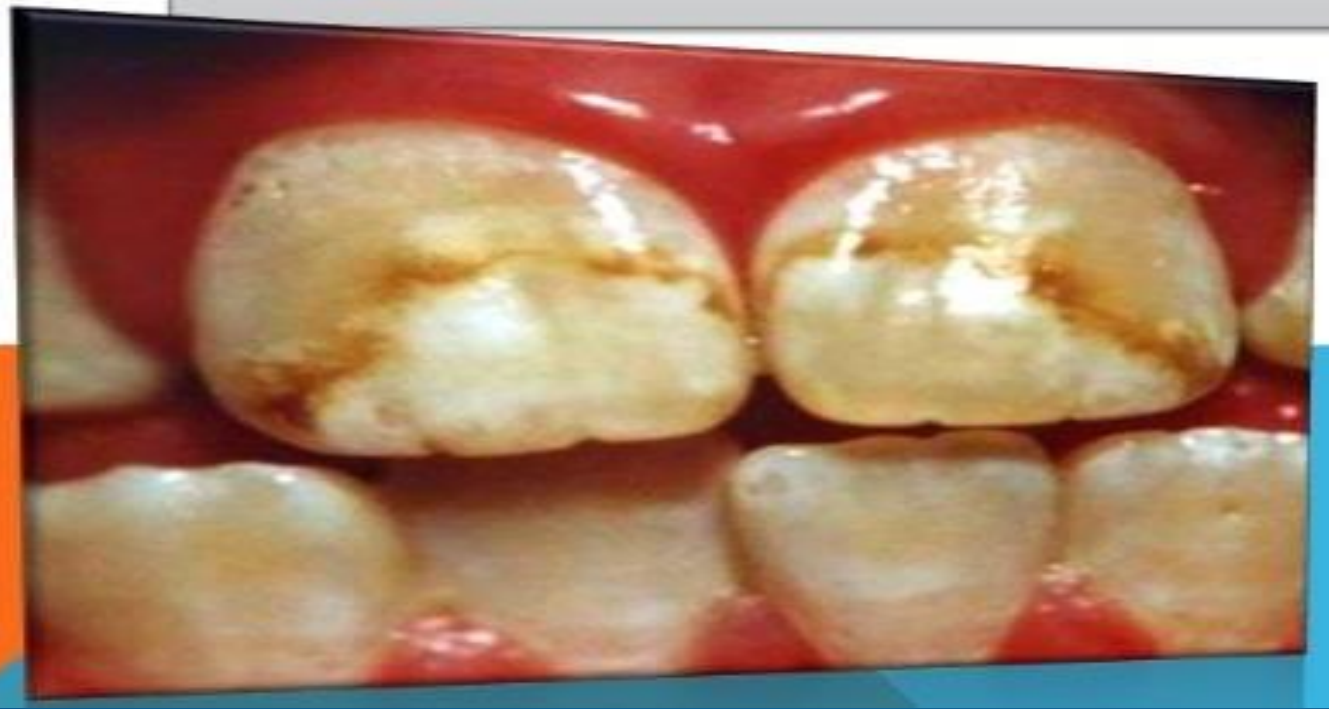
TYPE 1

Type of dentinogenesis imperfecta with **similar dental formalities** usually an *autosomal dominant trait* with variable expressivity but can be *recessive* if the associated **teogenesis imperfecta** is ecessive type.

TYPE 2

Occurs in people *without* other inherited disorders (i.e. **Osteogenesis imperfecta**).

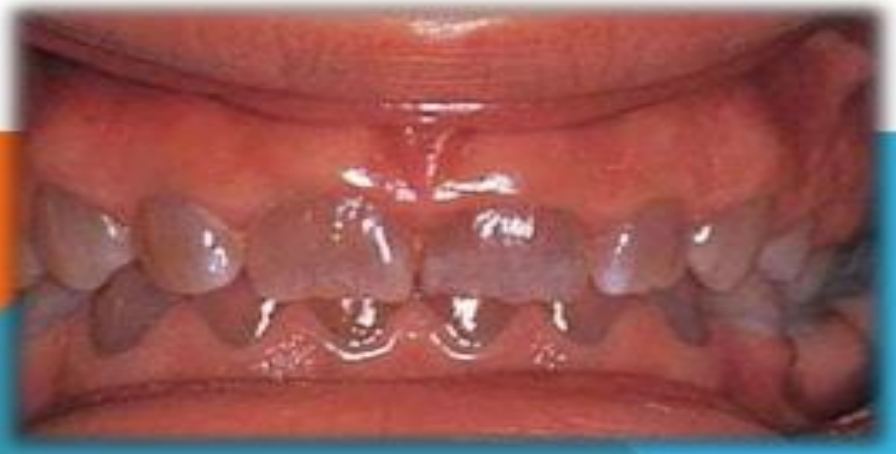
It is an autosomal dominant trait. A few families with type II have progressive **hearing loss** in addition to dental abnormalities.



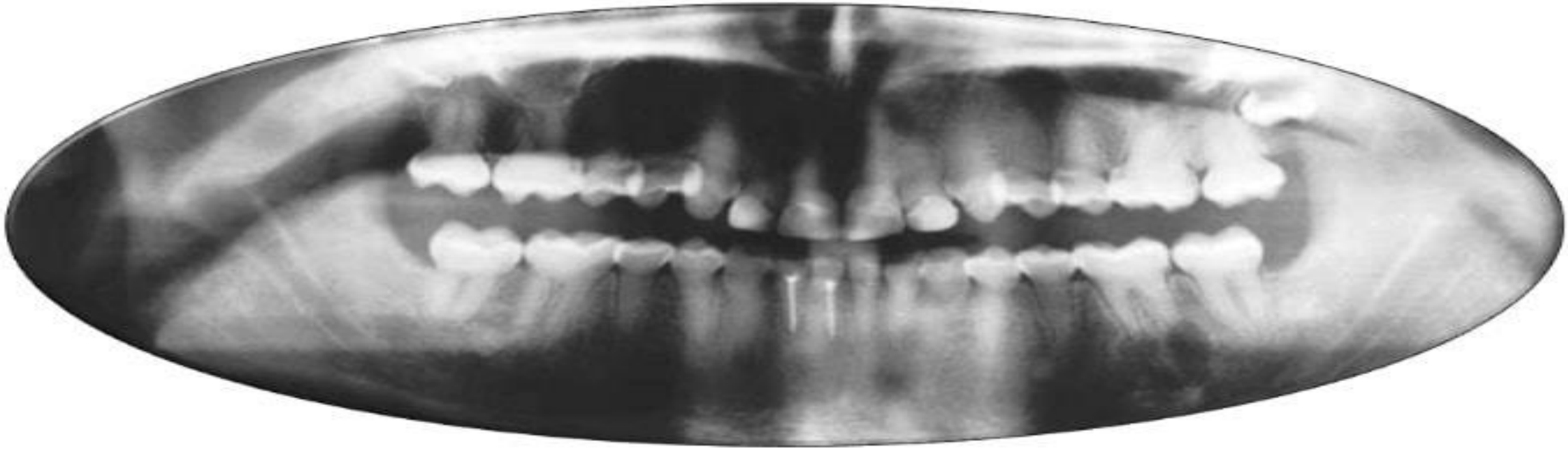
CLINICAL FEATURES

- Clinical appearance is variable.
- The teeth usually involved and more severely affected are deciduous teeth in type 1; whereas in type 2 both the dentitions are equally affected.
- The teeth may be gray to yellowish brown. They exhibit translucent or opalescent hue.
- Enamel is usually lost early due to loss of scalloping at the DEJ.

*However, the teeth are not more susceptible to dental caries than normal ones.



RADIOGRAPHIC FEATURES



- Type I and II show total **obliteration of the pulp chamber.**
- Type III shows thin dentin and extremely enormous pulp chamber. These teeth are usually known as **Shell**

HISTOLOGY

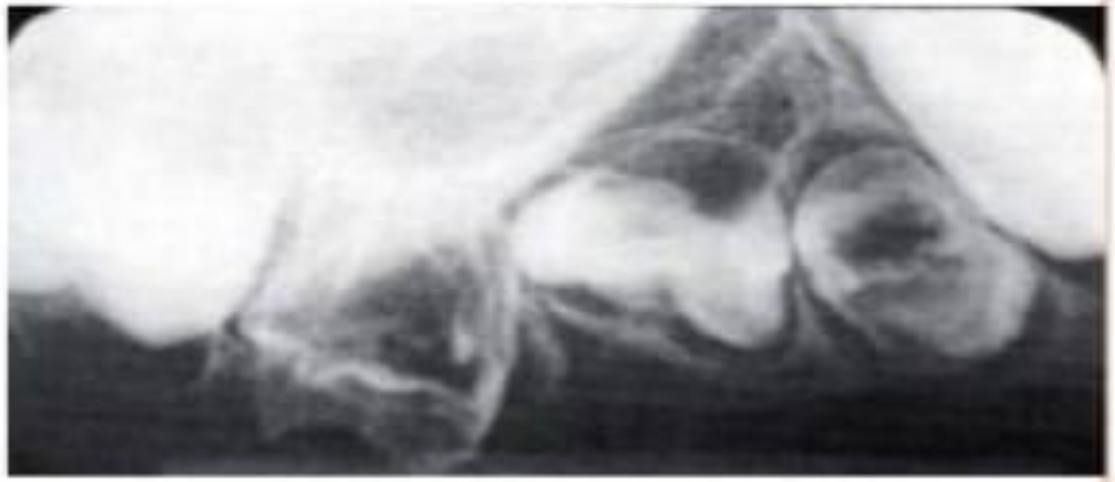
- **Dentinal tubules** are *irregular* and are *bigger* in diameter.
- Areas of **uncalcified matrix** are seen.
- Sometimes **odontoblasts** are seen in dentin.

Dentinogenesis Imperfecta

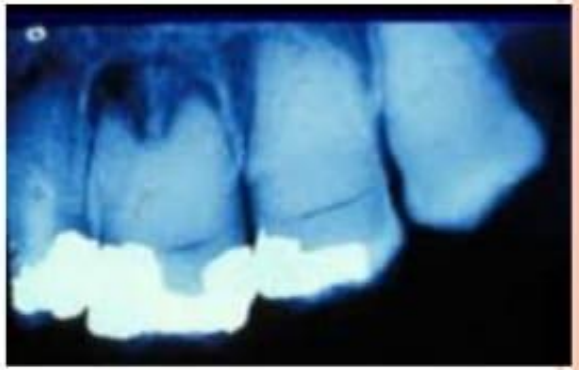
- Treatment:
  - ✓ prevent loss of enamel + subsequent loss of dentin through attrition
  - ✓ cast metal crowns on posterior
  - ✓ jacket crowns on anterior teeth

Dentin Dysplasia

- also known as "Rootless Teeth"
- rare disturbance of dentin formation
- normal enamel
- atypical dentin formation
- abnormal pulpal morphology
- hereditary disease

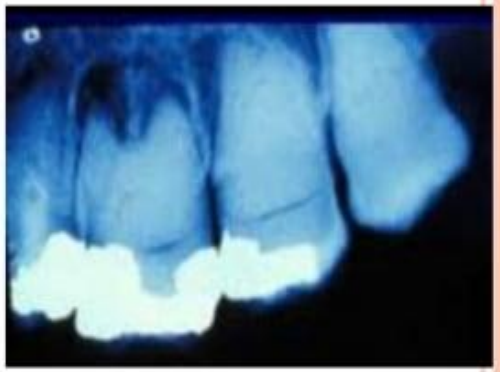


Type I (Radicular Type)



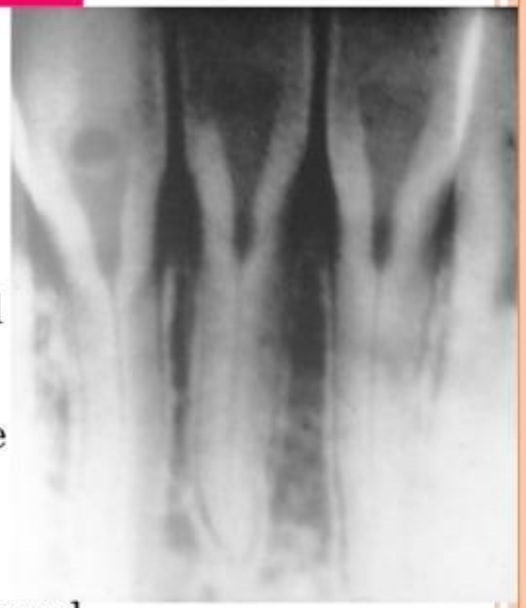
- both dentitions are of normal color
- periapical lesion
- premature tooth loss may occur because of short roots or periapical inflammatory lesions

Type I (Radicular Type)



- Radiographically:
  - ✓ roots are extremely short
  - ✓ pulps almost completely obliterated
  - ✓ periapical radiolucencies:
    - granulomas
    - cysts
    - chronic abscesses

Type II (Coronal Type)



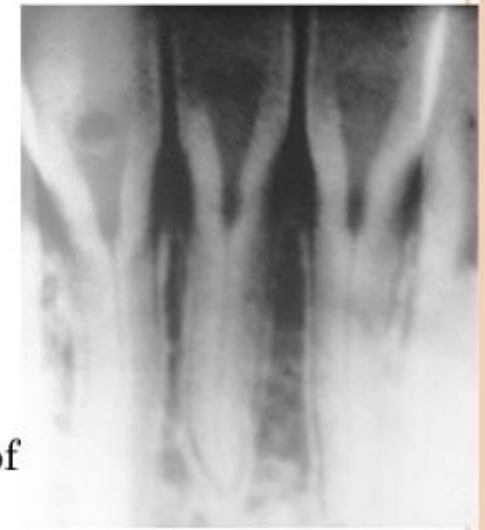
- color of primary dentition is opalescent
- permanent dentition is normal
- coronal pulps are usually large (thistle tube appearance)
  - ✓ filled with globules of abnormal dentin

Type II (Coronal Type)

- Radiographically:

(Deciduous)
- ✓ roots are extremely short
- ✓ pulps almost completely obliterated

(Permanent)
- ✓ abnormally large pulp chambers in coronal portion of tooth



3- Regional Odontodysplasia: (Ghost Teeth)

Regional odontodysplasia is a dental abnormality that involves the hard tissues derived from both epithelial (enamel) and mesenchymal (dentin and cementum) components of the tooth-forming apparatus.

The teeth in a region or quadrant of the maxilla or mandible are affected to the extent that they exhibit **short roots, open apical foramina**, **and enlarged pulp chambers**.

The thinness and **poor mineralization** quality of the enamel and dentin layers have given rise to the term ***ghost teeth***.

Regional Odontodysplasia

- one or several teeth in a localized area are affected
- maxillary teeth are involved more frequently than mandibular area
- etiology is unknown

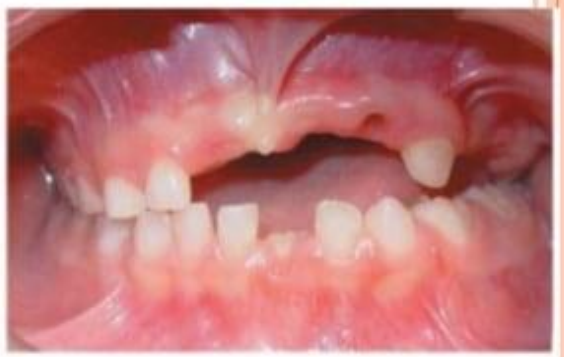
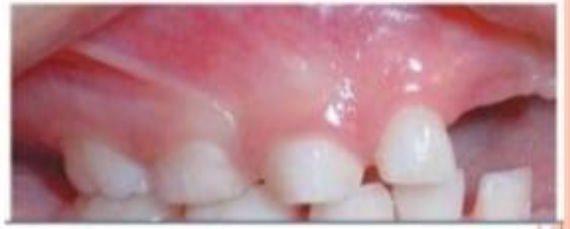


FIGURE 2



Regional Odontodysplasia

- teeth affected may exhibit a delay or total failure in eruption
- shape is altered, irregular in appearance

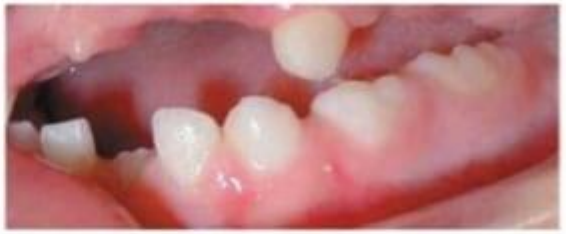
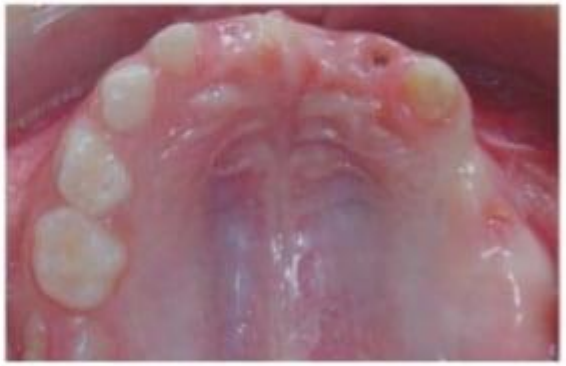


FIGURE 4



Regional Odontodysplasia

- Radiographically:
  - ✓ marked reduction in radiodensity
  - ✓ teeth assume a "ghost" appearance
  - ✓ both enamel + dentin appear very thin
  - ✓ pulp chamber is exceedingly large

Regional Odontodysplasia

- Treatment:
  - ✓ poor cosmetic appearance of teeth
  - ✓ extraction with restoration by prosthetic appliance

REGIONAL ODONTODYSPLASIA

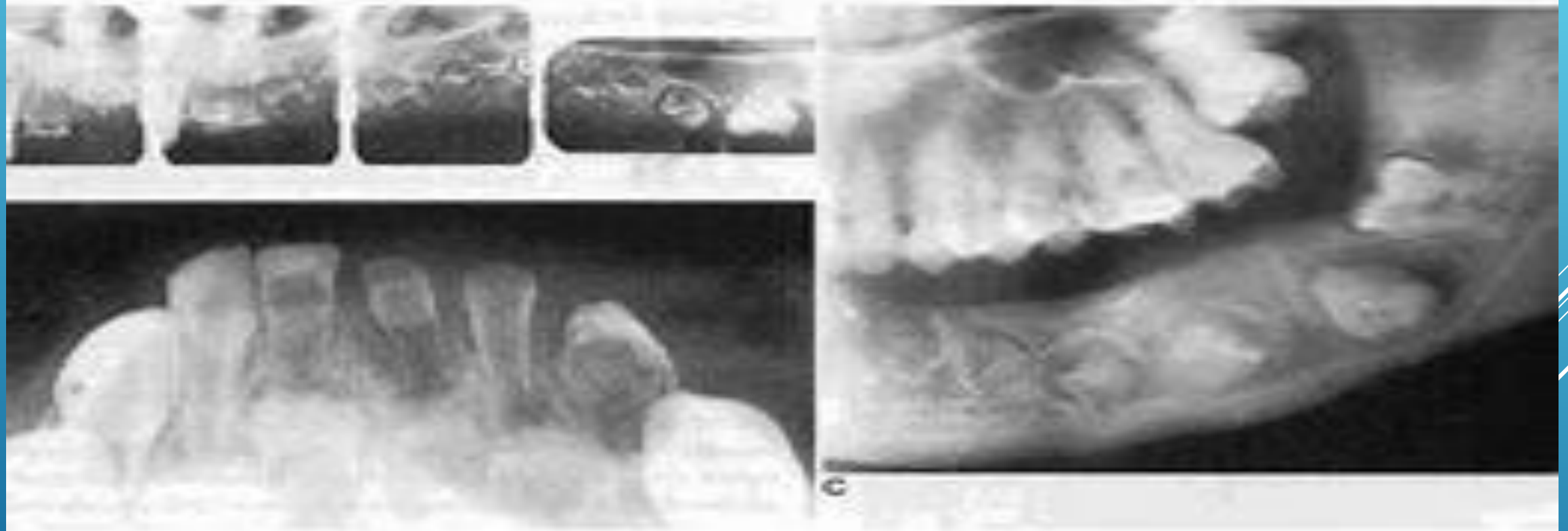


FIG. 18-10 Regional odontodysplasia revealing poor mineralization of both enamel and dentin. A, Involvement of the left maxillary dentition. B, Involvement of the primary incisors and cuspids. C, Involvement of the left mandibular cuspids and first and second molars; note the lack of eruption and hypoplasia of enamel and dentin expressed mainly as short roots.

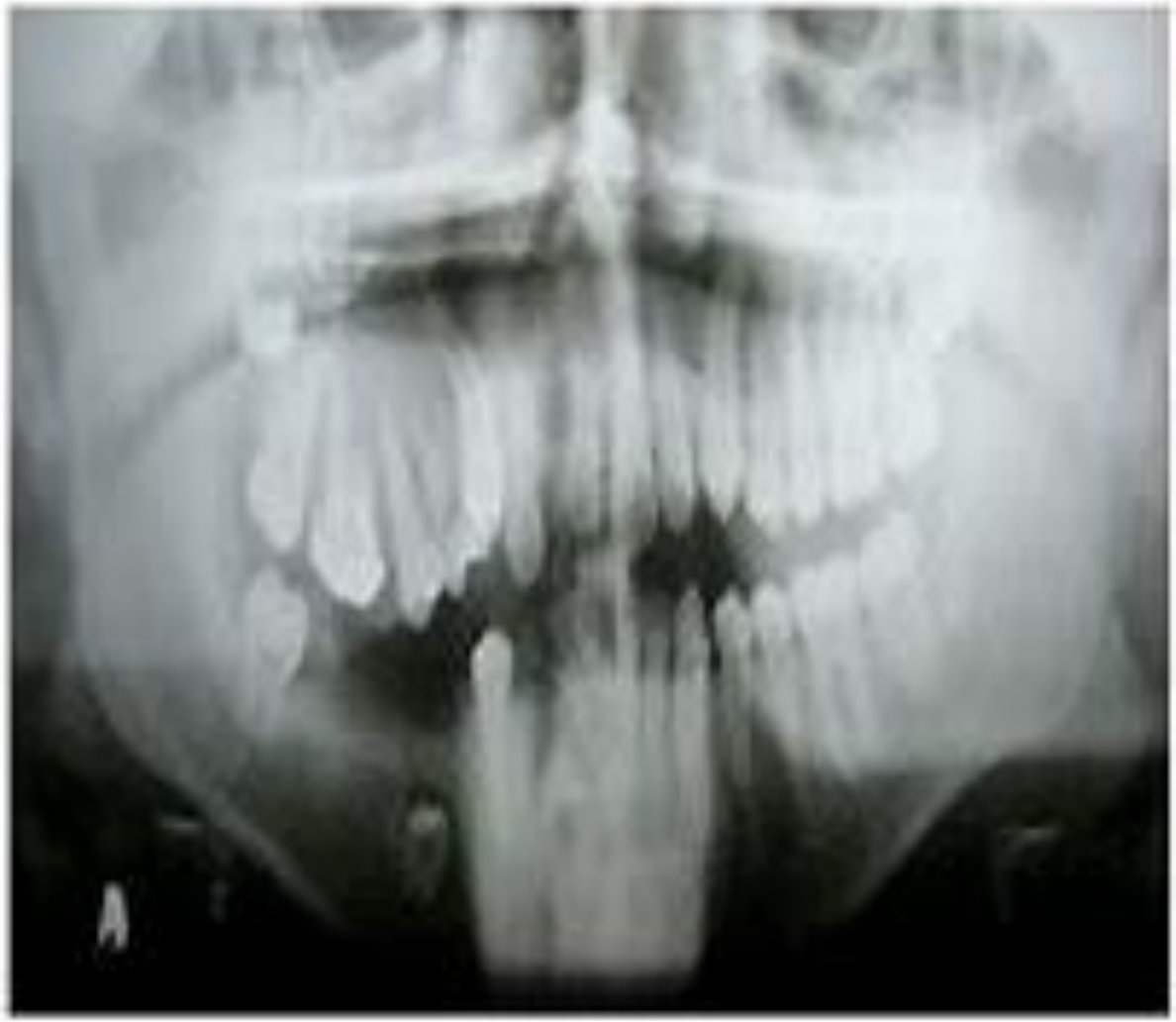


Fig. 3. Panoramic radiograph showing a dysplastic maxillary central incisor and teeth with a "ghostlike" appearance in the mandibular right quadrant.

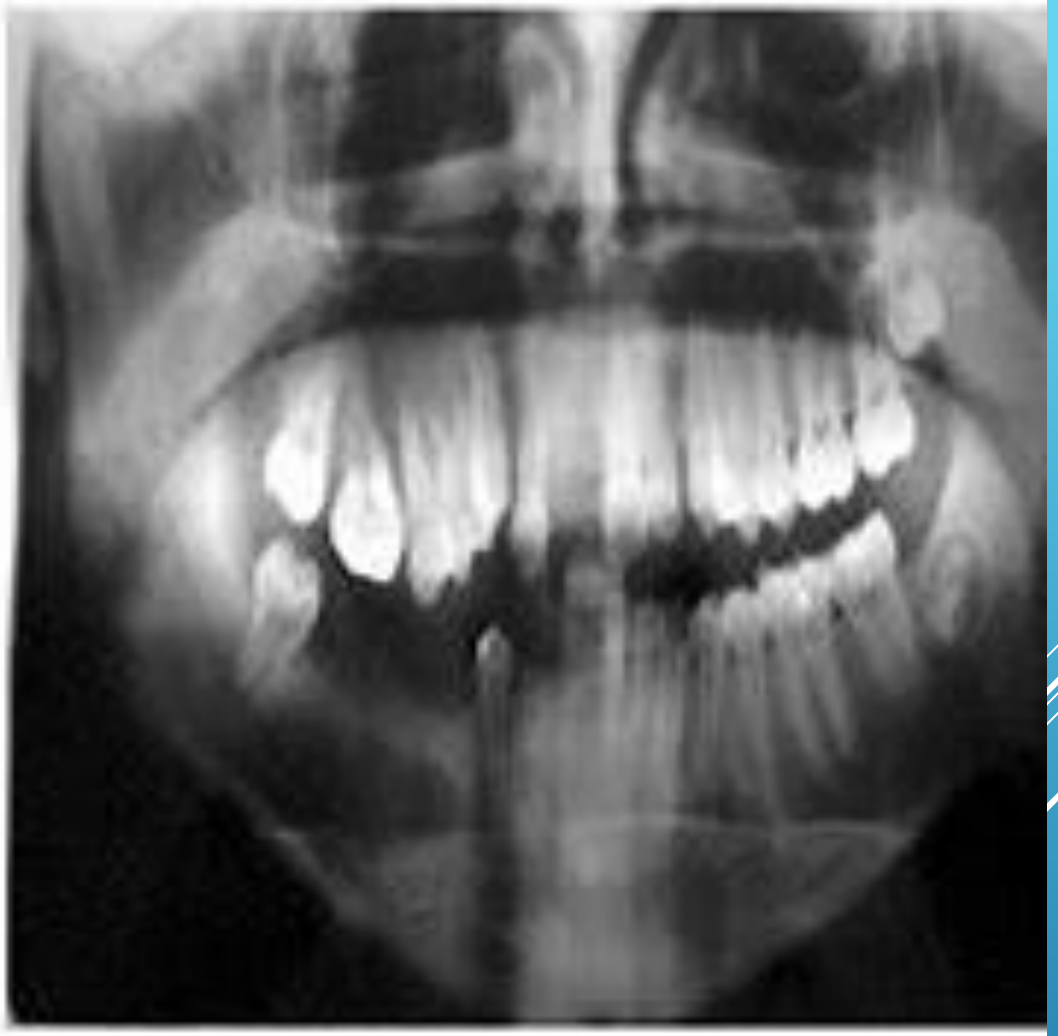


Fig. 6. Panoramic radiograph after initial treatment.

Shell Tooth

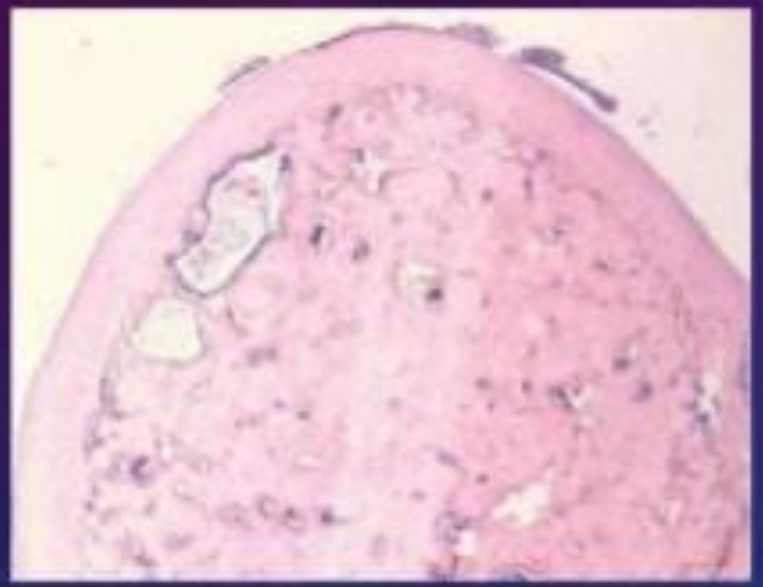
- normal thickness enamel
- extremely thin dentin
- enlarged pulps
- thin dentin may involve entire tooth or be isolated to the root
- most frequently in deciduous

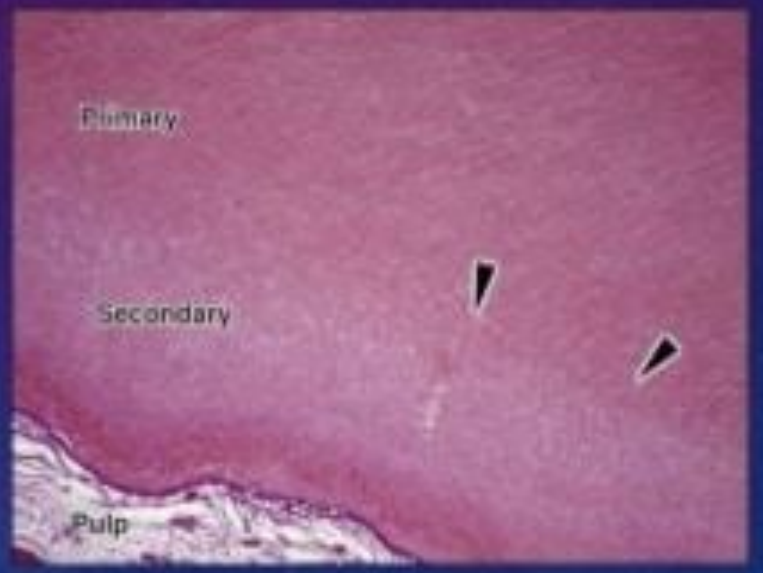
Histopathologically

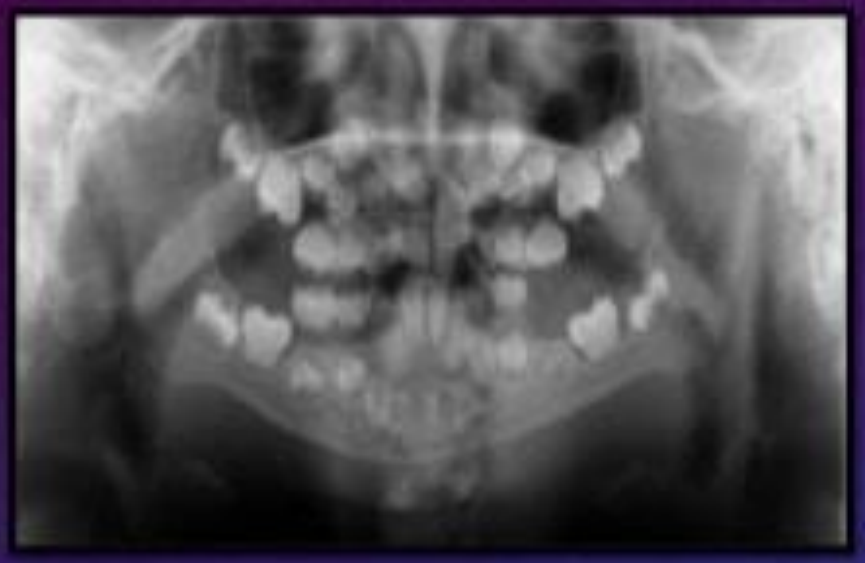
The enamel and the immediately subjacent dentin appear normal.

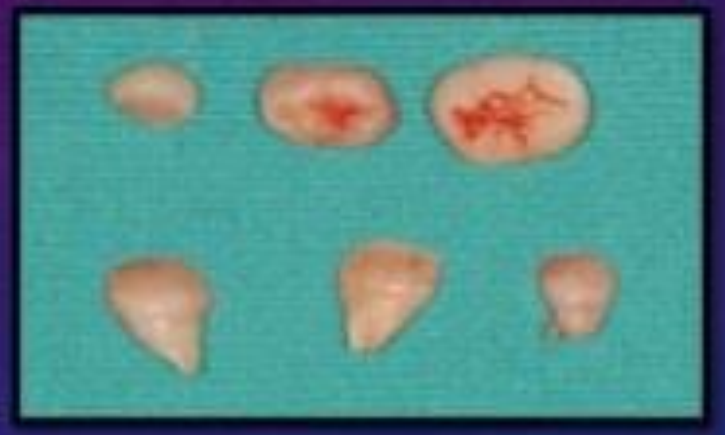
Deeper layers of dentin show an atypical tubular pattern with an amorphous, atubular area, and irregular organization.

Normal dentinal tubule formation appears to have been blocked so that new dentine forms around obstacles and takes on the characteristic appearances described as "lava flowing around boulders"




Primary
Secondary
Pulp





The radiograph revealed features of dentine dysplasia type I with normal appearance of crown but no root development

Environmental Discoloration of Teeth

Exogenous Stains

located on the *surface of the tooth*,(enamel)

Stains on the surfaces of teeth that can be removed with polishing are known as ***exogenous* or *extrinsic stains***.

Extrinsic dental stains are caused by predisposing factors and other factors such as poor oral

hygiene as dental plaque and calculus, foods, tobacco, chromogenic bacteria, and topical medications.

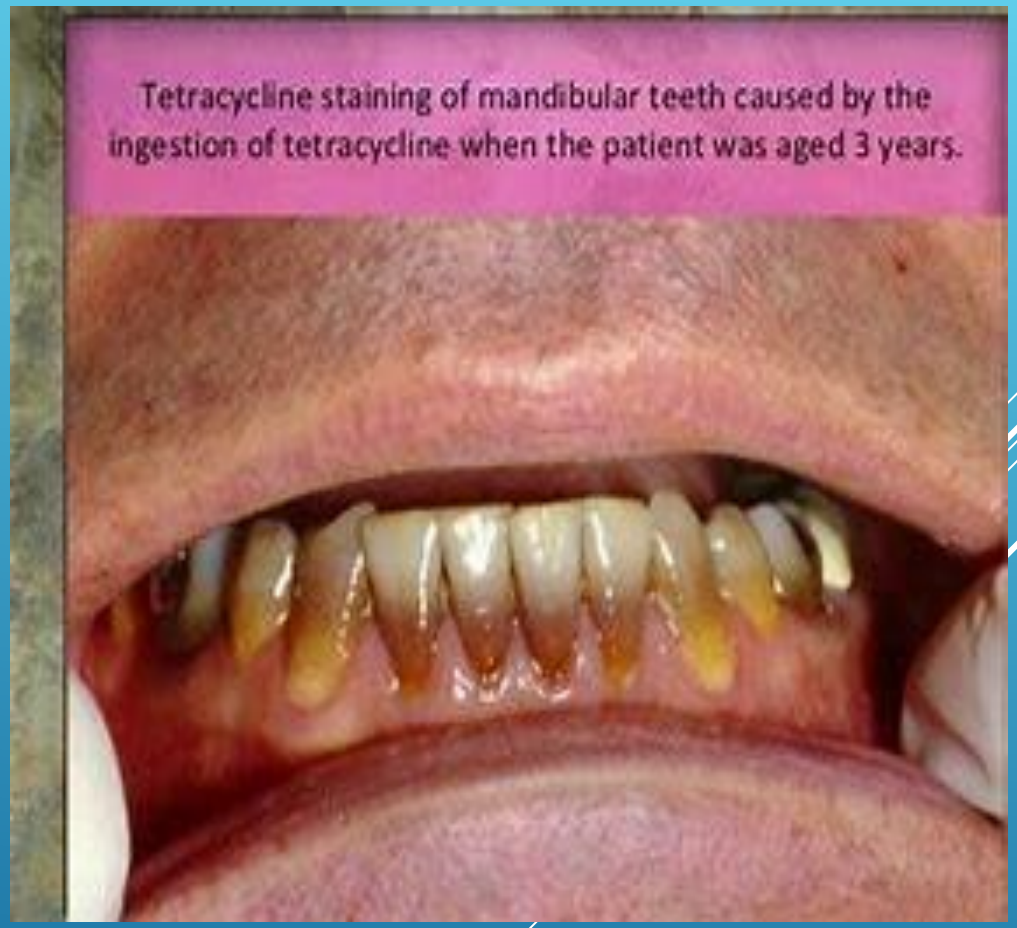
Extrinsic stains can present as green, orange, brown, yellow, or black.

Green and orange stains are typically found in patients with poor hygiene when certain chromogenic bacteria are present.

Endogenous Stains

Discoloration of teeth resulting from deposits of systemically circulating substances during tooth development is defined as ***endogenous* or *intrinsic staining*** that cannot removed by scaling and polishing, Systemic ingestion of ***tetracycline*** during tooth development is a well-known cause of endogenous staining of teeth. Tetracycline binds calcium and therefore is deposited in developing teeth and bones. The bright yellow color of the drug is reflected in subsequently erupted teeth.

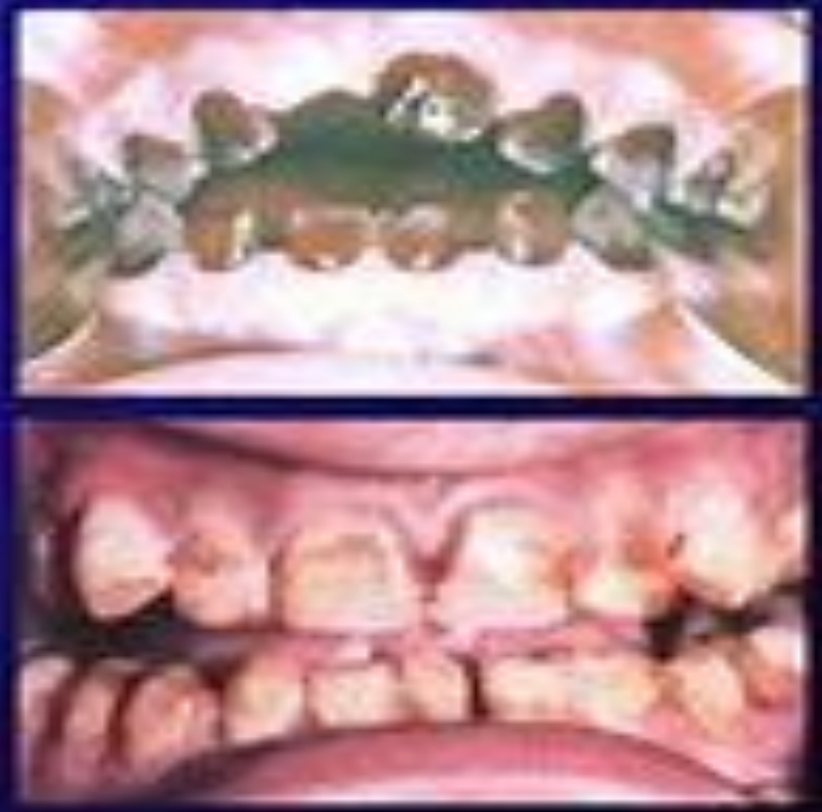
Intrinsic stains are those that affect the inner layer of the teeth.



Alterations in Color

Endogenous stain

- Congenital porphyria (hereditary)
  - Errors in porphyrin metabolism
    - Deposition of porphyrin in developing teeth which appear red to brown
- Liver disease, biliary atresia & neonatal hepatitis
  - It may cause discoloration of the primary teeth




Thank you !