Pathology

Osteodystrophy

Learning Outcomes

By the end of the lecture, you will be able to:

- Define osteodystrophy
- Identify types of osteodystrophy-
- Clarify the underlying pathogenesis.
- Describe gross and microscopic pictures of osteod
- Recognize the effects and complication of different variant of osteodystrophy

Agenda

- Definition of osteodystrophy
- Types of osteodystrophy
- Pathogenesis of each type
- Gross and microscopy of each type
- Clinical effect and fate of each type

Osteodystrophy

Definition:

It is disturbance of bone growth.

Types of osteodystronhies:

(A)Congenital
(B) Acquired

Types of Osteodystrophy

Hereditary

Osteogenesis Imperfecta

Osteopetrosis

Achondroplasia

Acquired

- Fibrous dysplasia.
- Paget's disease
-Osteoporosis
- Radiation osteodystrophy .
-Renal osteodystrophy.
- Endocrinal osteodystrophy.
- Digital clubbing

Vitamin D deficiency

Osteomalacia Rickets

Osteogenesis Imperfecta

- It is a rare hereditary condition.
- There is a defect in connective tissue development or maturation. Bone cortex is formed mainly of woven bone and small areas of lamellar bone.

Clinically: - Patient is short

-Bone weakness (fractures with mild trauma).

-Patient has blue sclera (the partial visibility of choroid through the thin sclera).

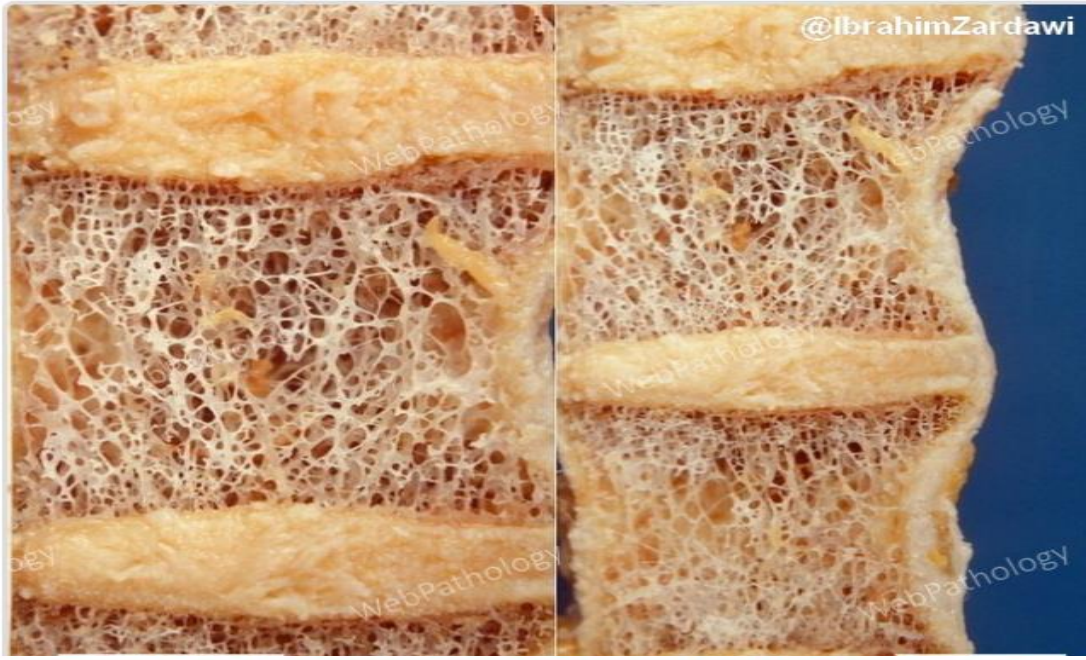
- Looseness of joints due to laxity of the ligaments.

- It may be associated with dentogenesis imperfecta (brown short teeth)

Osteogenesis Imperfecta

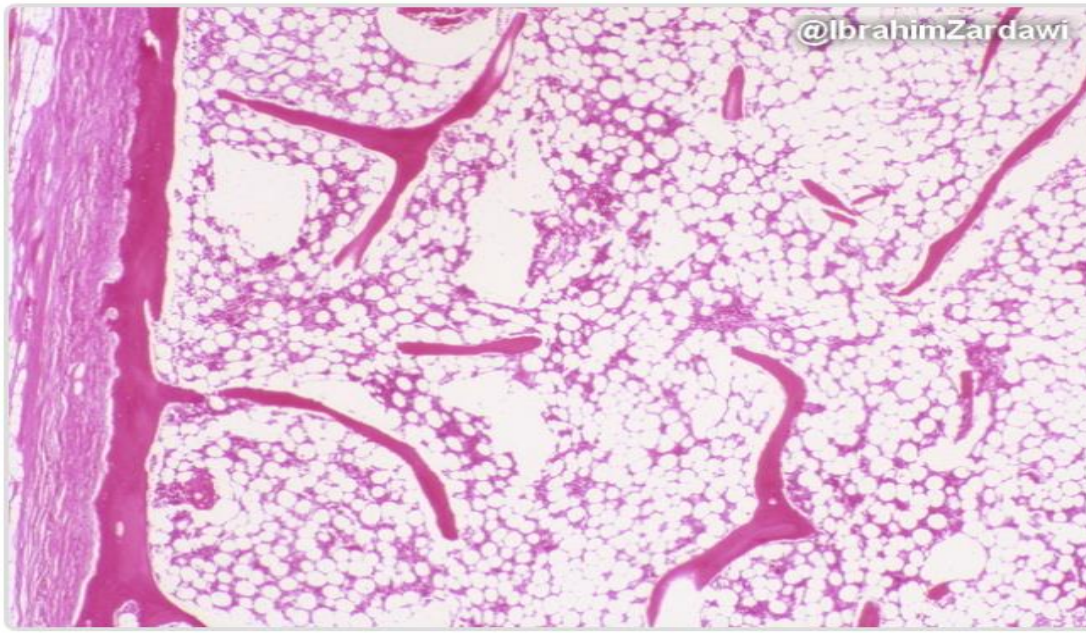
N/E:

- Osteoporotic vertebra show thinning and dropout of bone spicules.
- Horizontal trabeculae are thinner and there is loss of interconnectivity.



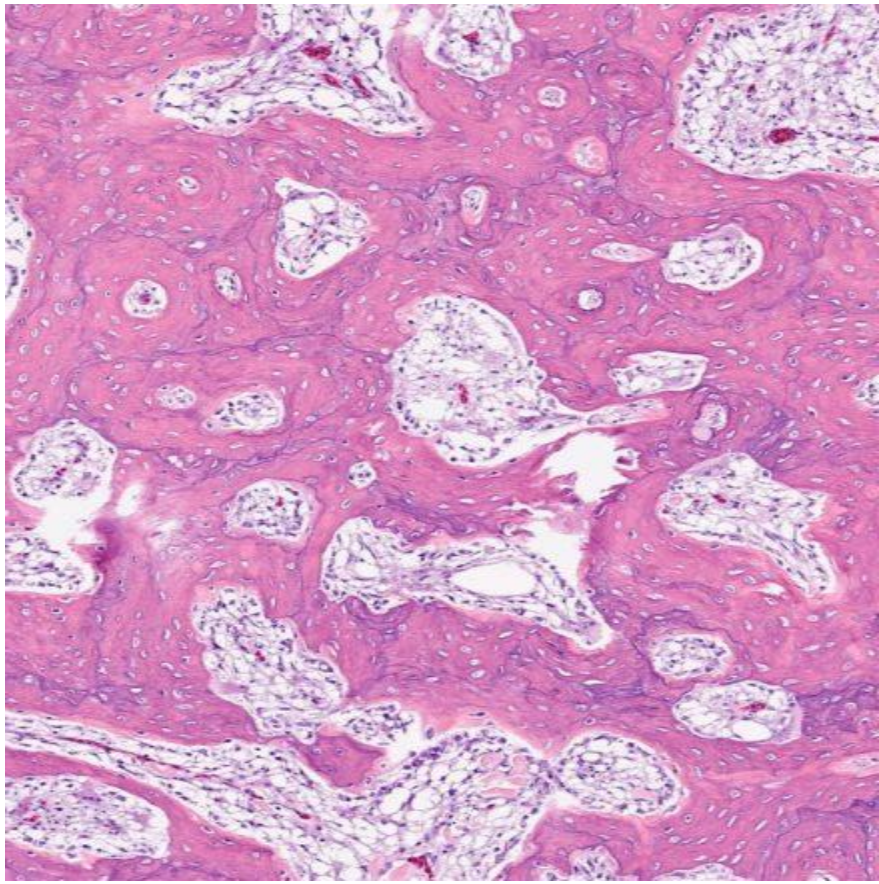
M/E:

- Thinning of the cortex .
- severe loss of cancellous bone
- markedly decreased interconnections between the bony trabeculae



Osteopetrosis

- It is a rare hereditary condition.
- Called Marble Bone Disease.
- Failure of the osteoclastic activity, thus no remodelling of the bone.
- Obliteration of the marrow cavities.
- Development of secondary anemia.
- The bone is fragile, has poor mechanical properties, thus fractures very easily.



Achondroplasia

- It is a disorder of bone growth.
- A common cause of disproportionate dwarfism
- A hereditary condition resulted in abnormally short stature with short limbs and relatively long narrow trunk.
- The head is large with prominence of the forehead, low nasal bridge.

Acquired Osteodystrophy

- Fibrous dysplasia.

- Paget's disease

-Osteoporosis

- Radiation osteodystrophy .

-Renal osteodystrophy.

- Endocrinal osteodystrophy.

- Digital clubbing

Vitamin D deficiency

Osteomalacia Rickets

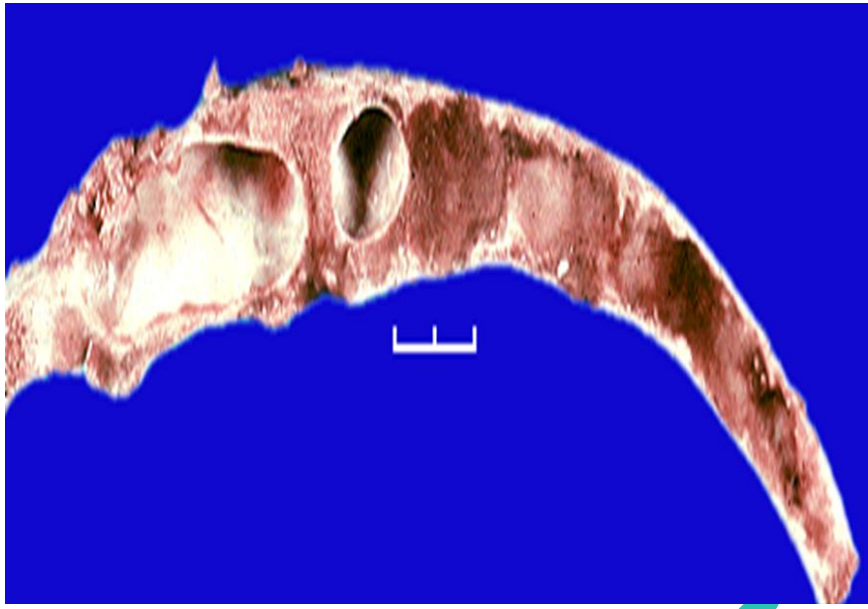
Fibrous dysplasia

An acquired disturbance of bone growth characterized by disorganized mixture of fibrous and osseous elements

Associated conditions: Skin pigmentation, endocrine dysfunction (Acromegaly, Cushing syndrome and hyperparathyroidism).

N/E

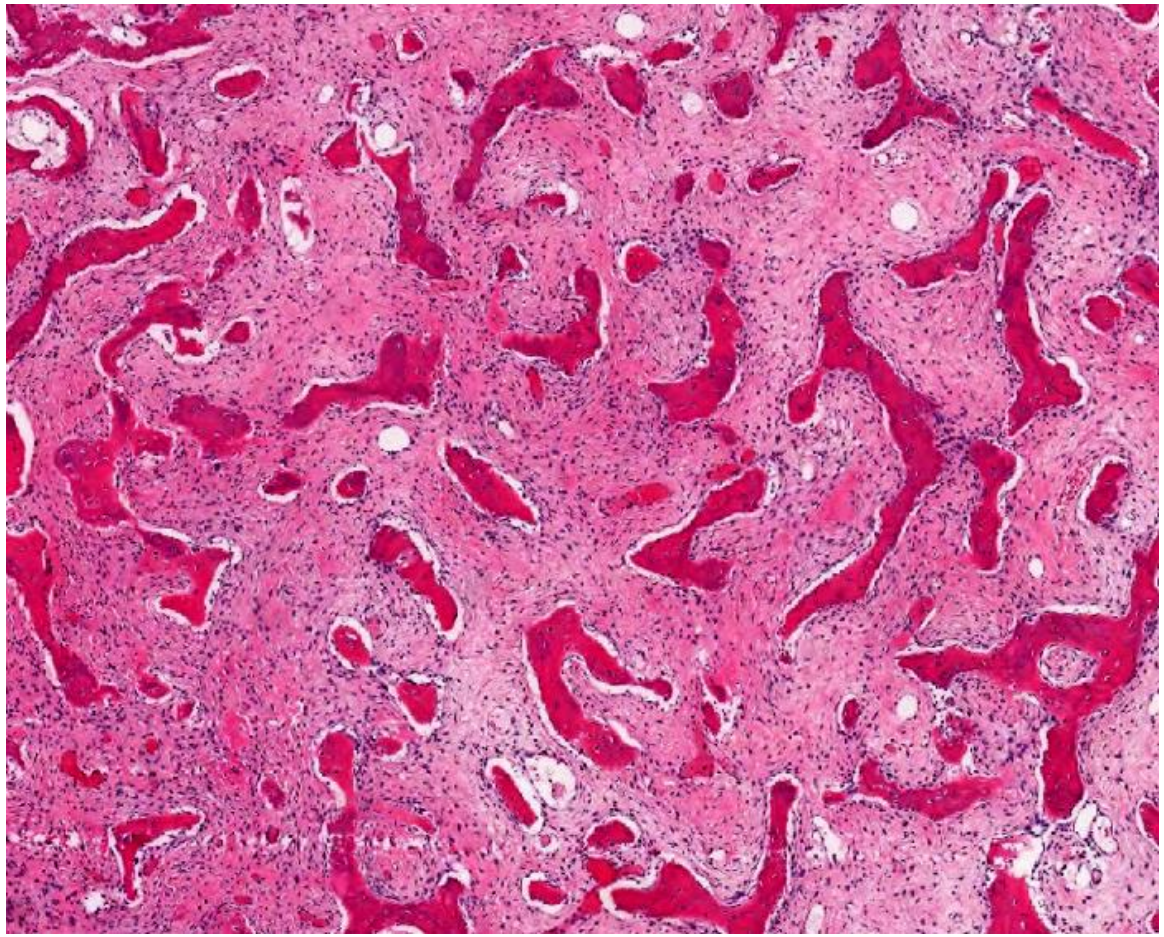
- It may be either polyostotic or monostotic.
- Site: In the medulla.
- Color: Grey white.
- Consistency: Gritty with cysts



Fibrous dysplasia

M/E: Irregular bone trabeculae embedded in a loosely arranged fibrous stroma. The bony trabeculae are formed of woven bone that lack osteoblastic rimming.

Complications: Deformities of bone and fractures.



Paget's disease of Bone

Acquired disturbance of bone growth characterized by bone resorption & new bone formation called Osteitis deformans

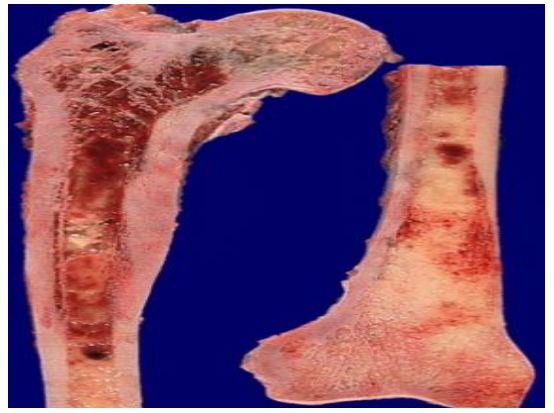
Clinically: age: after age of 40 years.

Sex: more in males.

Pathogenesis: It passes by 3 stages:

1. **Hot Stage:** Osteoclasts are over activated by a Virus > +++ bone resorption. So bone becomes rarified and easily fractured.
2. **Mixed stage:** Osteoblasts are activated to replace the destructed bone > +++ new bone formation which is irregular thick woven bone.
3. **Cold stage:** This stage is characterized by little cellular activity.

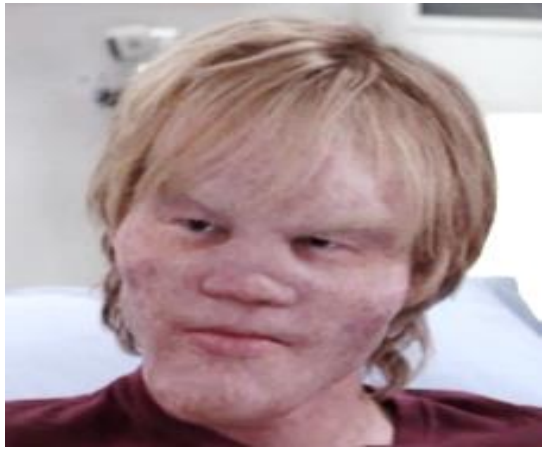
Paget's disease of Bone



N/E: It may be polyostotic or monostotic.

Sites: Skull, vertebrae and long bones

The bones show alternating coarse thickened areas and other lytic areas filled with blood



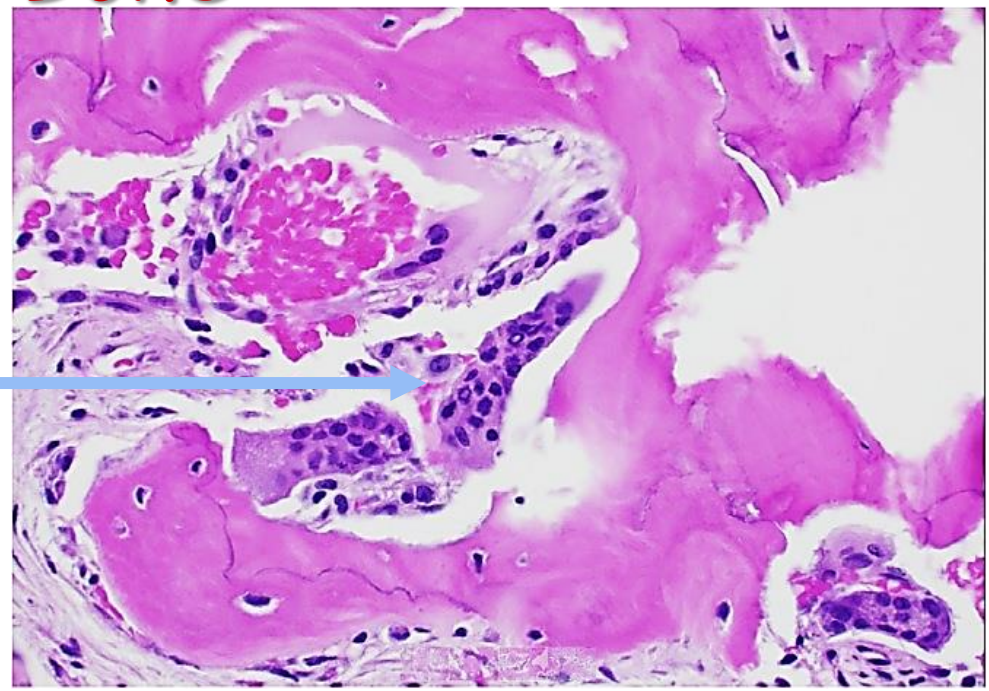
Causes bone deformeties:

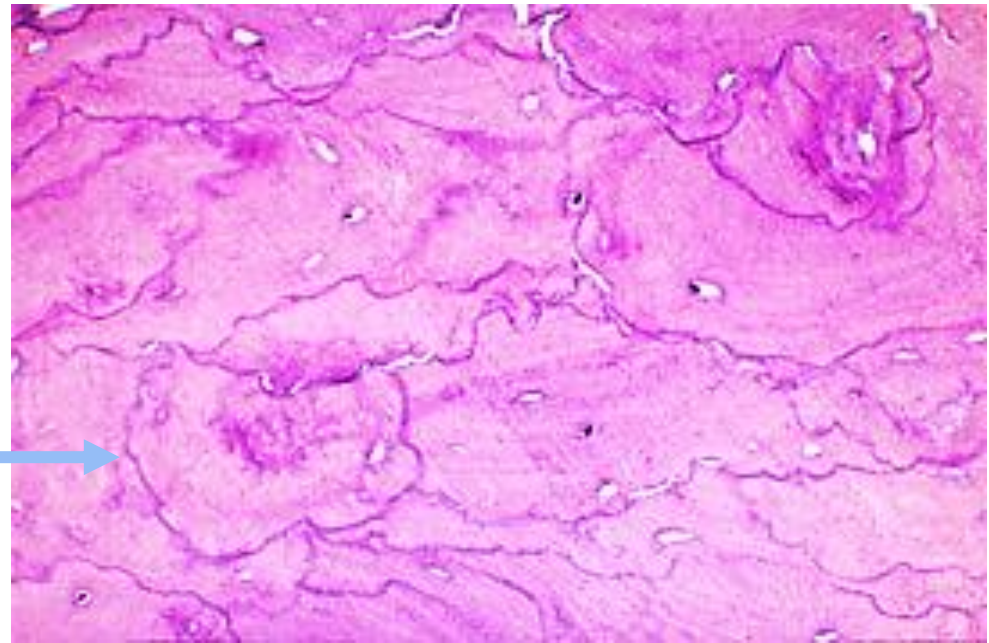
- lion face deformety of skull (leontiasis ossea)
- Kyphosis due to anterior collapse of vertebrae
- Forward bowing of legs.

Paget's disease of Bone

M/E:

- **thick, irregular, plate-like bony trabeculae** with multiple osteoclasts at the margin with several **resorption cavities**. The **marrow** space is filled with **fibrovascular tissue** containing scattered osteoclasts.
- The thickened bony trabeculae show mosaic appearance





Paget's disease of Bone

Complications:

1 - Fractures.

2- High output heart failure due to local increase of bone blood flow.

3- Malignancy: Mainly osteosarcoma (of old age).

Other malignancies: Fibrosarcoma, chondrosarcoma and giant cell tumor.

Vitamin D Deficiency
Rickets

Defect in bone mineralization in childhood.

Age: It starts after 6 months (after exhaustion of calcium stores formed during intrauterine life).

Etiology: Deficiency of vitamin D, C and phosphorous due to:

- Insufficient exposure to sunlight.
- Decreased intake or absorption of vitamin D.
- Pre-maturity due to increased demands of Ca and due to defective hydroxylation of vitamin D in the liver.

Rickets

Pathogenesis of rickets:

- Defective calcium level → cartilage is not calcified → cartilage cells don't degenerate, continue to proliferation & lay down chondroid matrix → ticking of ends of long bones
- New bone → irregular & poorly calcified

N/E of Ricket:

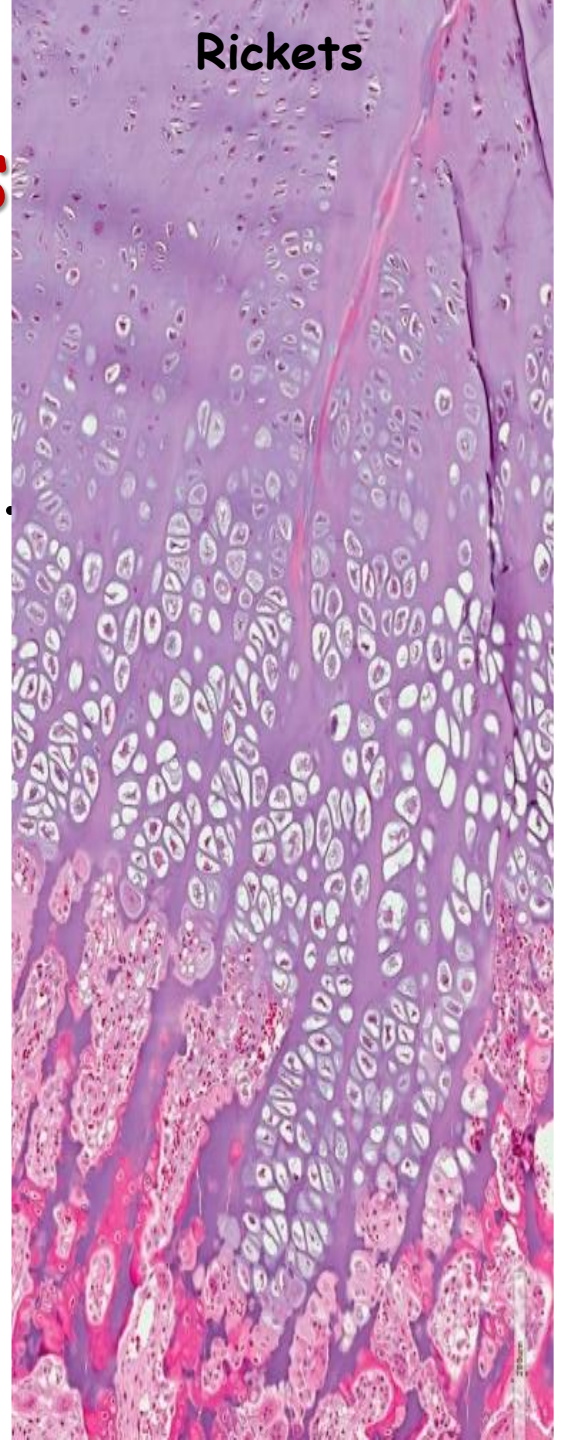
- Enlarged soft epiphysis (cut with knife).

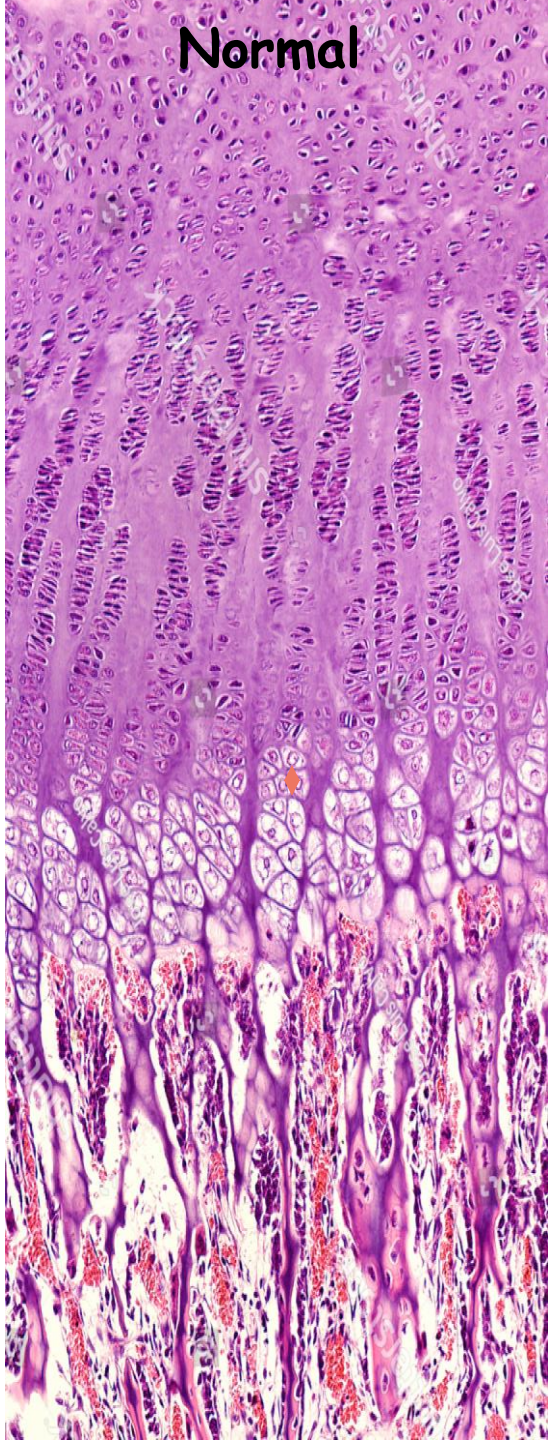
-The line between epiphysis and diaphysis is irregular.

Rickets

M/E of the cartilage plate in cases of rickets:

- The resting zone is relatively normal.
- The proliferating zone is widened and disorderly arranged.
- The zone of provisional calcification is poorly defined.
- Minimal amount of bone is formed.
- Areas of proliferating cartilage extend on the expense of metaphysis.





Rickets

Clinical picture:

General lesions:

1- Protrusion of the abdomen

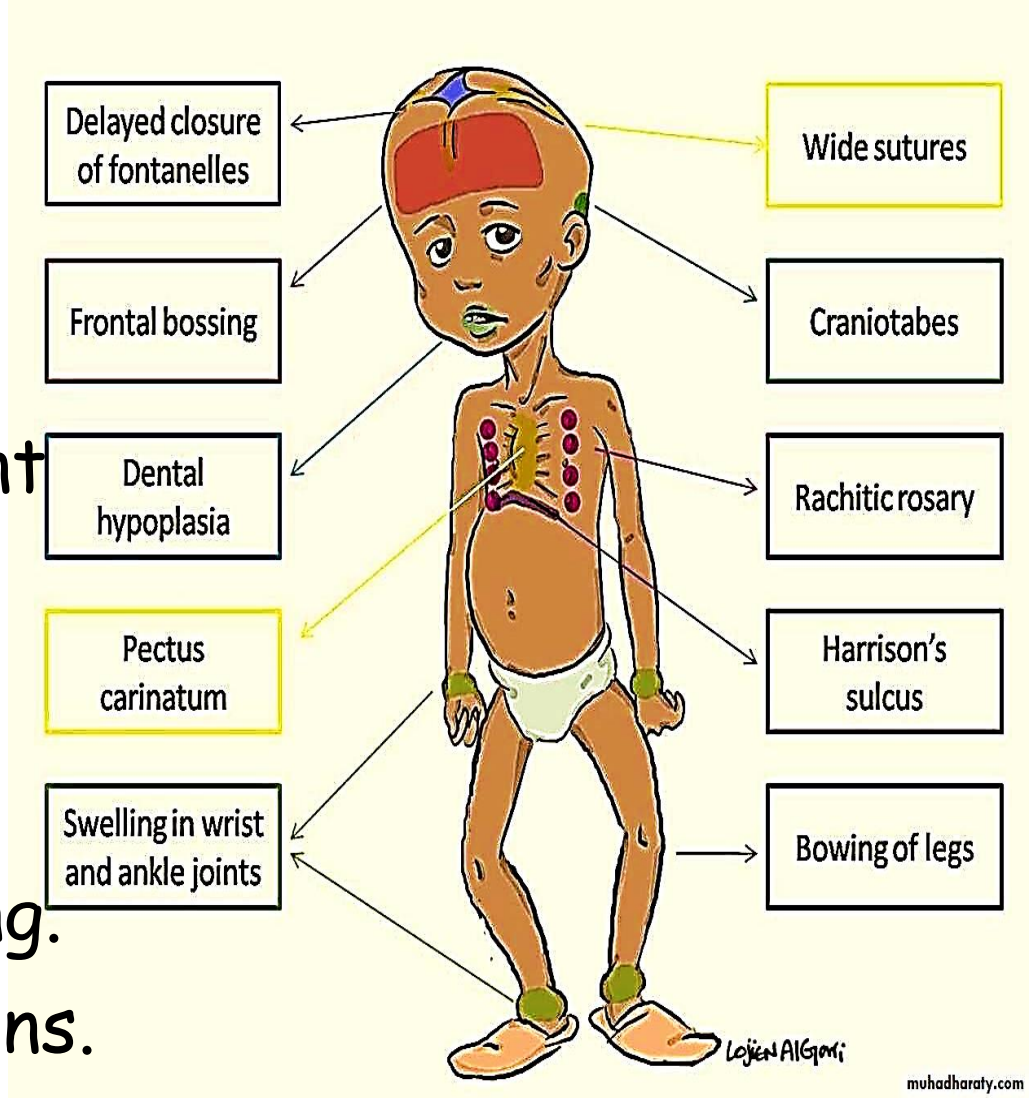
2- over weight due to limited movement

3- Generalized lymphadenopathy and splenomegaly.

4- Anemia

5- Delayed standing, sitting and walking.

6- Increased susceptibility to infections.



Rickets

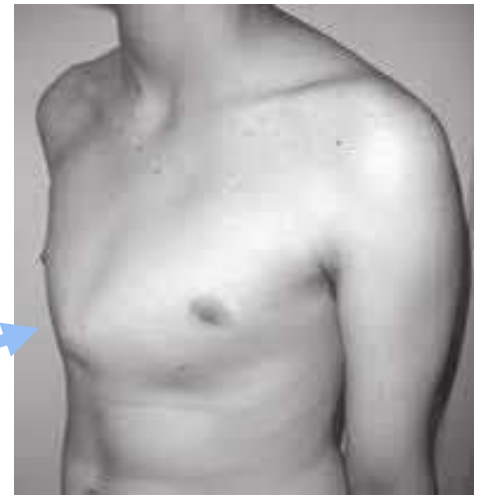
Skeletal changes :

1 -Skull:

- Delayed closure of fontanelles and sutures.
- Craniotabes (flat occipital bone due to pressure).
- Square shaped head and frontal bossing.
- Delayed dentition.

2- Chest:

- Rosary chest (prominence of the costochondral junctions).
- Pigeon chest (Flattening of the sides and protrusion of the sternum).
- Harrison's sulcus (a horizontal groove at the lower part of the chest at the site of attachment of the diaphragm).

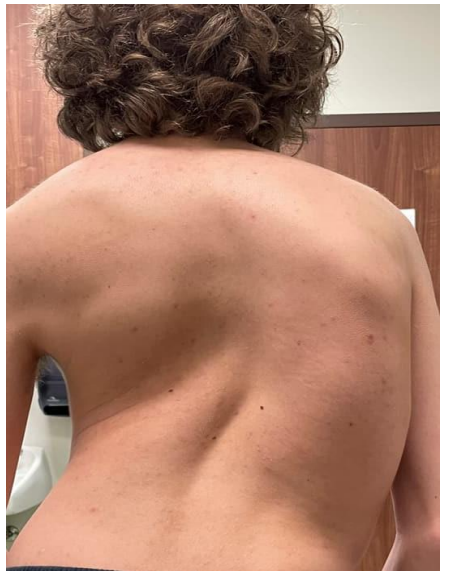


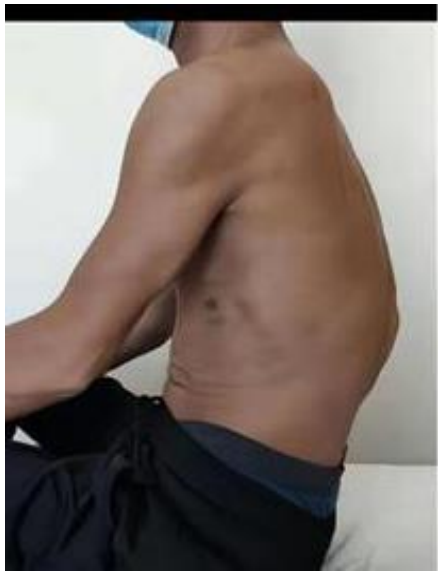
Rickets

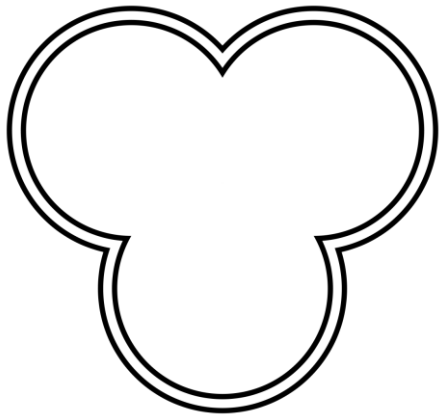
3- Vertebrae: Kyphosis, lordosis or scoliosis

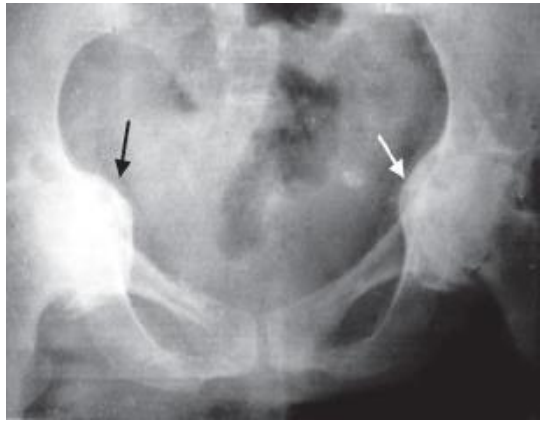
4- Long bones: Bowing of legs and prominence of epiphyseal cartilage.

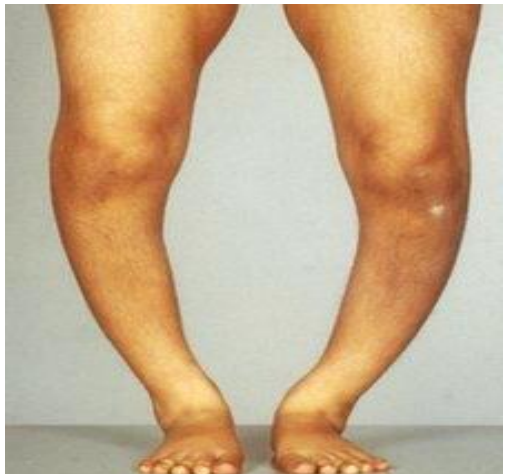
5- Pelvis: Trifoil pelvis.

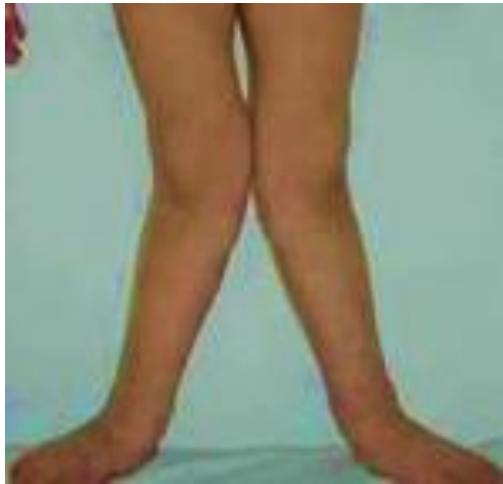












Osteomalacia

Definition: Defective bone mineralization in adult.

Causes: Vitamin D deficiency.

Insufficient exposure to sunlight

Increased demands (repeated pregnancies & lactation)

Pathogenesis:

Failure of calcification of bone matrix resulting in bone softening.

Clinical picture:

Lumbar lordosis

Bending of femur and tibia.

Contracted pelvis

Osteoporosis

Definition: It is reduction of bone mass to a level below that required for normal bone support. However, mineralization is normal.

Causes:

*Localized: Prolonged immobilization (paralyzed limb).

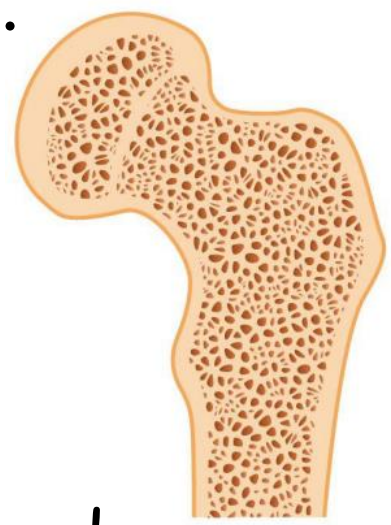
*Generalized:

1- Senile : Occurs in postmenopausal women and elderly men.

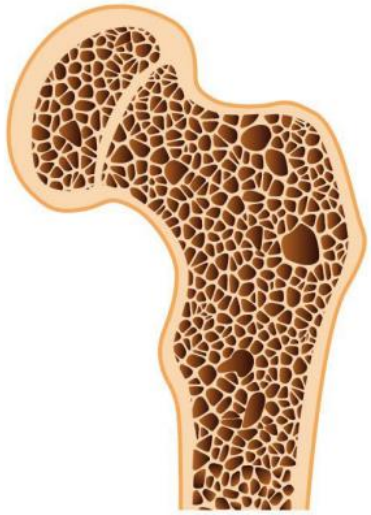
2- Scurvy: Vitamin C is essential for the synthesis and proper structure of collagen

3- Collagen disturbances (Osteogenesis Imperfecta)

4- Endocrinal.



Healthy bone



Osteoporosis

Radiation osteodystrophy

Exposure of the growth cartilage plate to radiation field leads to premature closure of epiphysis.

Renal osteodystrophy

- Damaged glomeruli lead to Phosphate retention
- Tubular damage lead to decreased 1,25-dyhydroxy vitamin D
- With renal failure there is decreased intestinal absorption of Ca : hypocalcemia

Endocrinal osteodystrophy

- Steroids lead to increased rate of bone resorption.
- Hyperthyroidism > increase rate of bone resorption.
- Hyperparathyroidism lead to mobilization of calcium from bone.

Digital clubbing

- It affects mainly the fingers. Nails are elongated, curved, raise and cyanotic.
- Phalanges are thickened with subperiosteal new bone formation.

Causes: Hypoxia, toxaemia, chronic lung diseases.

Now….Answer this

Which of the following characterizes Osteogenesis imperfecta?

a) Thickening of cortex.
b) Thinning of cortex.
c) Increased interconnected bone trabeculae.
d) Acquired osteodystrophy.
e) Normal bone development and maturation.

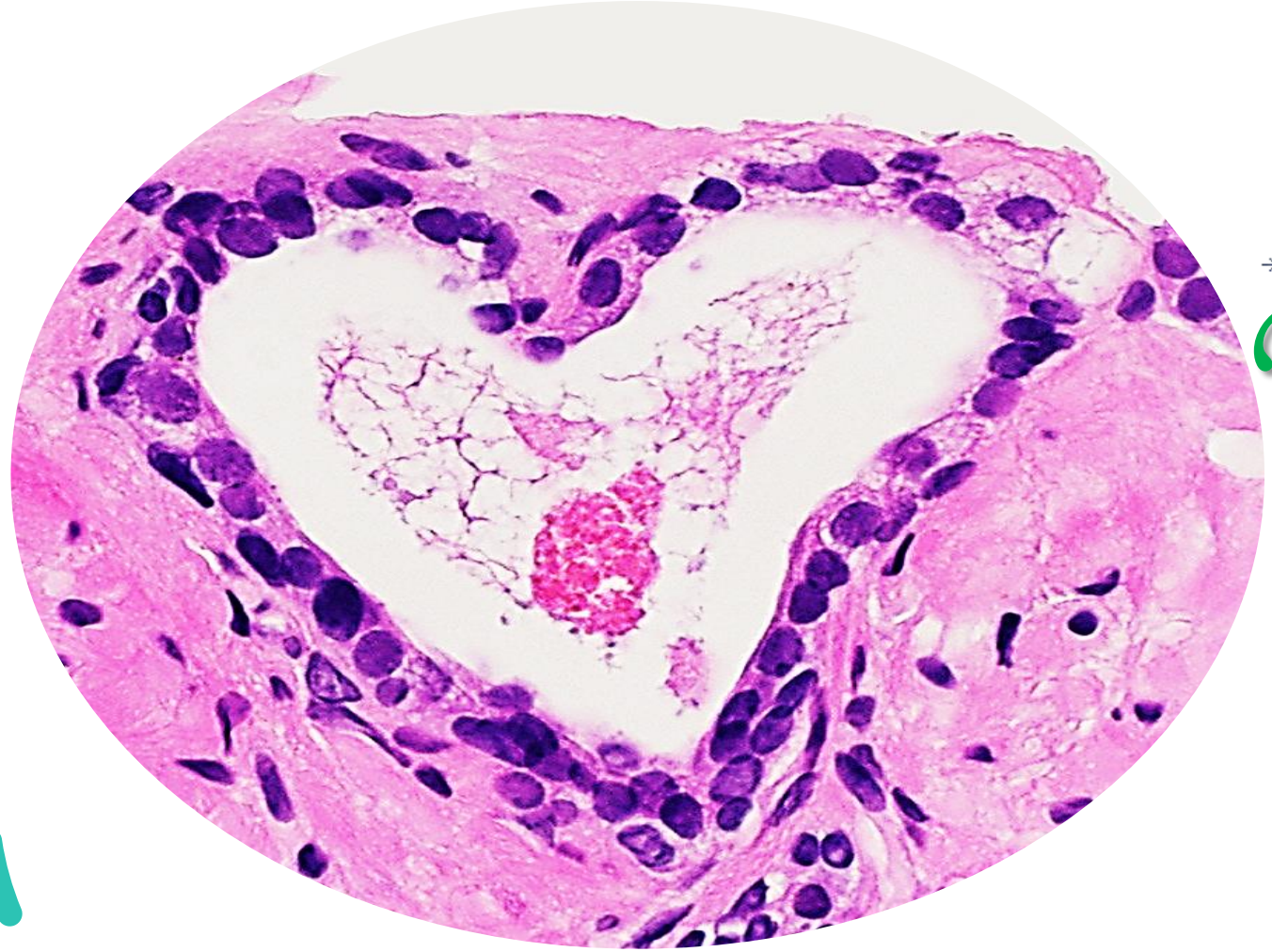
Discussion & Feedback

References & recommended readings

1. Robbins & Cotran Pathologic Basis of Disease, (Robbins Pathology), 2018 ISBN: 978-0-323-35317-5, Edition: 10th
2. Webpath:

   https://webpath.med.utah.edu/webpath.html

   https://www.pathologyatlas.ro/index.php



Thank you