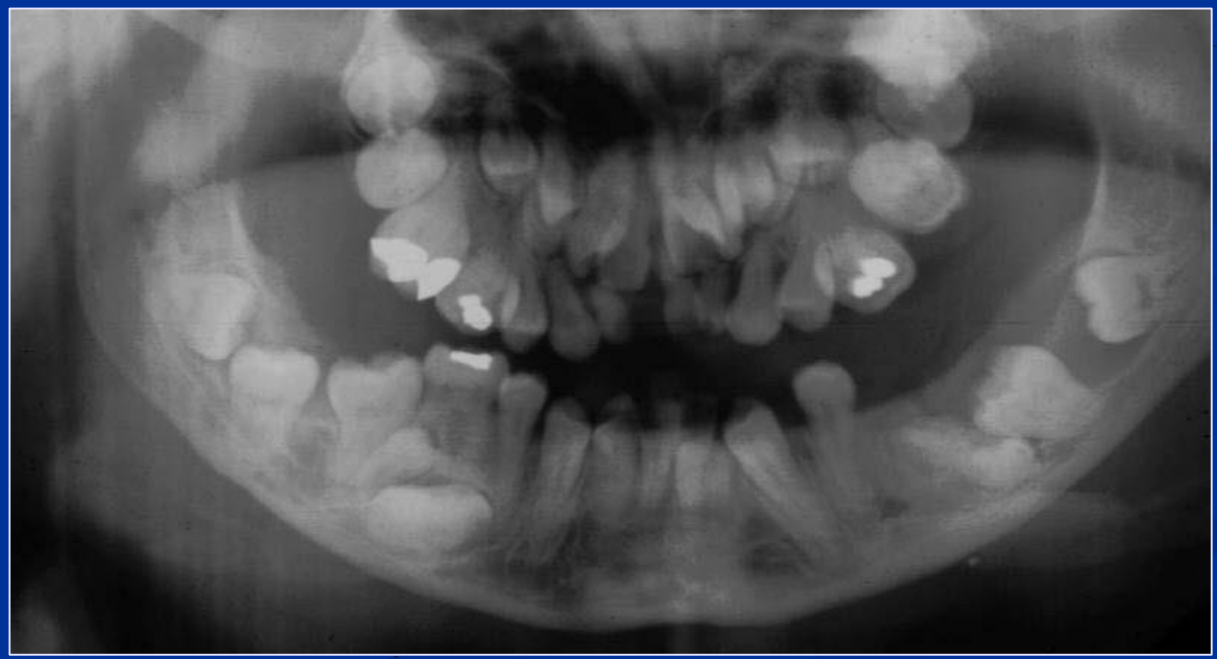
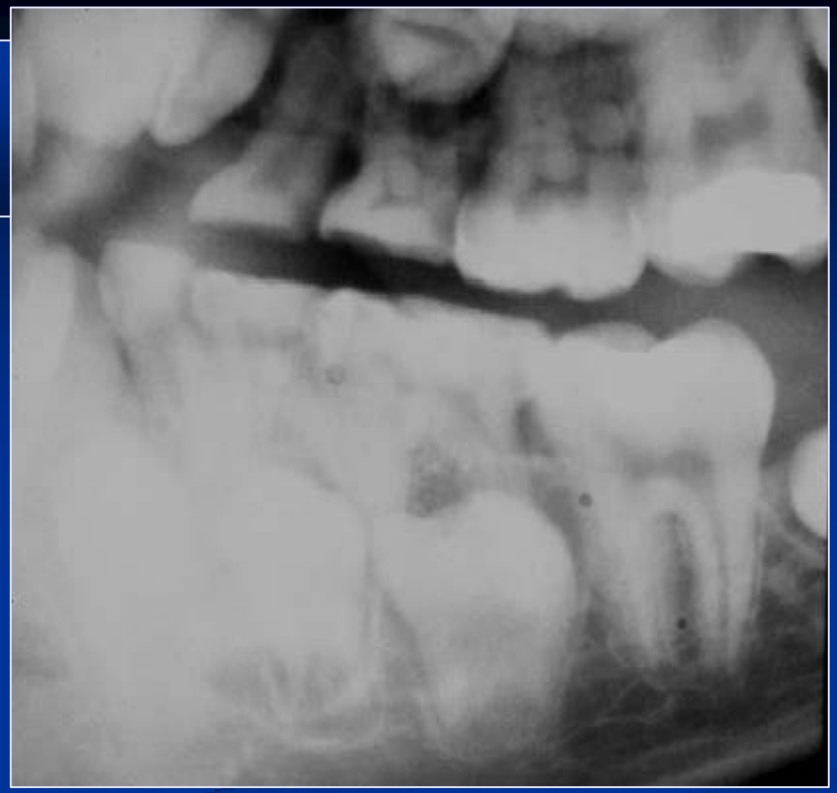
DISEASES OF BONE AFFECTING ORAL CAVITY.

DEPARTMENT OF ORAL AND MAXILLOFACIAL PATHOLOGY &
ORAL MICROBIOLOGY

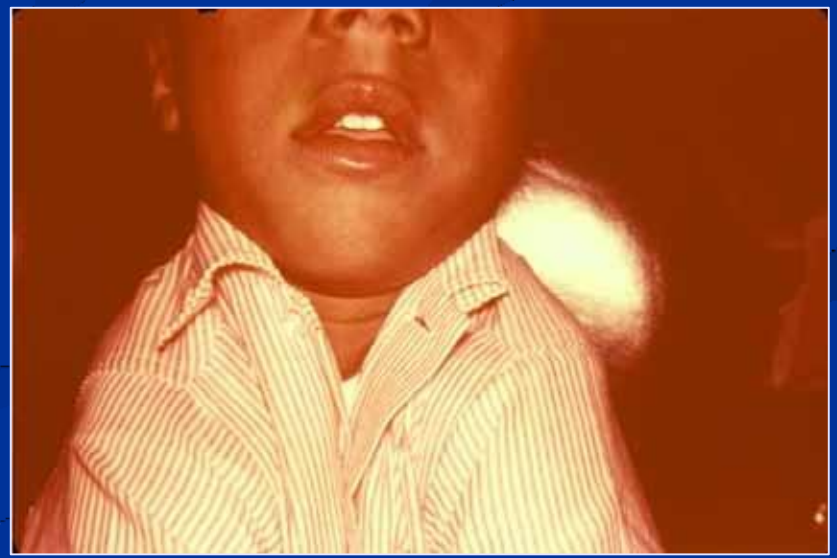
Cleidocranial Dysplasia



Cleidocranial Dysplasia
Cleidocranial Dysostosis

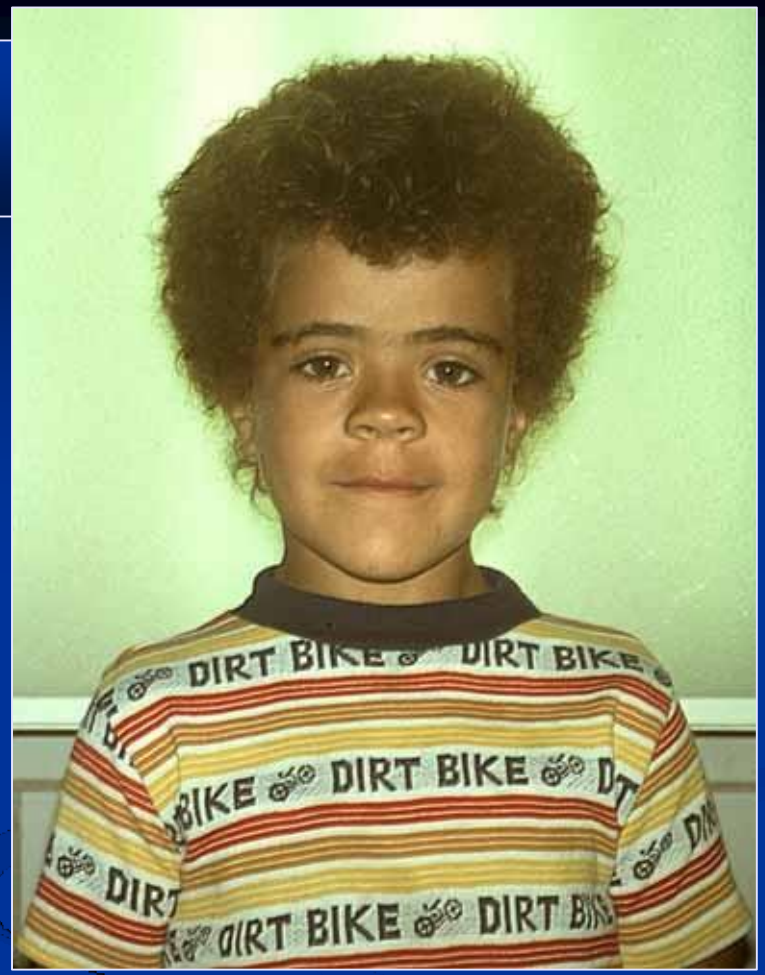


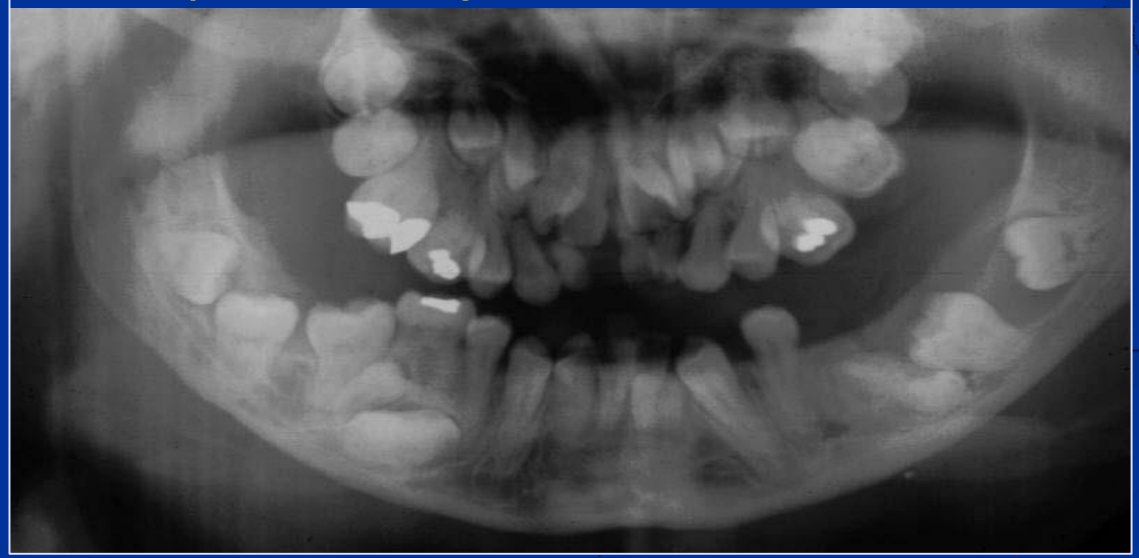
- Missing clavicles
- Supernumerary teeth
- Defective CBFA1
  - -- Chromosome 21
  - -- Guides osteoblastic differentiation
  - -- Guides appropriate bone formation
- AD inheritance
  - -- 40% spontaneous mutations
- GENDER,AGE,LOCATION PREDILECTION:
  - -- Gender:M=F
  - -- Childhood and teenage years
  - -- Clavicles, jaws
  - -- Rare

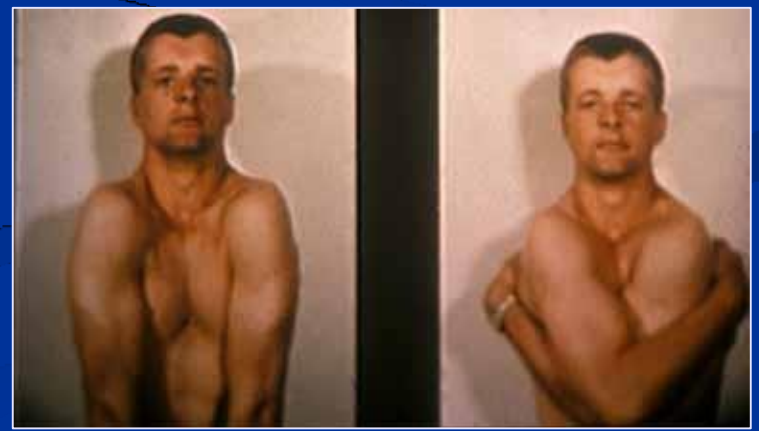


Cleidocranial Dysplasia
Clinical Features

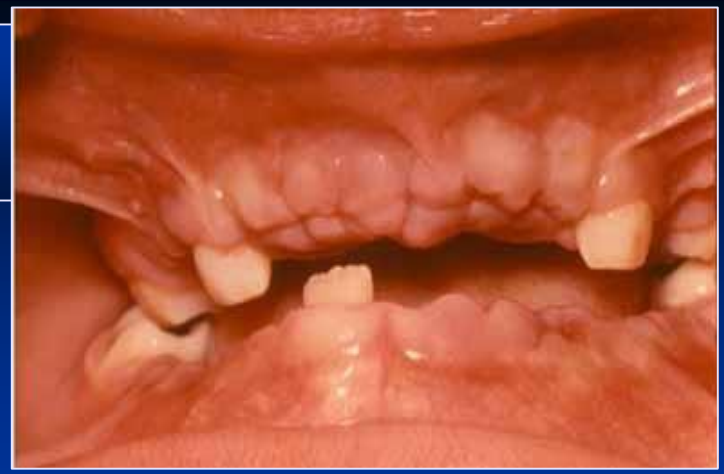
- Missing or partially missing clavicles
- Drooping shoulders
  -- Maybe can touch shoulders together
- Frontal and parietal bossing
- Delayed closure of skull sutures
  -- Wormian bones
- Prolonged retention of deciduous teeth.
- Underdeveloped maxilla and high arch palate.
- Supernumerary teeth





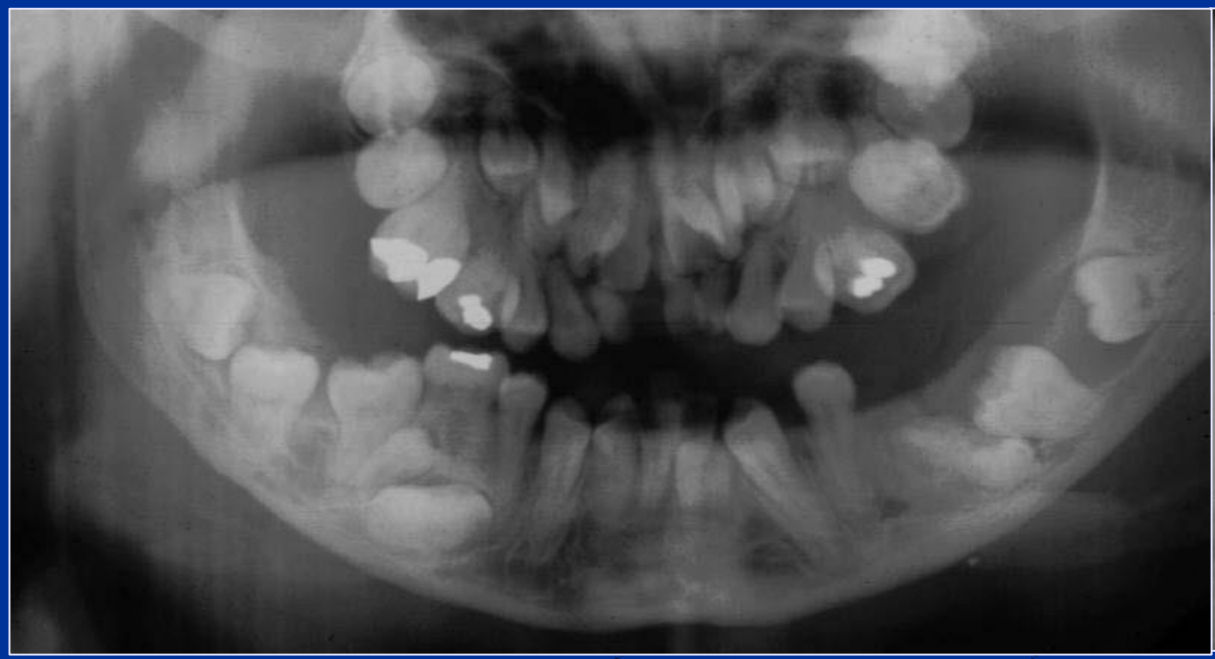


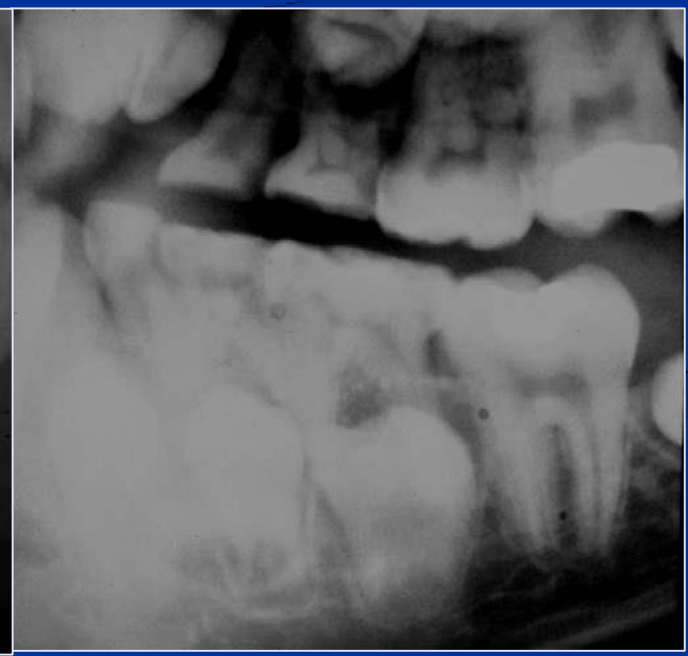
Cleidocranial Dysplasia
Histopathology, Treatment



- Micro: Bone looks normal
- Permanent teeth lack secondary cementum

- Treat: Surgical exposure and orthodontic repositioning of teeth
- Extraction of teeth with denture construction

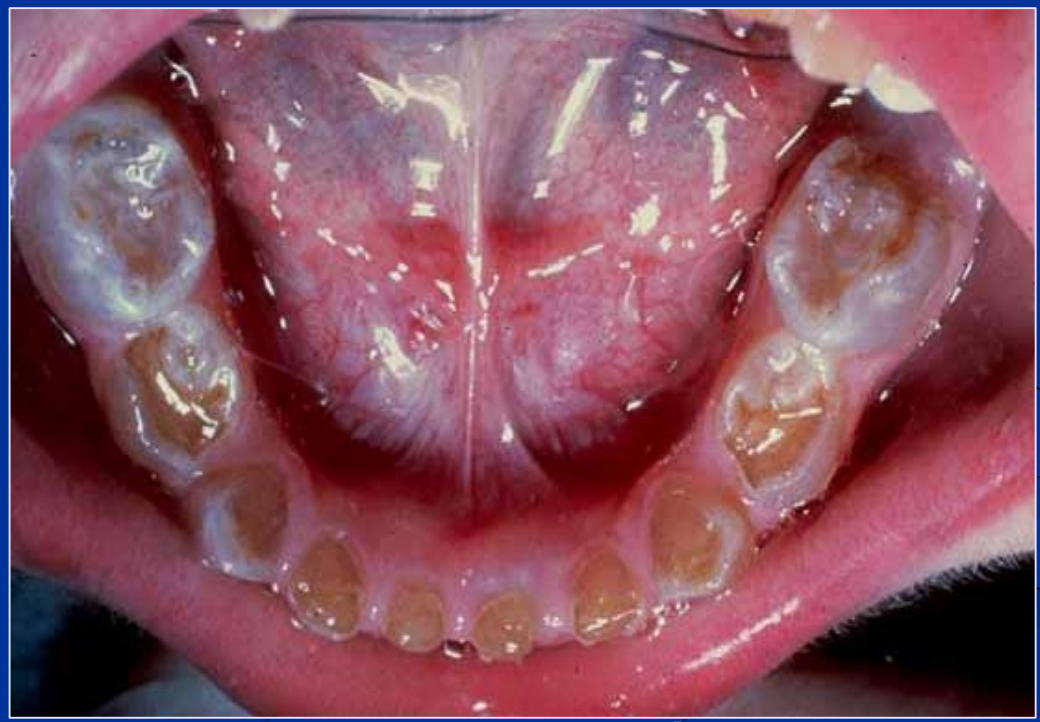




Atypical Bone Production

Name	Abnormality
Osteogenesis imperfecta	Abnormal collagen production
Osteopetrosis	Lack of osteoclastic activity
Fibrous dysplasia of bone	Marrow fibrosis, then excess bone
Cherubism	Marrow fibrosis, giant cells
Paget's disease of bone	Excess osteoblastic activity
Cemento-osseous dysplasias	Marrow fibrosis, excess bone, cementum

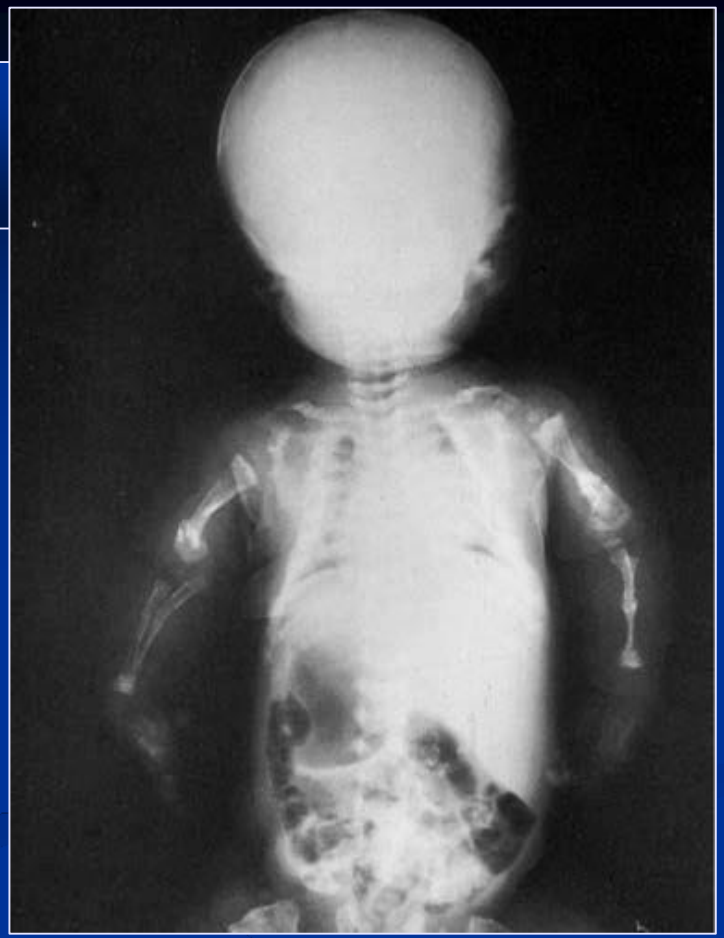
Osteogenesis Imperfecta

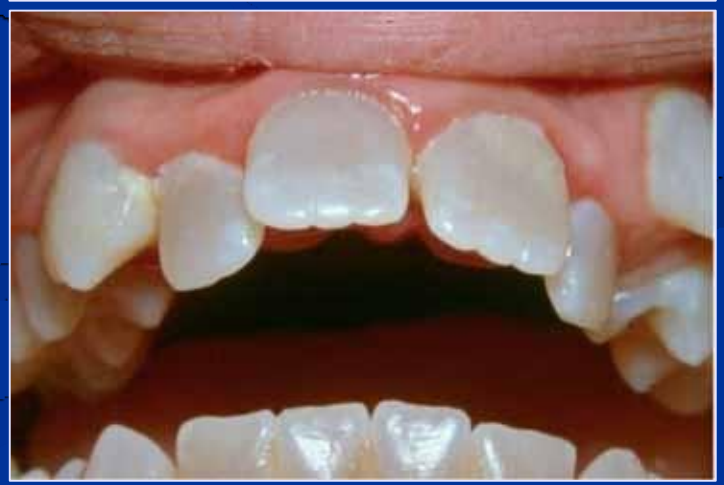


Osteogenesis Imperfecta
Brittle Bone Disease

- Mutation in genes for type I collagen
- AD inheritance, some AR

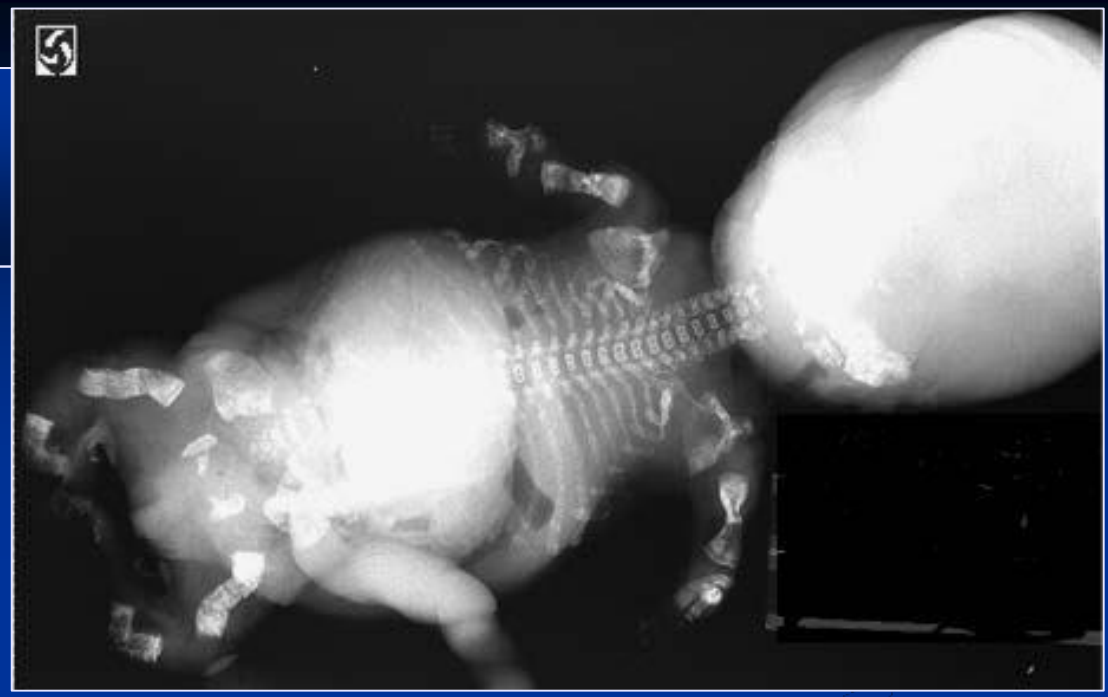
- GENDER,AGE,LOCATION PREDILECTION:
  -- None
  -- Infant and young child
  -- Bone, teeth (opalescent dentin)
  -- 1/8,000 births

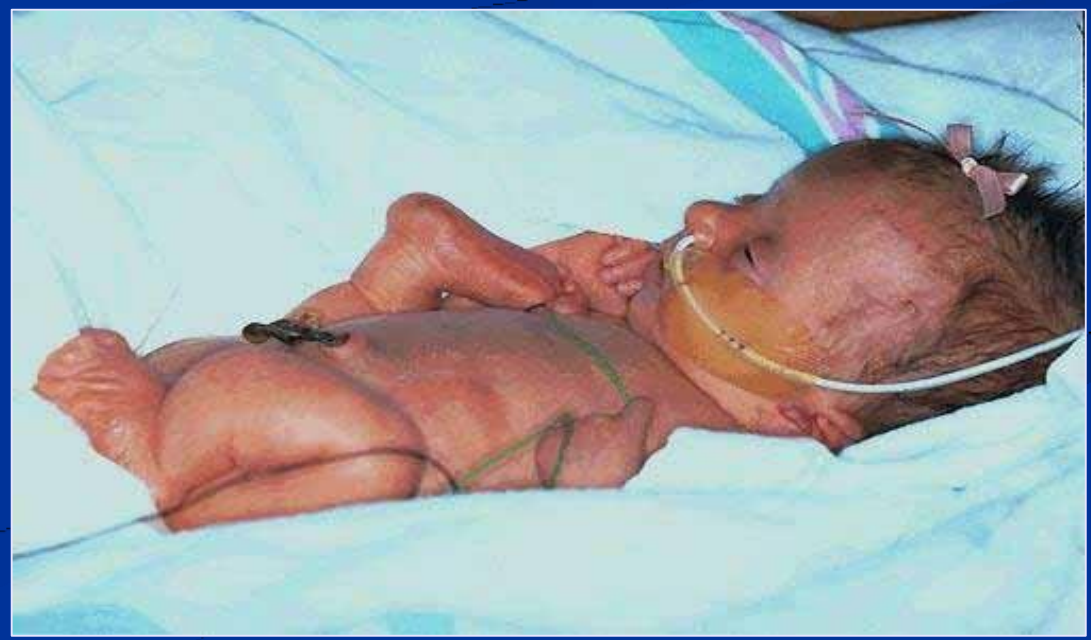




Osteogenesis Imperfecta

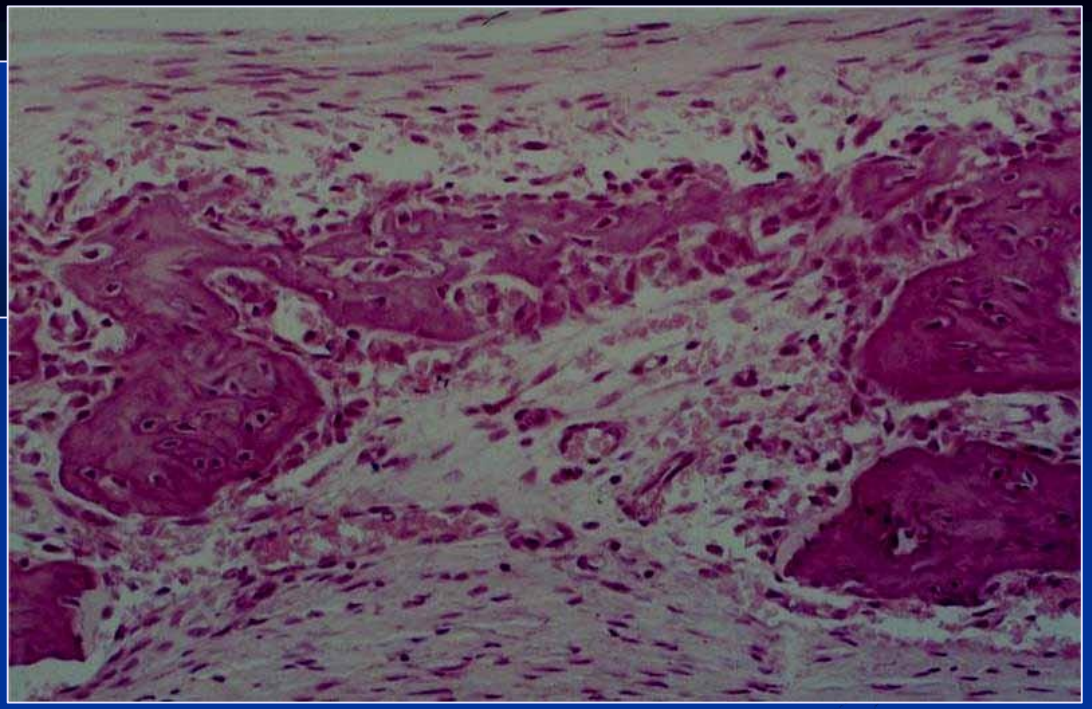
- Fragile & porous bones
- Deformity of long bones
- Pathologic fracture
- Hypermobile joints
  -- “Double-jointed”
- Capillary fragility
  -- Pathologic bleeds
- Osteopenia

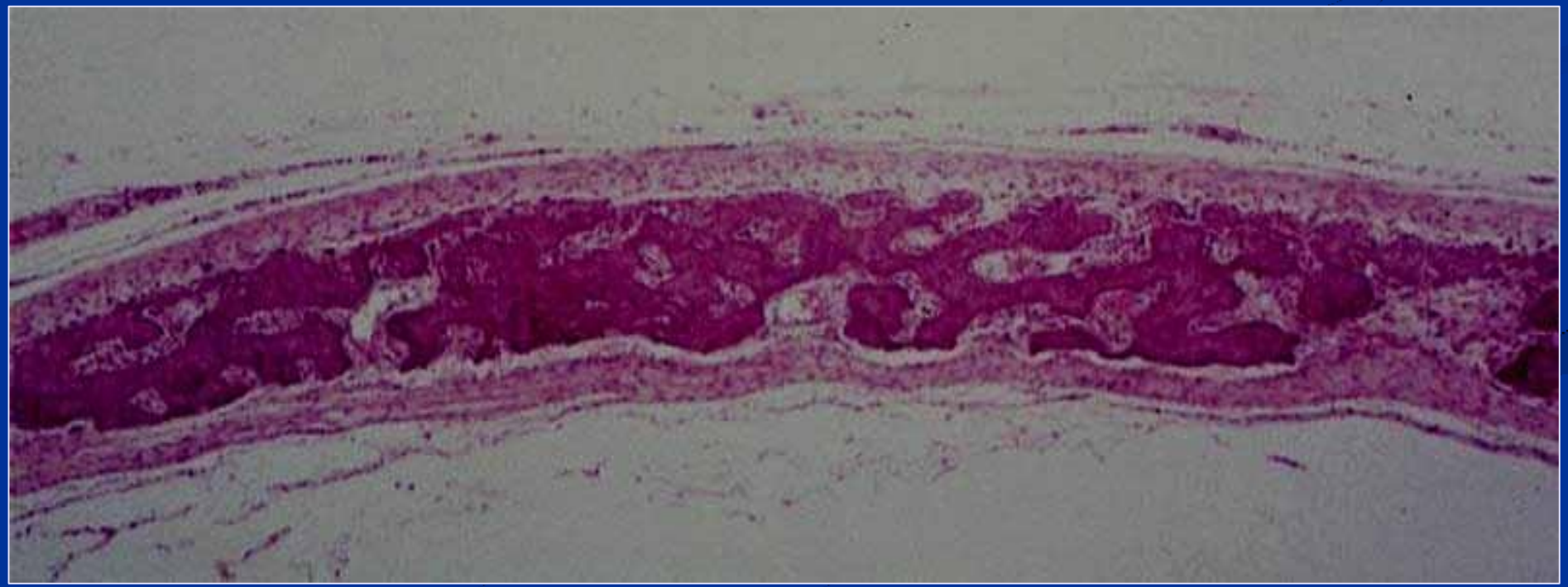




Osteogenesis Imperfecta
Histopathology

- Immature, irregular trabeculae
- Fibrous background
- Diminished amount of marrow
- Teeth have small pulps
- Shell teeth have large pulps

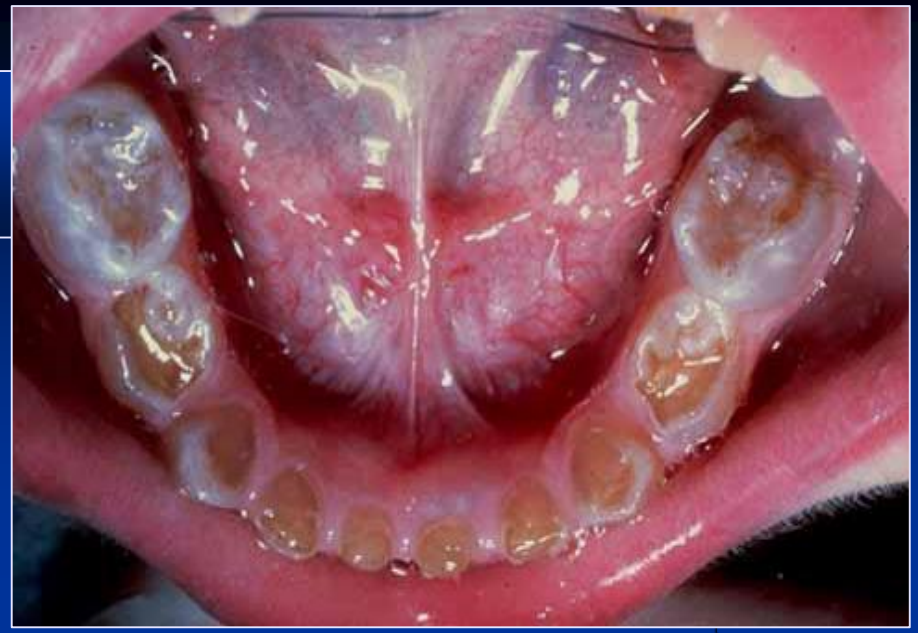


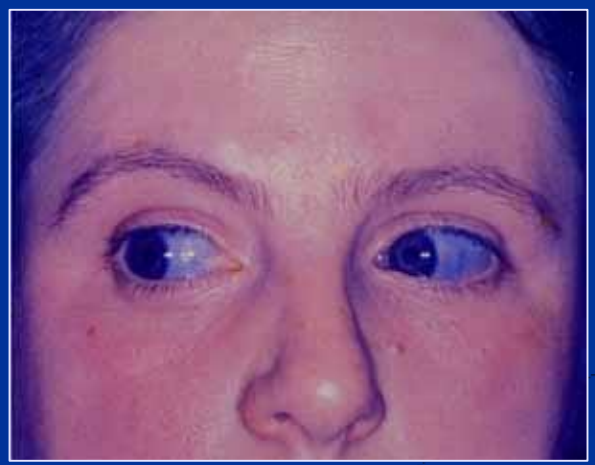


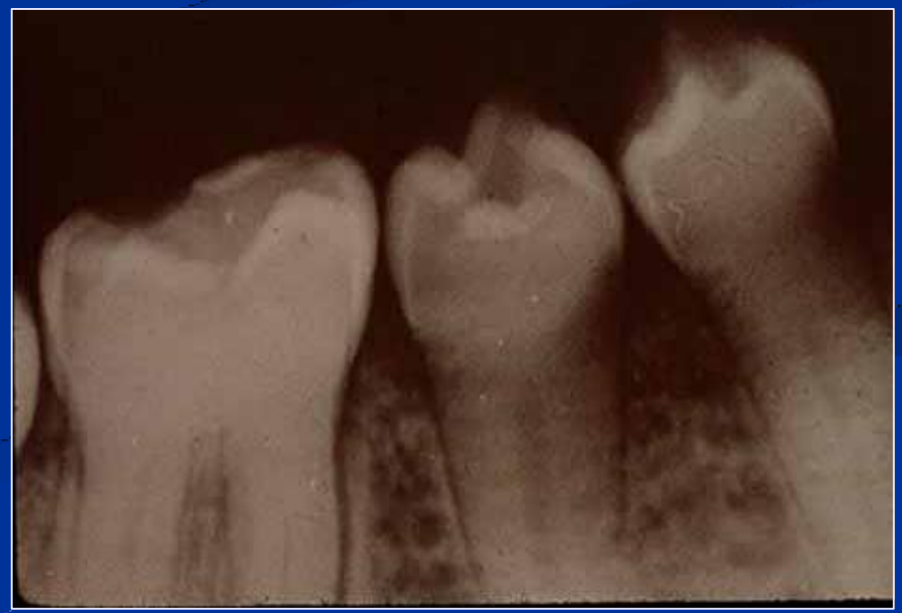
Osteogenesis Imperfecta

Clinical Features – Head & Neck

- Hearing deficits
- Wormian skull
- Blue sclera
- Maxillary hypoplasia
- Rarely: mixed radiolucent/opaque areas of jaws
- Blue/gray “translucent” teeth
  - -- Opalescent dentin
  - -- Like dentinogenesis imperfecta
  - -- Maybe shell teeth
  - -- Fracture of crown

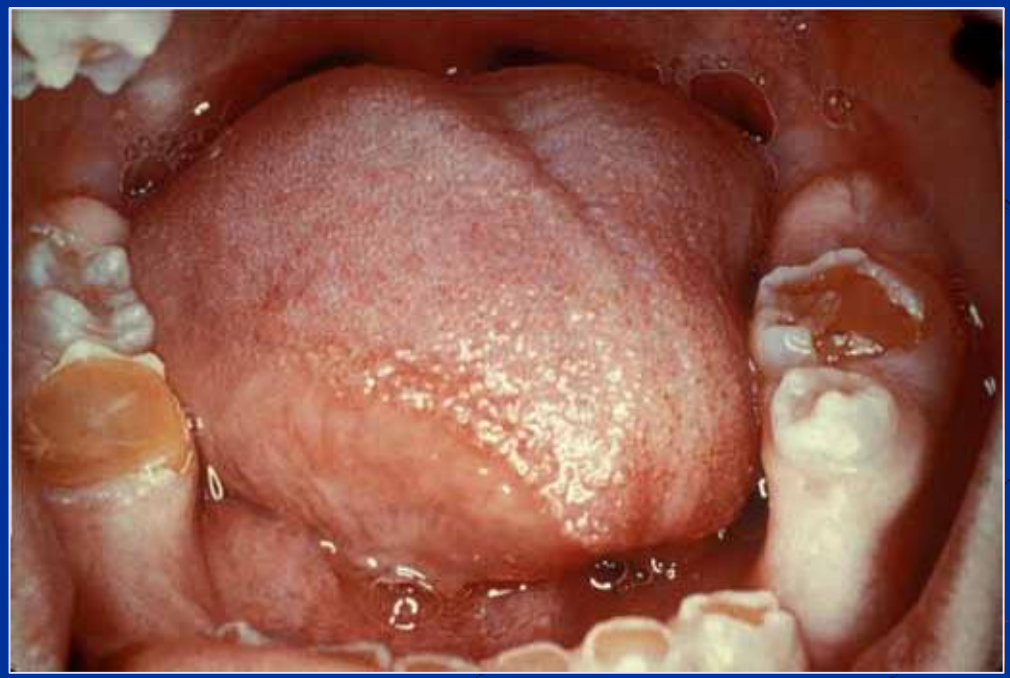


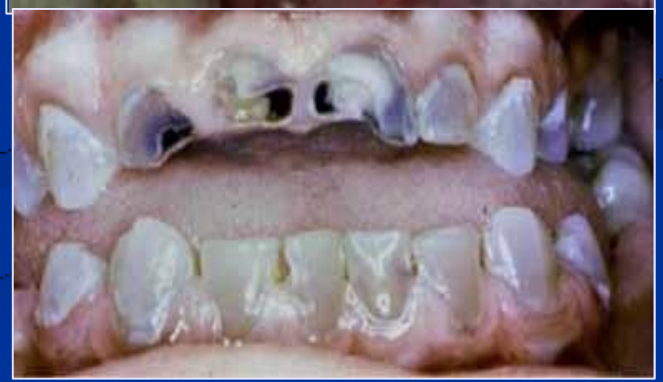




Osteogenesis Imperfecta
Pathophysiology, Treatment

- Risk of broken bones
- Anemia (less marrow)
- Tooth fractures
- Treat: Cautious behavior to prevent fracture
- C-section for birth
- Shorten dental crowns
- Overdenture

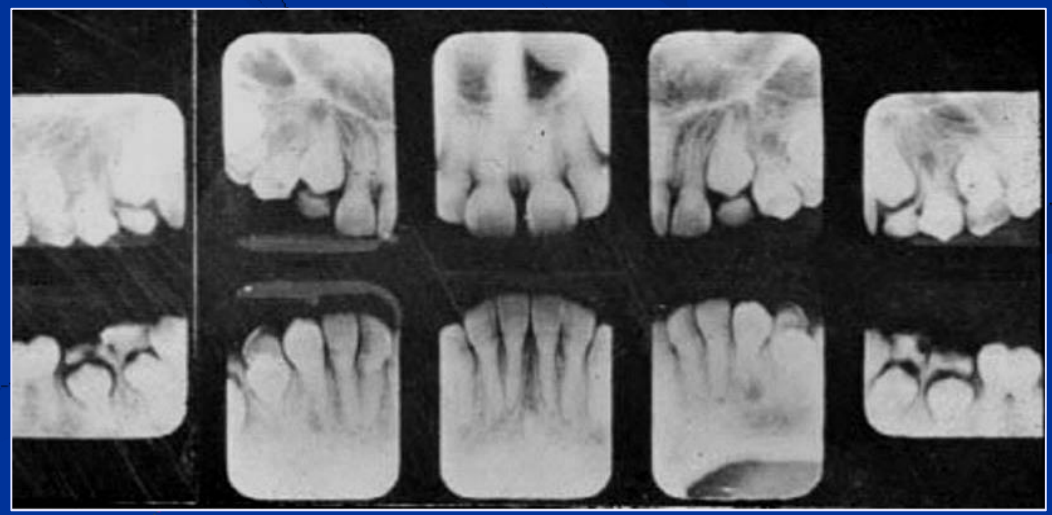




Osteogenesis Imperfecta
Types I - II

- Type I:
  - -- Most common
  - -- Mildest form
  - -- Blue sclera throughout life
  - -- AD inheritance

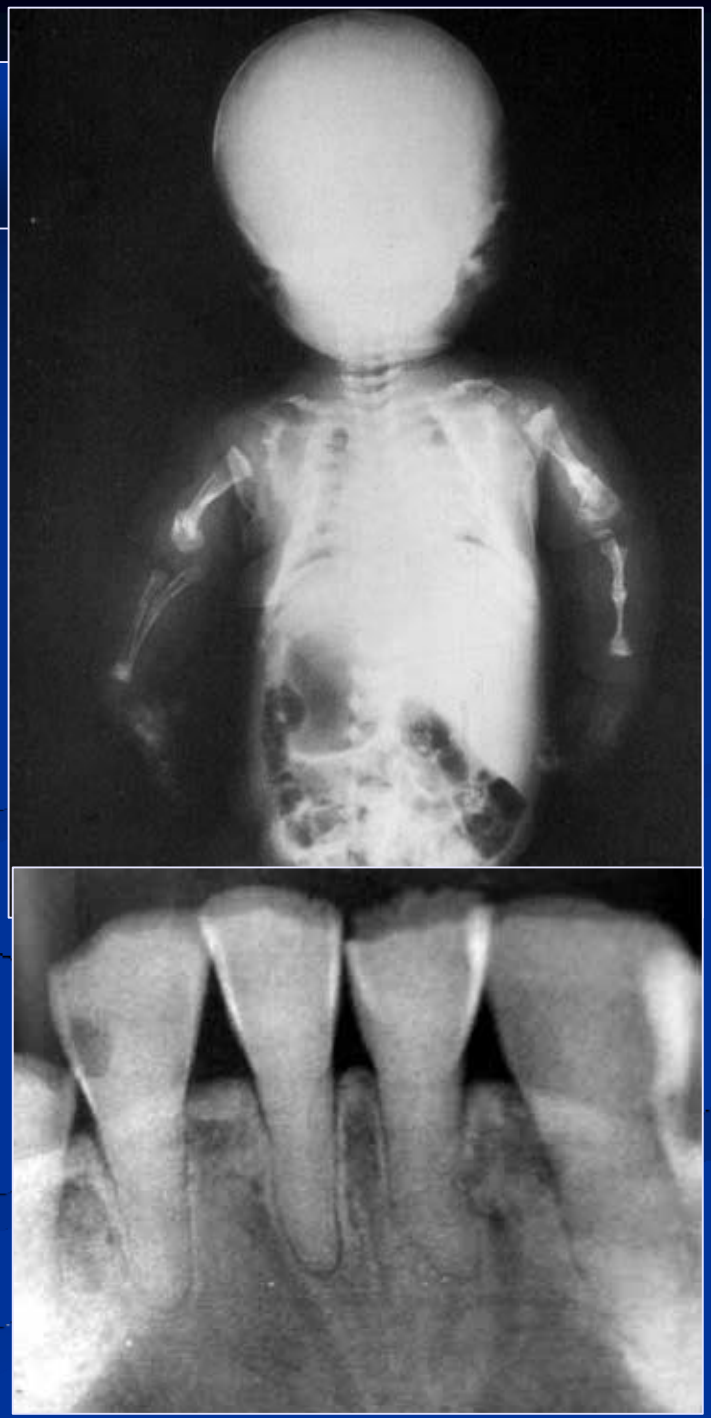
- Type II:
  - -- Most severe
  - -- 90% are stillborn or die shortly after birth
  - -- Both AD & AR inheritance is seen



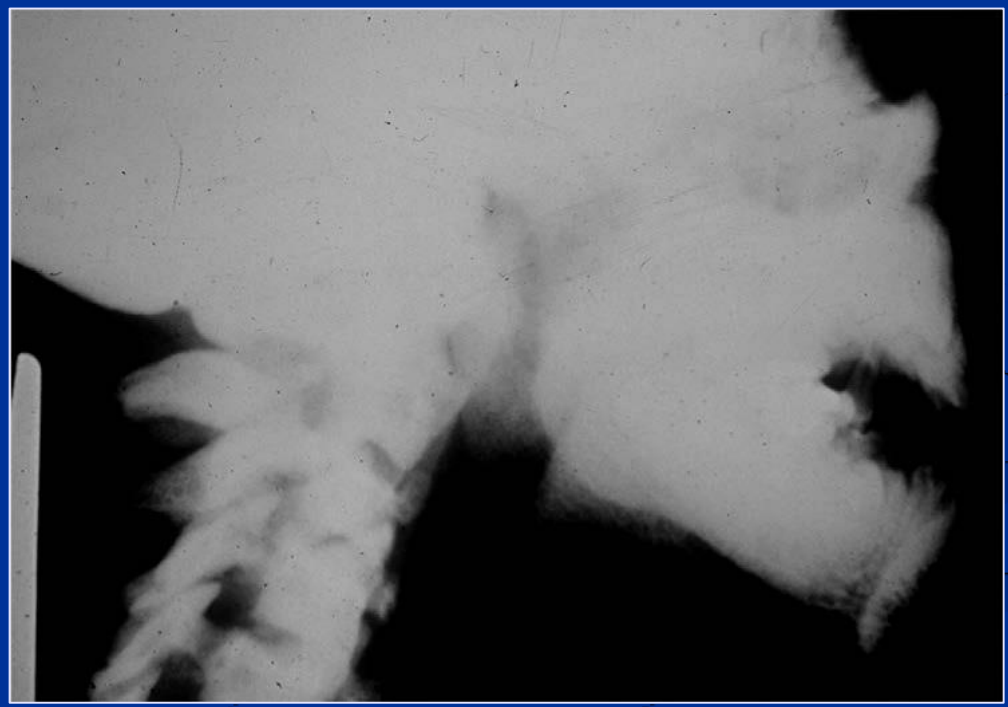
Osteogenesis Imperfecta
Types III - IV

- Type III:
  - -- Second most severe form
  - -- Usually noticed after 6 months
  - -- Sclera often normal
  - -- Most die before adulthood (usually from cardiopulmonary problems from kyphoscoliosis)
  - -- Both AD & AR inheritance

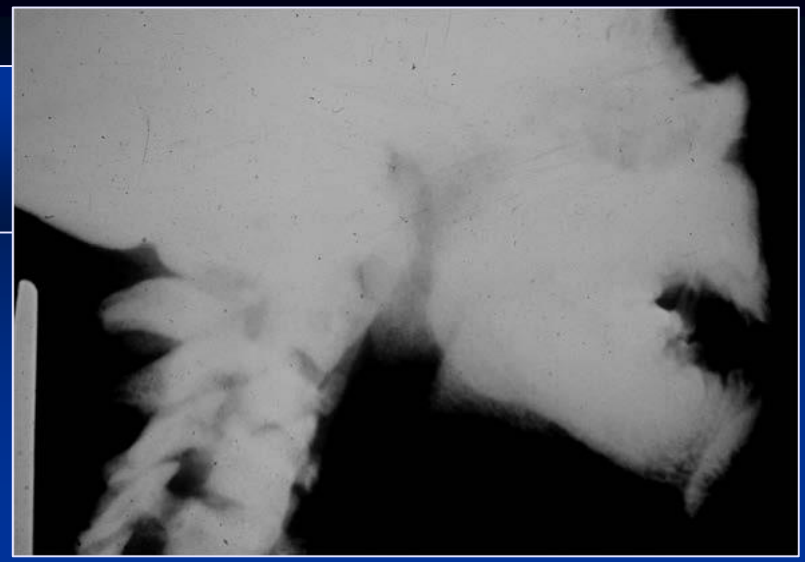
- Type IV:
  - -- Fractures in 50% at birth
  - -- Blue sclera usually fades with time
  - -- AD inheritance



Osteopetrosis

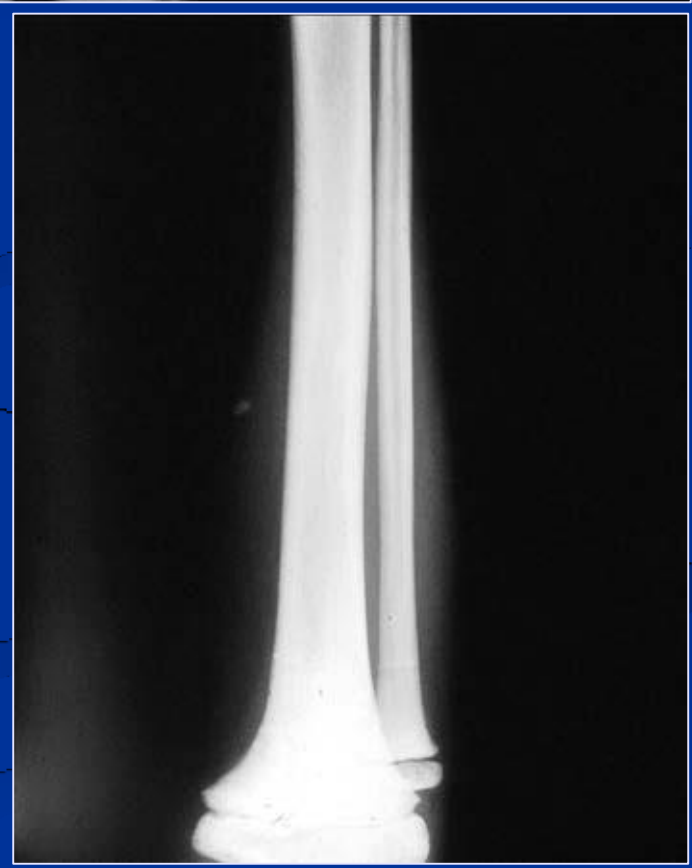


Osteopetrosis
Albers-Schönberg Syndrome



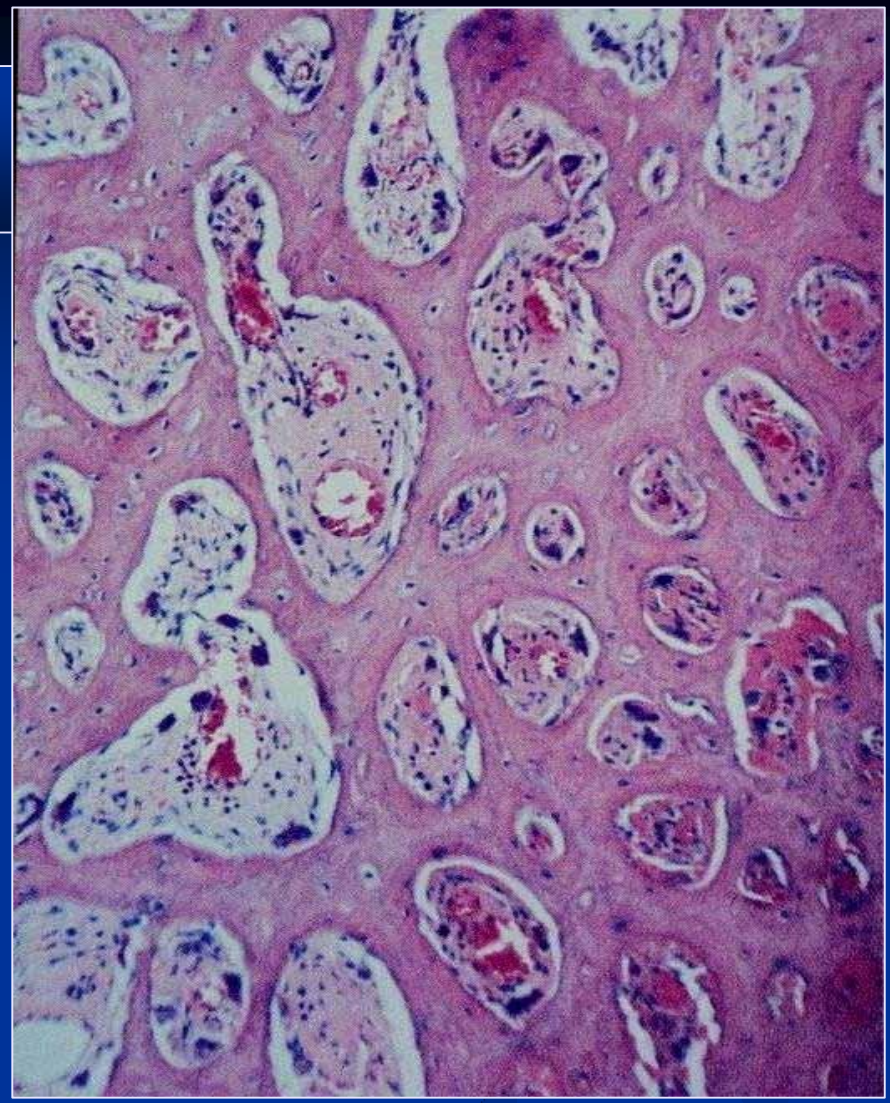
- Inherited failure of osteoclastic function
  -- AD or AR
- Number of osteoclasts is normal or high
  -- But no bone resorption
  -- Gradual thickening of trabeculae/cortex
  -- Gradual sclerosis of bone

- GENDER,AGE,LOCATION PREDILECTION:
  -- None
  -- Infancy (except adult onset type)
  -- None
  -- Prevalence rate: 1/100,000 persons

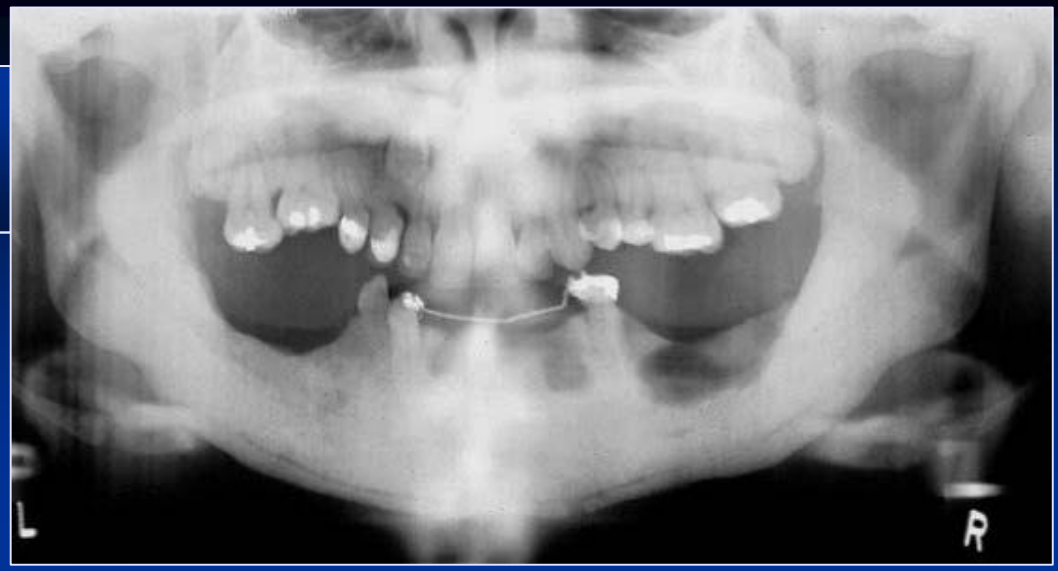


Osteopetrosis
Histopathology

- Thick lamellar bone
- Forms concentric rings as it fills marrow spaces
- Thick cortex & trabeculae
- Small marrow spaces, fibrosed
- Osteoclasts & osteoblasts
- Inflammation if also has osteomyelitis

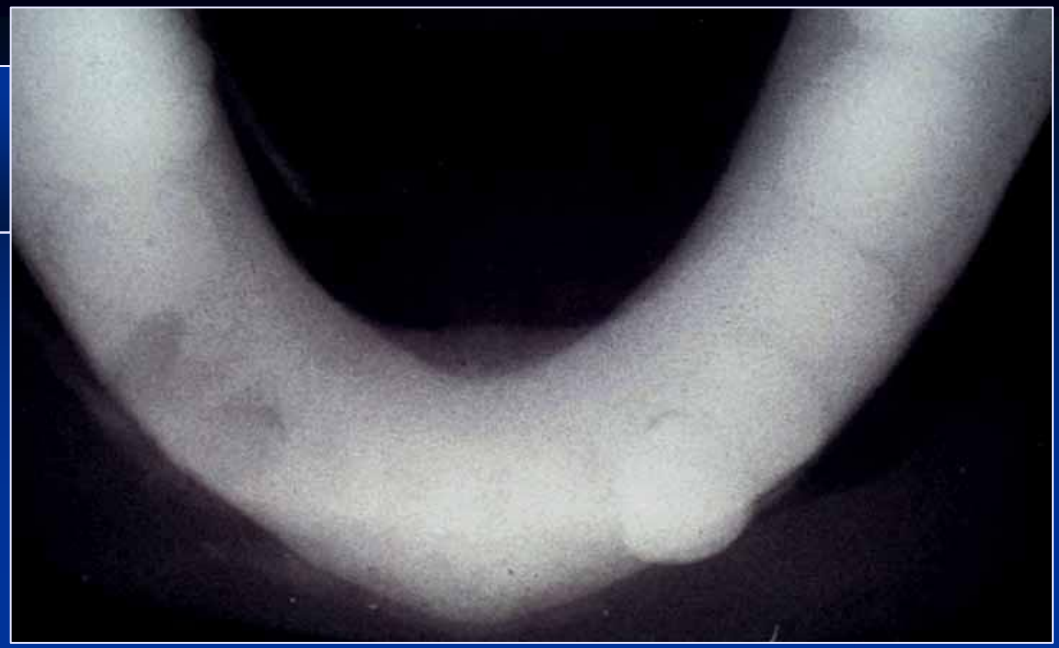


Osteopetrosis
Clinical Features



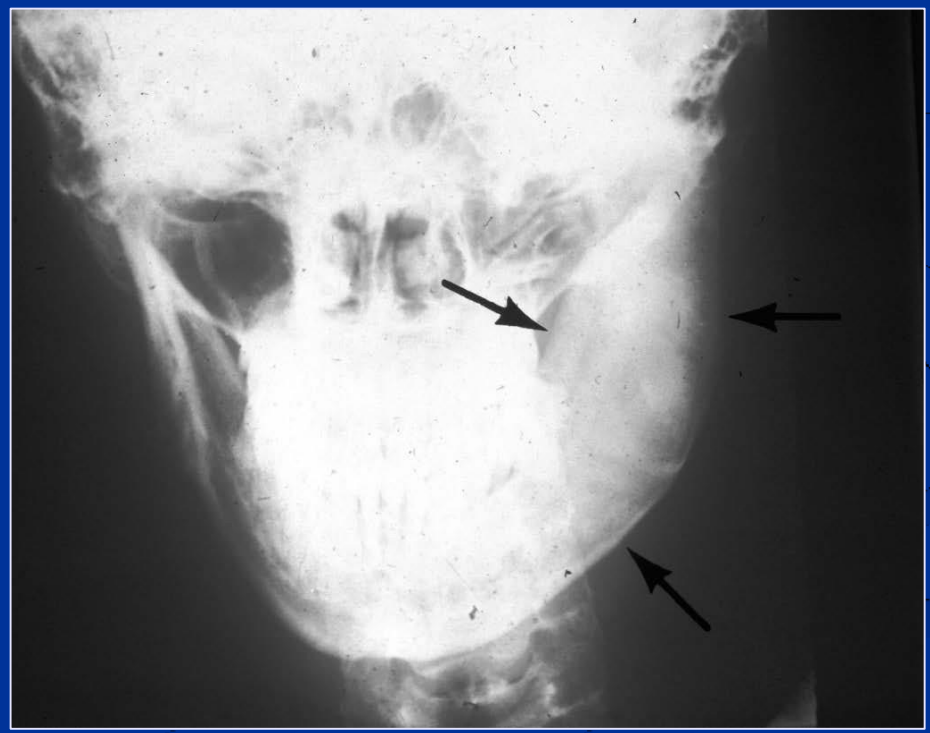
- Slow opacification
- Anemia
  -- Spaces fill with bone
- Reduced immune function
  -- Fewer hematopoietic cells
  in small marrow spaces
- Decreased blood flow to bone (ischemia)
  -- Maybe painful
- Pathologic fractures
- Often osteomyelitis, fails to heal
  -- Primarily a jaw problem
- Crimping of nerves in foramina:
  -- Hearing loss
  -- Vision loss
  -- Facial palsy
- Delayed/stopped tooth eruption

Osteopetrosis



- Infantile osteopetrosis
  - -- Malignant osteopetrosis
  - -- Severe
    (usually die in first decade)
  - -- Most common form
  - -- AR inheritance
- Adult osteopetrosis
  - -- Benign osteopetrosis
  - -- Not too severe
  - -- Usually no marrow deficit
  - -- Bone pain is common
  - -- AD inheritance
- Treat:
  - -- Antibiotics to counter fewer hematopoietic cells
  - -- Treat anemias if possible
  - -- Try to prevent dental infection
  - -- Hyperbaric therapy

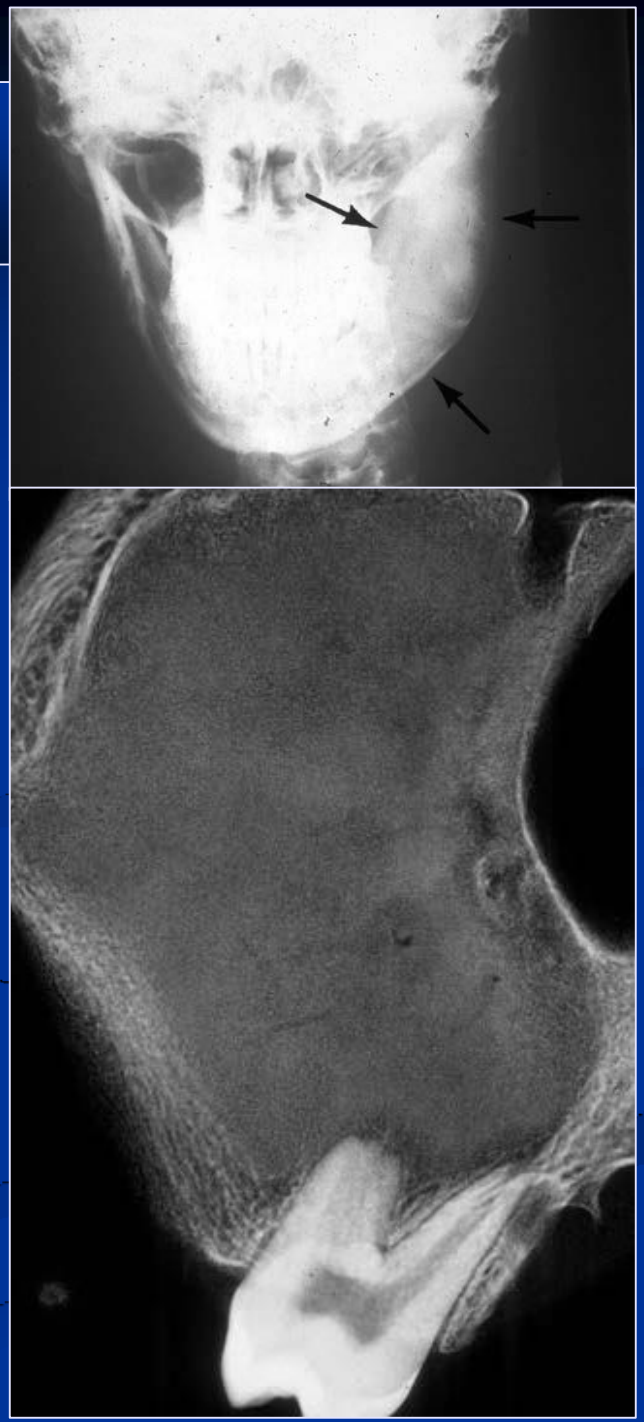
Fibrous Dysplasia of Bone



Fibrous Dysplasia of Bone

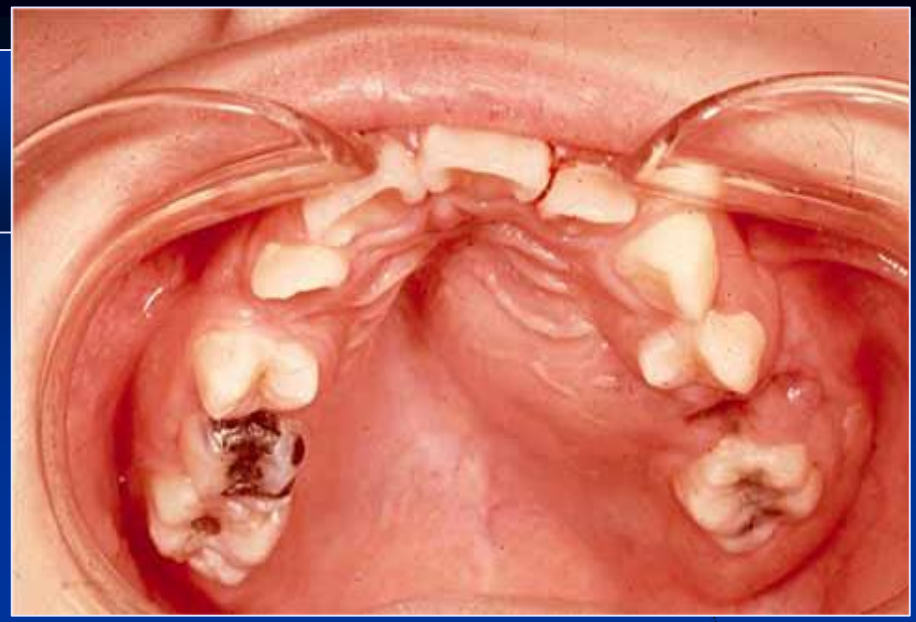
- Developmental anomaly
- *Postzygotic* mutation of GNAS 1 gene (guanine nucleotide-binding protein, alpha-stimulating activity polypeptide 1)
  -- Explains regional nature of disease

- GENDER,AGE,LOCATION PREDILECTION:
  --None
  -- 7-20 years of age
  -- Posterior maxilla
  -- 85% of cases
  -- Jaws: among most common sites

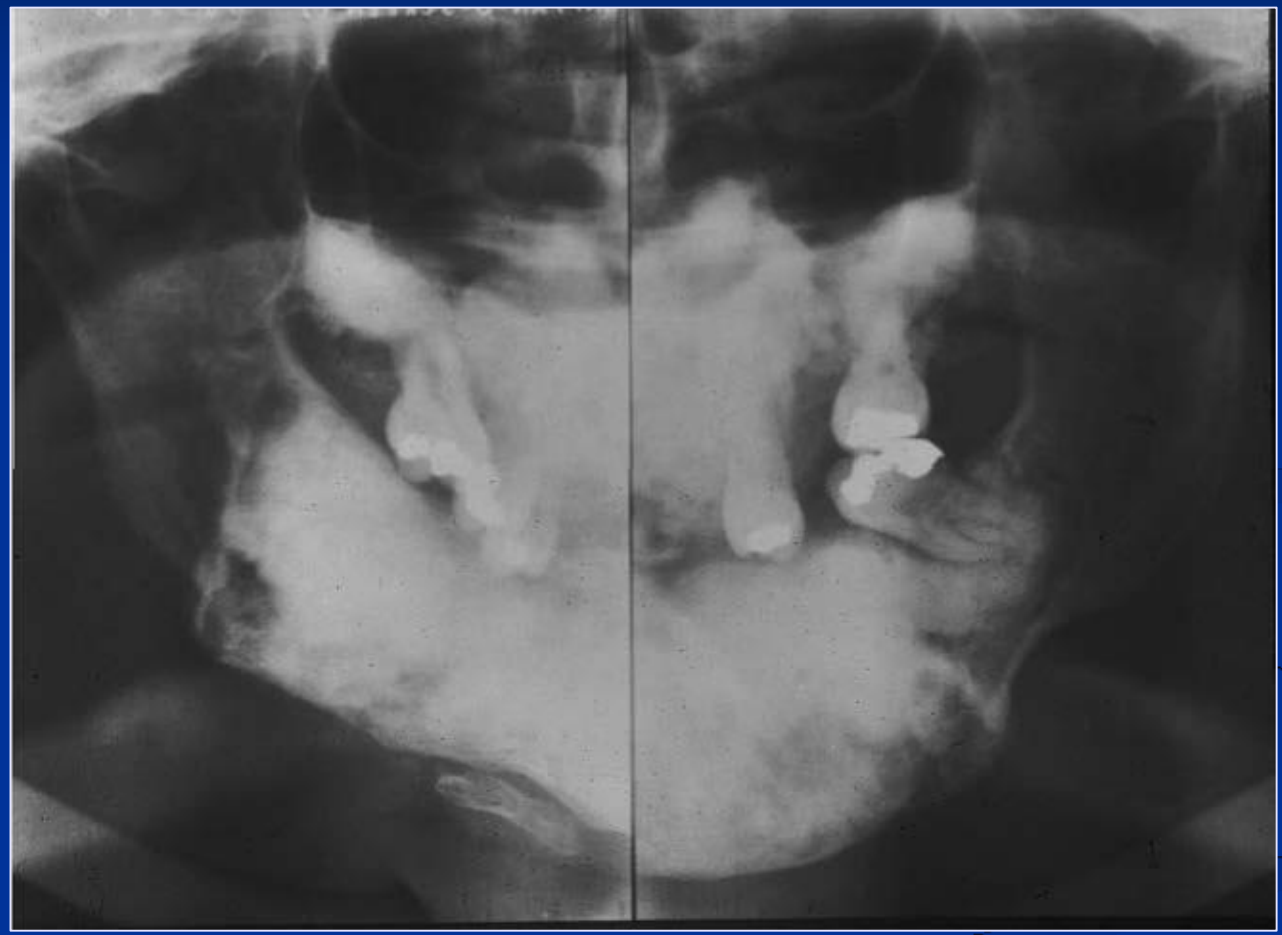


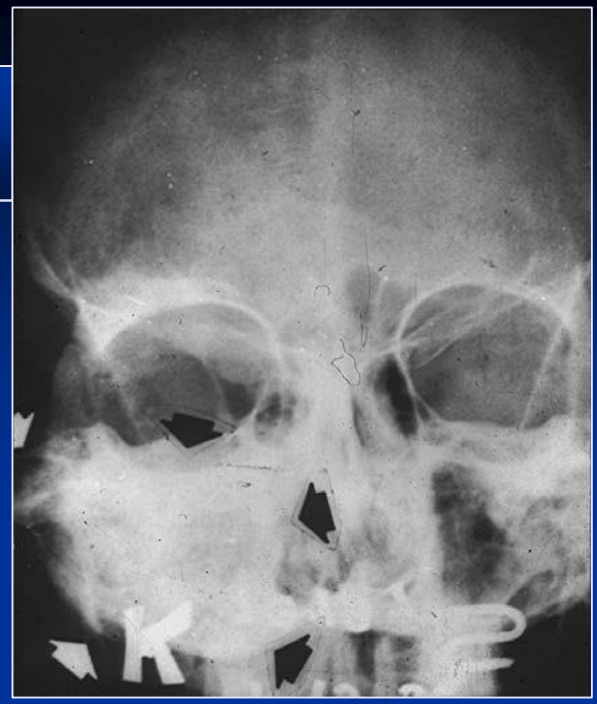
Fibrous Dysplasia of Bone
Clinical Features

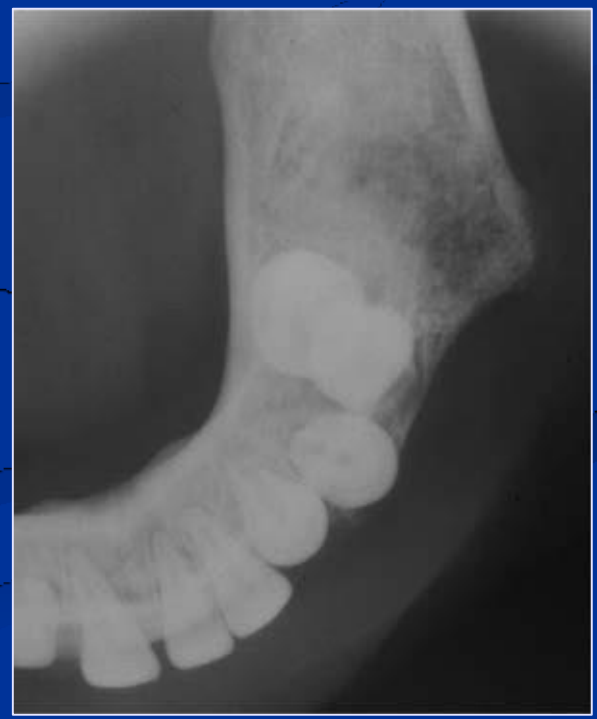
- Painless bony enlargement, diffuse
  -- Cortical expansion and thinning
- Ground-glass radiopacity
  -- Or irregular opacities
  -- Or mixed lucent/opaque
- Poorly demarcated
- Lamina dura:
  -- May be hard to see
- PDL may be thin
- Teeth become separated
  -- Remain viable
- Growth is slow



Fibrous Dysplasia of Bone



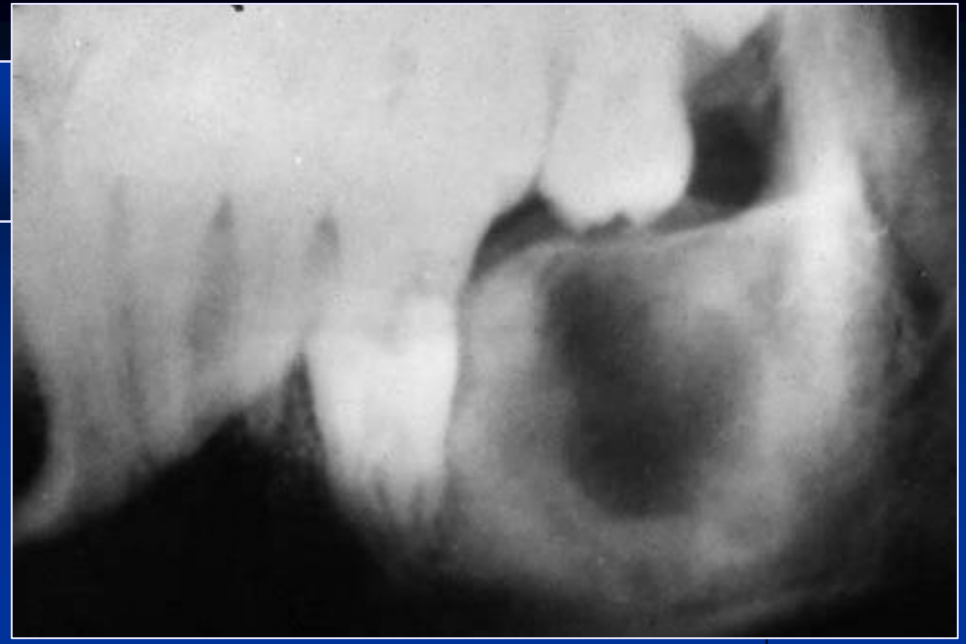


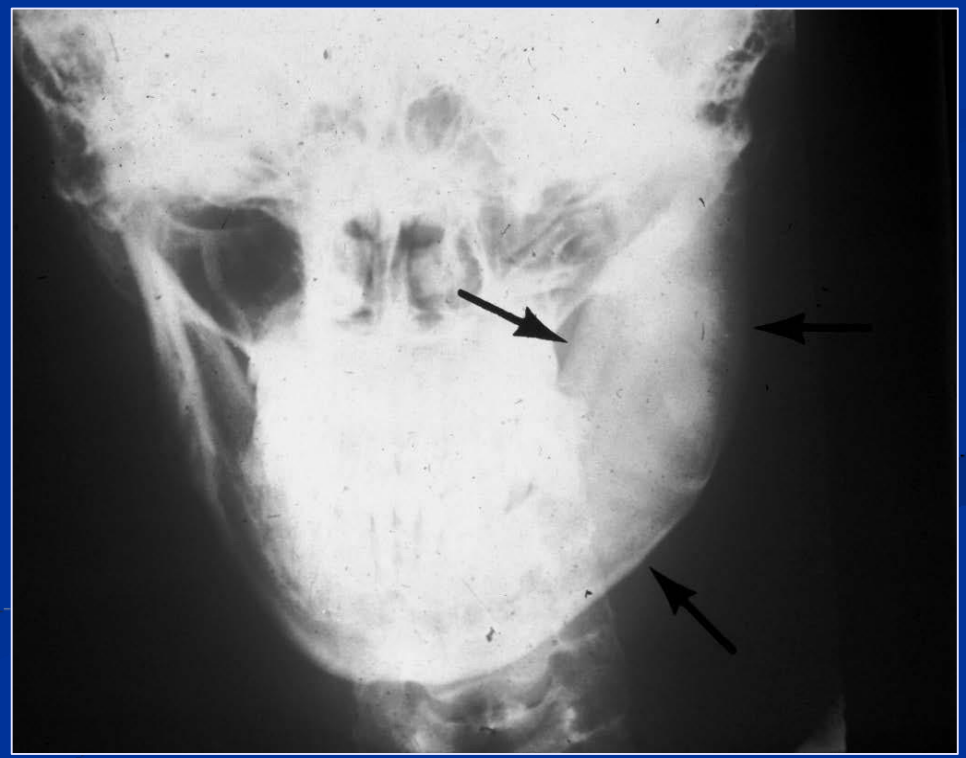


Fibrous Dysplasia
Clinical Variants

- Monostotic fibrous dysplasia
  - -- One bone involved
  - -- 85% of cases
  - -- Jaws: among most common sites

- Polyostotic fibrous dysplasia
  - -- Multiple bones involved
  - -- Jaffe-Lichtenstein syndrome
  - -- McCune-Albright syndrome

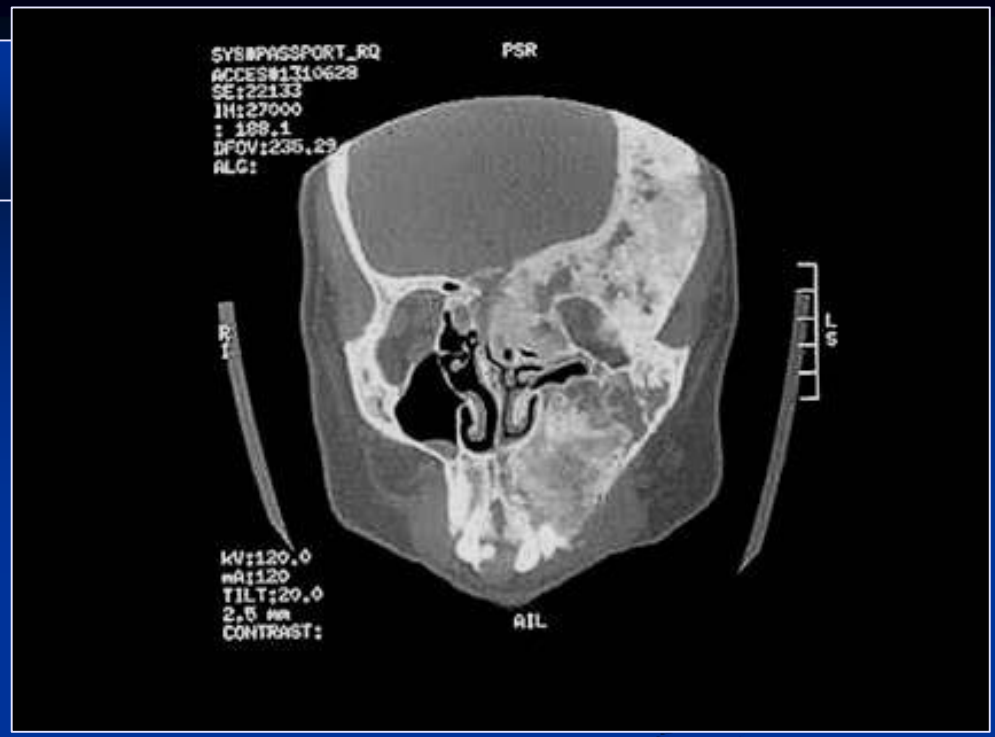


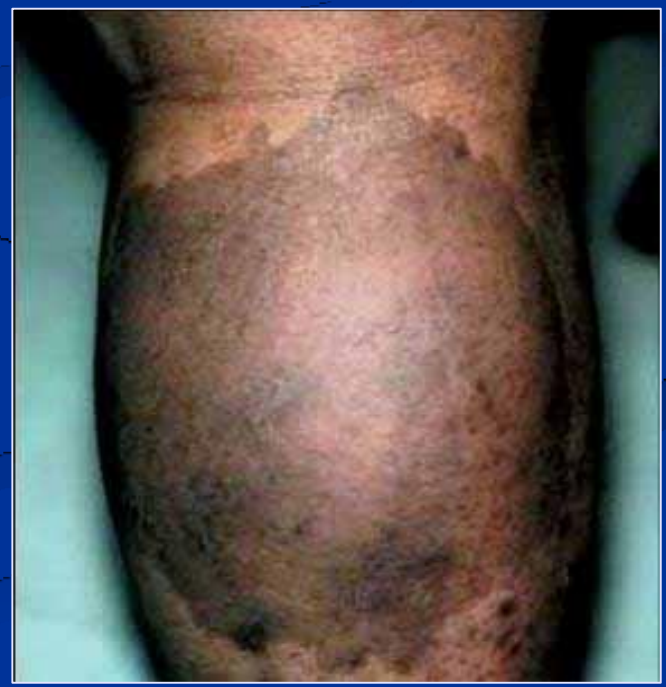


Fibrous Dysplasia
Clinical Variants

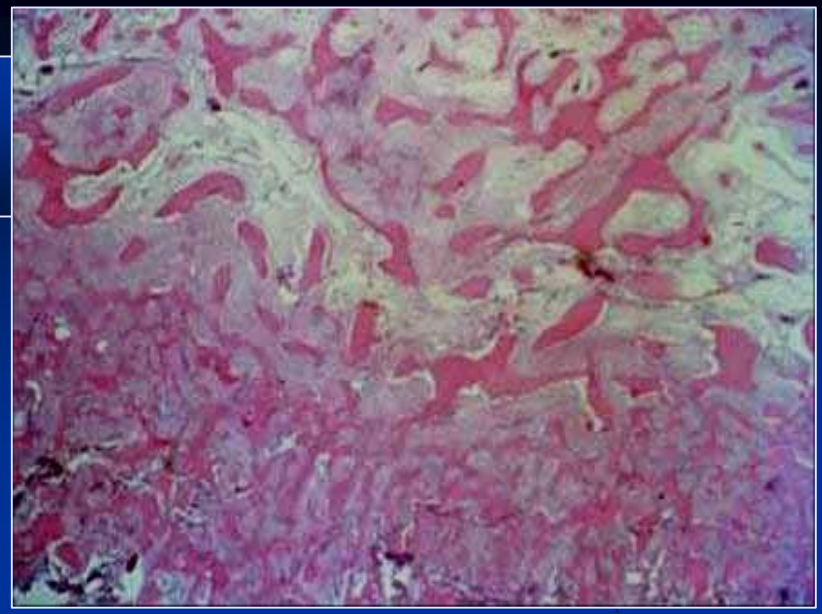
- McCune-Albright syndrome:
  - -- Relatively uncommon
  - -- May involve most of skeleton
  - -- Café au lait spots
  - -- Hockey stick deformity of hip

- Jaffe-Lichtenstein syndrome
  - -- Only café au lait spots
  - -- FD of bone

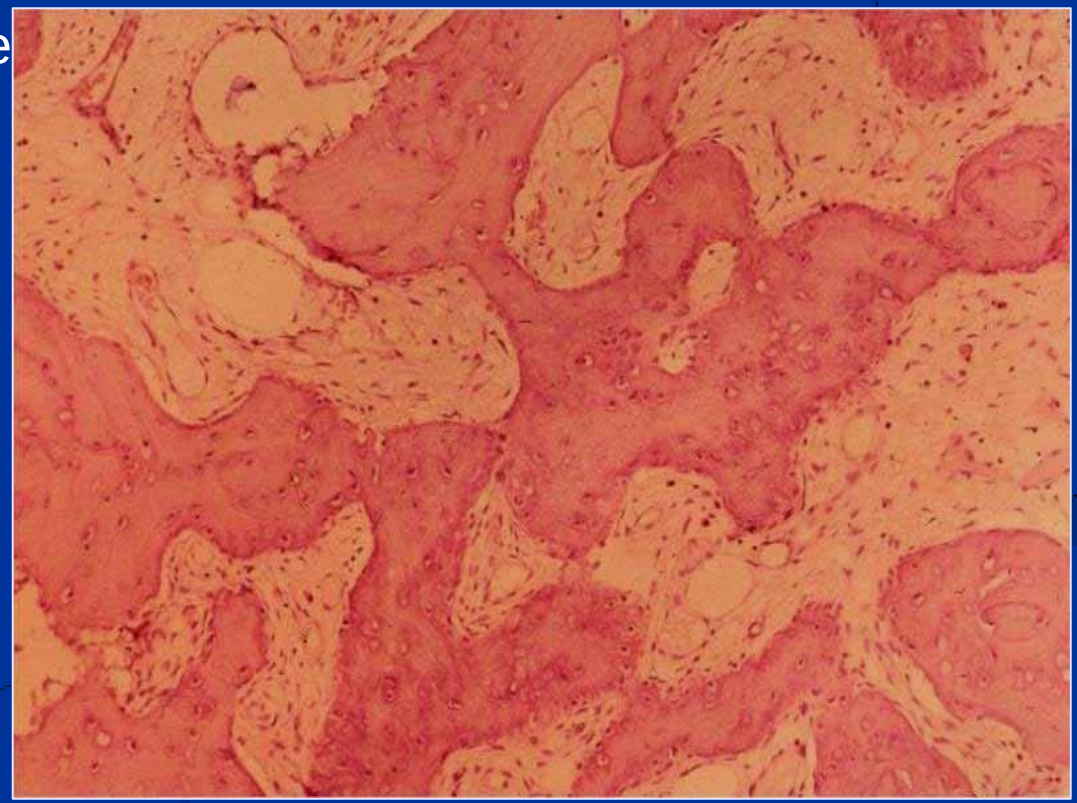




Fibrous Dysplasia
Histopathology



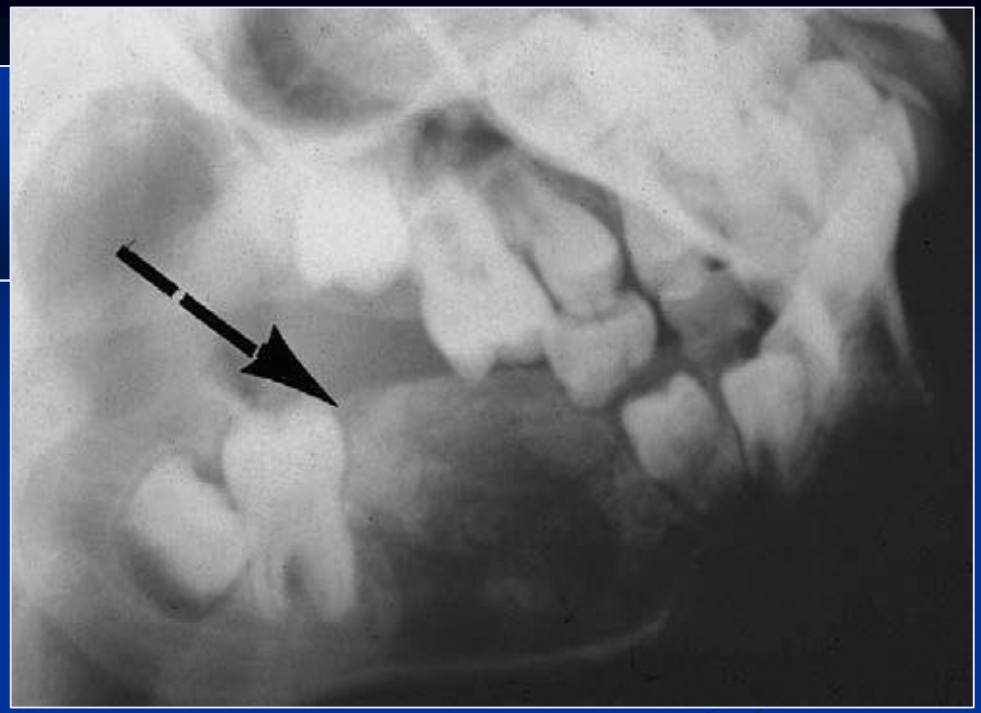
- Irregular in shape but evenly spaced woven immature coarse trabeculae
- Fibrous connective tissue
- Osteoclastic activity seen whe calcification extends surface of trabeculae
- Large osteocytes
- 'Chinese' letter like pattern of trabeculae and blending with sorrounding normal bone
- Becomes more ossified with time

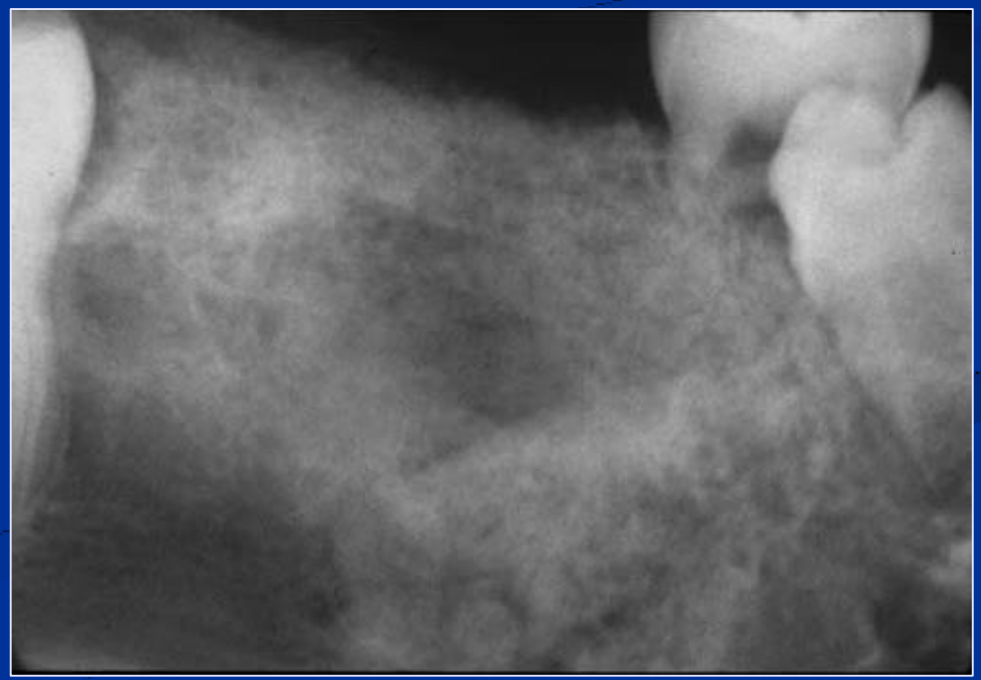


Fibrous Dysplasia of Bone

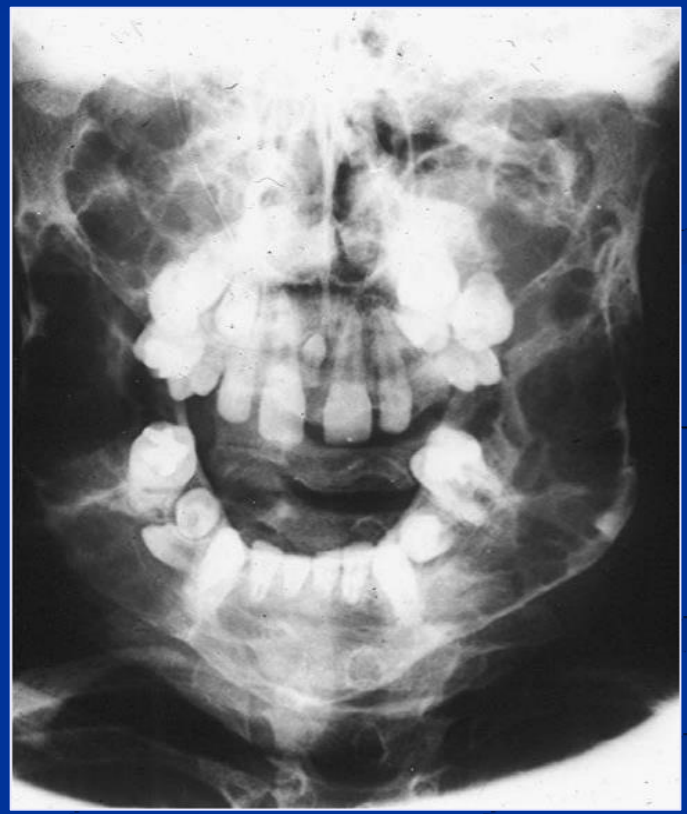
- Radiographic Features:
- Radiolucency surrounded by endosteal scalloping[Rinds Sign]
- Ground Glass appearance
- Displacement of teeth and distortion of nasal cavity

- Treatment : surgical recontouring
- Vit D and Bisphosphonates
- Rarely used modality is allograft/autograft





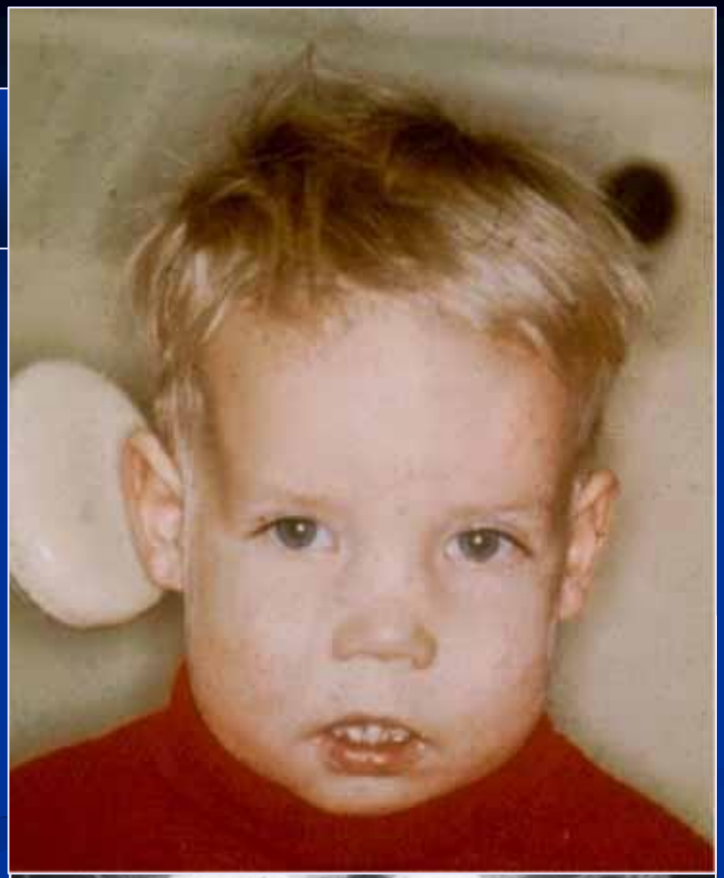
Cherubism

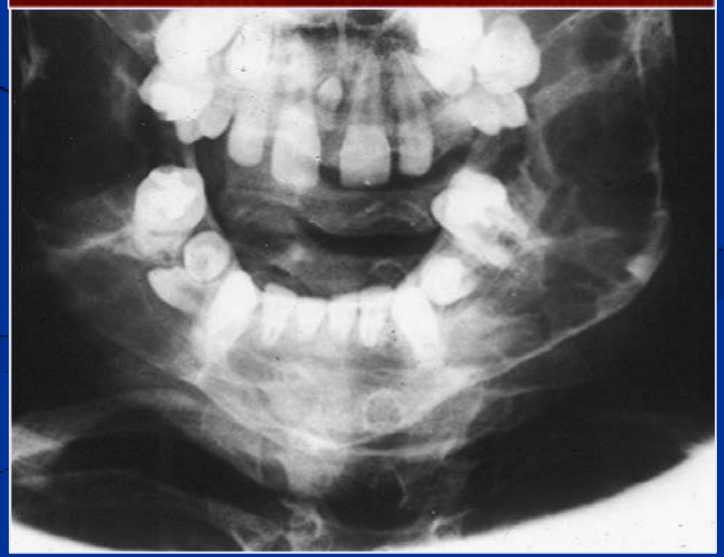


Cherubism
Familial Fibrous Dysplasia

- Non Neoplastic bony lesion with developmental anomaly having "cherubic look"
- AD inheritance
  -- Defective gene on chromosome 4p16.3

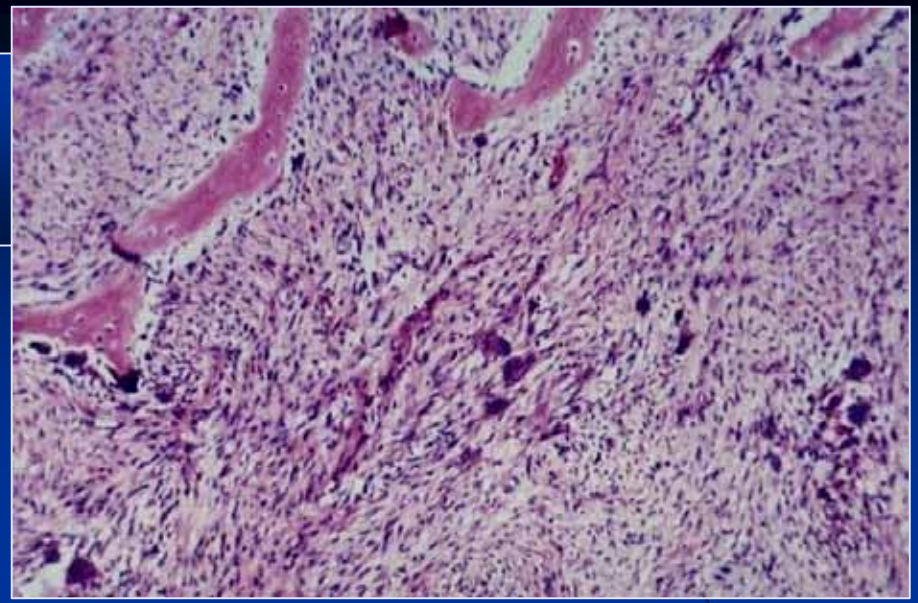
- Gender Age Location Predilection:
  -- None
  -- Radiographically evident till 3 years then bony enlargment rapidifies till 15 years and then remodlling occurs till 3 deecades after that it gets subtle
  -- Posterior jaws
  -- Maybe: ribs and humerus

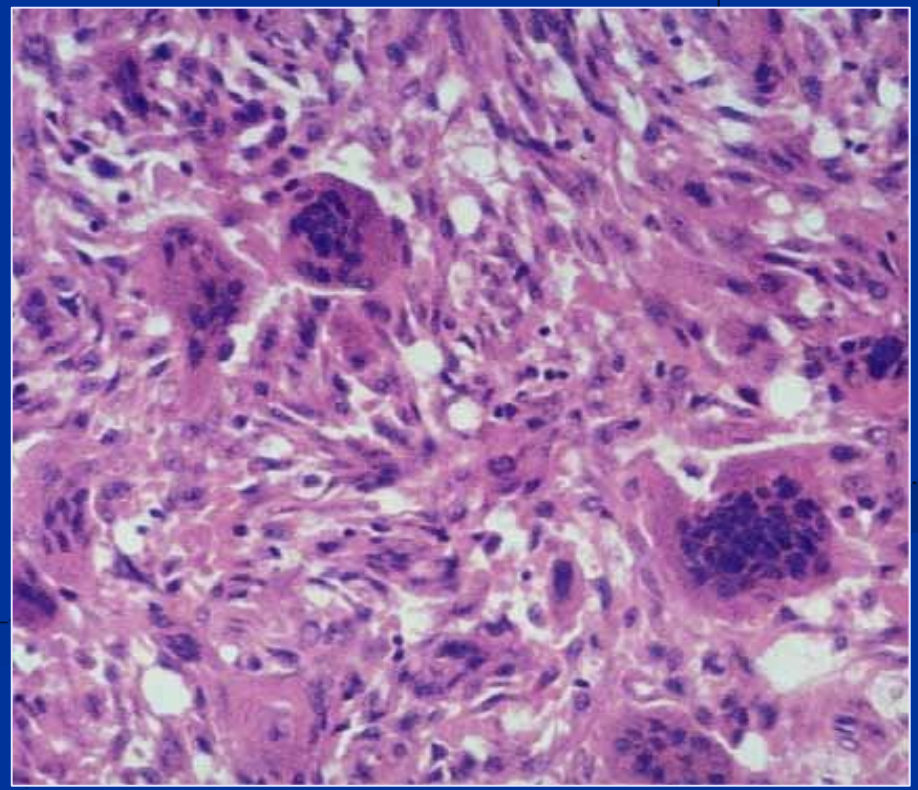




Cherubism
Histopathology

- Loose, immature fibrous stroma
- Scattered multinucleated giant cells
- Eosinophilic cuffing
  -- Around small vessels
- Sparse, immature bone
- Spindle shaped fibroblasts
- Like giant cell granuloma
- Old lesions:
  -- Densely fibrous
  -- Fewer giant cells
  -- More mature bone
  -- Maybe becomes normal?

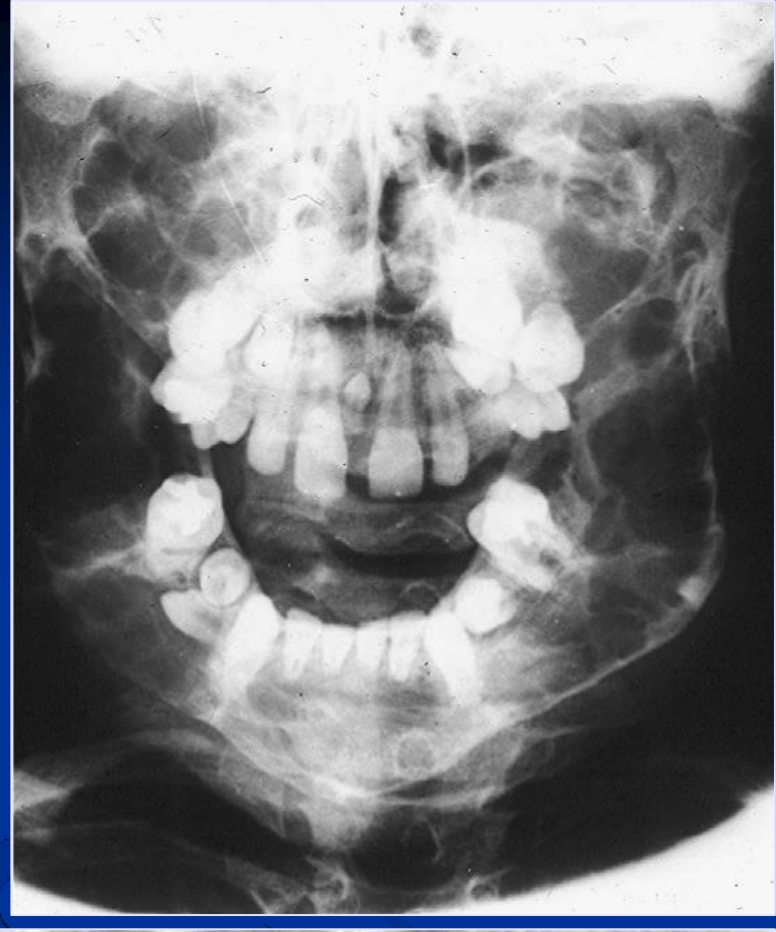


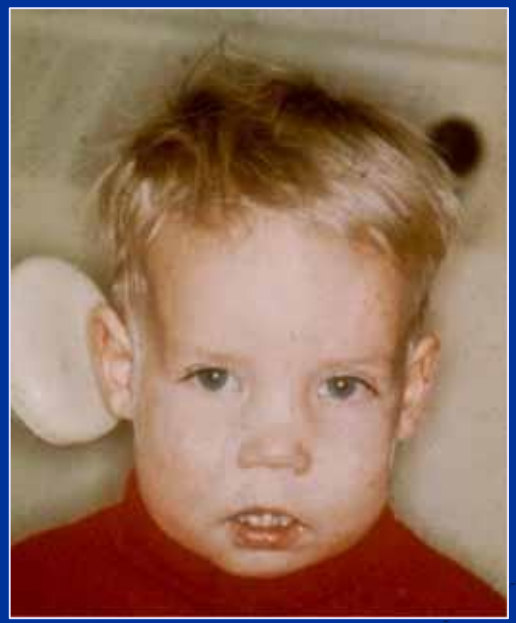


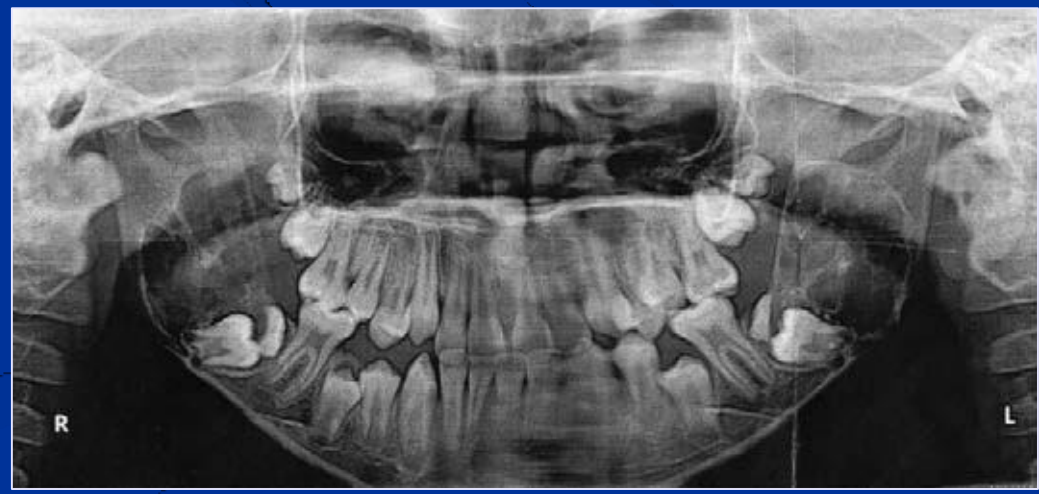
CHERUBISM

Clinical Features

- Bilateral Enlargment; maybe 4 quadrants
- Related to noonan syndrome
- Premature exfoliation of teeth and delayed eruption of teeth
- “Eyes toward heaven”
- Painless firm to palpation
- Tooth buds often pushed out of position
- Wide alveolus







❖**Grades of cherubism:**

1: Ascending rami of mandible

2: Ascending rami of mandible and maxillary tuberosity

3: whole maxilla and mandible except condyle and coronoid process

Radiographic features:

Multilocular expansion of jaw

Ground Glass appearance

Floating tooth syndrome

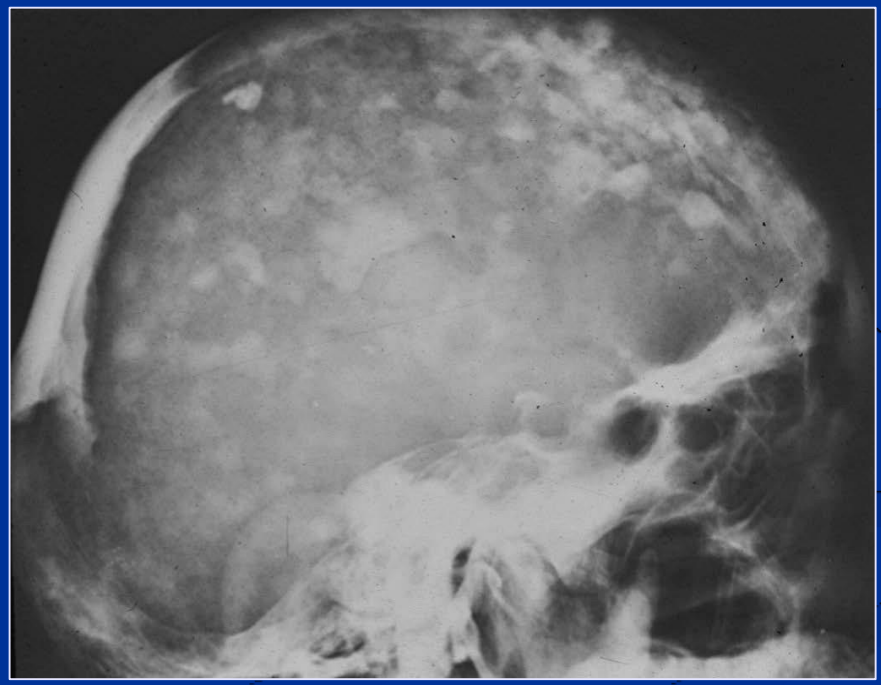
Cherubism

Prognosis; Treatment

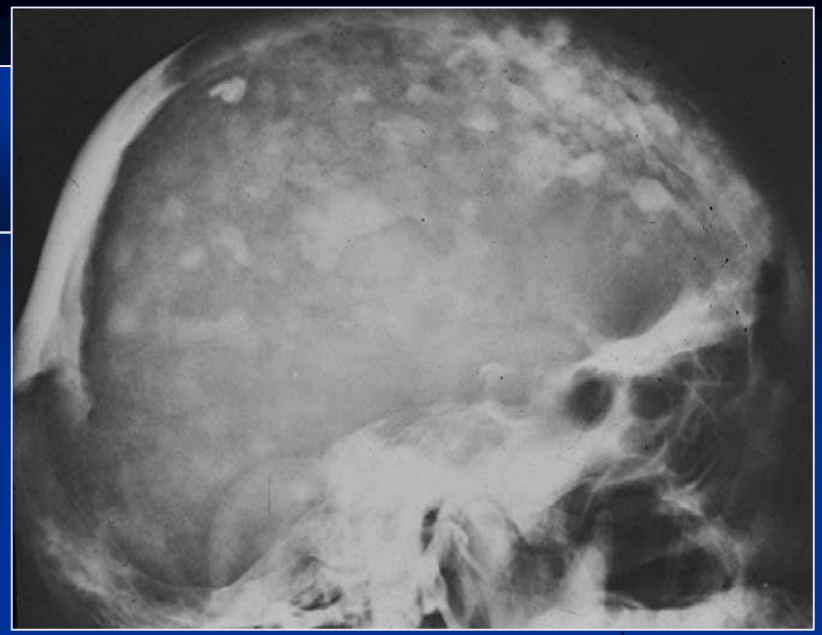
- Enlarges until puberty, then “burns out”
- Face is normal by 25 - 40 years of age
- Can push tooth buds great distances
- May develop central giant cell granuloma
- Maybe: pathologic fracture

- Treatment: none, unless pathologic fracture
  -- Surgical curettage can be performed
  -- Irradiation works, but risk of future sarcomas
  Rarely needed

Paget Disease of Bone

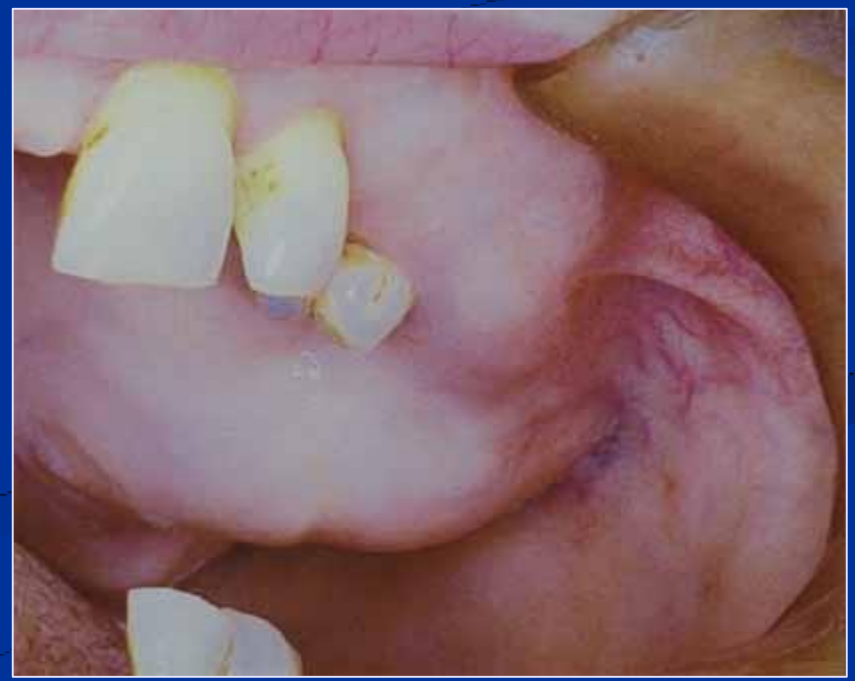


Paget Disease of Bone
Osteitis Deformans



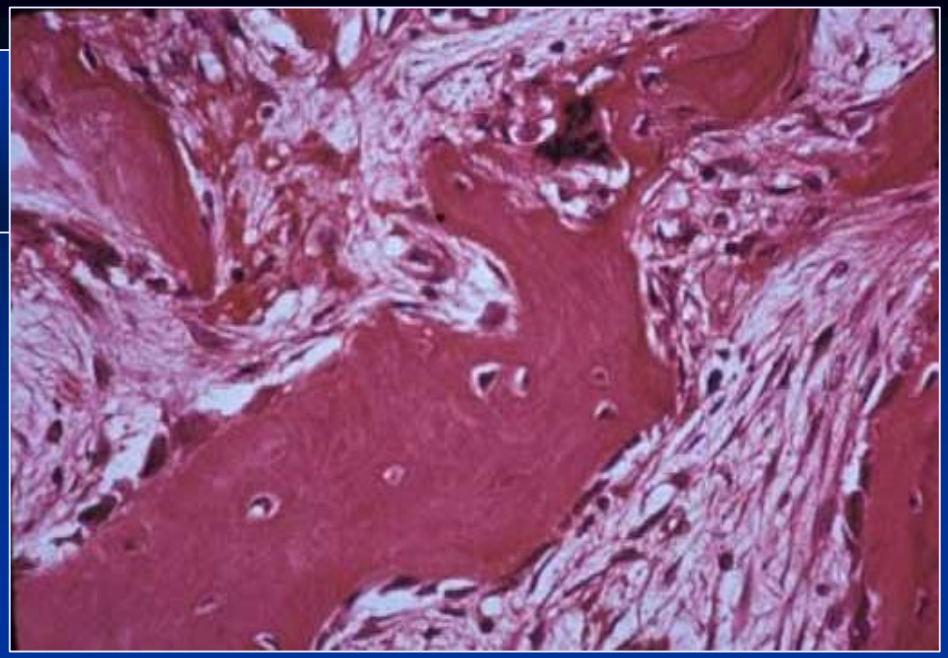
- Enhanced bone resorption & deposition
  -- Possibly from a slow virus
  And Parathyroid hormone related
  -- Paramyxovirus detected in osteoclasts

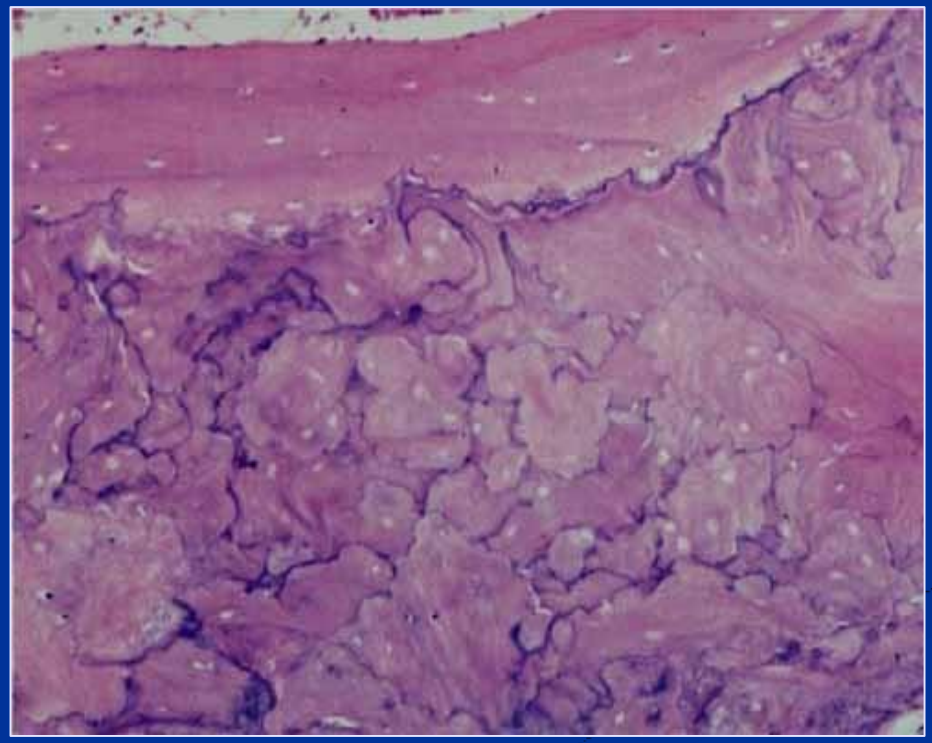
- Gender Age Location Predilection:
  -- Men : female ratio -1:1
  -- >50 years
  -- skull:25-65%
  Long bones:25-65%
  Pelvis and spine:30-75%
  -- 1/100 persons over 45 years of age
  (but most disease is subclinical)

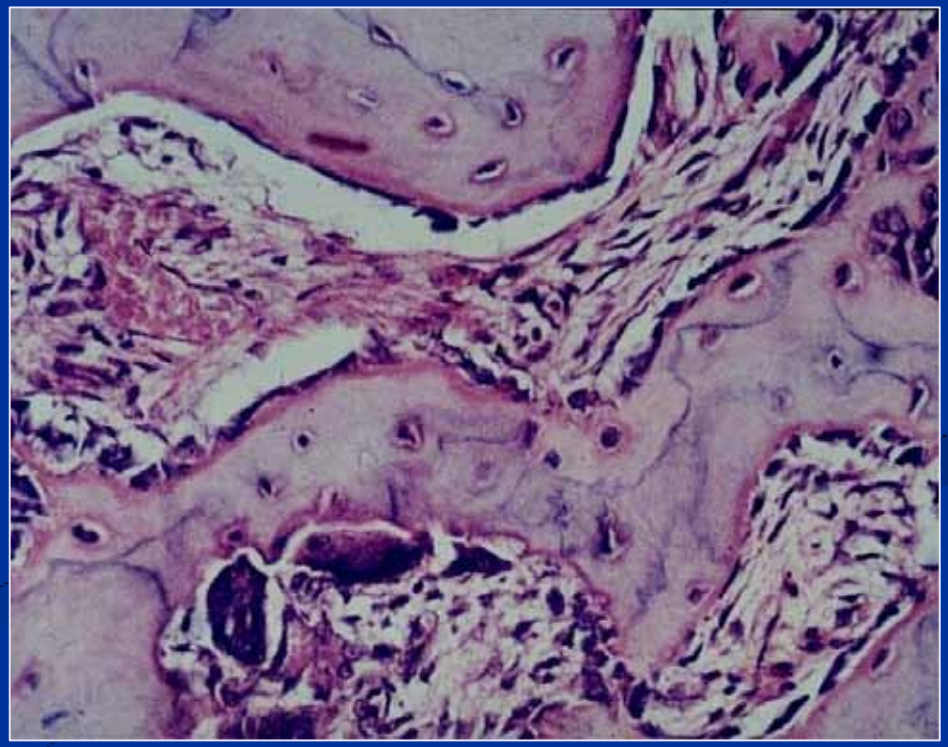


Paget Disease of Bone
Histopathology

- Immature trabeculae
- Earliest osteoclastic activity
- Then Abundant osteoblastic activity
- Fibrous background stroma
- Remodelling with many reversal/cement lines in the bone



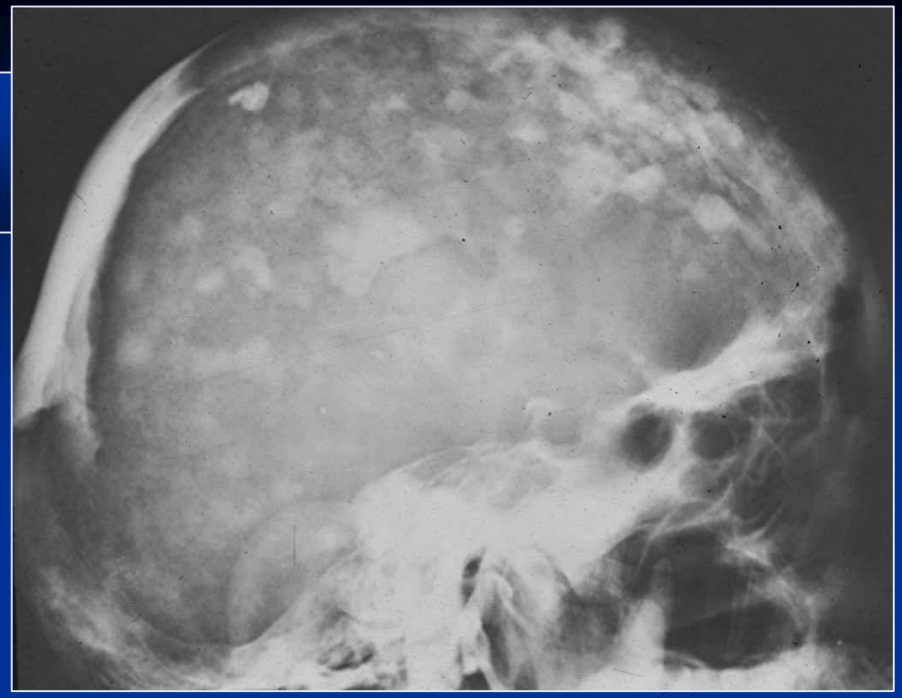


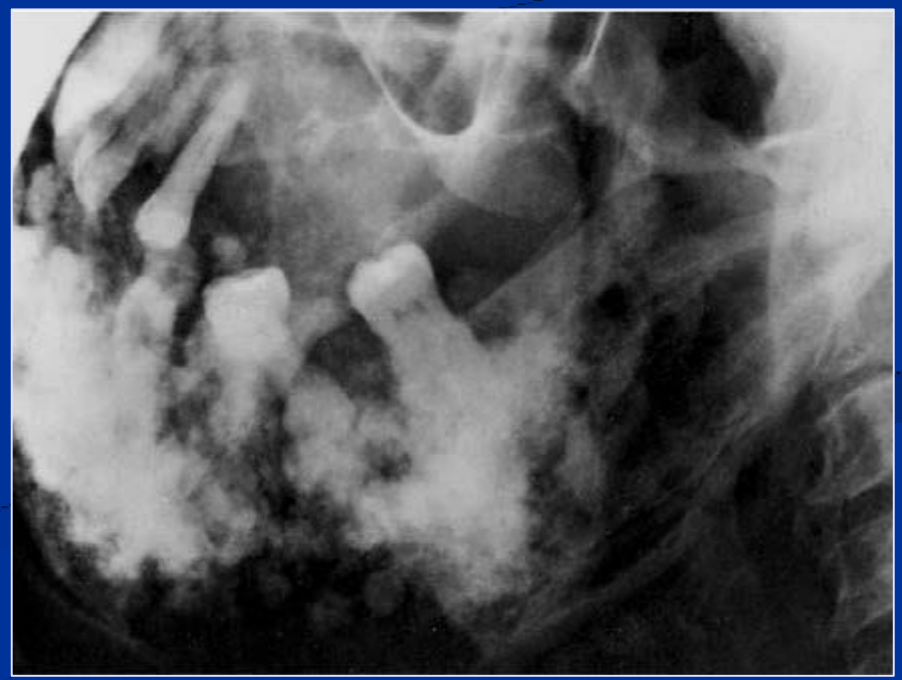


Paget Disease of Bone
Clinical Features

- Irregular radiolucency/radiopacity
- Bowed legs
- Maybe: hypercementosis
- Maybe: bone pain
- Early stage is radiolucent
  -- Osteoporosis circumscripta
- Become more radiopaque over time
  -- Cotton wool appearance
  -- Cloud-in-the-sky appearance
- May get anemia and bleeding
  -- No hematopoietic tissue/platelets
  -- Immune deficiency
- Deafness and visual loss
  -- Pinched nerves in foramina

More seen in maxilla then mandible
Maxillary enlargment with flat palate
Displaed tooth and unable to close mouth



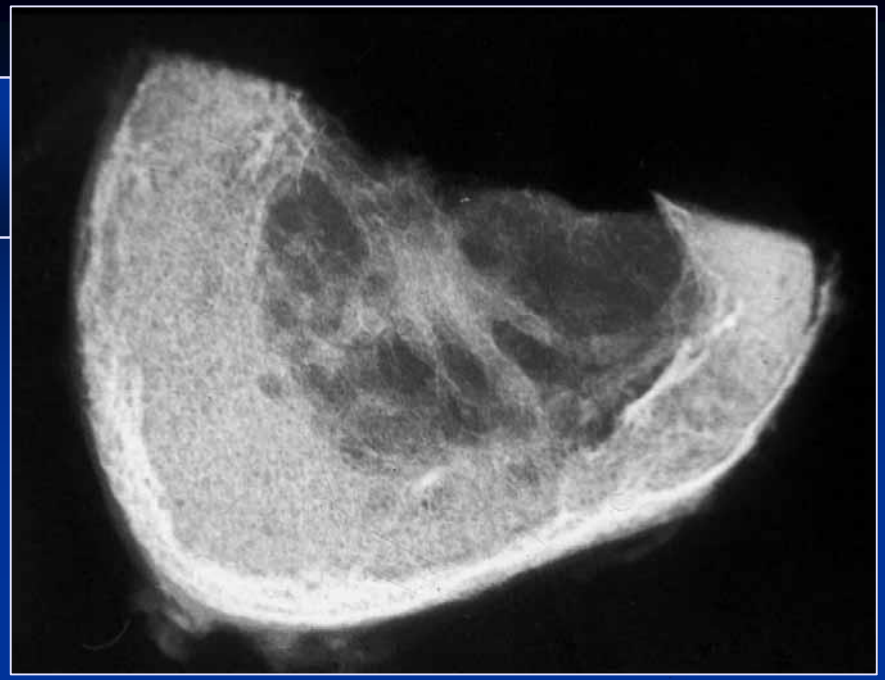


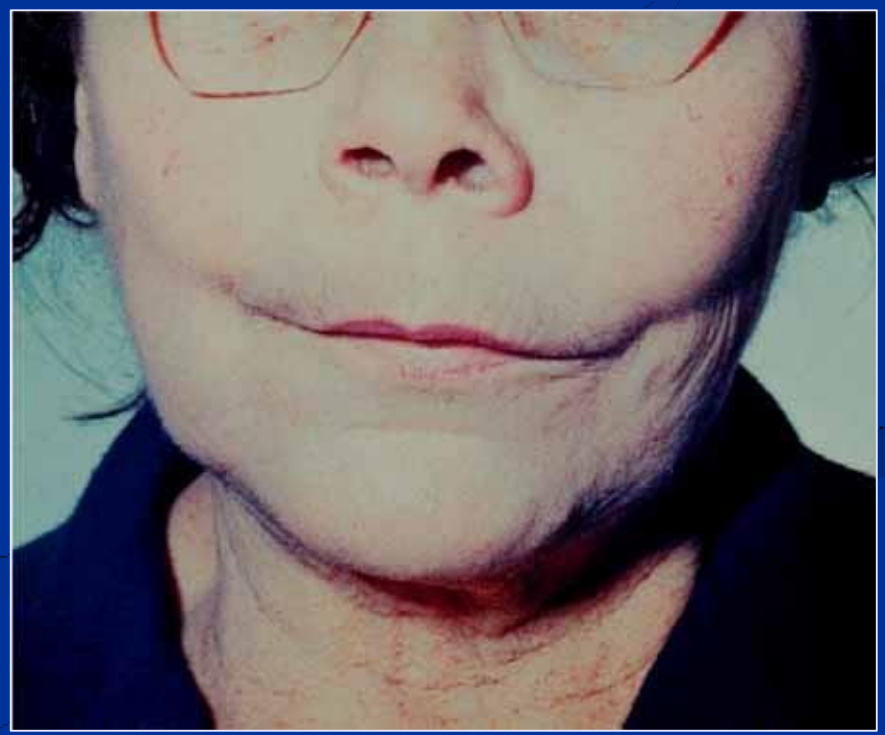
Paget Disease of Bone
Clinical Features

- Thickened cortex
- Enlarged bone
  -- Denture no longer fits
  -- Hat no longer fits
  Characteritic Platybasia and Leontiasis Facia

Laboratory Findings:

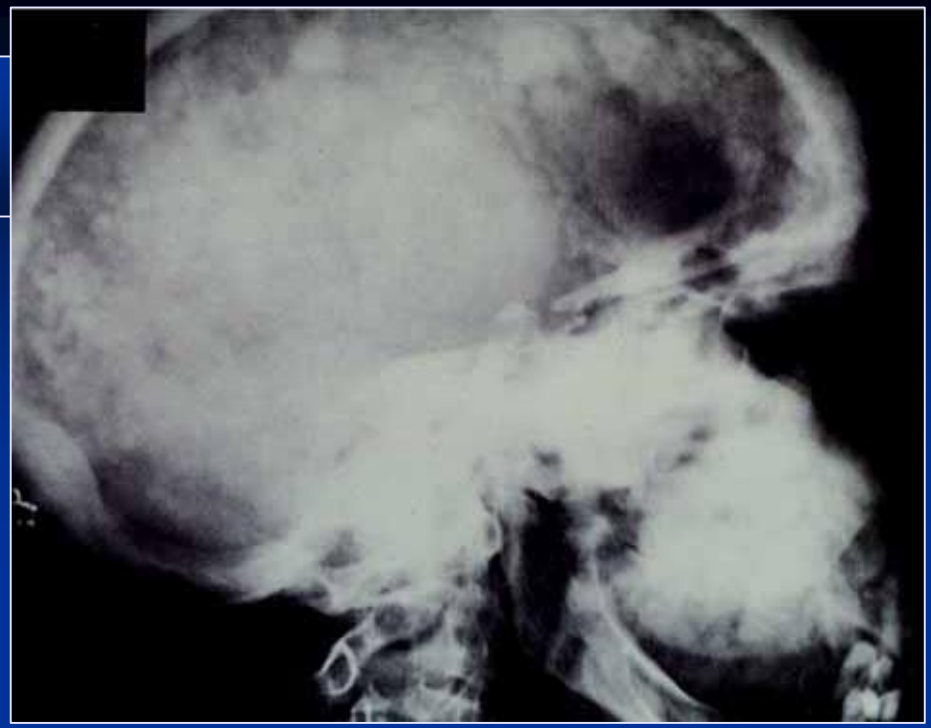
- Elevated alkaline phosphatase
  -- 25%+ above normal
- Normal calcium and phosphorus
- Urinary hydroxyproline
- Osteoarthritis

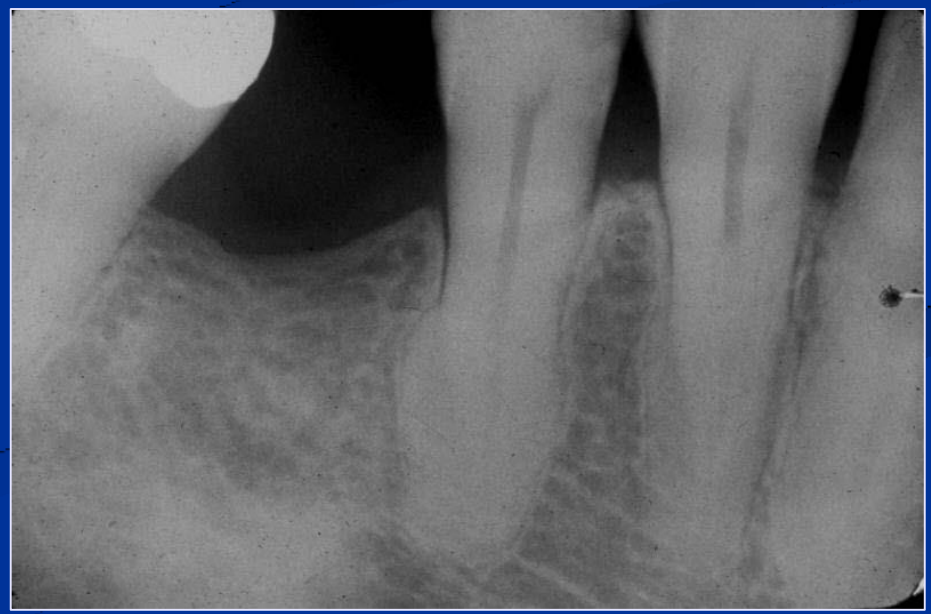




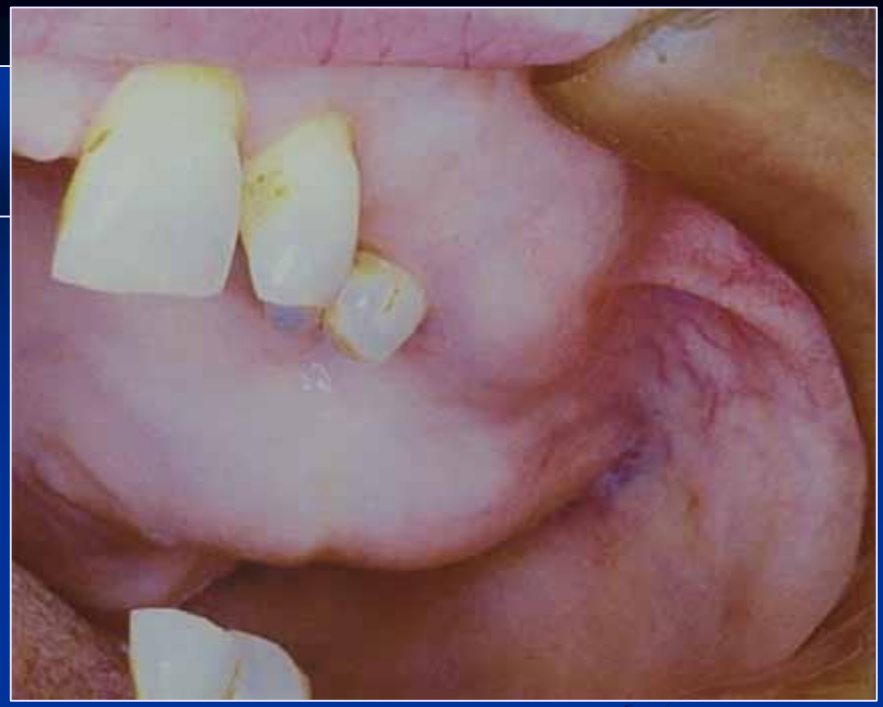
Paget Disease of Bone
Clinical Features

- Lincoln's beard (black beard)
  - -- Technetium bone scan (scintigraphy)
  - -- Hot (black) mandible
- Small sinuses
- Bone pain may mimic toothache





Paget Disease of Bone
Prognosis; Treatment



- Seldom causes death
- Parathyroid hormone antagonists
  -- e.g. calcitonin, bisphosphonates
  -- To reduce bone turnover
- Cytotoxic antibiotics, e.g. plicamycin
  -- Inhibit osteoclastic activity
  (used only in severe cases)
  Calcitonin replacement
- Aspirin for pain
- Antibiotics for osteomyelitis (sclerotic, late phase)
- New dentures or bridgework may be required as maxilla expands
- May do skull base surgery to relieve nerves, vessels
- **Caution**! 1 - 13% risk of osteosarcoma or giant cell tumor
  -- The former seldom in jaws
  -- The latter often in jaws
  COMPLICATIONS: Factures and Osteosarcoma

Cemento-Osseous Dysplasia

-- Commonest fibro-osseous lesion
-- Occurs in the tooth bearing area & form excessive cementum like matrix
-- Unknown , reactive or dysplastic process
-- Lesion arise from PDL or due to defect in the extra PDL bone remodelling
-- Influenced by local & systemic factors

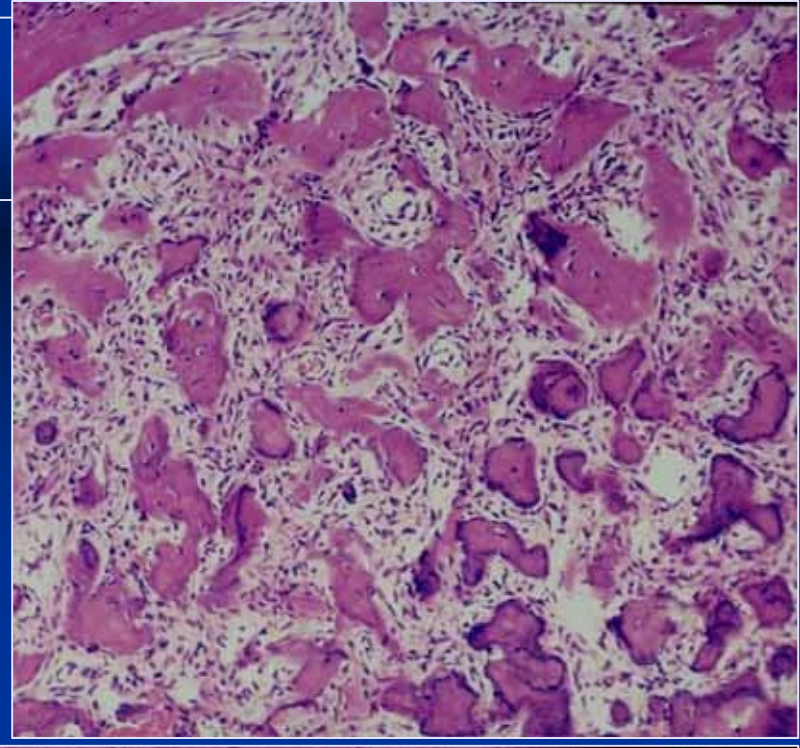
Subtypes:

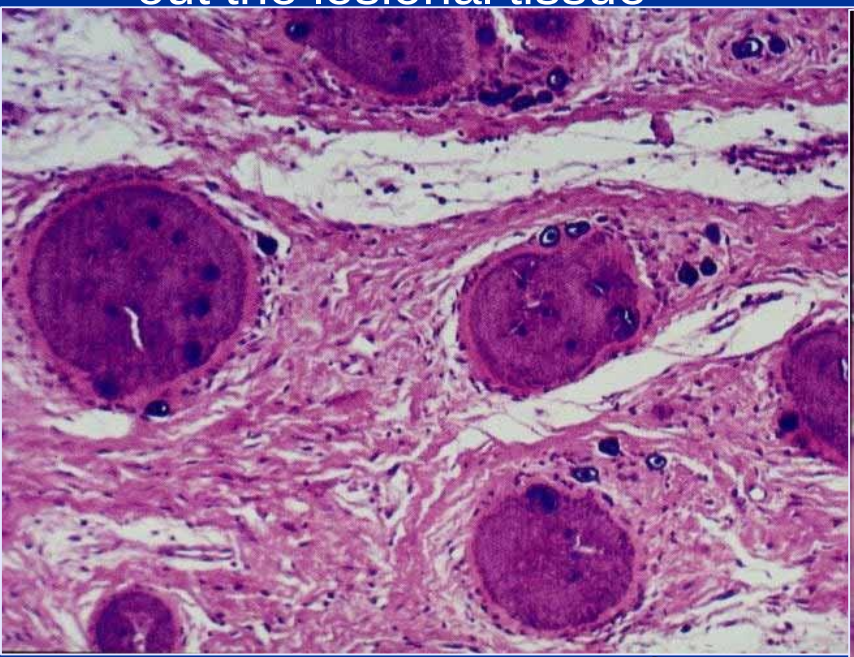
- Periapical cemento-osseous dysplasia
- Localized cemento-osseous dysplasia
- Florid cemento-osseous dysplasi

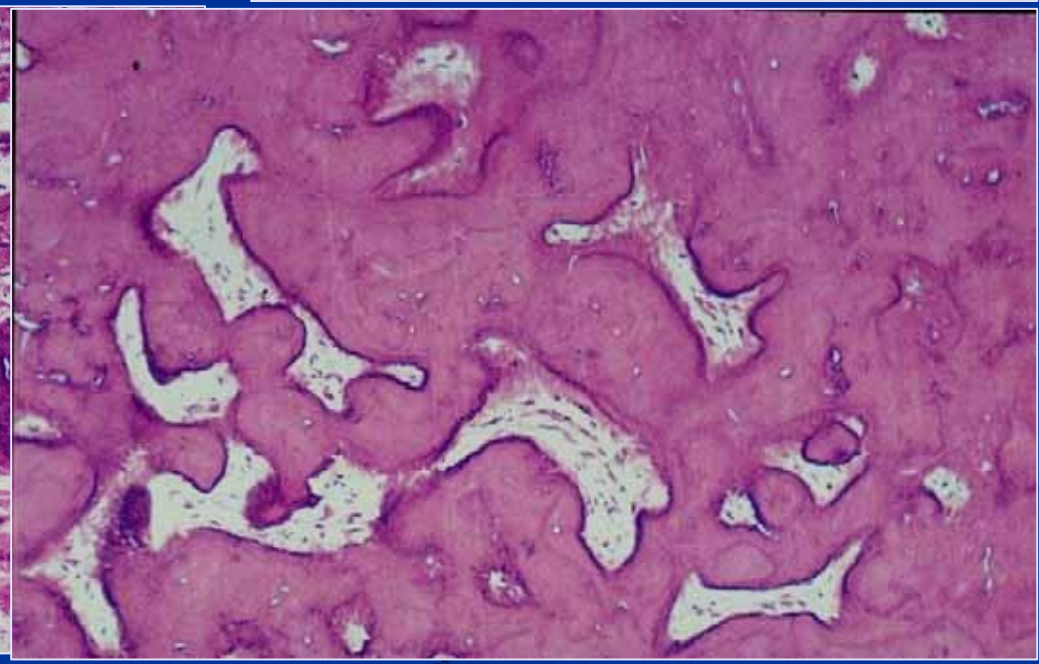
Cemento-Osseous Dysplasia

Histopathology = Same for All Subtypes

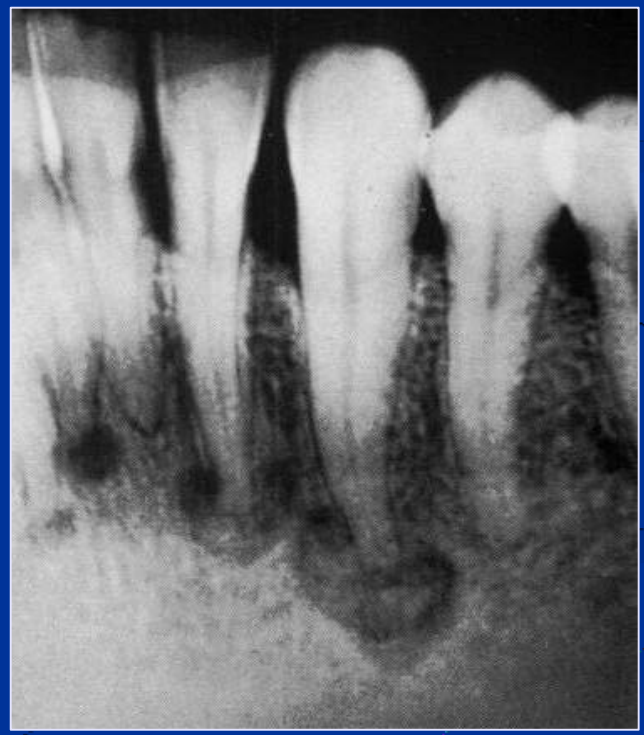
- Immature bone and globular cementum
- Fibrous stroma, maybe immature
- Fibrous capsule, often
- Relatively avascular
- No inflammatory cells
- More calcification with time
- Haemorrhage typically noted through-out the lesional tissue







Periapical Cemento-Osseous Dysplasia



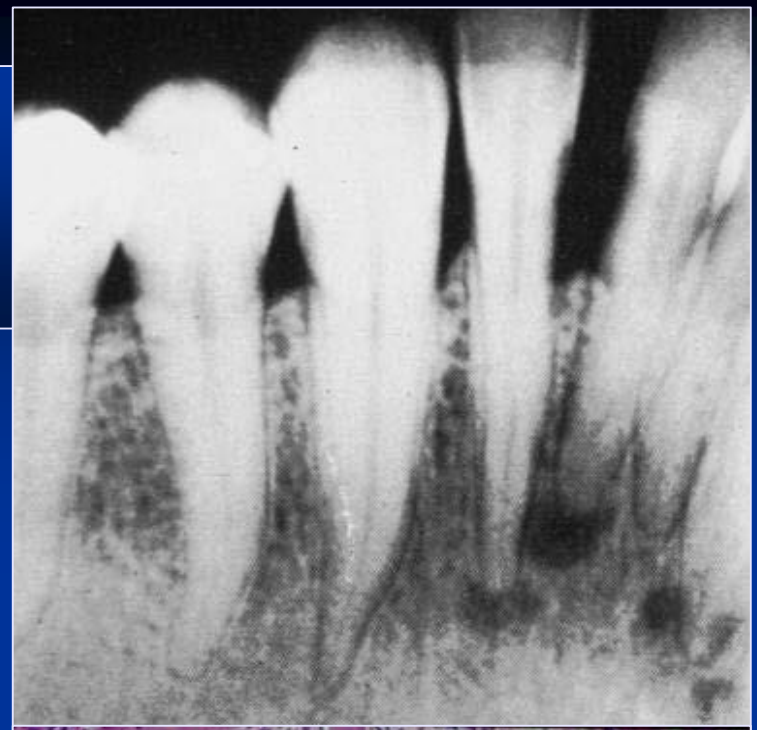
Periapical Cemento-Osseous Dysplasia

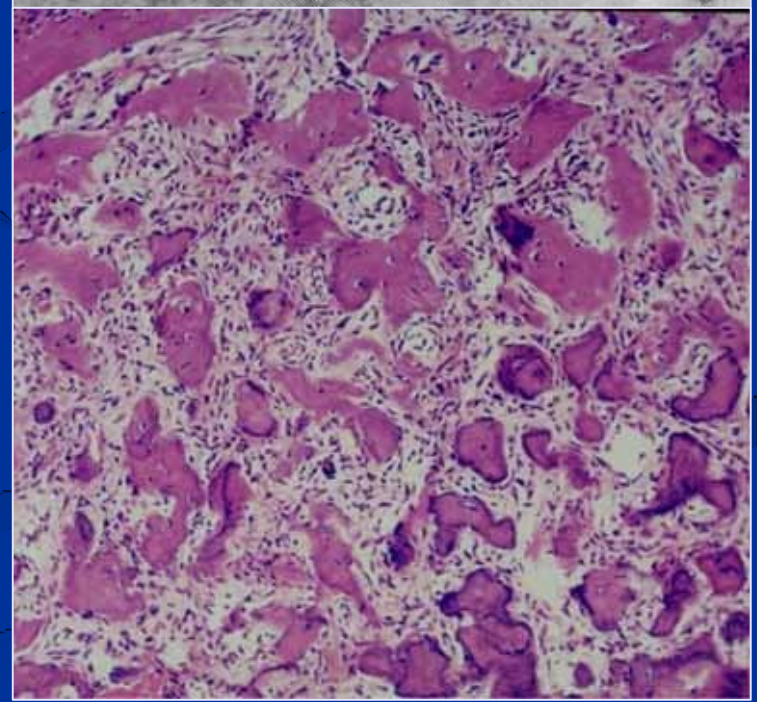
Periapical Cemental Dysplasia

- Developmental anomaly
- Unknown etiology (inherited?)

- GENDER,AGE,LOCATION PREDILECTION:
  - -- 14:1 female;male ratio
  - -- 70% in blacks
  - -- 30-50 years old
  - -- well defined
  - -- Anterior mandible
  - -- Apices of teeth
  - -- seldom exceed 1cm in

May be part of florid cemento-osseous dysplasia

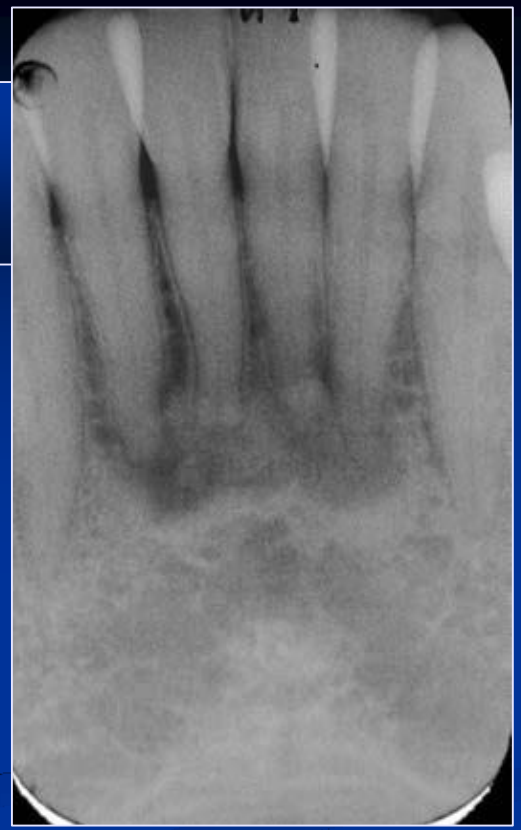


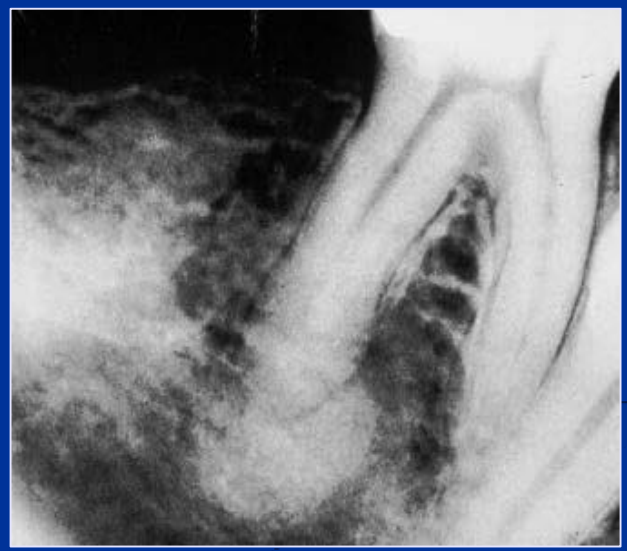


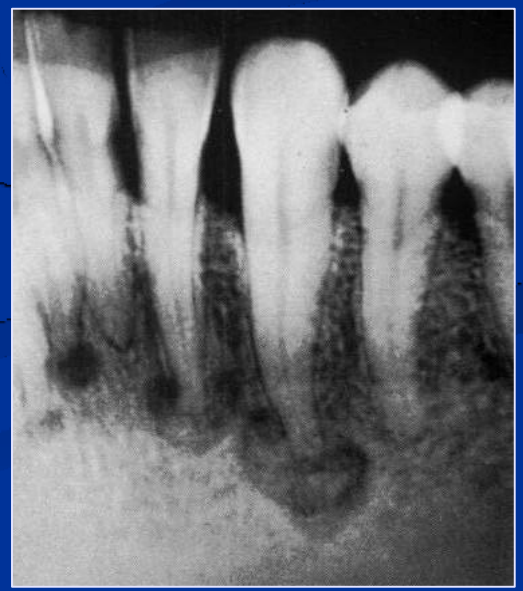
Periapical Cemento-Osseous Dysplasia

Periapical Cemental Dysplasia

- Radiolucencies at apices
- Maybe: central opacity
- Viable teeth
- Center = 1-2 mm below apex
- Irregular capsule at periphery
- Intact PDL
- Asymptomatic, usually
  -- May be tender, aching
- Nonexpansile



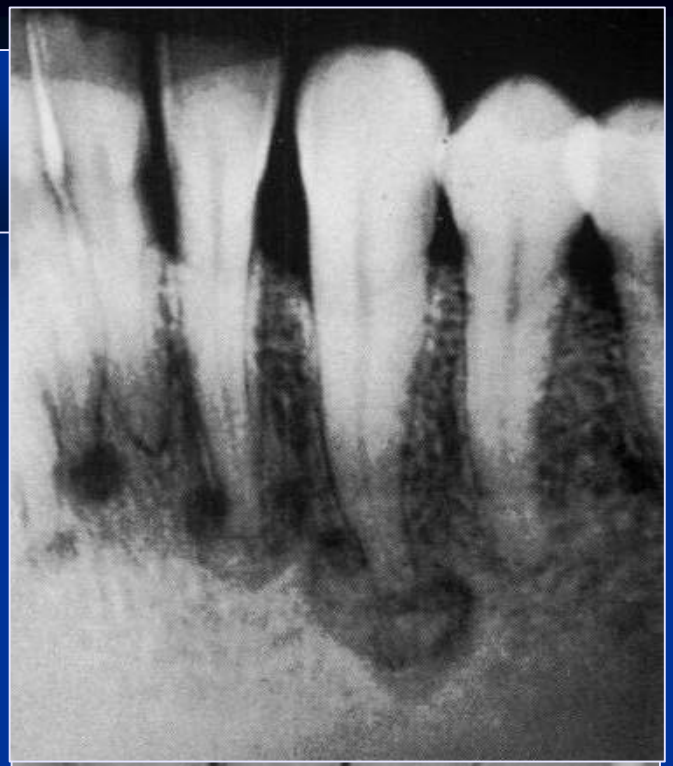


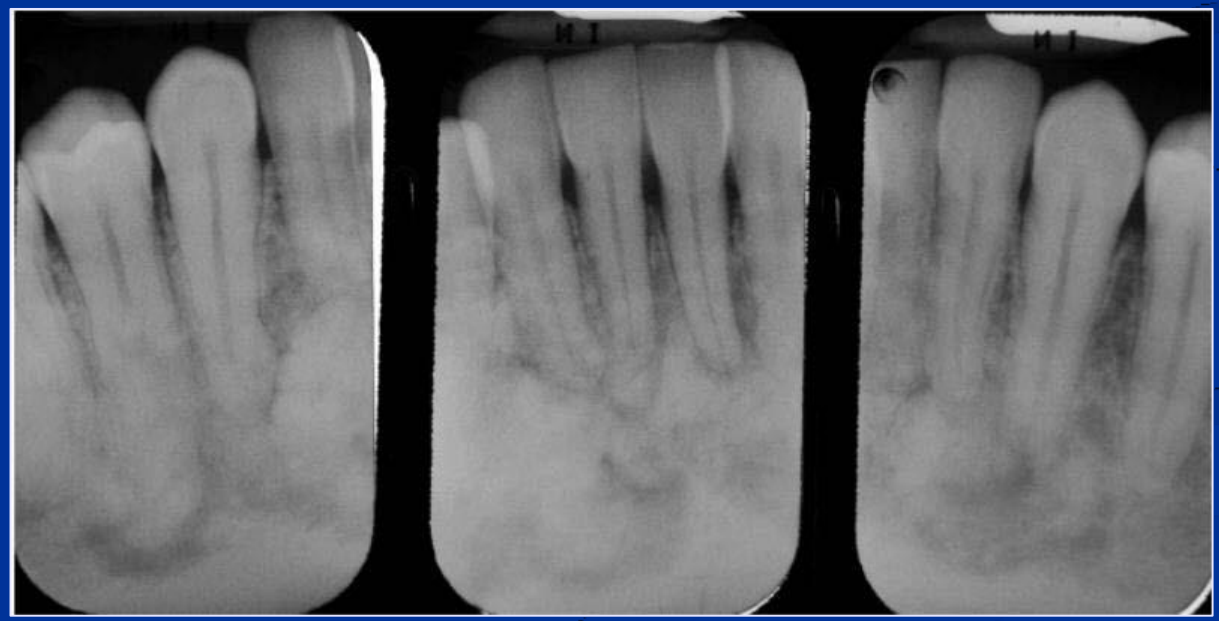


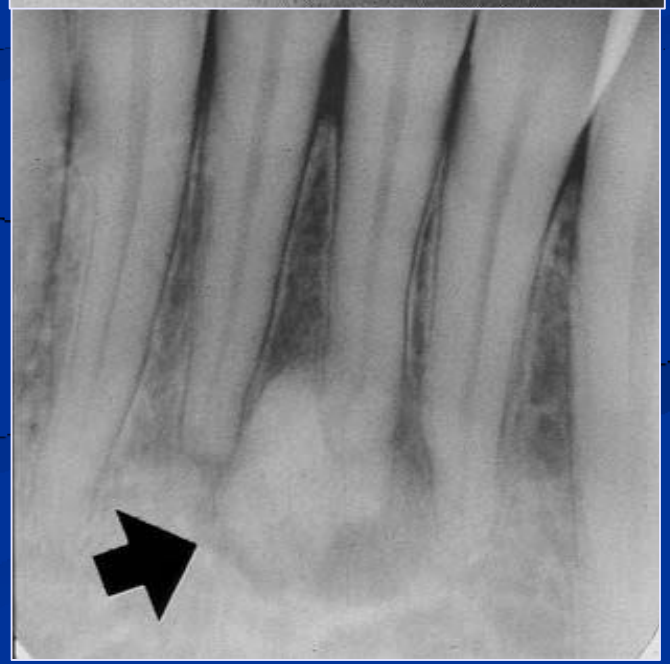
Periapical Cemento-Osseous Dysplasia

Periapical Cemental Dysplasia

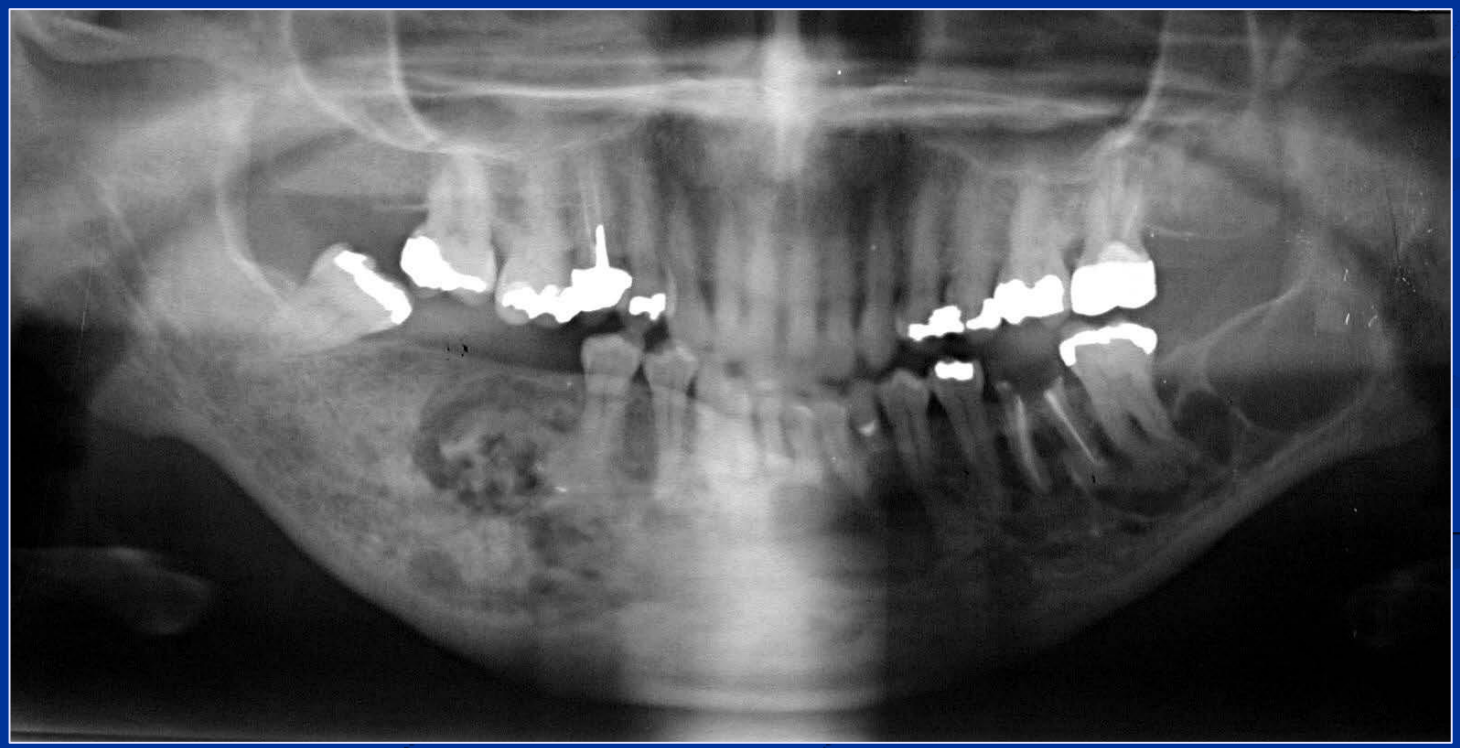
- More radiopaque over time (years)
  -- Calcified from center outward
- Treat: none required
- If symptomatic: curettage
- Can extract the tooth
  -- Lesion is not attached
- Prevent infections (lesion is avascular)







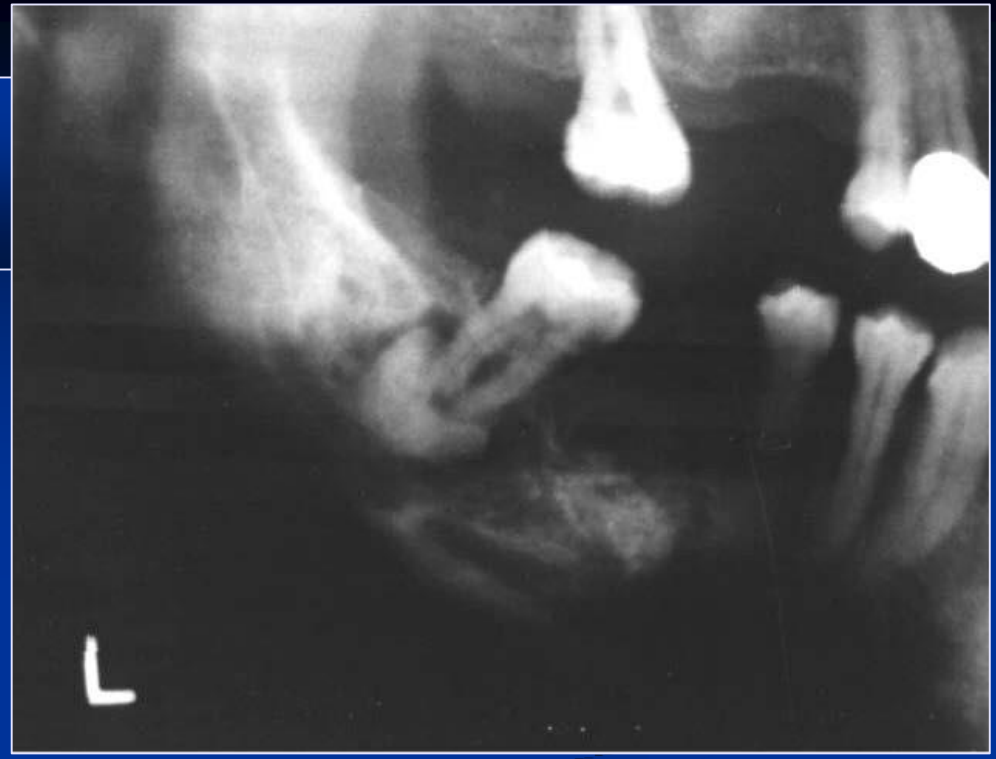
Focal Cemento-Osseous Dysplasia

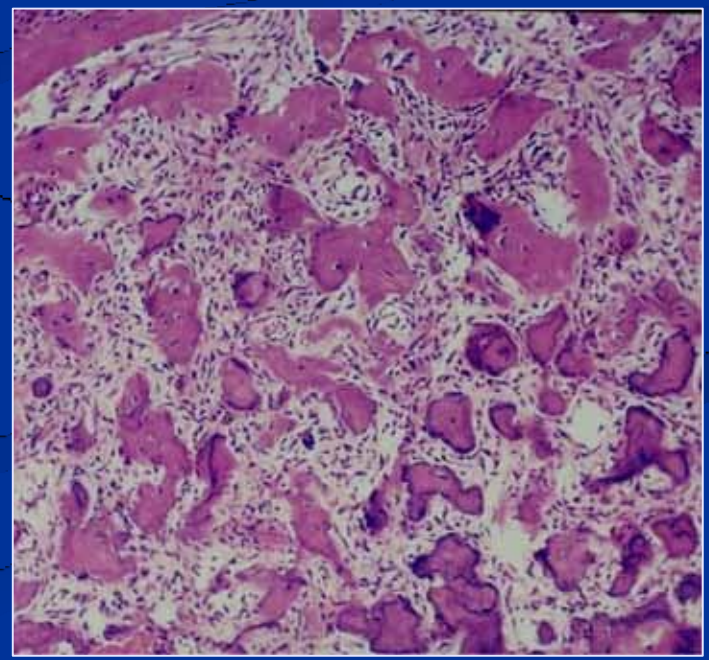


Focal Cemento-Osseous Dysplasia

- Developmental anomaly?
- Etiology: unknown

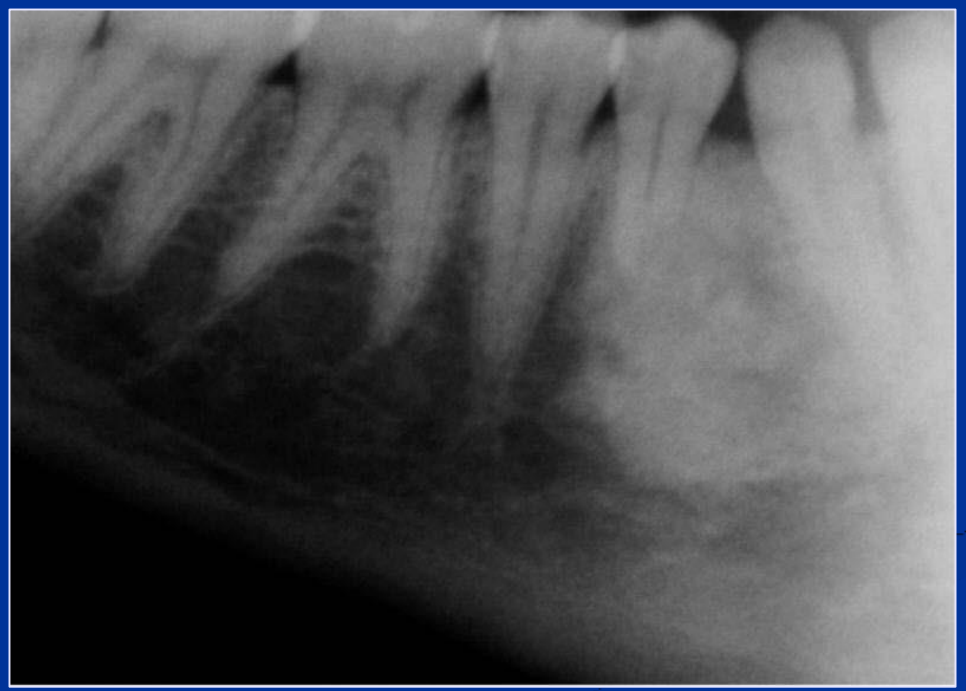
- GAL:
- 90% females
- Middle aged females[mid 30]
- Especially in african & american
- Posterior mandible
  -- Single site involved

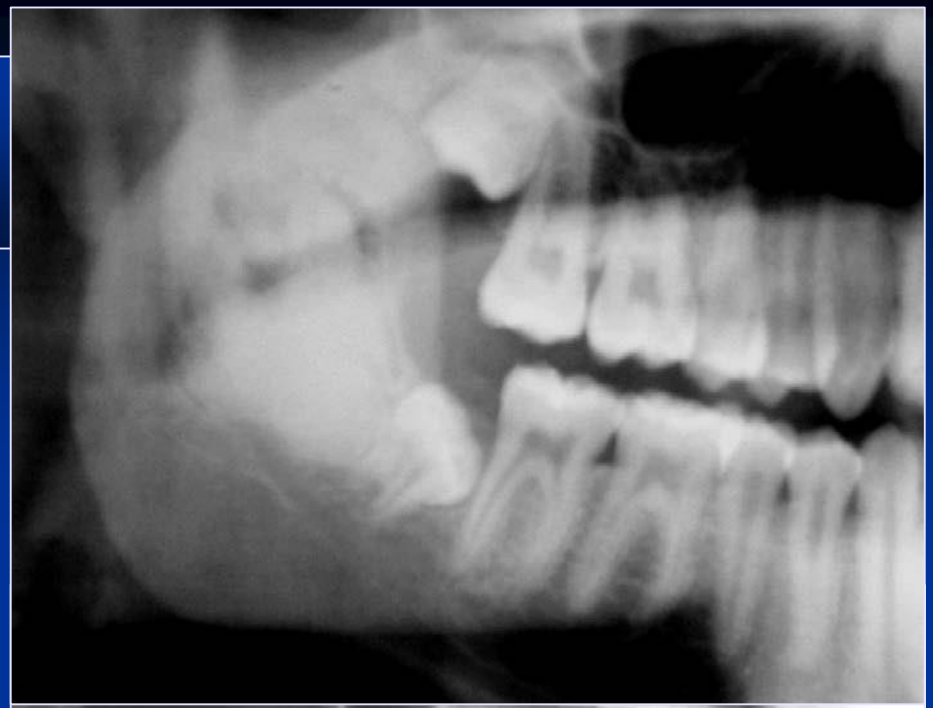


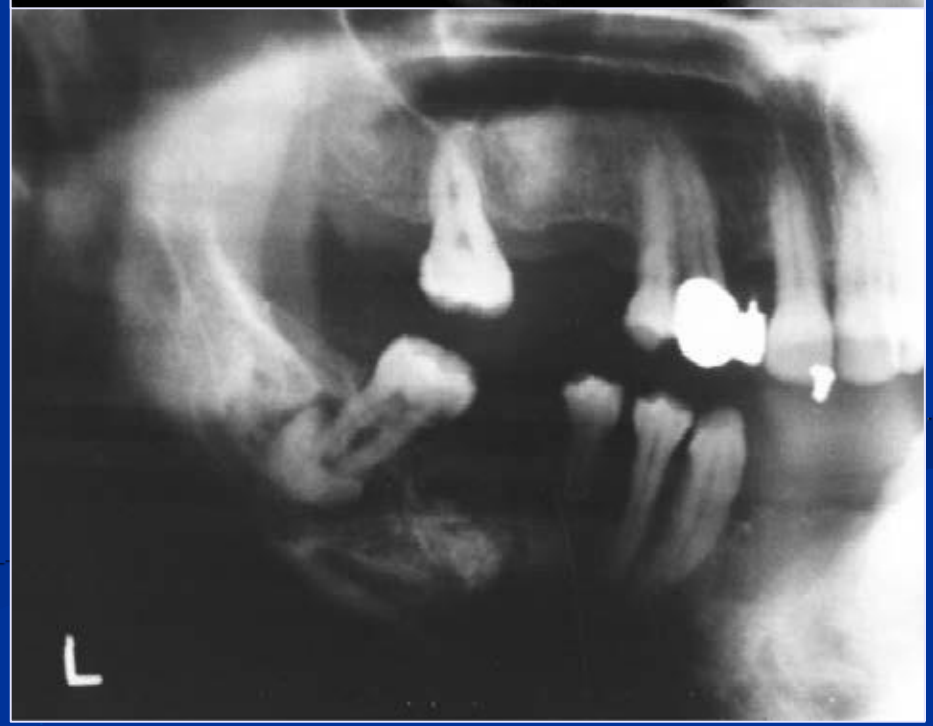


Focal Cemento-Osseous Dysplasia

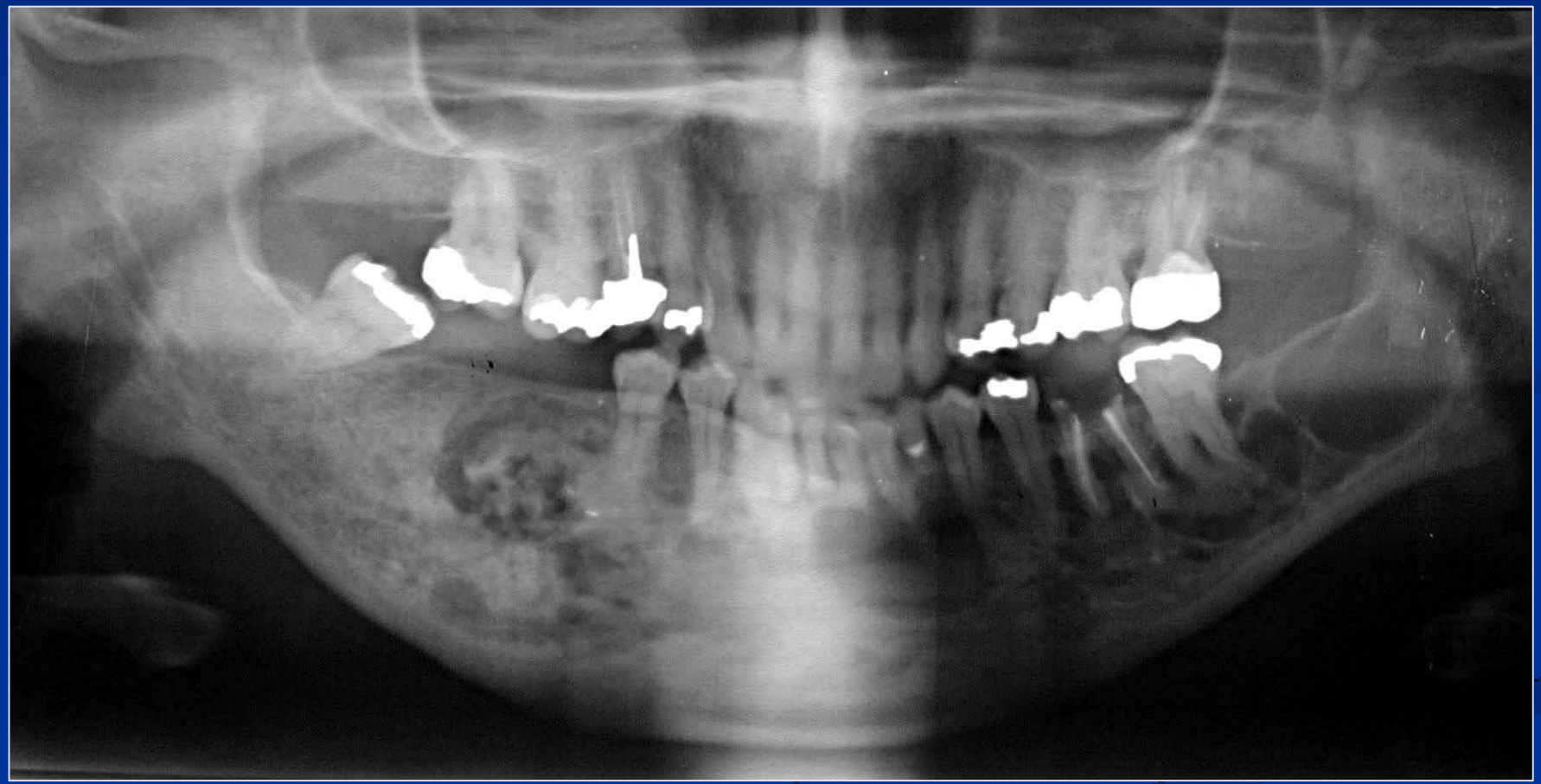
- Asymptomatic, usually
- May be tender, aching
- Nonexpansile
- Radiolucent/opaque areas
- < 1.5 cm. diameter
- Moderately well demarcated
- More sclerotic over time





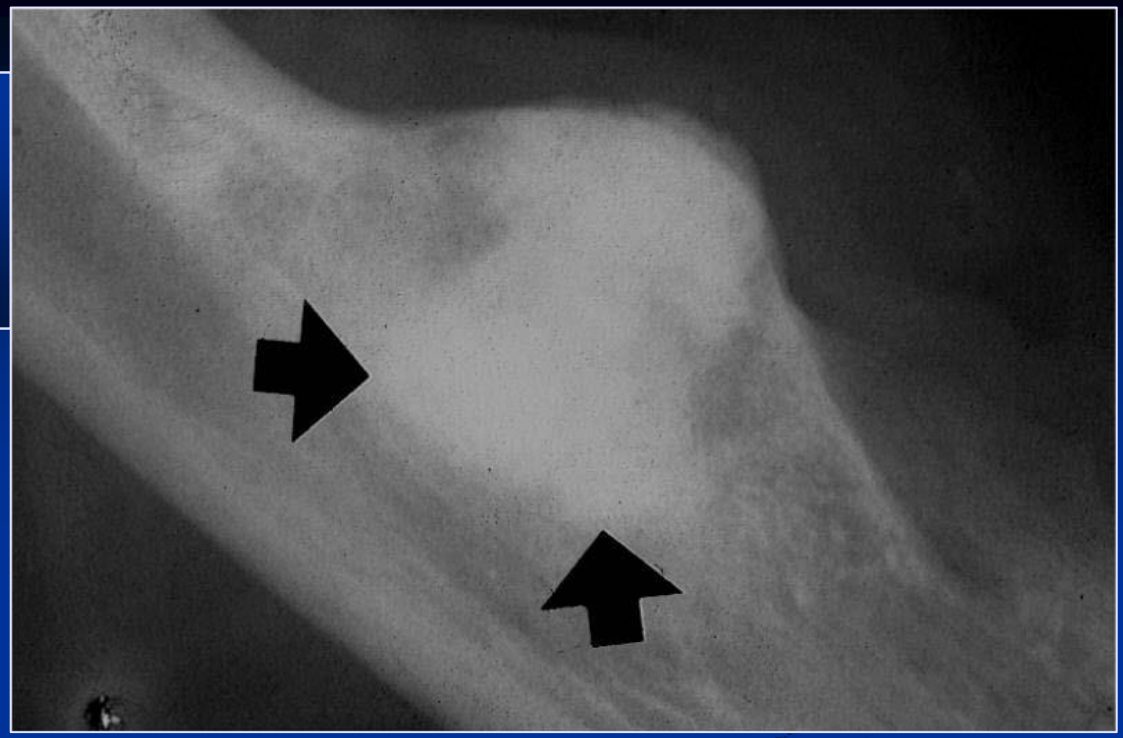


Focal Osseous Dysplasia
With Traumatic Bone Cyst



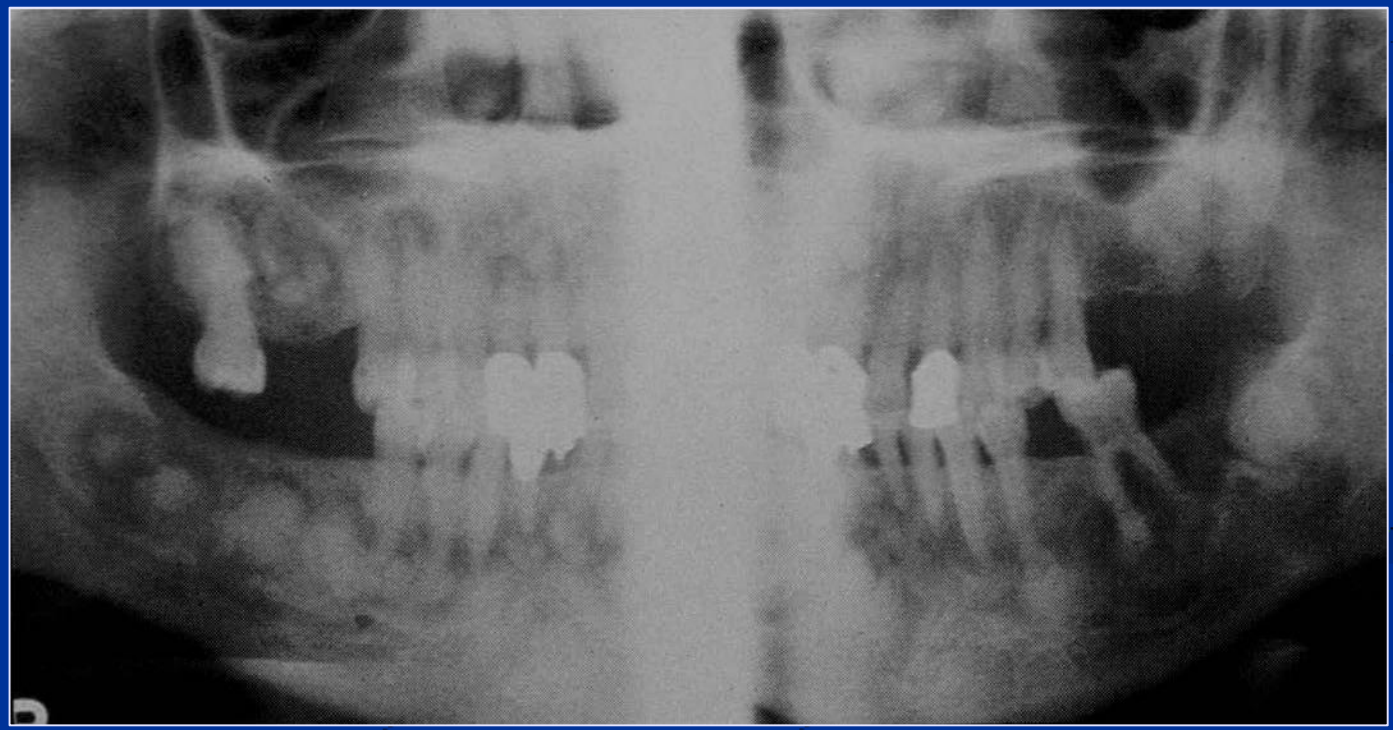
Focal Cemento-Osseous Dysplasia

Prognosis; Treatment

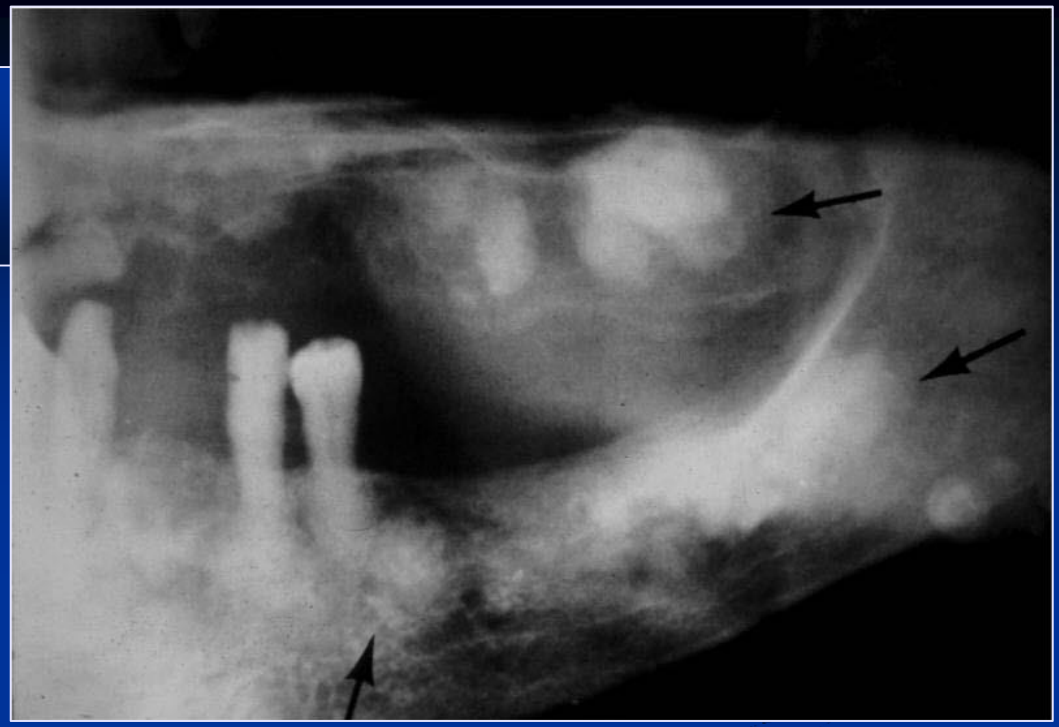


- Limited growth
- Will not resorb with alveolar ridge
  -- Under denture
- Treat: none required
- If symptomatic: curettage
- Can extract adjacent teeth
  -- Lesion is not attached
- Prevent infections
  -- Lesion is avascular

Florid Cemento-Osseous Dysplasia



Florid Cemento-Osseous Dysplasia

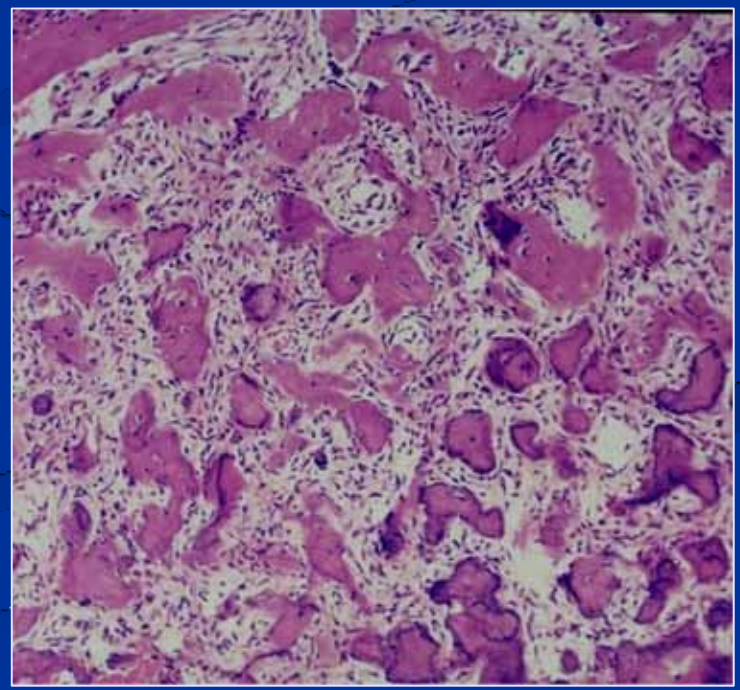


- Developmental anomaly?
- Some familial cases

GENDER,AGE,LOCATION PREDILECTION:

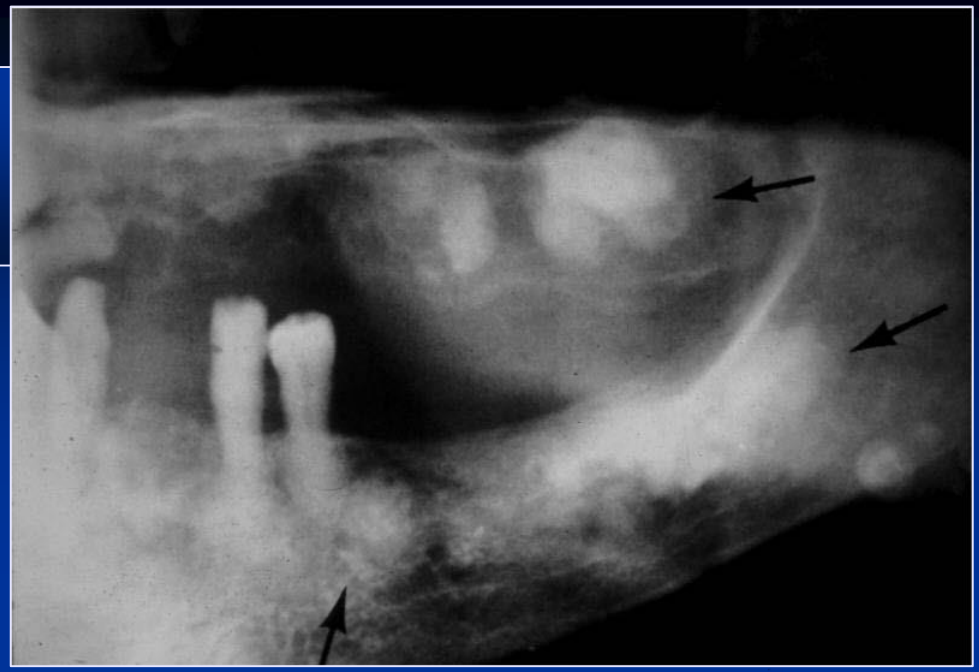
Females

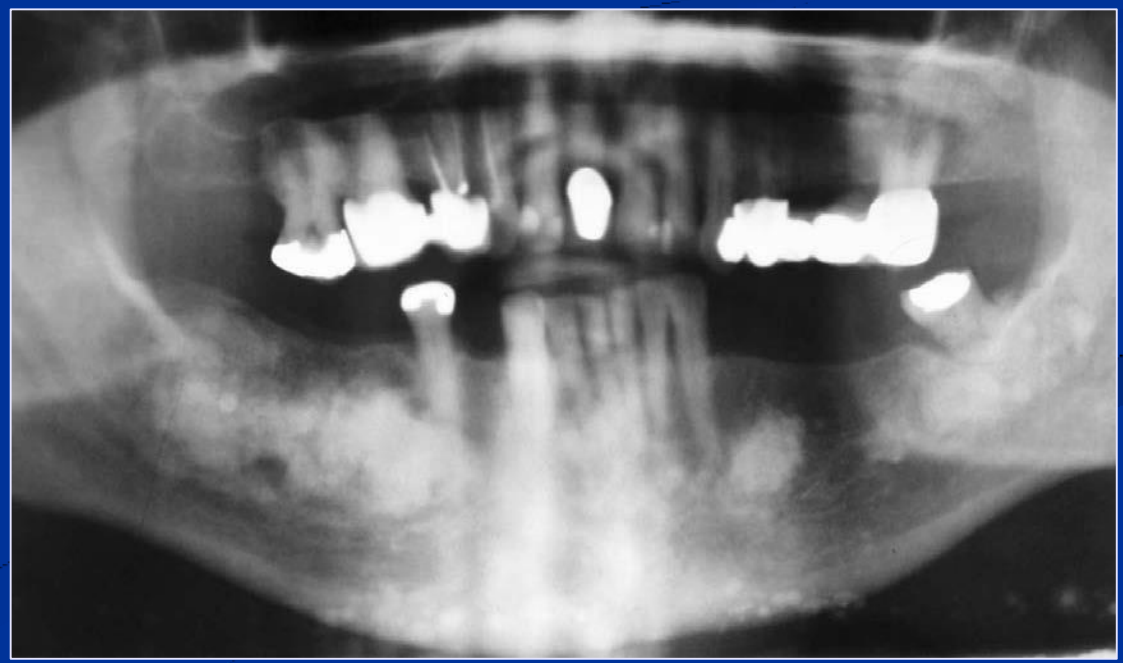
- Middle-aged
- Blacks
- Usually mandible
- Multiple quadrants involved



Florid Cemento-Osseous Dysplasia

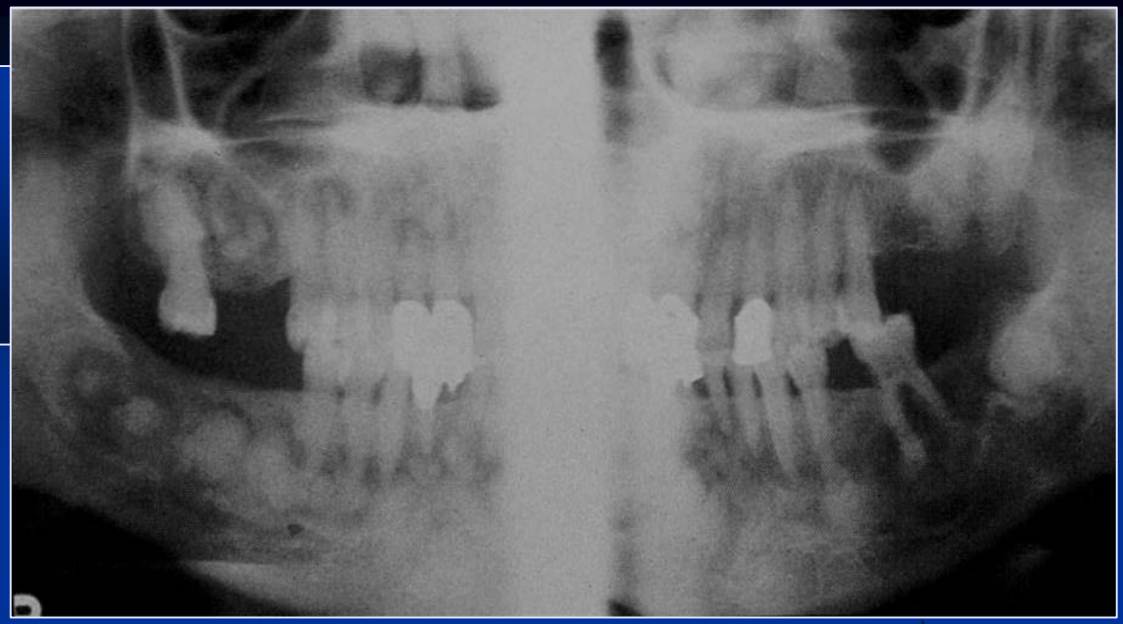
- Radiolucent/radiopaque
  -- Irregular “capsule” around it
- Multiple quadrants
- Maybe: expanded cortex
- Maybe: tenderness/pain
- Larger than focal/periapical types
- Usually some periapical lesions
- Maybe: surface exposure
  -- Rather avascular
- Maybe:
  -- Traumatic bone cyst(s)





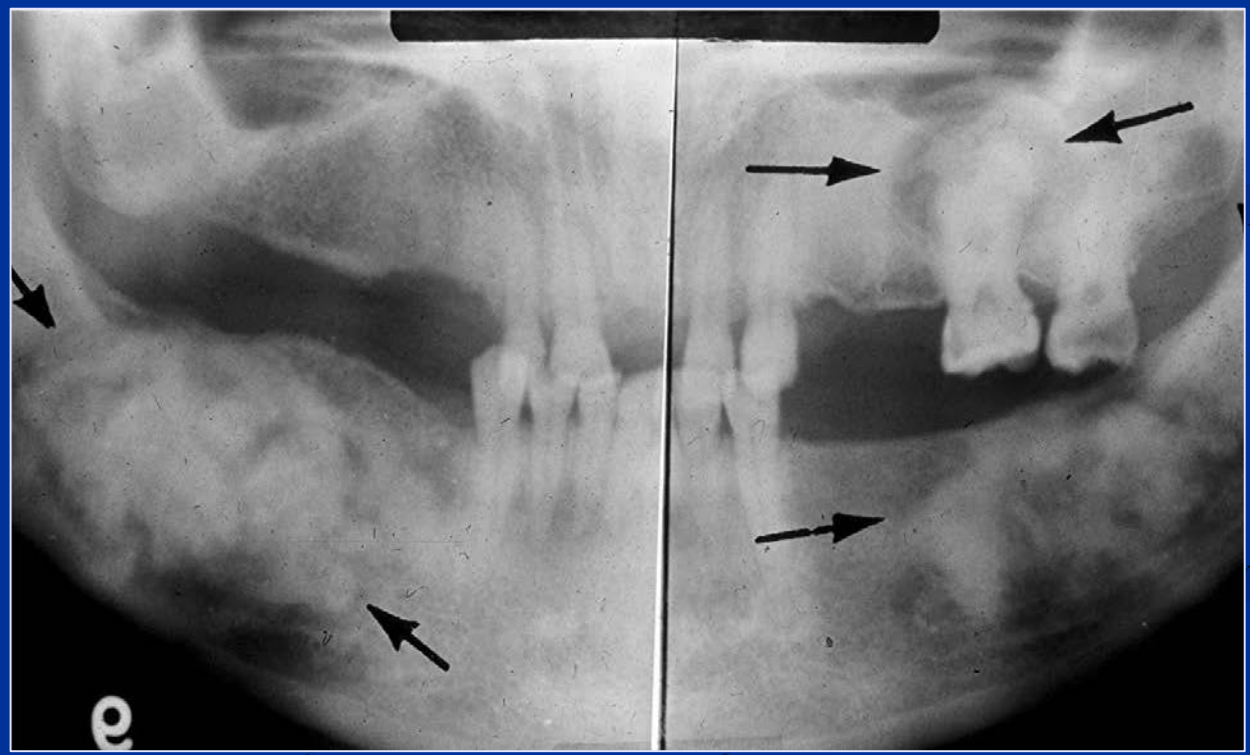
Florid Cemento-Osseous Dysplasia

Prognosis; Treatment



- Treat: none required
- If symptomatic: curettage
- Can extract the tooth
  -- Lesion is not attached
- Prevent infections
  -- Lesion is avascular

Familial Gigantiform Cementomas

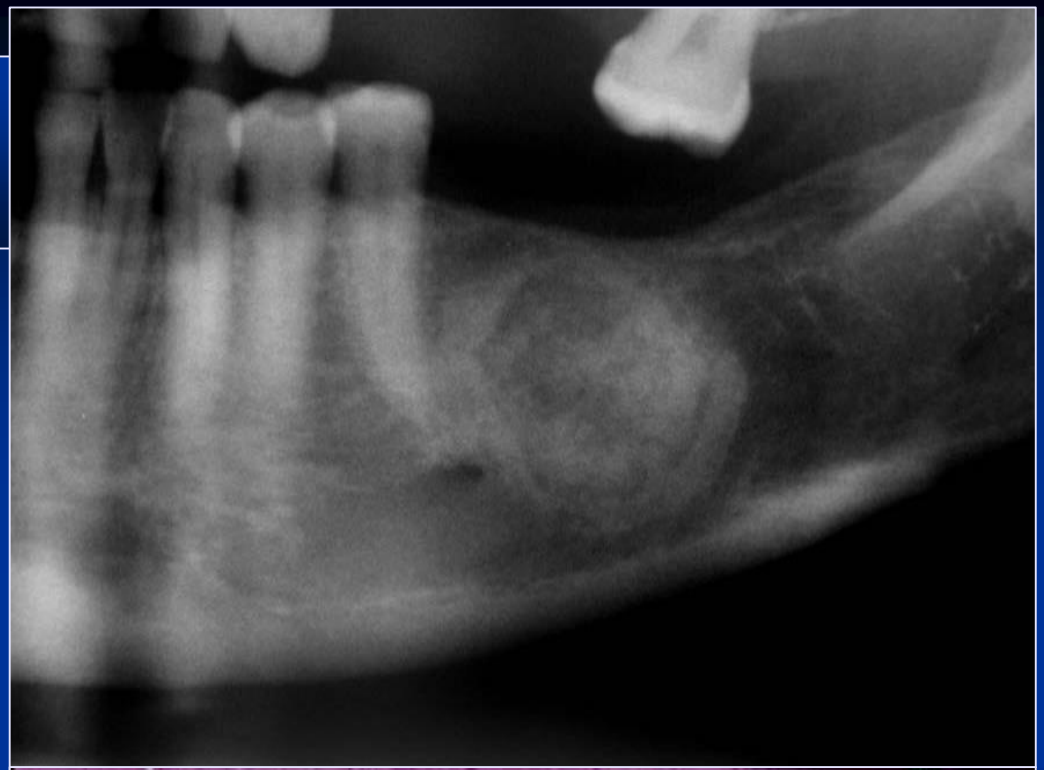


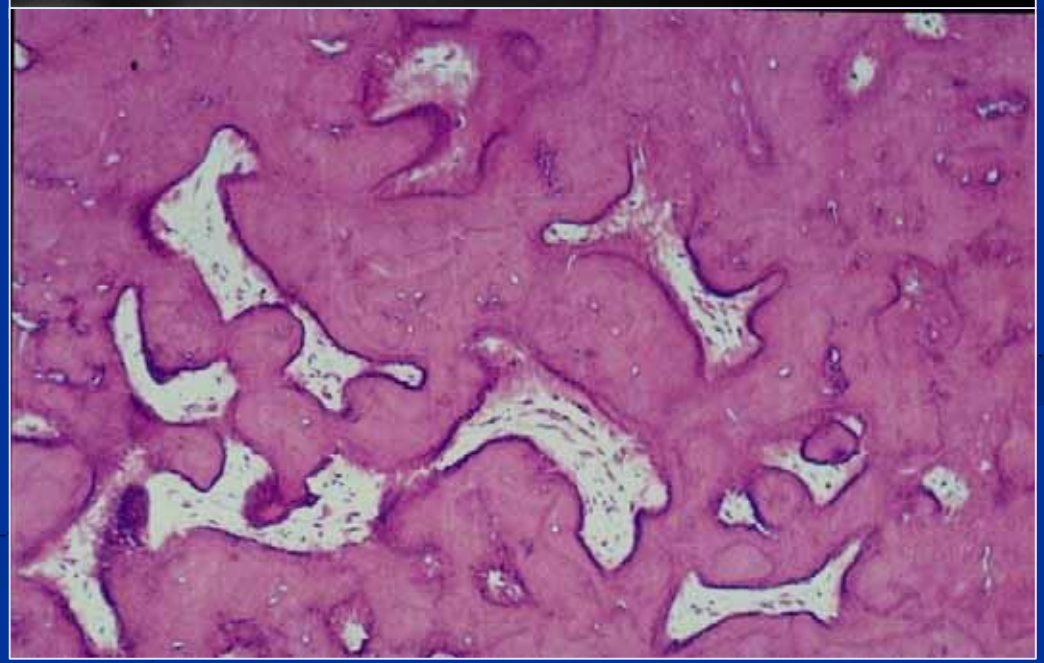
Familial Gigantiform Cementoma

- Benign fibrosseous lesion of jaw bone

Clinical features:

-- None
-- First and second decades
-- Posterior mandible
--Predominantly in Black females
--Slow growing,multifocal swelling involving multiple quadrantsof jaw
--Facial deformity
--Excessive cementum deposition





❑ Etiology :

-- After extraction or minor surgical procedures

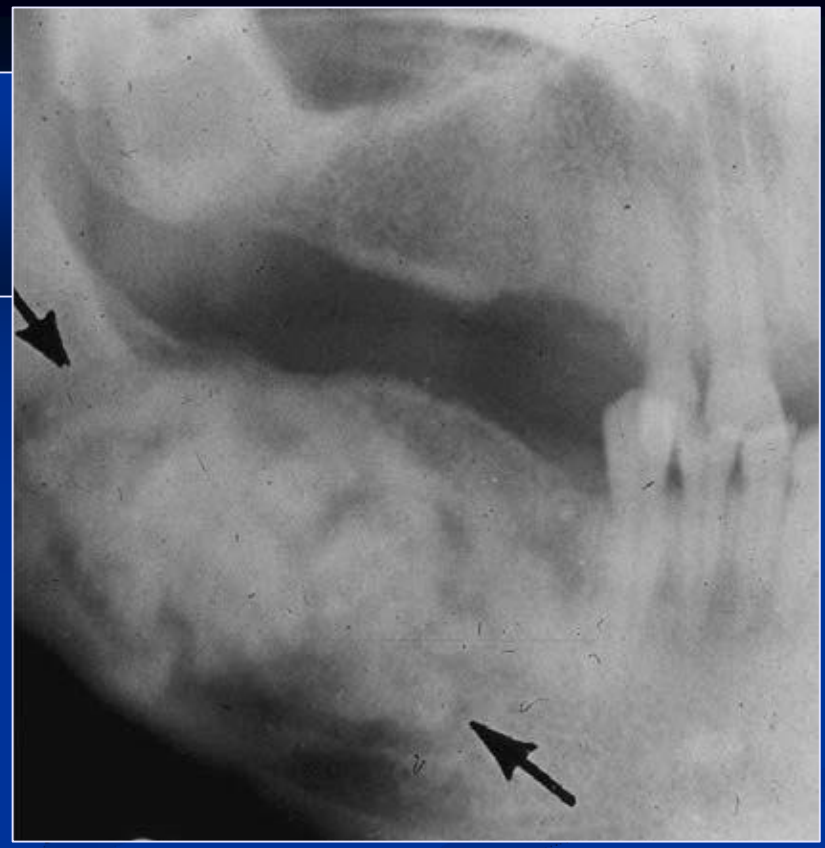
❑ Histopathologic features :

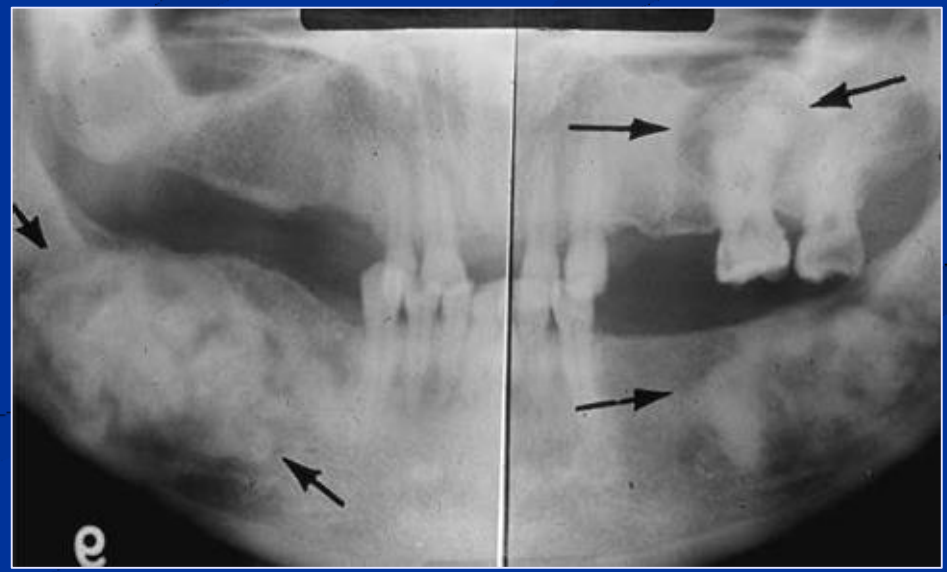
-- Dense cementum like, acellular, mineralised tissue in fibrous connective tissue

-- Limited amount of bone formation in some cases

Familial Gigantiform Cementoma

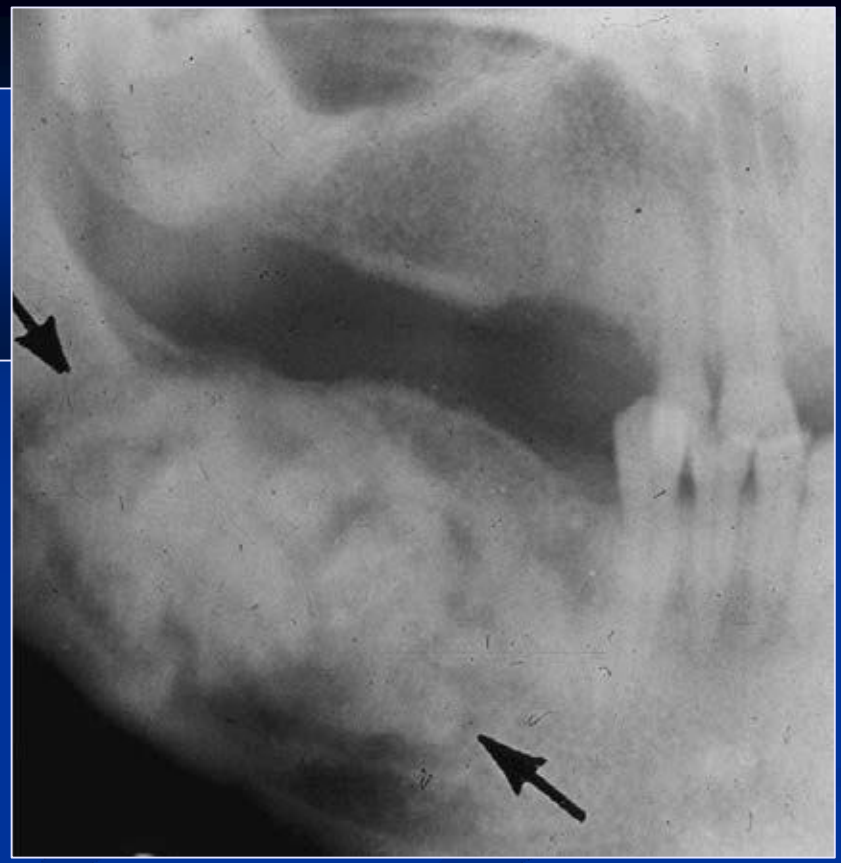
- Radiolucent/radiopaque
  -- Early: more radiolucent
  -- Late: more radiopaque
- Well demarcated
- Irregular “capsule”
- Asymptomatic, usually
- May be very large
- Impaction of teeth





Familial Gigantiform Cementoma

Prognosis; Treatment



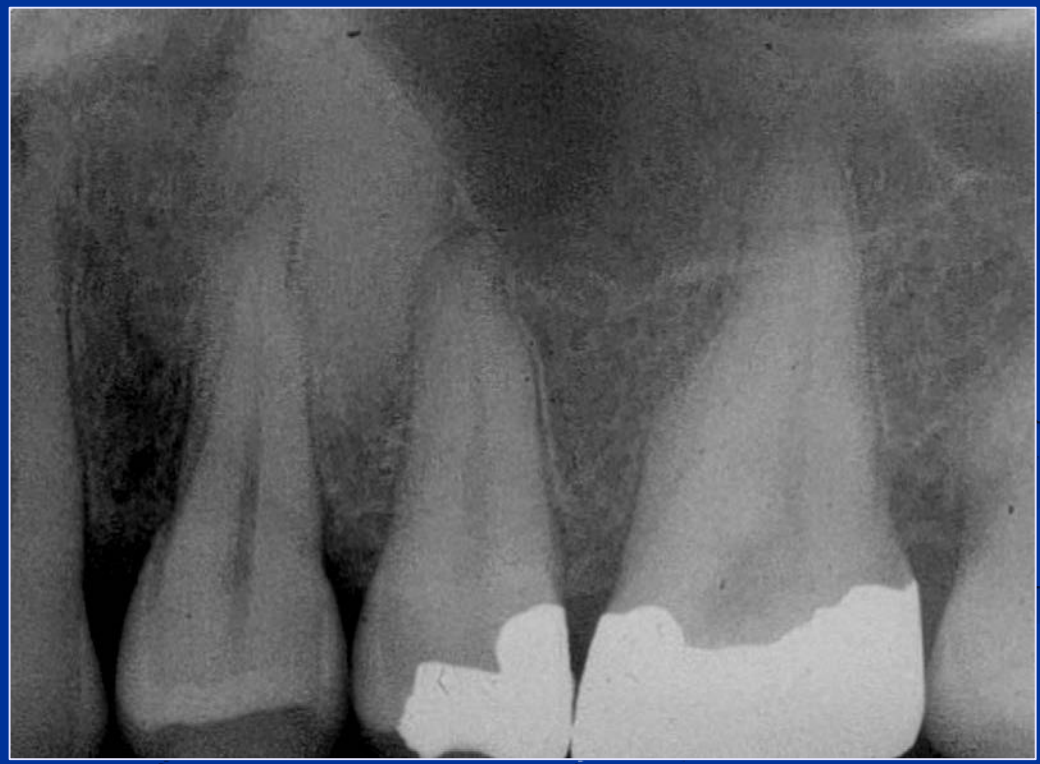
- Continues to enlarge until fifth decade
- Caution: watch for infection
  -- It's rather avascular

- Treatment: when predominantly radiopaque
  -- Surgical resection
  -- Recontouring
  -- May have to repeat

Inflammatory/Ischemic Bone Disorders

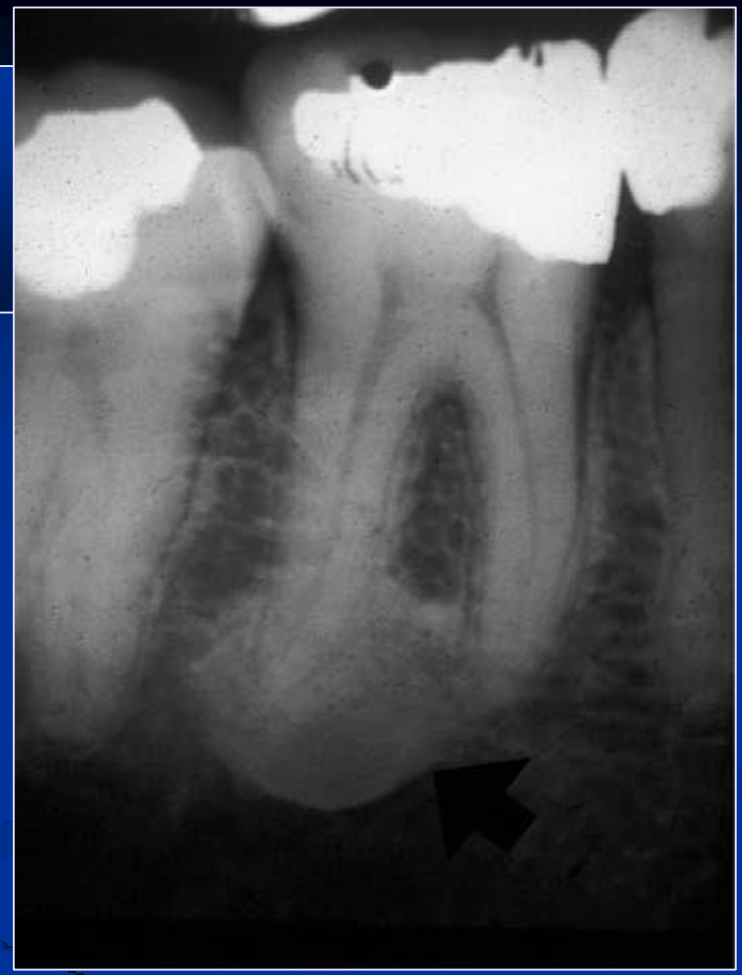
Name	Abnormality
Infantile cortical hyperostosis	Excess mandibular cortical bone
Bone scar	Exuberant bone response to infarction
Condensing osteitis	Exuberant bone response to infection
Osteitis	Inflamed bone
Acute osteitis	Dry socket, poor bone healing
Osteomyelitis, acute/chronic	Inflamed bone marrow or cortex
Ischemic osteonecrosis	Poor blood flow through marrow spaces

Condensing Osteitis



Focal Chronic Sclerosing Osteomyelitis

Condensing Osteitis; Ischemic Osteosclerosis?



- Etiology: exuberant bone “healing”
  -- After infection, trauma

- GENDER,AGE,LOCATION PREDILECTION:
- None
- Teenagers & young adults
- Most common mandibular first molar
- Usually painless
  -- Sometimes painful or tender

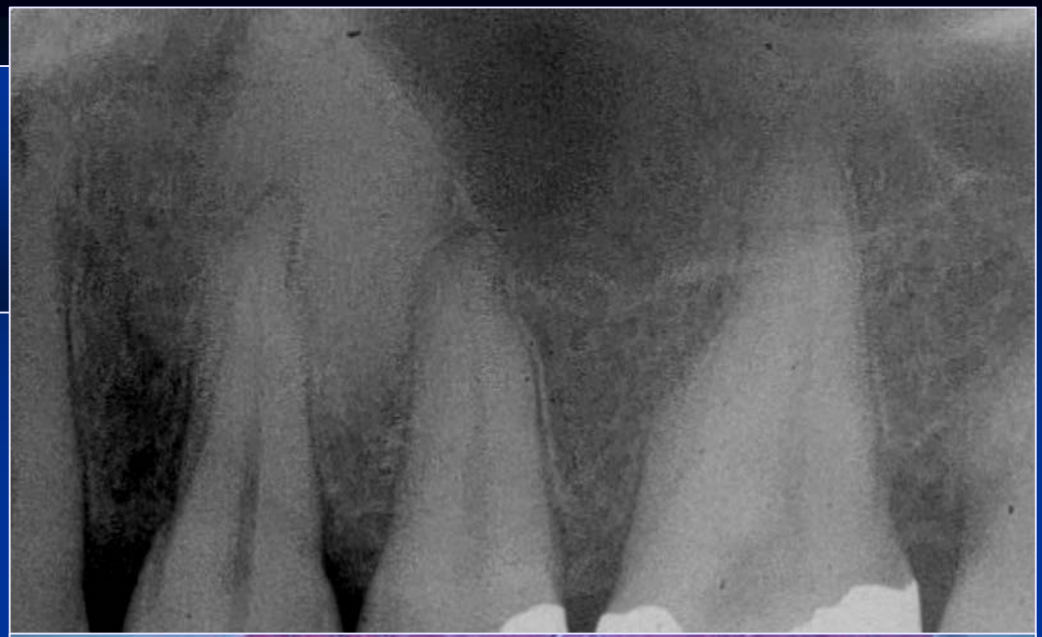
Focal Chronic Sclerosing Osteomyelitis
Condensing Osteitis

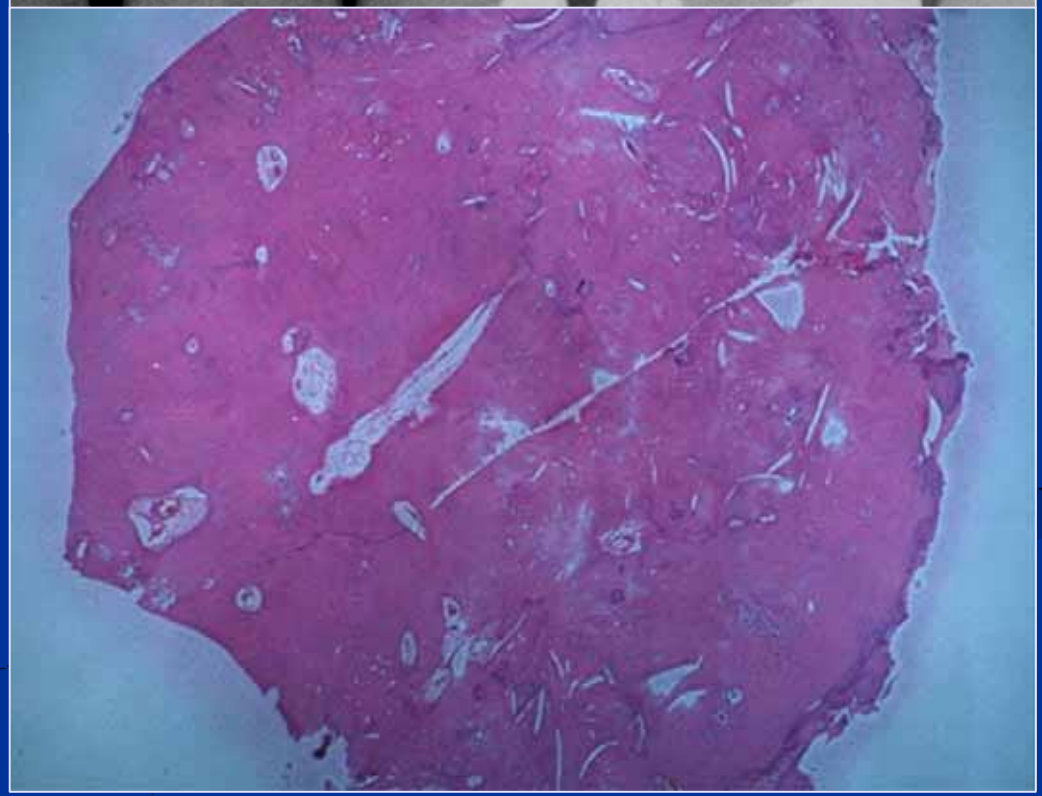
- Histopathologic features

-- Dense mass of bony trabeculae
-- Osteolytic lacunae are empty
-- Reversal and resting lines giving pagetoid appearance

- Radiographic features :

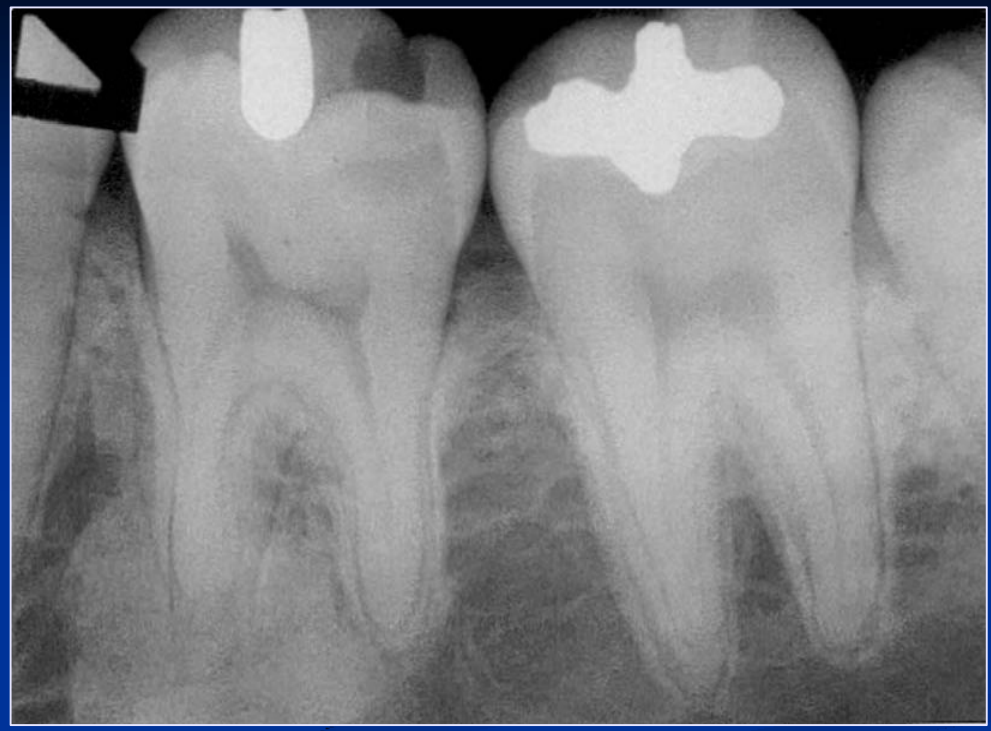
-- well-circumscribed radiopaque sclerotic mass of bone
-- intact lamina dura
-- location: apex of one or both roots





Focal Chronic Sclerosing Osteomyelitis

Condensing Osteitis



-- PDL widening is distinguishing feature from benign cementoblastoma

--Remains indefinitely once formed

--Bone scar

- Treatment :

--No treatment need unless painful

--If painful: surgical curettage