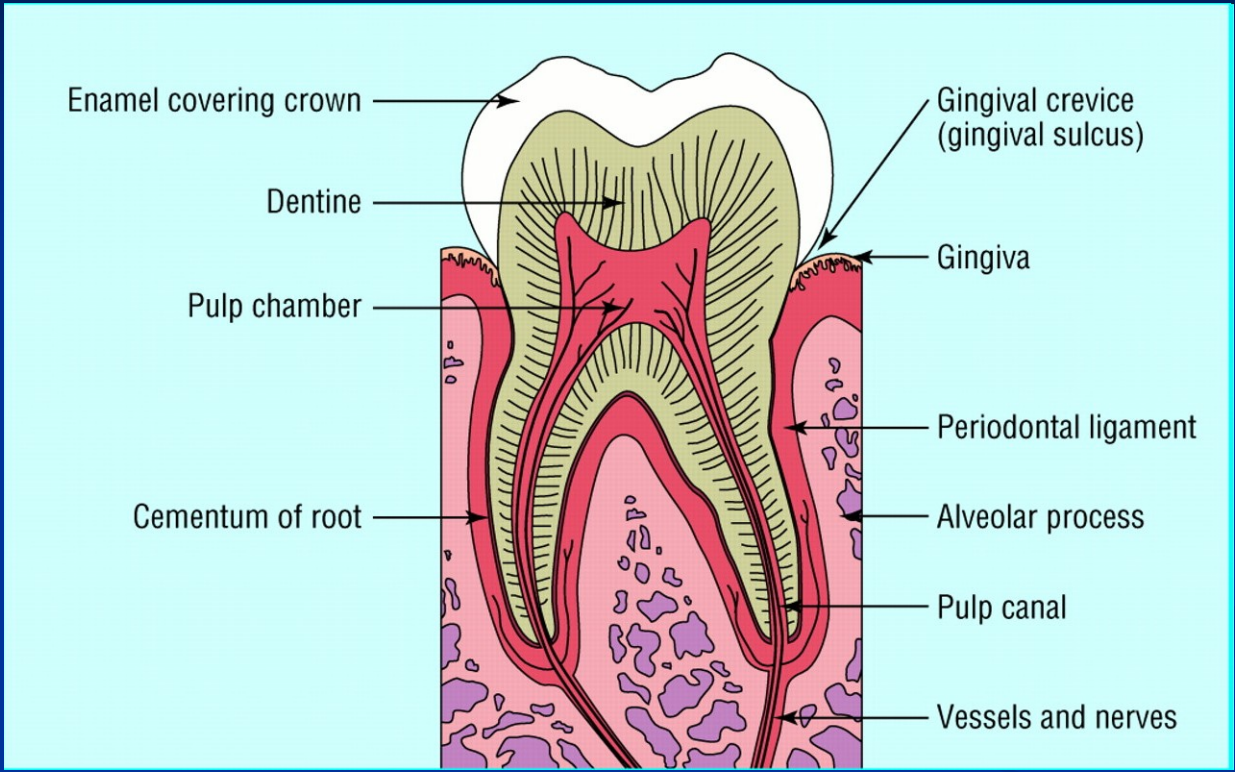
Disorders of development of teeth and craniofacial anomalies.

Markéta Hermanová

Disorders of development of teeth.



- Disturbances in number of teeth
- Disturbances in size of teeth
- Disturbances in form of teeth
- Disturbances in structure of teeth
- Craniofacial anomalies

Disorders of development of teeth.

- Prenatal
- Postnatal

- Inherited
- Aquired

Disturbances in number of teeth.

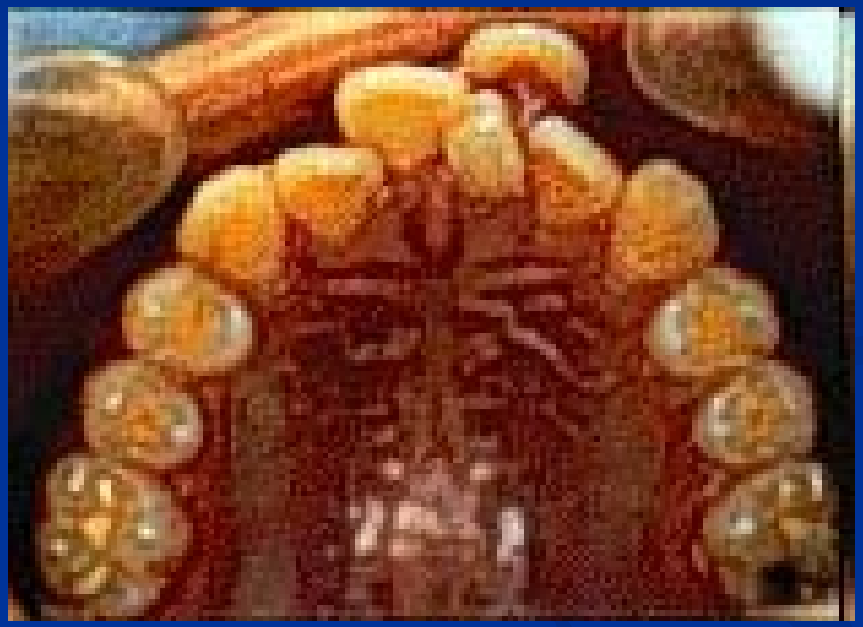
- Hypodontia, anodontia, and associated syndromes
- Hypohidrotic ectodermal dysplasia
- Hyperdontia (supernumerary teeth)

- **Hypodontia: congenital absence of teeth**
  - More common in the permanent dentition (2-10 % in populations)
  - In primary dentition <1 %; assoc. with the absence of permanent succesional tooth
  - Racial and geographic differences
  - Symetric/asymetric
  - 3rd molar, permanent maxillary lateral incisors, mandibullary 2nd premolarsy
  - A role of control and regulating genes in the development of teeth
  - Assoc. wih other craniofacial anomalies and syndromes
- **Anodontia:** complete absence one or both dentitions

- **Hypohidrotic ectodermal dysplasia**

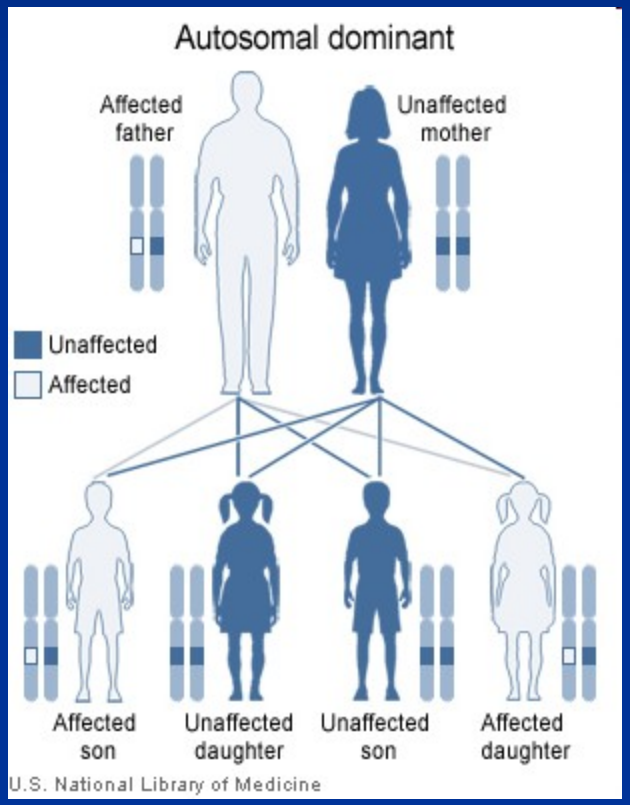
- Congenital absence of ectodermal structures
- X-linked (GR), mutation in EDA gene (signalling molecule), failure of interactions between epithelial and mesenchymal tissues; rarely AR
- Smooth dry skin, scanty hairs, partial or total absence of sweat glands (hyperthermia)
- Severe hypodoncia (teeth retarded in eruption, deformed teeth, conical crowns of teeth)
- Female carriers - minimal hypodontia

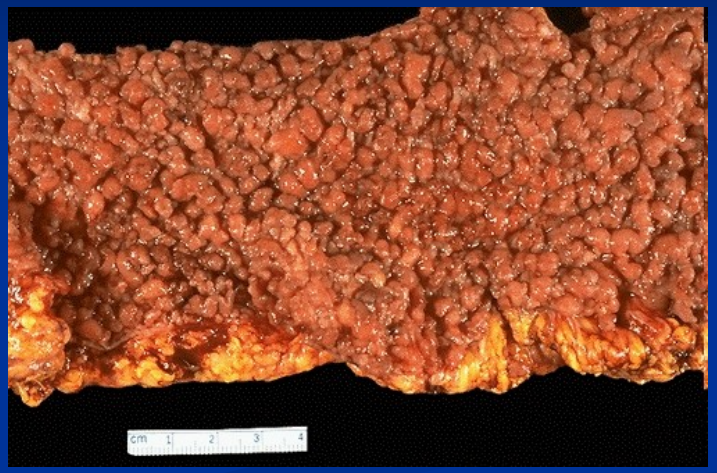
- **Hyperdontia (supernumerary teeth)**
  - maxilla (anterior and molar regions)
  - assoc. with cleft palate and cleidocranial dysplasia
  - F>M
  - Unusual in deciduous dentition
  - Prevent the eruption, causes malposition, resorption of adjacent teeth, developlment of dentigerous cysts if unerupted
  - *Mesiodens:* between maxillary central incisors
  - *Paramolar:* alongside the maxillary molars, usually buccaly placed
  - *Distomolar:* distally to a 3rd molar

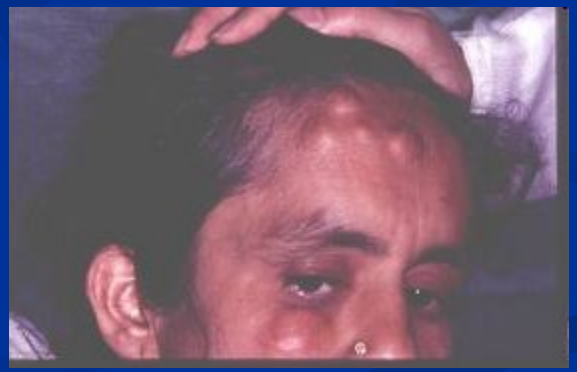


Syndrom/anomaly	Associated features
Hypodontia	
Cleft lip/palate	Deafness, cranial and skeletal abnormalities
Crouzon syndrome (FGFR gene)	Craniosynostosis, maxillary hypoplasia, hypertelorism
Down syndrome (trisomy 21)	Multiple, e.g. mental retardation, macroglossy, maxillary hypoplasia, anomalies of heart
Hypohidrotic ectoderma dysplasia	Hypotrichosis, hypohidrosis, saddle-nose
Ellis-van Creveld syndrome	Dwarfism, polydactyly, cardiac malformations
Oro-facial digital syndrome	Cleft palate, hypoplasia of nose, digital malformations
Hyperdoncie	
Cleft lip/palate	Deafness, cranial and skeletal abnormalities
Cleidocranial dysplasia (RUNX2 gene)	Aplasia of clavicles, delayed ossification of fontanelles, enlargement of cranium
Gardner syndrome (APC gene)	Osteomas of jaws, skin cysts and fibromas, intestinal polyposis-carcinomas
Sturge-Weber angiomatosis	Venous angiomatosis (also facial and oral), cerebral angiomatosis
Oro-facial digital syndrome	Cleft palate, hypoplasia of nose, digital malformations

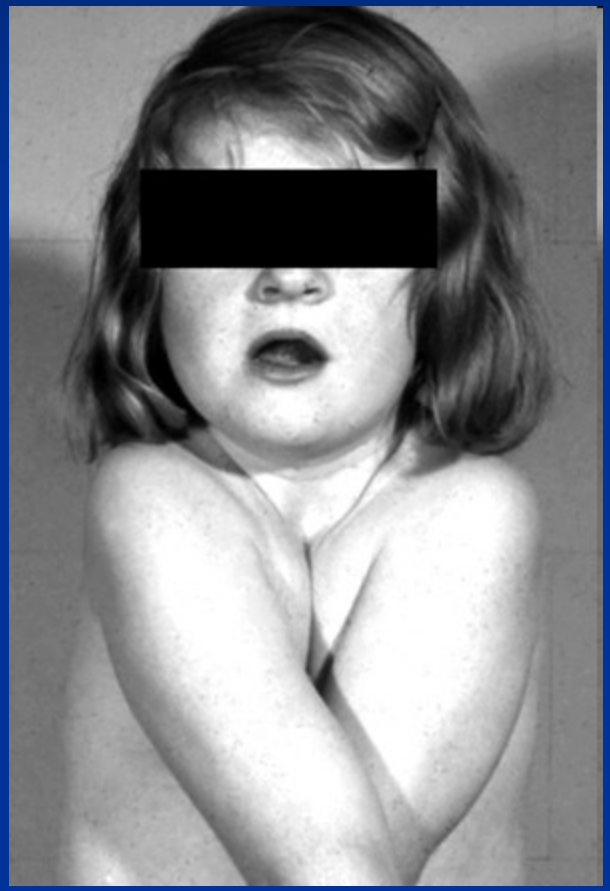
Gardner syndrome

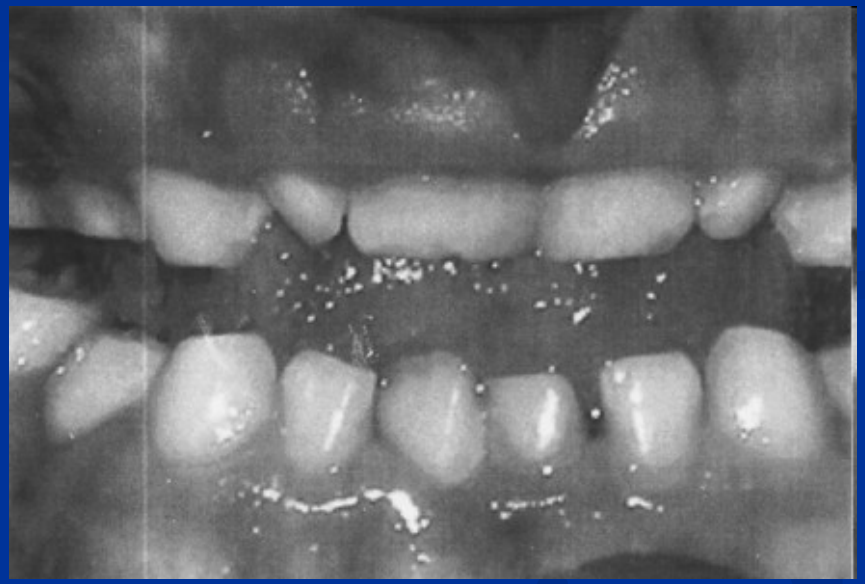




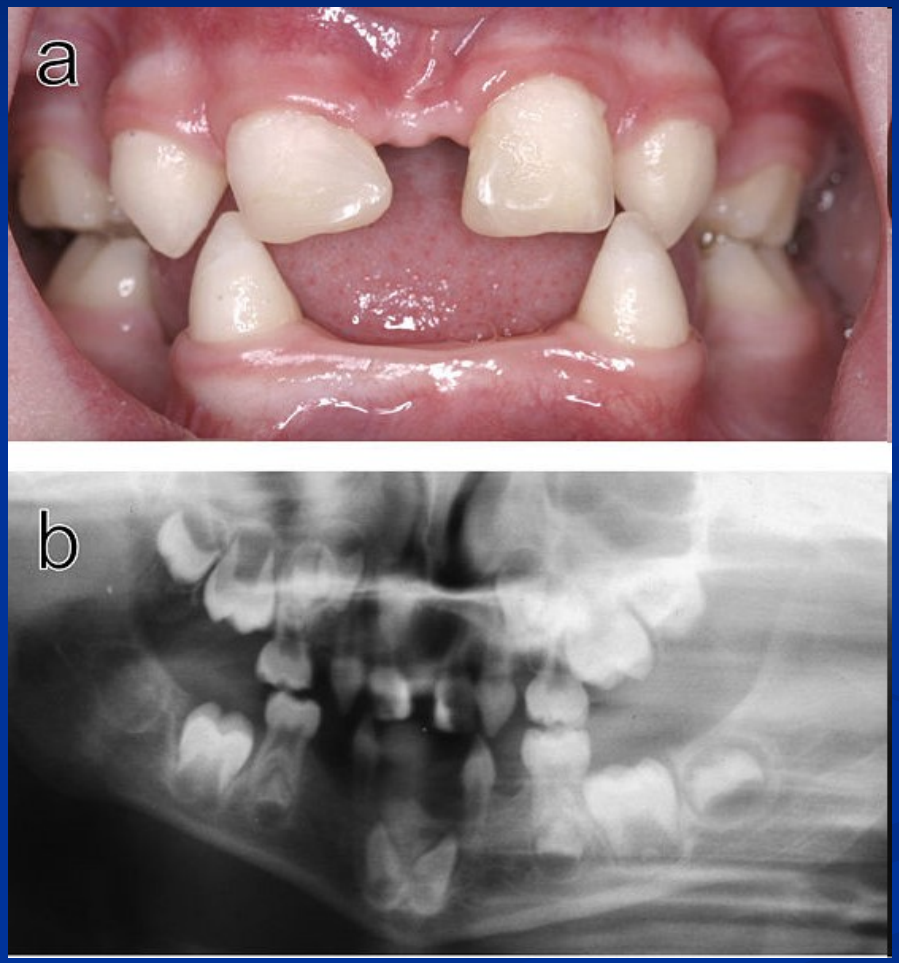


Cleidocranial dysplasia.





Hypodontia.



Orofacial clefts

- In combination with over 300 syndromes
- 70 % non-syndromic
- 1/500-1000 births
- Clefts of the lip and palate (45 %)>clefts of the palate (30 %) >clefts of the lip (25 %)
- Multifactorial causes

- **Cleft lip**: defective fusion of the medial nasal process with maxillary process
- **Cleft palate**: failure of palatal shelves to fuse

- **Lateral facial cleft** (isolated or with mandibulafocial dysostosis): lack of fusion of the maxillary and mandibullary processes; uni- or bilateral

- **Oblique facial cleft**

  (from upper lip to the eye, +CP; failure of fusion of the lateral masal process with the maxillary process or caused by amniotic bands)

- **Median cleft of the upper lip**

  (failure of fusion of the medial nasal processes; in several syndromes, in holoprosencephaly)

- **Median maxillary anterior alveolar clefts**

  (bony defect in the midline of the maxilla between incisors)

Disturbances in size of teeth

- **Macrodontia**
- **Microdontia**

- **Genetic factors**

(microdontia in Down syndrome, in congenital heart diseases)

- **Environmental factors**
- **May involve the entire dentition**

Disturbances in form of teeth.

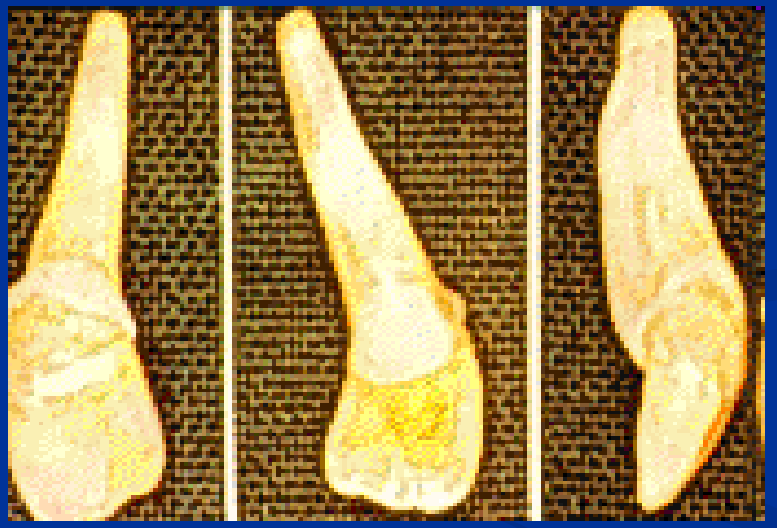
- **Dilaceration**
  - Tooth severely bent along its long axis, trauma
  - Maxillary incisor

- **Taurodontism**
  - Pulp chamber higher, with no constriction in amelocemental junction
  - Failure of Hertwig´s sheath invaginate at the proper horizontal level
  - Sporadic or assoc. with Klinefelter and poly-X chromosomes syndromes

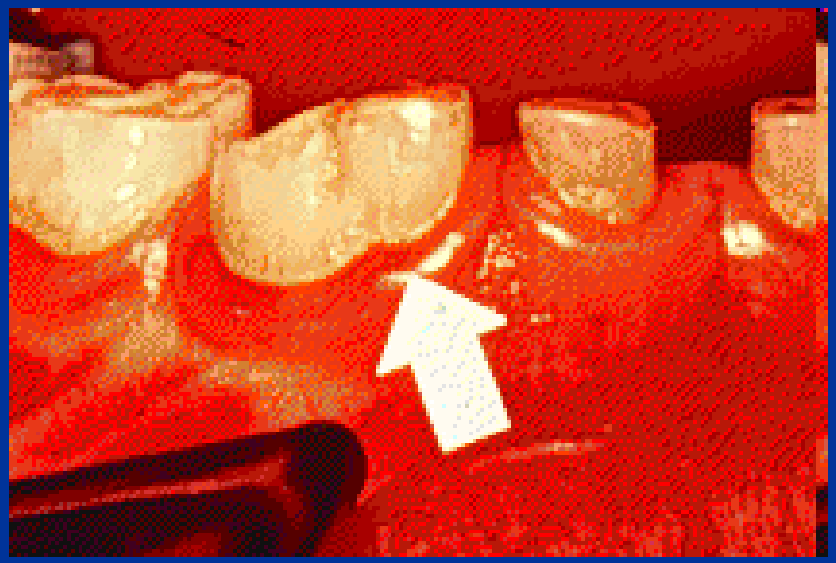
- **Double teeth**
  - Developmental anomaly, teeth joined together (crowns, roots, or both (with/without joining of the pulp)
  - More often in primary dentition
  - Fusion (the union of two or more separate developin)
  - Gemination (incomplete division of teeth)

- **Concrescence**
  - Acquired disorder, affects more often permanent dentition
  - Teeth united by cementum (anatomically close teeth (2nd and 3rd maxillary molar, hypercementosis in inflammation)

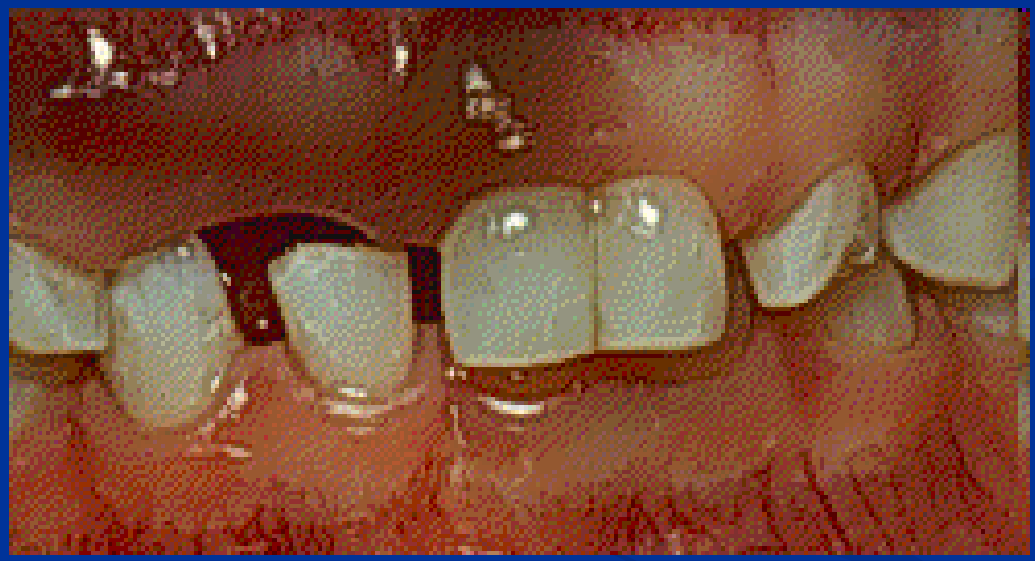
Taurodontism



Dilaceration



Fusion



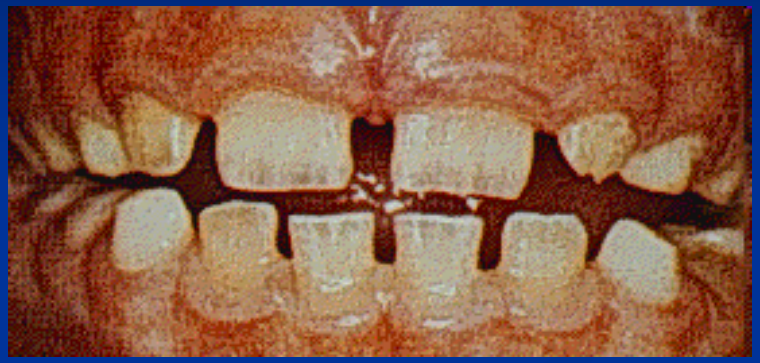
Gemination

Disturbances in structure of teeth

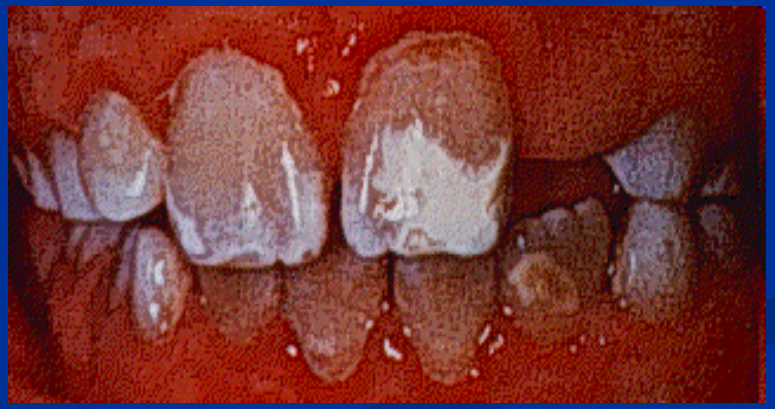
- Disturbances in structure of enamel
- Disturbances in structure of dentine
- Disturbances in structure of cementum

Amelogenesis
Secretory phase
Secretions of enamel matrix proteins by ameloblasts: amelogenin, enamelin, ameloblastin, tuftelin
Enamel matrix proteins – maturation iniciation
Crystallites growing mainly in length, little in width or thickness
Amount of matrix produced determines thickness of enamel and crown morphology
Maturation phase
Secretion of matrix protein ceases
Growth in length of crystallities is terminated
Secretion of proteolytic enzymes and degradation of matrix proteins
Crystallites growing in width and thickness

- **Defective matrix production – *enamel hypoplasia***



- **Defective maturation/mineralisation – *hypomineralized enamel***



Local causes of developmental abnormalities of enamel	
	Infection, trauma, radiotherapy. Idiopathic.
General causes	
Environmental/systemic causes (chronological dysplasias)	
Prenatal	Infections: rubeolla, syphilis,… Maternal diseases Excess fluoride ions
Neonatal	Hemolytic disease of newborn Hypocalcaemia Premature birth/prolonged labour.
Postnatal	Infections (viral exanthemata) Heart diseases, endocrinopathies, GIT diseases Avitaminosis (D) Chemotherapy Excess fluoride ions
Genetic causes	
Teeth affected	Amelogenesis imperfecta
+ generalized defects	Ectodermal dysplasia syndromes , Down syndrome

Genes encoding enamel proteins.

- Amelogenin
- Enamelin
- Ameloblastin
- Tuftelin

Amelogenesis imperfecta.

- **2 types:**

- **hypomineralized/hypomaturation type**

(normal tooth morphology when first erupt, soft chalky enamel easily lost, exposing dentine)

- **hypoplastic type**

(enamel of normal hardness, variable thickness)

- AD most often; rare XR (amelogenin)

Local causes of developmental abnormalities of dentine

Trauma, radiotherapy, Turner teeth (due to trauma/infection of primary teeth)

General causes of developmental abnormalities of dentine

Dentinogenesis imperfecta

Typ I	assoc. with osteogenesis imperfecta
Typ II	Teeth only affected, AD, both dentitions affected, discoloration (amber like), obliteration of pulp
Typ III	Racial isolate in USA, type II like

Dentinal dysplasia

Typ I	Radicular (rootless teet)
Typ II	Coronal

Environmental/systemic causes

Avitaminosis D

Hypophosphatemia

Hypophosphathasia

Juvenile hypoparathyreoidism

Other mineral deficiences, drugs, chemotherapeutics,…

- **Regional odontodysplasia („ghost teeth“)**
  - Unknown etiology
  - Abnormalities of enamel, dentin, pulp, dental follicle
  - Both dentition affected
  - Delayed eruption of abnormally formed tooth
  - Reduced radioopacity of the teeth with lost of distinction between enamel and dentine („ghostly“ appearance)

Disturbance in structure of cementum.

- Coronal third covered by a narrow layer of acellular (primary) cementum
- apical 2/3 covered by an additional thicker layer of cellular (secundary) cementum

- **Hypercementosis**

- Idiopathic or known causes
- Ancylosis, concrescence
- causes: periapical inflammation, mechanic stimulation, functionless/unerupted tooth, Paget´s disease of bone

- **Hypocementosis**

- In hypophosphatasia, in cleidocranial dysplasia,…..

Causes of macroglossia
Congenital and hereditary
Vascular malformations
Hemihyperplasia
Cretenism
Beckwith-Wiedemann syndrome (omphalocele, visceromegaly, gigantism, hypoglycemia)
Down syndrome
Mucopolysaccharidoses
Neurofibromatosis
Multiple endocrine neoplasia, type 2B
Acquired
Edentulous patients
Amyloidosis
Myxedema
Acromegaly
Angioedema
Tumors

- **Microglossia**
- **Aglossia**

(in oromandibular-limb hypogenesis syndrome)

- **Ankyloglossia (tongue-tie)**

(short, thick lingual frenum)

Thanks for your attention……..