DEVELOPMENTAL DISTURBANCES OF THE TEETH

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FUSION OF THE TEETH

- **Fusion** represents the union of two independently developing **primary** or **permanent teeth**.
- **Fusion leads to** the formation of **a single large tooth and reduces the total number** of visible teeth in the affected arch by one (or more).

- **Fused teeth** will have **separate pulp chambers and separate pulp canals**

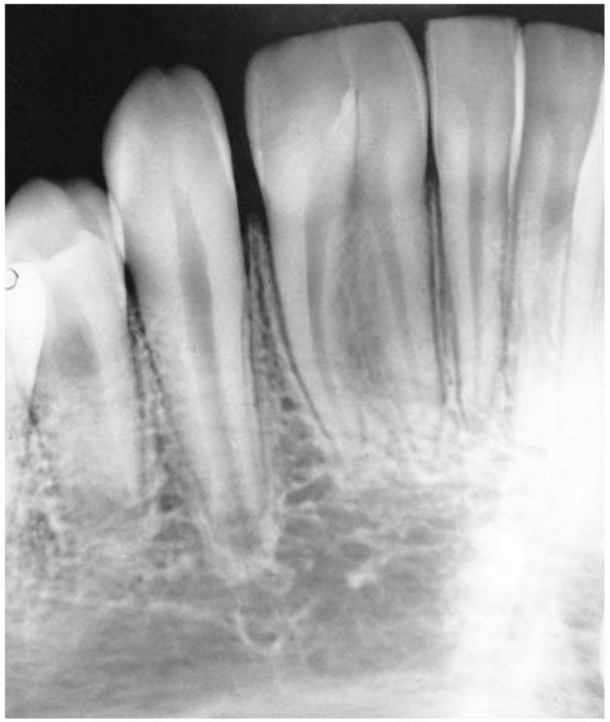


Fig. 3.5 Fusion of a permanent central and lateral incisor.

- **Dental fusion** is usually **localized** to the **anterior of the mouth**, with the **maxillary central and lateral incisors** being the most frequently affected teeth
- a hereditary pattern
- Fused teeth are at **higher risk of developing dental caries along the line of crown fusion** necessitating the **placement of a restoration**

- a **frequent finding** when **two primary teeth fuse** is the **developmental absence of one of the corresponding permanent teeth**

- patients with fused teeth often require a **multidisciplinary approach** for their dental care **involving pediatric dentistry, endodontics, surgery, restorative dentistry, and orthodontics**

- **Surgical sectioning** and **separation of fused teeth** may be possible.
- and although dentin is exposed, such teeth are easily moved orthodontically without the risk of ankylosis

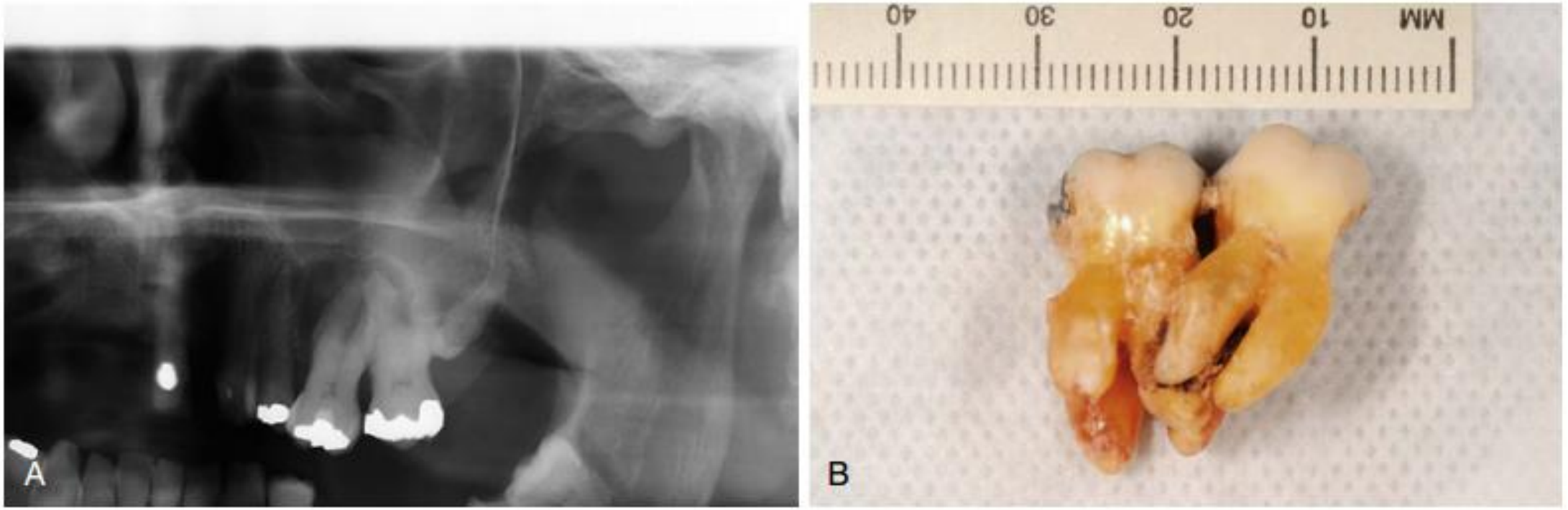


Fig. 3.6 Concrescence illustrated in (A) Cropped panoramic film left maxilla. Noted concrescent of first and second molars and (B) Concrescent left maxillary first and second molars. (Photo post extraction. *Palermo D, Davies-House A. Unusual finding of concrescence. Case Reports 2016; 2016: bcr2016214597.*)

- Fusion of teeth involving **only cementum** is termed **concrescence**

- **concrescent teeth only** coalesce **at the root level** (involving solely the **cementum**)
- this condition results in a **normal number of crowns** being visible in the arch
- The most **commonly affected teeth** are the **maxillary molars**
- with **the joining** of a **second and third molar**, or a **third molar joining with a supernumerary tooth**,
- although it has also been reported in **the mandible**
- Although concrescence **typically impacts** the development of **only two teeth,** there have been **rare cases noted of triples**.

- It has been speculated that localized **trauma**, **excessive occlusal force**, **restriction of space to grow** during dental follicle development, and/or **localized infection** during and/or after development may play a role

- **Indications that concrescence** may be **present include incomplete (or difficulty with) tooth eruption,** associated **occlusion problems leading to cheek biting** and/or the **formation of traumatic ulcers** in the region, **localized periodontal inflammation**, and **fracture of the maxillary tuberosity** and/or the **maxillary sinus floor**

GEMINATION

- the **geminated tooth** appears clinically as a **bifid crown on a single root**
- The **crown is usually wider than normal, with a shallow groove** extending from the incisal edge to the cervical region
- This dental anomaly frequently occurs in a **unilateral fashion** within the **anterior portion** of the mouth and can be seen in **both primary and permanent** dentitions;

- although it probably appears more frequently in primary teeth.

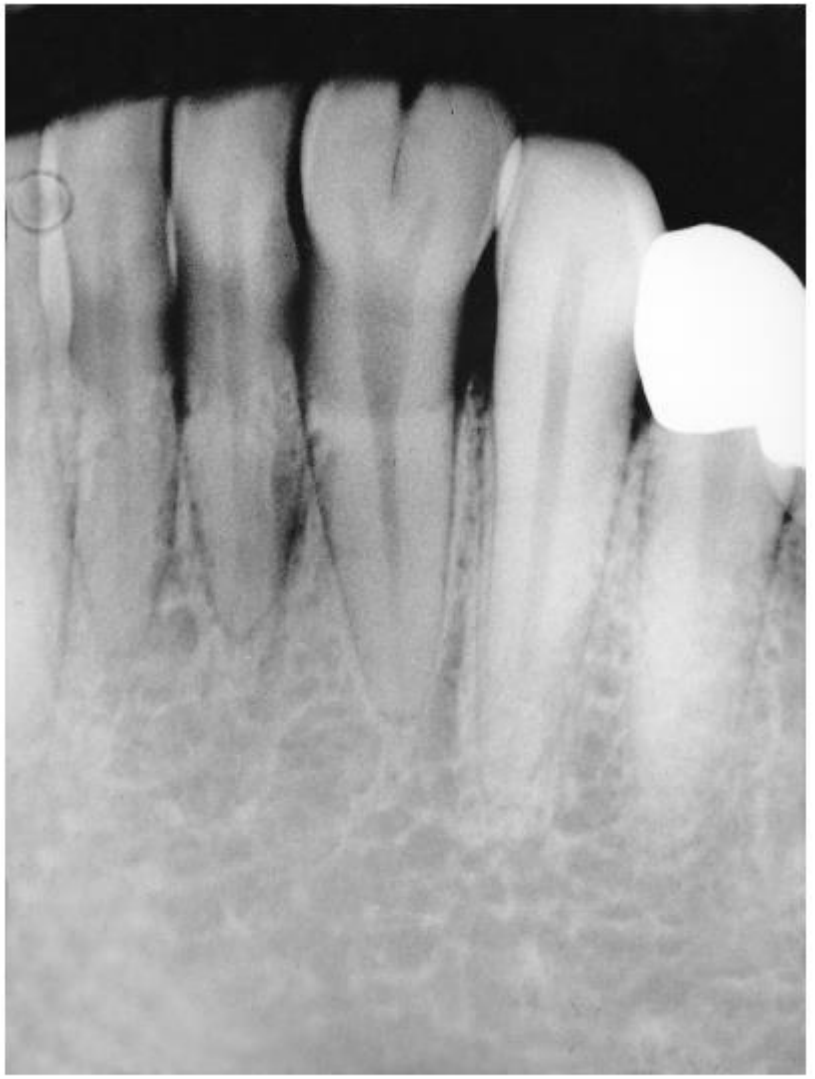


Fig. 3.7 Gemination of a mandibular lateral incisor. The crown has a groove on the labial surface and is wider than normal.

- geminations can show **a familial tendency** but are also likely to be influenced by **environmental factors**

- In contrast to gemination, "**twinning**" occurs when there is a **complete division of a single tooth germ** during the proliferation stage which leads to the development **of two separate teeth** from a single tooth bud, thereby creating an **extra tooth** within the affected arch.

- gemination many include **poor esthetics**, **dental crowding**, a heightened risk of **caries**, and **periodontal problems**

- The **treatment of a permanent anterior geminated tooth** may involve **reduction of the mesiodistal width of the tooth** to allow for **normal development of the occlusion**. **Periodic "disking"** of the tooth is recommended when the crown is not excessively large
- as is eventual preparation of the tooth for **restoration if dentin is exposed**.
- However, **if the crown is extremely large**, the tooth size may be unable to be masked, **necessitating removal** and a **combined orthodontic and prosthodontic approach** to rehabilitation.

DENS INVAGINATUS (DENS IN DENTE)

- Dens invaginatus is a developmental irregularity where the **enamel organ invaginates into the dental papilla** prior to the mineralization of the tooth.

- Dens invaginatus (or a tooth within a tooth) **can be diagnosed radiographically**
- The invagination is thus **lined with enamel** and a **foramen cecum**, with the probability of **communication or proximity between the cavity of the invagination and the pulp chamber**

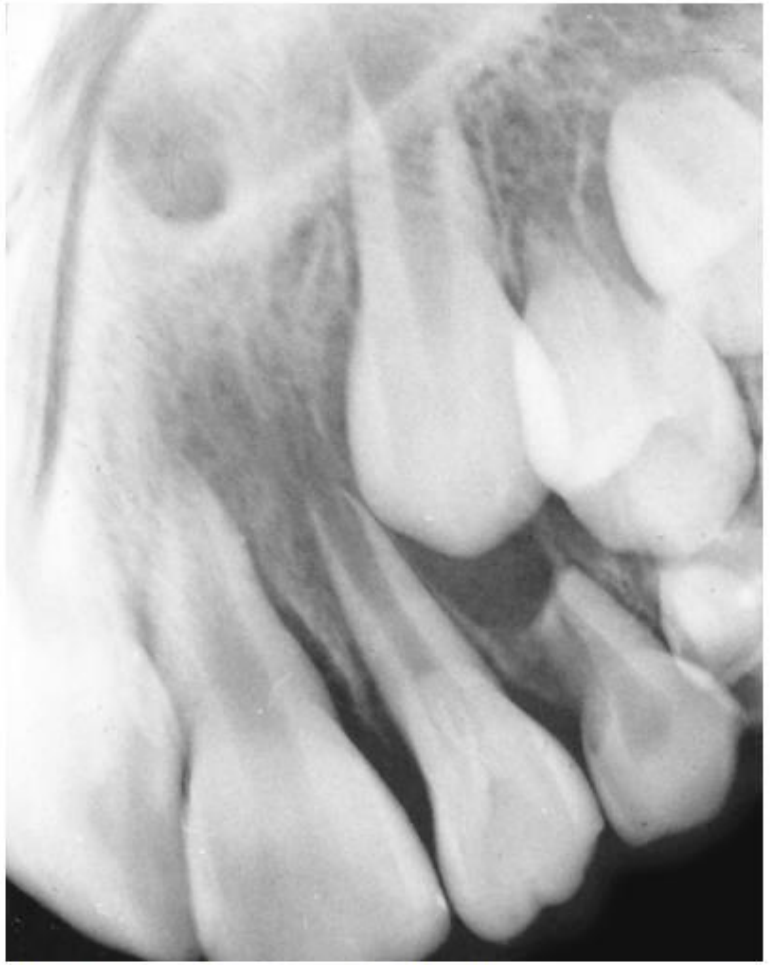


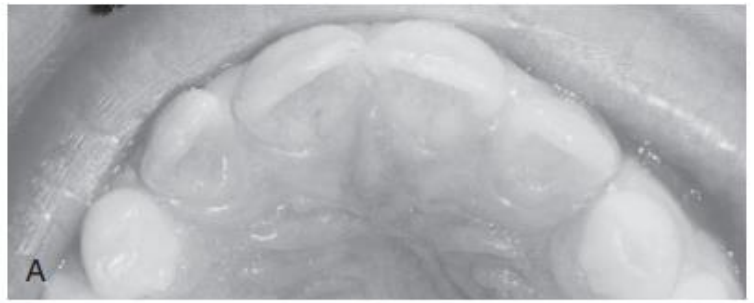
Fig. 3.9 Dens in dente in a maxillary lateral incisor. A communication between the invagination and the pulp chamber apparently caused pulpal necrosis.

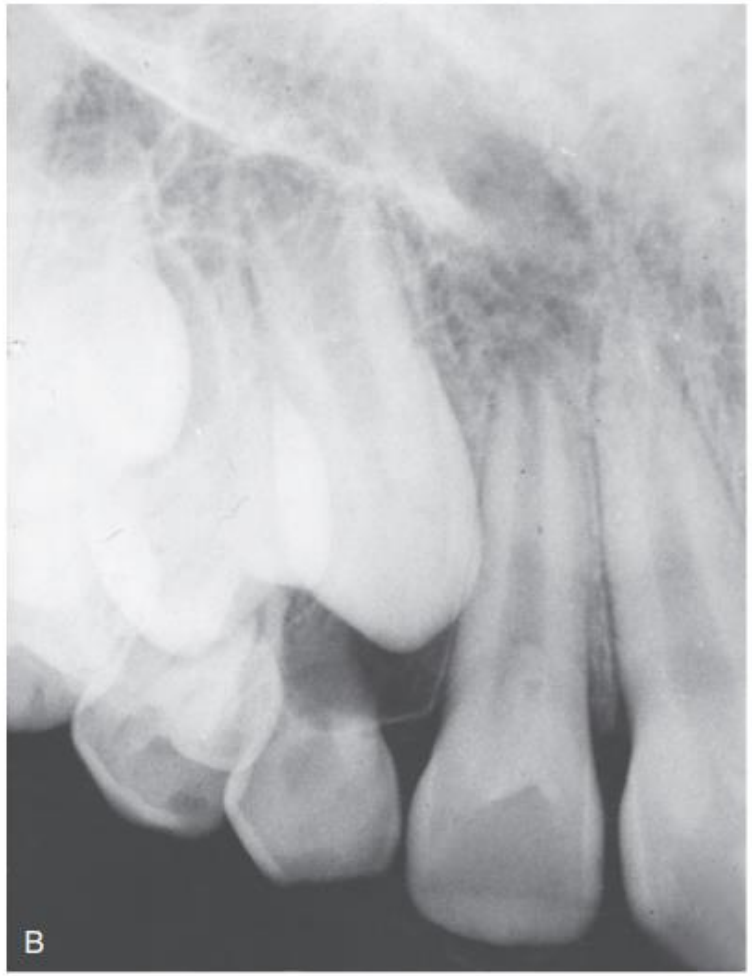
- Although a **greater incidence** in **males** has been reported for **the primary dentition,**
- **females** have been reported to be more commonly affected in the **permanent dentition**,
- or that there is **no gender diff**

- lingual invagination of the enamel, and can occur in **primary and permanent** teeth
- It is **most** often seen in the **permanent maxillary lateral incisors**, followed by **maxillary canines** or other **posterior maxillary teeth**, and **rarely in mandibular teeth**
- **autosomal dominant** inheritance
- Although **usually a nonsyndromic trait**, dens invaginatus has been **associated with Williams syndrome** in one case and other syndromic disease.

- **Anterior teeth** with dens invaginatus are usually of **normal shape and size**

- In **other areas of the mouth**, however, the tooth can have an **anomalous appearance**





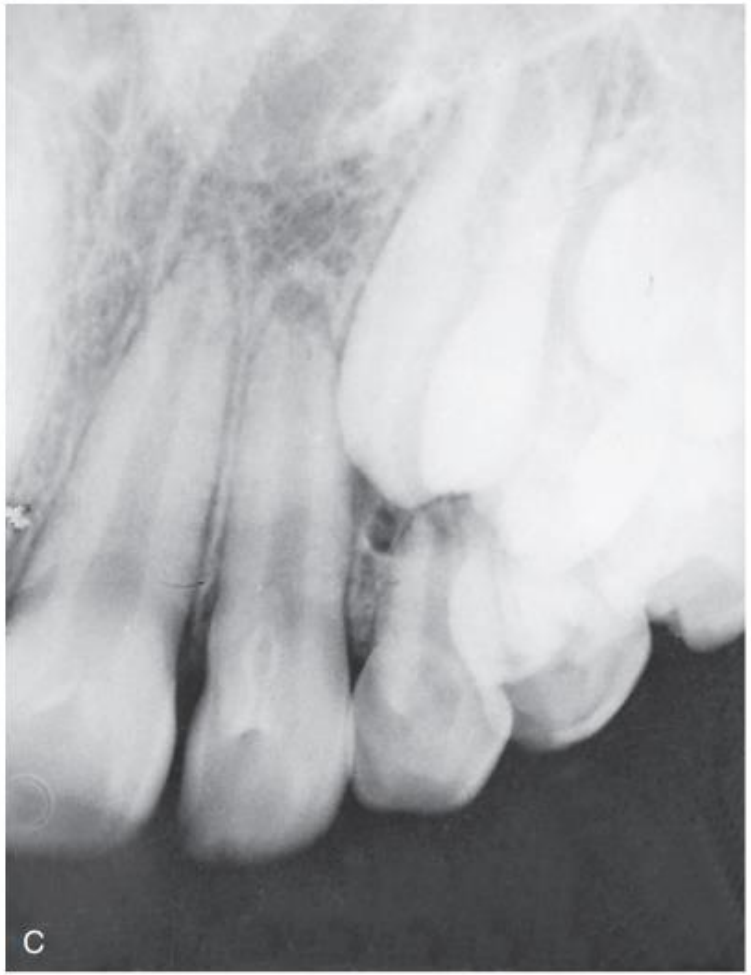


Fig. 3.8 (A) Small, "nonsticky" pits on the lingual surfaces of the maxillary lateral incisors are the only clues to the dens in dente condition of the teeth revealed radiographically in (B) and (C).

- Application of a **sealant or a restoration in the opening of the invagination** is the recommended treatment **to prevent pulpal involvement.**

- If the condition is detected **before complete eruption** of the tooth, **gingival tissue must be removed** **to facilitate cavity preparation and restoration**.
- A common presentation may be **tooth necrosis**, and the child may have an **acute abscess**.

- The **prognosis** of such teeth **depends** on **pulp morphology and restorability of the crown**

DENS EVAGINATUS AND TALON CUSP

- **Dens evaginatus** is a **dental anomaly** in which an **accessory cusp forms** on the **occlusal surface** on teeth in the **posterior**.
- **A dens evaginatus** that develops on an **anterior tooth** is termed **a talon cusp** due to its typical shape resembling the talon of an **eagle's claw**
- 15% of **Native Alaskan Eskimos and North American Indians** present with dens evaginatus

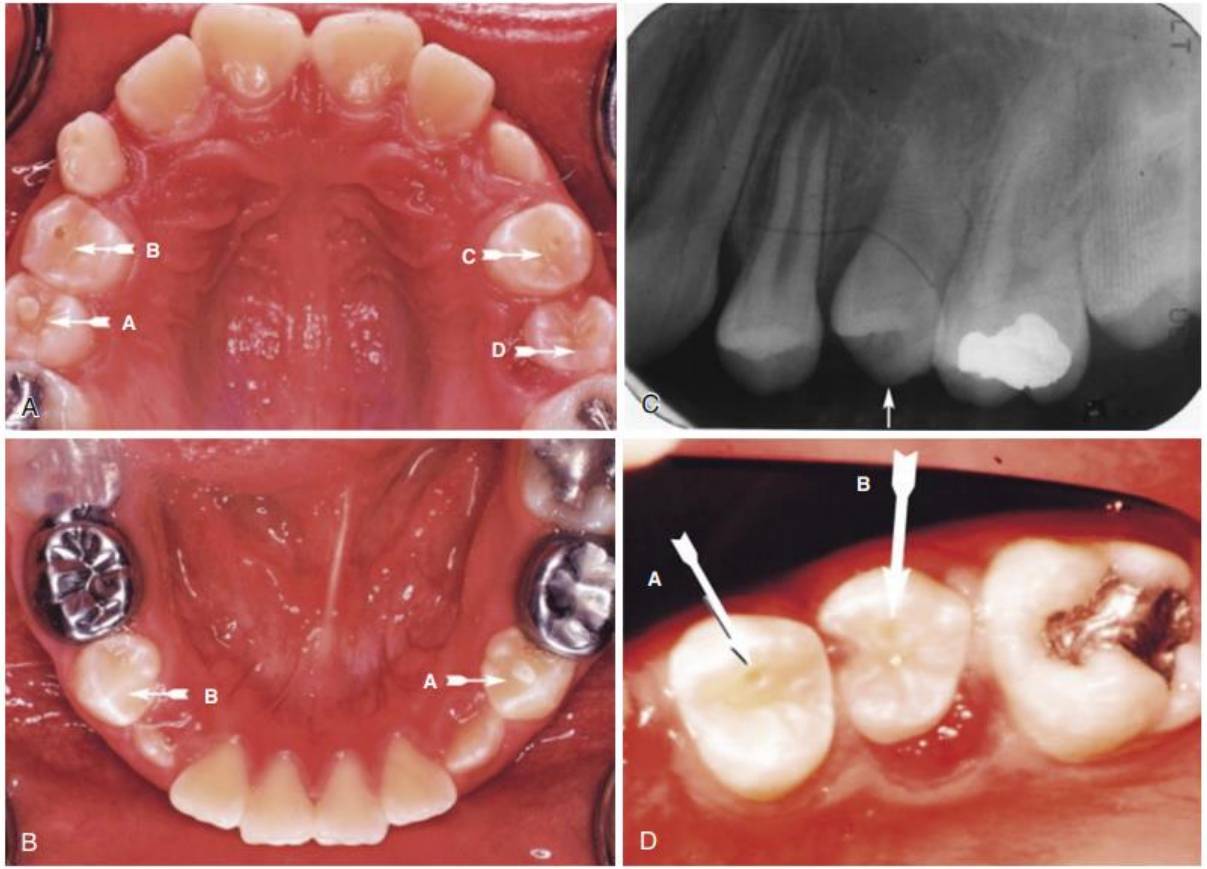
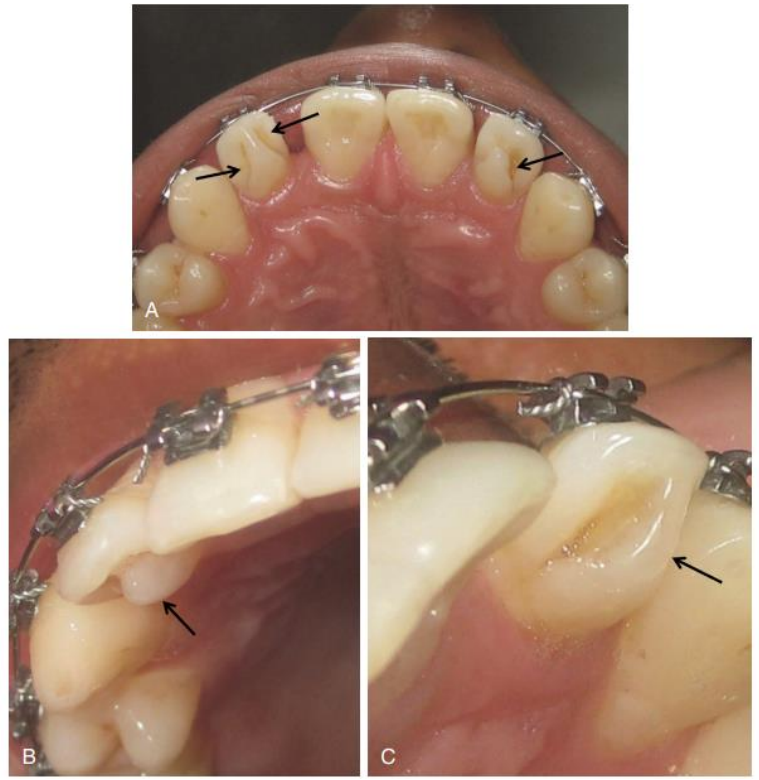


Fig. 3.10 Dens evaginatus observed in the dentition of a female Hispanic patient (9 years and 4 months old). (A) Photographic evidence of dens evaginatus on tooth no. 4 (arrow at A), intraoral appearance of occlusal caries on teeth nos. 5 and 12 at the site of fractured tubercle (arrows at B and C), and white halo on tooth no. 13 (arrow at D). (B) Dens evaginatus presenting as an exaggerated enlargement of the transverse ridge on tooth no. 21 (arrow at A) and a slight enlargement of the transverse ridge on tooth no. 28 (arrow at B). (C) Periapical radiograph of tooth no. 13 (arrow) revealing abnormal root development and periodontal defect on the mesial aspect. (D) The site of occlusal caries (arrow at A) on tooth no. 12 left after the tubercle fractured and the white halo (arrow at B) on tooth no. 13 representing the site of the fractured tubercle. *(Reproduced from The Journal of the American Dental Association, Vol 133 (Issue 2), Shelly Stecker and Anthony J. DiAngelis, Dens evaginatus: A diagnostic and treatment challenge, pages 190-193, Copyright (February 2002) with permission from Elsevier.)*

- **Talon cusps** develop on the **lingual (or buccal) tooth** surfaces of the **anterior portion of the maxilla more** often than the mandible
- .
- **Talon cusps developing** in the **adult dentition** on the **lingual tooth surface**(s) (about 68%) are more common than those forming on the buccal/facial surface (30%).
- Although **rare**, it has been documented that talon cusps form on **both the lingual and buccal sides** of a single tooth.
- While **unilateral talon cusps** occur with the **greatest frequency** in the **mandible**, approximately **one-fifth** of all cases are **bilateral**.
- "**Double teeth**" occur concurrently **with mandibular talon cusps** approximately half of the time.



- The presence of dens evaginatus may be **unaesthetic and interfere with occlusion.**

- If **unerupted**, it may **resemble** **a compound odontoma** or a **supernumerary tooth** and lead to **unnecessary surgery.**

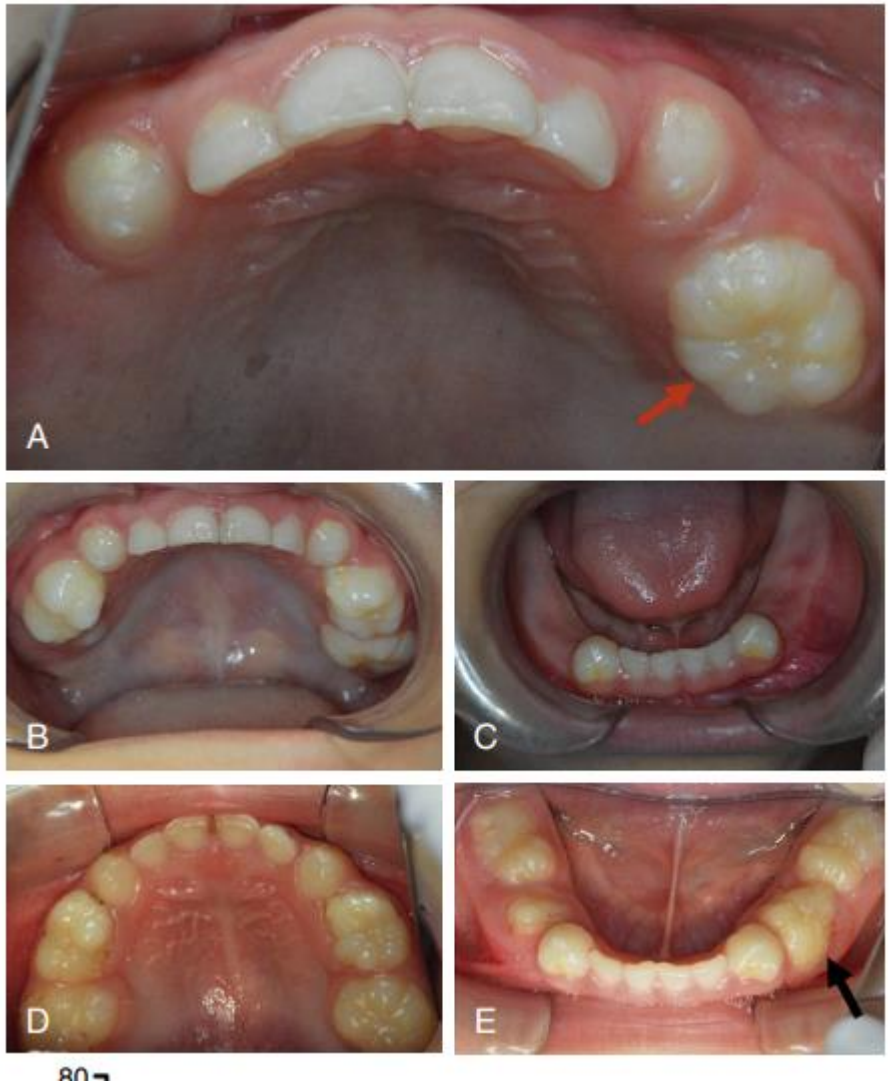
- If the **groove**s(between the talon cusp and the rest of the tooth)are **deep**, they should be **sealed** **and** monitored for caries development.

- topical **fluoride** application
- **endodontic** treatment
- **Orthodontic** treatment may be considered
- the **extra cusp** or opposing tooth surface

GLOBODONTIA

- Globodontia is a trait that affects teeth in the **molar and canine** fields.
- Affected **posterior teeth** are **enlarged** and have **a globular or bulbous appearance** often **lacking** any discernable **cusps** or major **occlusal grooves.**

- Affected **canines** can also exhibit **hypoplastic enamel**. Incisors appear to be unaffected by this condition.
- **primary and permanent** dentitions



- Globodontia is a **hallmark feature** of a condition called **otodental syndrome**
- the **primary** **dentition** tends to be **more seriously** affected.
- In addition, **globodontia** has been associated with **enflamed or enlarged gingiva and delay in tooth eruption**

- **Otodental syndrome** is a rare autosomal dominant disorder presenting with **tooth shape and size irregularities** in the **posterior dentition** (**bulbous, pumpkin-shaped teeth**), and high-frequency sensorineural **hearing loss**.

TAURODONTISM

- This anomaly is characterized by a tendency for the **body** of the tooth to **enlarge** at the **expense of the roots**.
- The **pulp chamber is elongated** and **extends deeply into** the region of the **roots**
- A similar condition is seen in the teeth of **cud-chewing animals** such as the bull (Latin, taurus).
- with syndromes such as **tricho-dento-osseous (TDO) syndrome** , **otodental dysplasia** , and **X-chromosome aneuploidies**
- The inheritance may also be **polygenic**

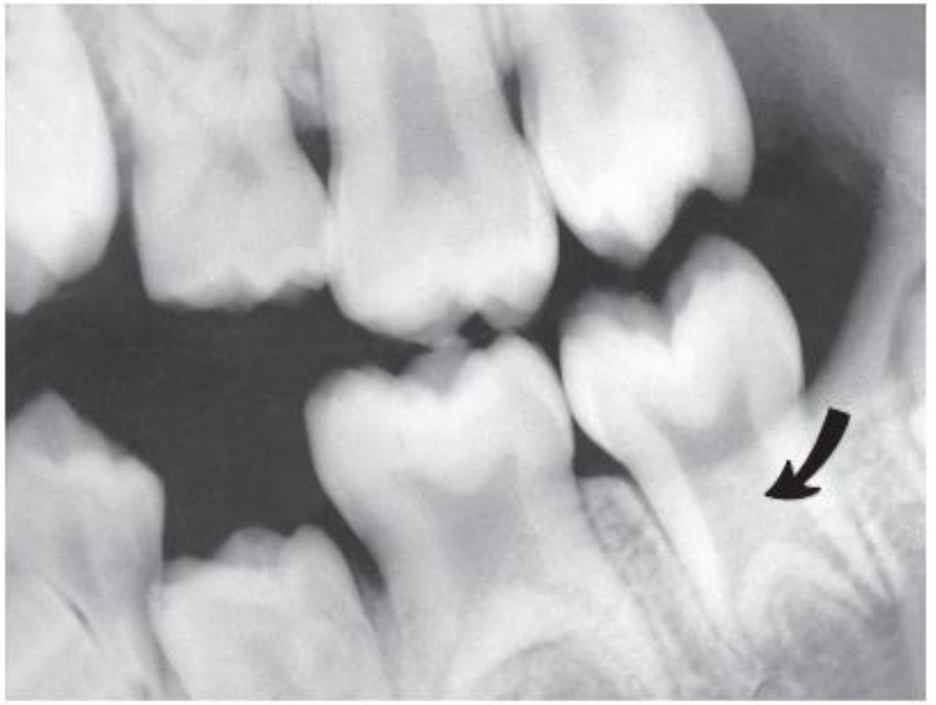


Fig. 3.33 Taurodontism. Notice the elongated pulp chamber and short root canals *(arrow)*.

INHERITED DEFECTS OF DENTIN

- **Two** broad categories of **heritable dentin defects**, **dentinogenesis imperfecta and dentin dysplasia,** are identifiable, each with **distinct subtypes**.
- .

DENTINOGENESIS IMPERFECTA (HEREDITARY OPALESCENT DENTIN)

- an isolated **autosomal dominant** trait
- Witkop suggested that there are **two distinct diseases** and recommended the terms **dentinogenesis imperfecta** for the disease that occurs in conjunction **with osteogenesis imperfecta and hereditary opalescent dentin** for the disease that occurs **as an isolated trait**.
- **Shields** et al. proposed a new **classification:** (**Shields) type I** dentinogenesis imperfecta and (**Shields) type II** dentinogenesis imperfecta, respectively

- In addition, the dentin defects seen in the **isolated Brandywine triracial population** in **southern Maryland** were termed (Shields) **type III dentinogenesis imperfecta.**
- These latter defects consisted of variable expression of the **features of (Shields) type I (without osteogenesis imperfecta) and type II, shell-like teeth, and multiple pulp exposures**

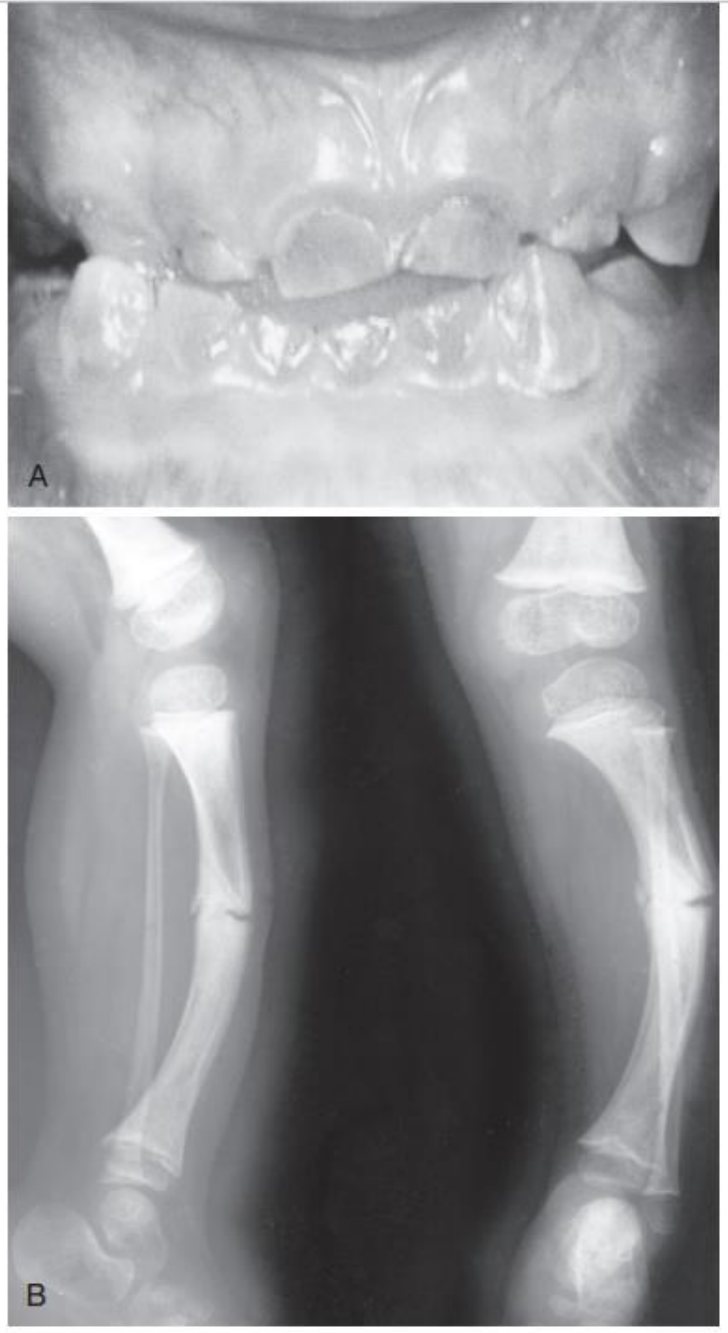


Fig. 3.34 (A) A 5-year-old girl with dentinogenesis imperfecta and osteogenesis imperfecta. The child had sustained numerous fractures of the long bones. (B) A fracture of the tibia is evident in the radiograph.

- **normal dentin** formation is confined to **thin layer next to the enamel** and **cementum**, followed by a layer of **disorderly** dentin containing a few tubules.

- **The roots of shell teeth are short**, and the **primary teeth** may be **exfoliated prematurely.**

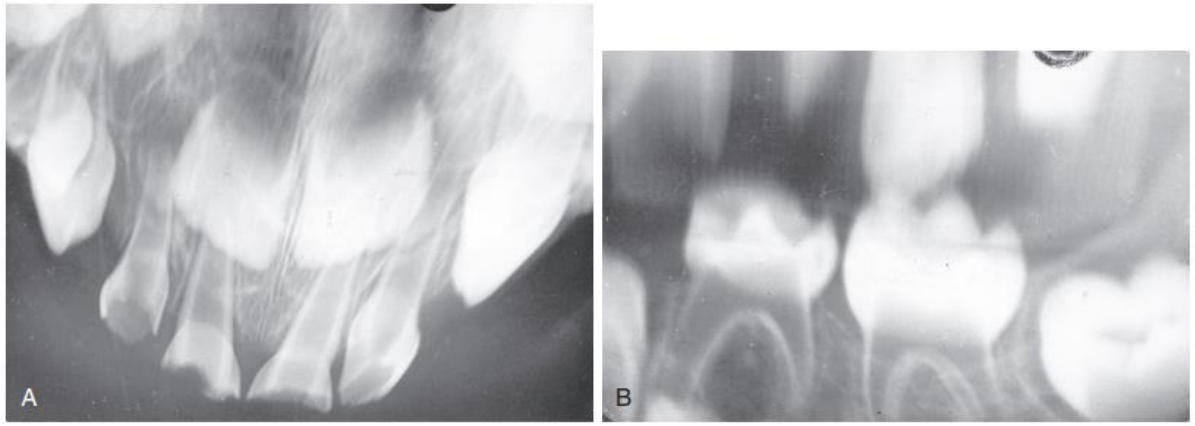


Fig. 3.35 Shell teeth. The large size of the pulp cavities indicates the nonexistence of secondary dentin.

- Xiao et al. and Zhang et al. have found **mutations in the DSPP gene**, which **codes** for the two **major noncollagenous dentin matrix proteins dentin sialoprotein (DSP)** and **dentin phosphoprotein** (also known as **phosphophorin**), in patients with (Shields) type II dentinogenesis imperfecta.

- The **clinical picture of dentinogenesis imperfecta** is one in which the primary and permanent teeth are a characteristic **reddish-brown to blue-gray opalescent color**.

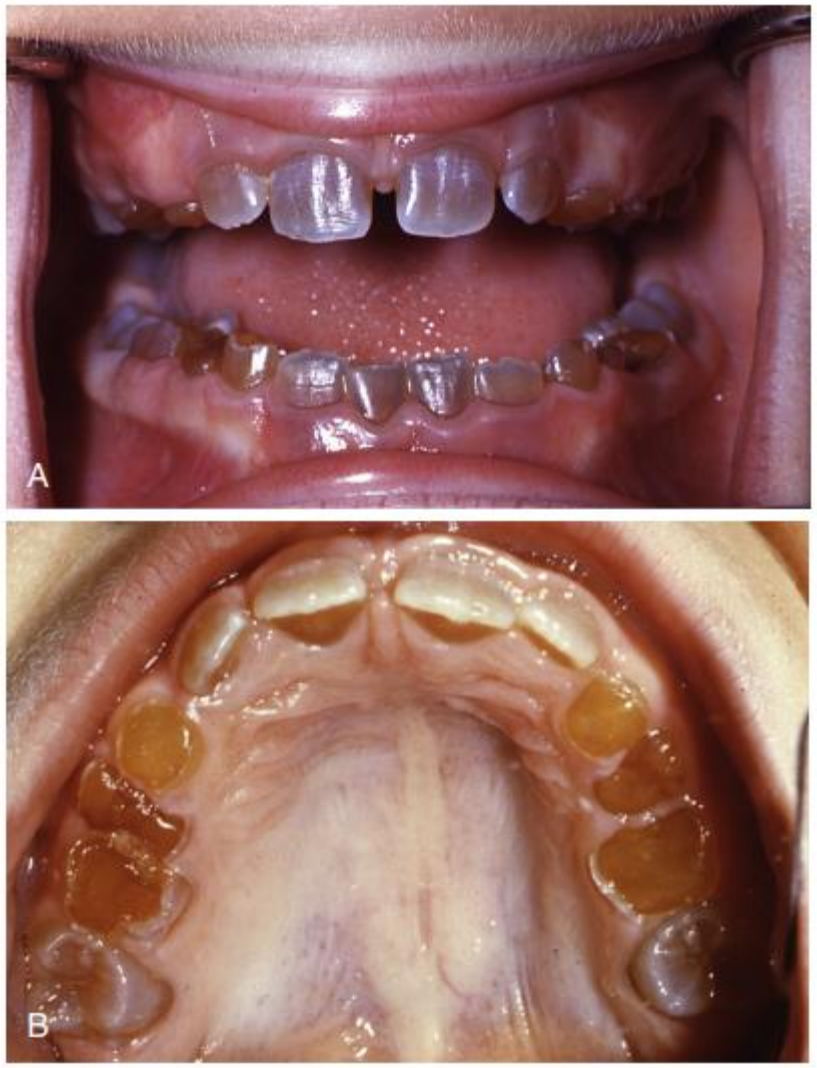


Fig. 3.36 Dentinogenesis imperfecta. The primary teeth are severely abraded. Enamel is breaking away from the incisal edge of the lower permanent central incisors.

- Soon after the primary dentition is complete, **enamel is worn** and often **breaks away** from the incisal edges of anterior teeth and the occlusal surfaces of posterior teeth.

- The **exposed soft dentin abrades rapidly**,

- **the smooth, polished dentin surface** is continuous with the **gingival tissue**

- **Radiographs** show **slender roots and bulbous crowns**. The **pulp chamber is large initially and undergoes obliteration**

- **Periapical rarefaction** in the **primary** dentition is observed only **occasionally**

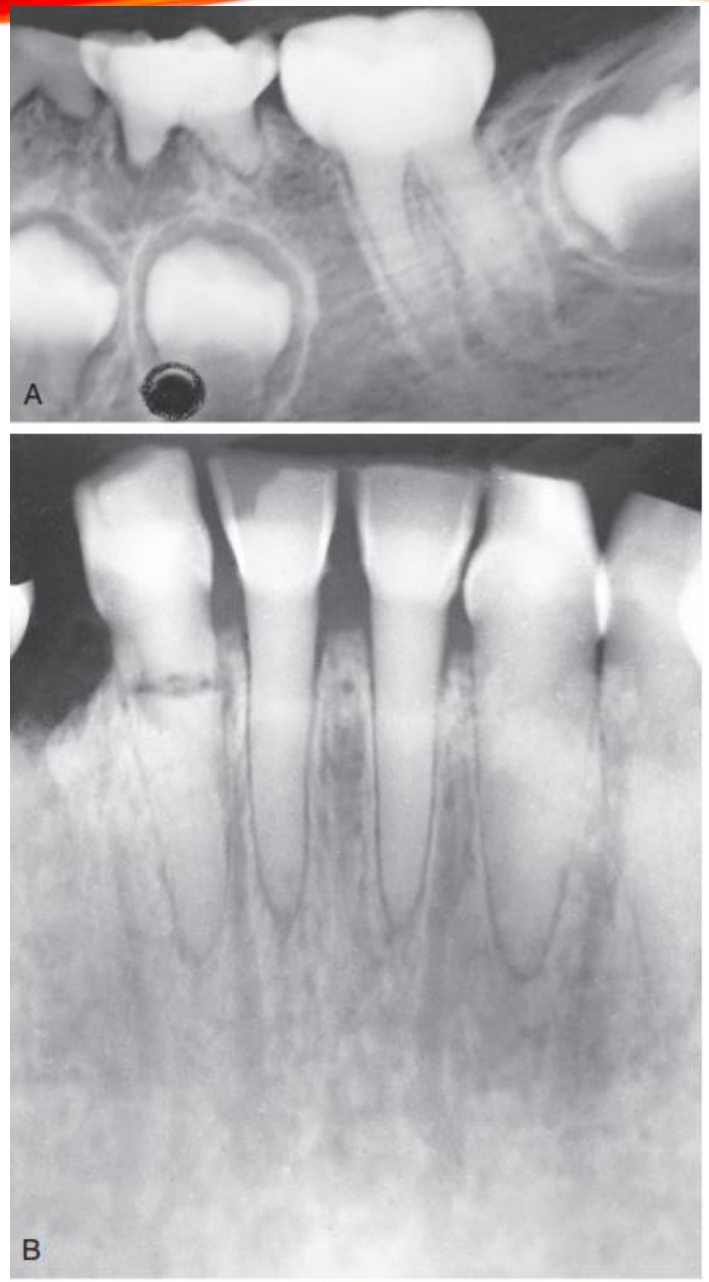


Fig. 3.37 (A) Slender roots with ribbon-like pulp canals and bulbous crowns are characteristic of dentinogenesis imperfecta. The primary molars show periapical rarefaction. (B) Root fractures are common in older patients.

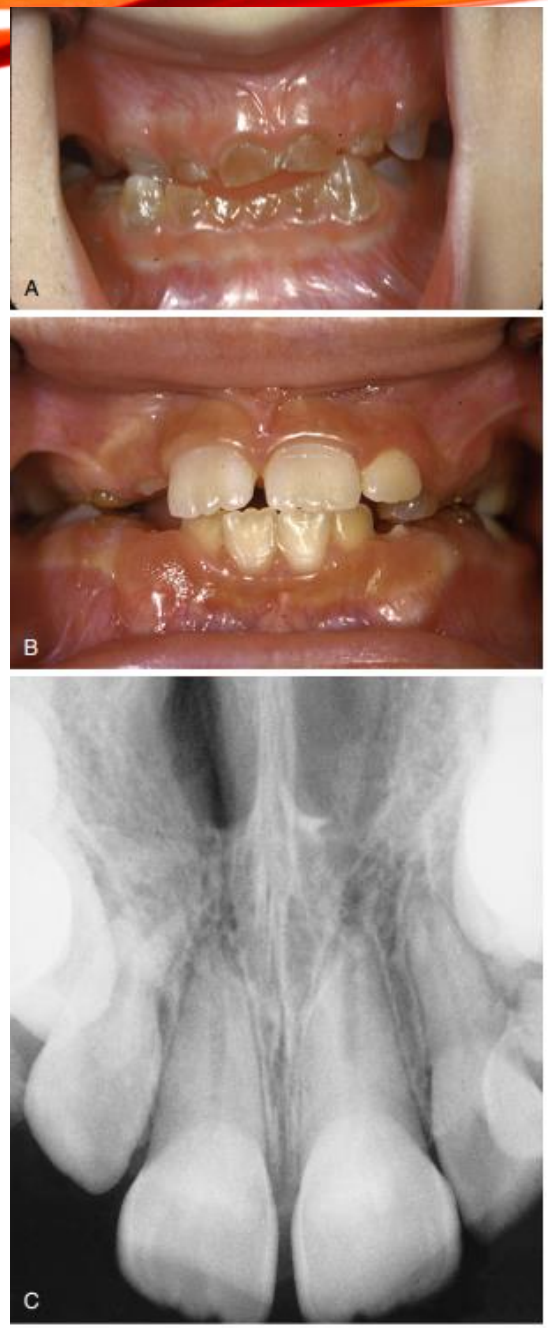


Fig. 3.38 (A) Four-year-old child with dentinogenesis imperfecta. (B) The permanent teeth, in contrast to the primary teeth, are normal in color. (C) The radiograph shows typical dentinogenesis imperfecta.

- **treatment**
- The placement of **stainless-steel crowns** on primary posterior teeth
- **Full-coverage restorations** may be placed on the permanent
- **Bonded veneer restorations** on anterior teeth
- Unfortunately, the **long-term prognosis** of these teeth is **poor**;
- despite best efforts, they are usually lost in early adulthood.

- **Teeth with periapical rarefaction and root fracture** should be **removed**. **Extraction** of such teeth is **difficult** because of the brittleness of the dentin.

DENTIN DYSPLASIA

- Dentin dysplasia is a **rare disturbance of dentin formation** that Shields et al
- categorized into **two types**: **radicular dentin dysplasia (type I)** and **coronal dentin dysplasia (type II).**

- Both **primary and secondary dentitions** are affected in **dentin dysplasia type I**, which is inherited as an **autosomal dominant trait**.
- **Radiographically**, the **roots are short** and may be **more pointed** than normal. Usually, the **root canals and pulp chambers are absent except** for a **chevron-shaped remnant in the crown.**

- The **color and general morphology of the crowns** of the teeth **are usually normal**, but they may be slightly opalescent and blue or brown.

- **Periapical radiolucencies** may be present at the **apices** of affected

- Investigation of individuals with **dentin dysplasia type I associated with extreme microdontia and aberrant crown morphology** disclosed homozygosity for a splice-site **mutation in the SMOC2 gene**. Since parents and siblings heterozygous for the mutation were not affected, the SMOC2 gene may not be involved in dentin dysplasia type I without microdontia and aberrant crown morphology

- On noting the **phenotypic similarity** of Shields **type II dentinogenesis imperfecta** to that in **the primary dentition** in **dentin dysplasia type II,** Dean et al

- **Dentin dysplasia type II** is inherited as an **autosomal dominant trait** in which the **primary dentition** appears **opalescent and radiographically** shows **obliterated pulp chambers similar to** those in **dentinogenesis imperfecta**.

- **Unlike** dentinogenesis imperfecta, however, **in dentin dysplasia type II, the permanent dentition has normal color and**

radiographically exhibits a thistle tube pulp configuration with pulp stones.

AMELOGENESIS IMPERFECTA

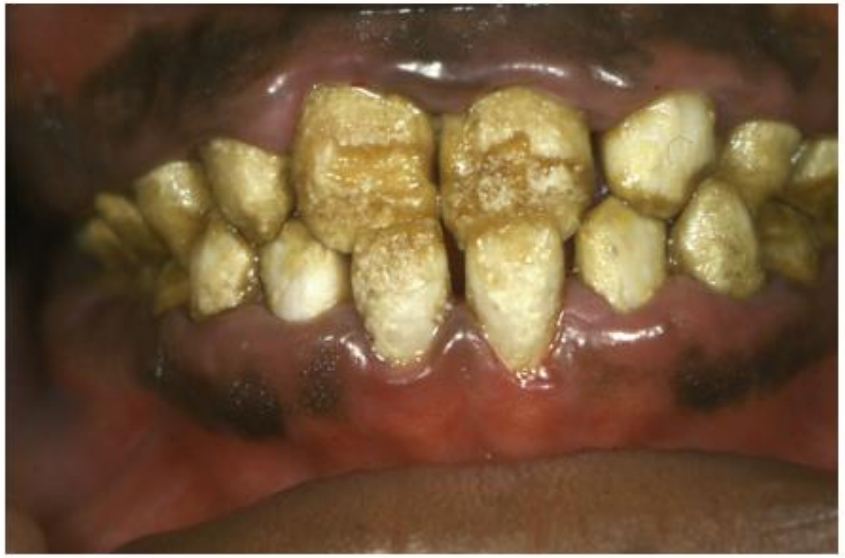


Fig. 3.39 Both the primary and permanent teeth are affected by the hereditary anomaly amelogenesis imperfecta. The enamel is pitted but hard.

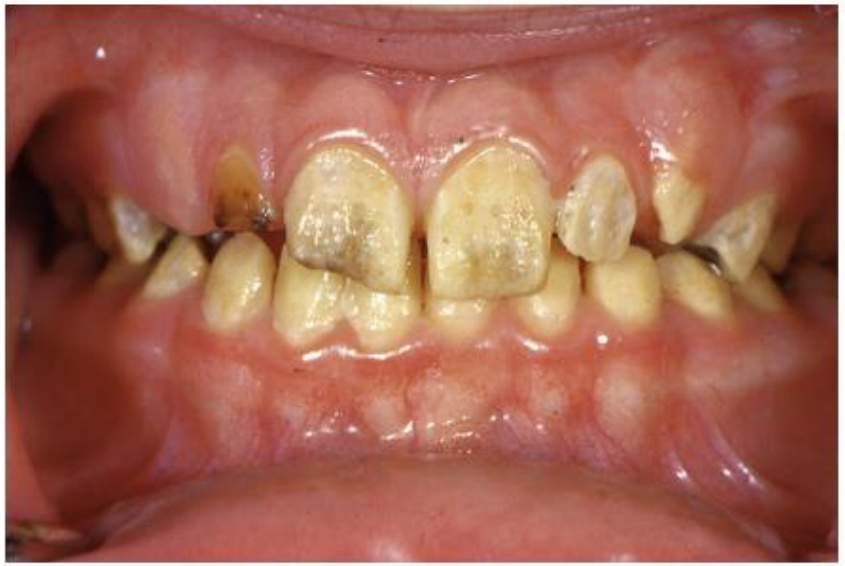


Fig. 3.40 Hypocalcification type of amelogenesis imperfecta. The primary teeth were similarly affected. The enamel surface is soft.

- a developmental defect with a **heterogeneous etiology** that affects the **enamel** of both **primary and permanent dentition**.
- **Amelogenesis imperfecta** has a wide **range of clinical appearances**, with **three broad categories** observed clinically: the **hypocalcified type**, the **hypomaturation** type, and the **hypoplastic** type.

- Although amelogenesis imperfecta can occur as part of several syndromes

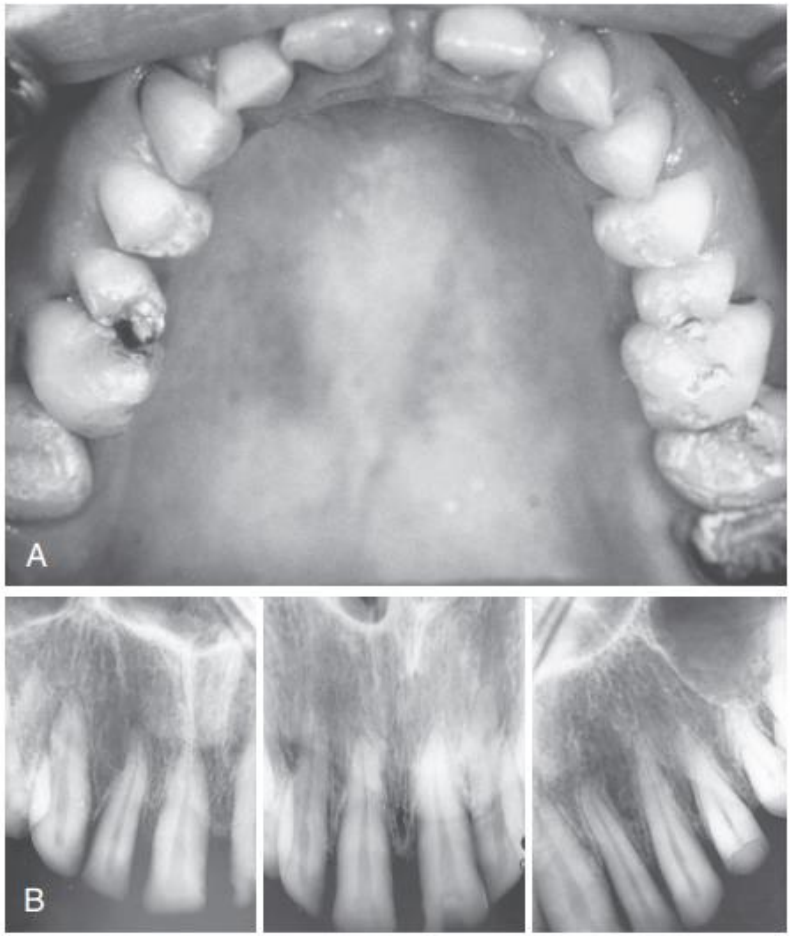


Fig. 3.41 (A) Case diagnosed as amelogenesis imperfecta. The permanent teeth have a thin covering of pigmented enamel. (B) The radiographs show essentially normal root morphology. The crowns have a thin covering of enamel.

- Aldred and Crawford proposed two separate phenotypes, namely enamel that is predominantly
- **hypoplastic** (i.e., **enamel** that is **either uniformly thin, with spacing between adjacent teeth, or irregular, giving rise to pits or grooves**)
- or
- predominantly **hypomineralized** (i.e., **with soft, poorly formed enamel with altered color and translucency)**

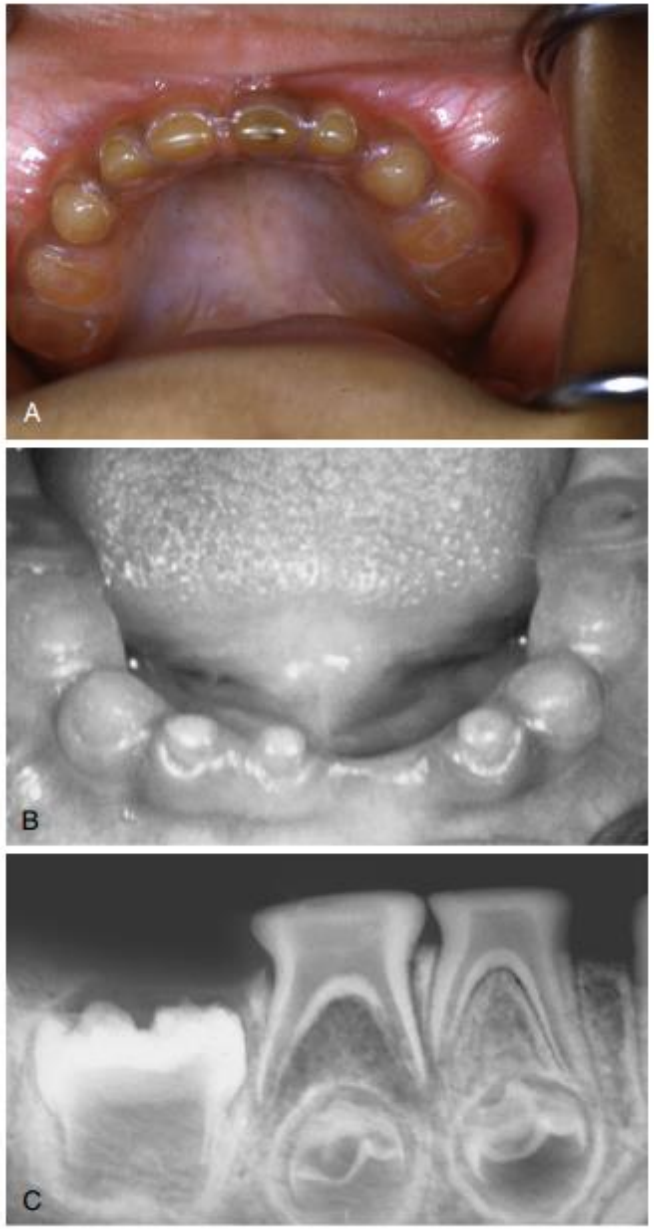


Fig. 3.43 (A and B) Severely abraded teeth are almost entirely devoid of enamel. The outline of a large pulp chamber can be seen through a thin covering of dentin. The mandibular second primary molars have pulp exposure. (C) Radiograph shows large pulp canals and large pulp chambers. Apical rarefaction is associated with pulp exposure of the second primary molar.

- **Hart et al.** recommended a **standardized nomenclature** for describing amelogenesis imperfecta that causes alterations at the genomic, complementary DNA, and protein levels.
- **Two clinically distinct forms** of **autosomal dominant amelogenesis imperfecta— smooth hypoplastic amelogenesis imperfecta and local hypoplastic amelogenesis imperfecta**—are associated with **mutations** in the enamelin **(ENAM)** gene located at 4q21.

- In addition, **autosomal dominant** amelogenesis imperfecta can be associated with **mutation in the Kallikrein-4 (KLK4) gene**, and **autosomal recessive pigmented hypomaturation amelogenesis imperfecta** with an **enamelysin (also termed Matrix Metallopeptidase-20, MMP-20) gene mutation,** illustrating the heterogeneity of the condition.

- An **X-linked form (AIH1)** has been found to be associated with as many as 14 mutations in the **Amelogenin X-Linked (AMELX) gene** located at Xp21.

- However, at least one family has had the trait linked to **another location on chromosome Xq22-q28.13**

- The **defective tooth structure** is limited to the enamel.
- On **radiographic examination**, the **pulpal outline** appears to be **normal**, and the **root morphology** is that of **normal** teeth.

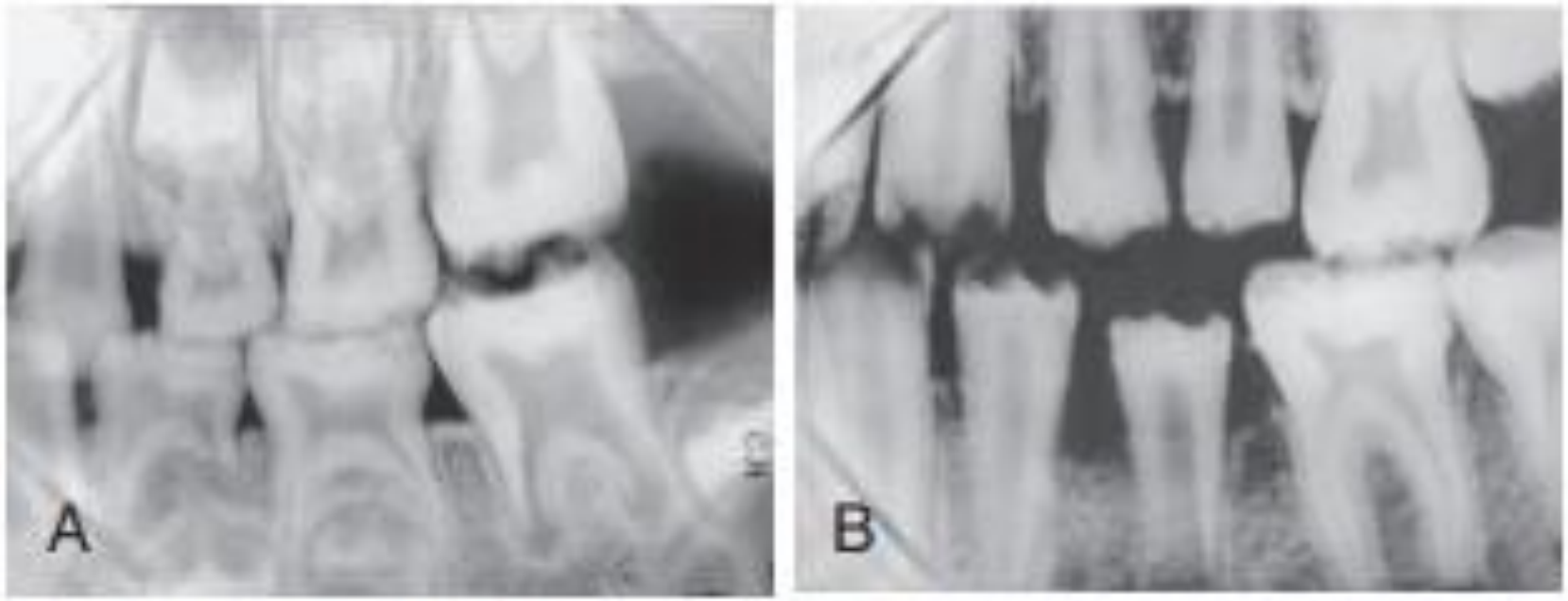


Fig. 3.42 (A and B) Left bite wing radiographs of a patient with amelogenesis imperfecta. Radiograph in B was made 6 years after radiograph in A and demonstrates the maintenance of a caries-free dentition despite the thin enamel.

- In the **hypoplastic type**, the **enamel matrix** appears to be **imperfectly** formed; although **calcification** subsequently **occurs in the matrix and the enamel is hard,** it is **defective** in **amount** and has a **roughened, pitted surface**

- In the **hypocalcified type**, **matrix formation appears to be of normal thickness, but calcification is deficient, and the enamel is soft**.
- In both of these more common types of the defect, the **enamel becomes stained** because of the **roughness** of the surface and increased **permeability.**

- In still **another variant of amelogenesis imperfecta**, **a thin, smooth covering of brownish-yellow enamel** is present.
- In this type, the **enamel does not** seem excessively **susceptible to abrasion or caries**

- Seow139 has suggested that some cases reported as **amelogenesis imperfecta with taurodontism** were actually **cases of TDO syndrome**

- **Amelogenesis imperfecta** may also be **associated with nephrocalcinosis syndrome**, also called **enamel-renal syndrome or Lubinsky syndrome**. Inheritance appears to be autosomal recessive. In addition to **hypoplastic enamel, the teeth often fail to erupt and undergo resorption due to the disappearance of the reduced enamel epithelium.** Histopathology of the **gingiva**, which **may be enlarged**, **reveals islands of odontogenic calcification**.
- Nephrocalcinosis may be the cause of renal impairment but only rarely end-stage renal failure, unless complicated by renal stones and recurrent infections

- The **treatment** **of teeth with amelogenesis imperfecta**– like defects depends on the **severity** of the condition and the demands for **aesthetic improvement.**
- full-coverage restorations
- the teeth can be prepared for **full-coverage restorations**.
- For some cases of the **hypoplastic types**, **bonded veneer restorations** may offer a more conservative alternative for the management of the aesthetic problem of the anterior teeth. Patel et al. have reported successful treatment with **porcelain laminate veneer restorations**.

ENAMEL AND DENTIN APLASIA

- **Teeth with** characteristics **of both dentinogenesis imperfecta and amelogenesis imperfecta**

- **Chaudhry** et al. reported such a case and called the condition **odontogenesis imperfecta**.

- Schimmelpfennig and McDonald observed a similar dentition and termed it **enamel and dentin aplasia**.

- The **primary teeth** were essentially **devoid of enamel**, and the **smooth, severely abraded dentin** was **reddish-brown**.
- **Radiographs** showed **normal alveolar bone** around the roots of the teeth. Two teeth had **pulp exposure** and pulpal degeneration (Fig. 3.43). **Radiolucent** areas were present at the apices of the two primary teeth

- The **pulp chambers and canals** in all the **primary teeth** were **extremely large**, with **no** evidence of becoming **obliterated**.
- In ground sections of the primary teeth, the **dentinal tubules** showed **little evidence of a normal growth pattern. They were few and irregular**, with a tendency toward branching.
- The **cementum appeared normal and was acellular**.
- **No evidence of secondary dentin formation** was found.
- A few fragments of **enamel adhering to the dentin appeared thinner than normal**, and few normal morphologic characteristics were present.
- The dentino-enamel junction was atypical in that it **lacked** the characteristic **scalloping.**

- The **permanent teeth**, when they erupted, were **partially covered with a thin, gray, poorly coalesced coating of enamel.**
- Brown dentin could be seen on the labial aspect of the central incisors and at the bases of the fissures of the first permanent molars.
- **Stainless-steel crown restorations** were placed even before complete eruption to protect the teeth from continued abrasion.