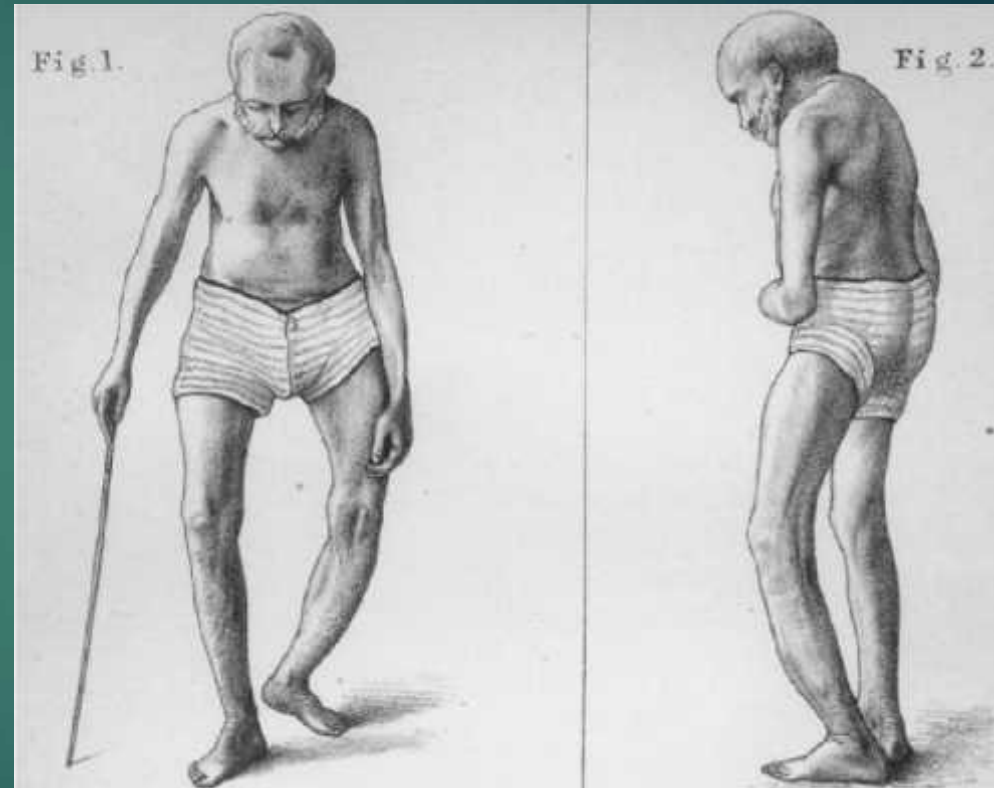


# PAGET'S DISEASE

ELIZA STAVRIDE MD, MSC  
RADIOLOGY DOCTOR



# Sir James PAGET 1814 - 1899

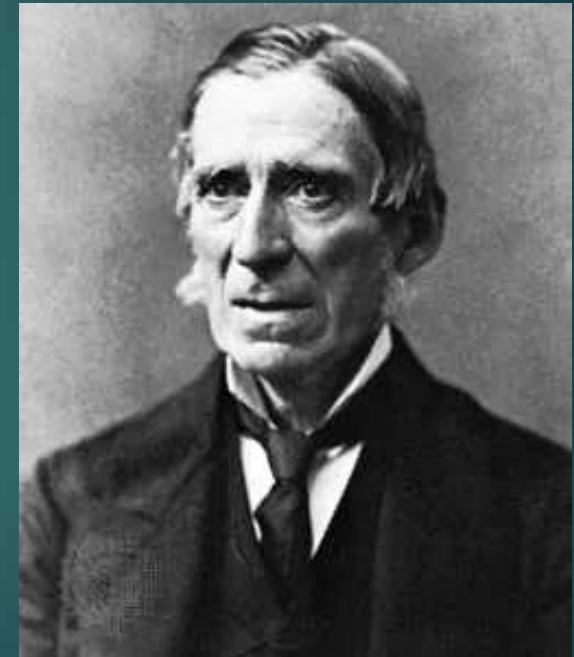
- ▶ Paget's disease of the bone (1877)



*Osteitis deformans*

1888 → Sir Jonathan Hutchinson

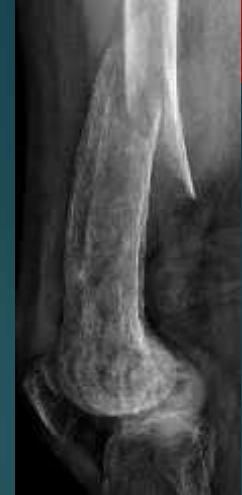
- ▶ Paget's disease of the breast (1874)
- ▶ Paget's - Schroetter Syndrome (1858)



# PAGET'S DISEASE



Abnormal remodelling of the bone  
↓  
Bone less organised and weaker than normal  
↓  
Deformity and Fracture



- Environmental factors
- Genetic factors
- Viral infection of bone cells



# Demographics

**Age** older than 40 – 50

**Sex** men > women

**National Origin** Great Britain, Central Europe, United States, Greece

rare → Scandinavia, Asia, Africa



# 3 Phases

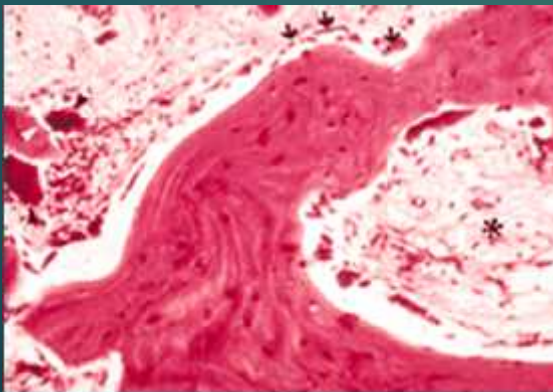
## Lytic phase



incipient – active



osteoclasts



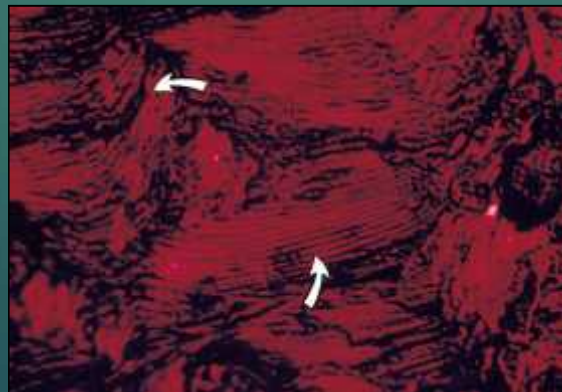
## Mixed phase



active



osteoclasts + osteoblasts



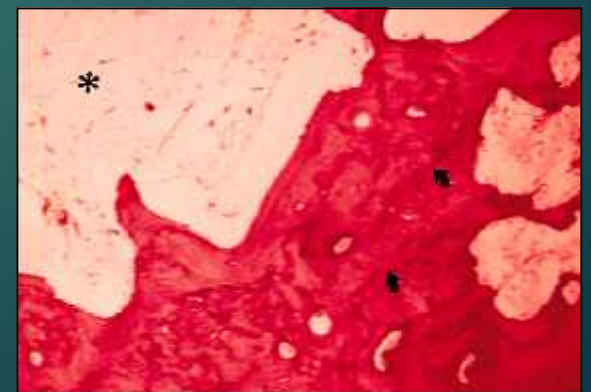
## Blastic phase



late – inactive



~~osteoblasts~~



# Distribution

- ❖ Pelvis ( 30 – 75% )
- ❖ Spine ( 30 – 75% )
- ❖ Skull ( 25 – 65% )
- ❖ Femur ( 25 – 35% )



## Monostotic Disease

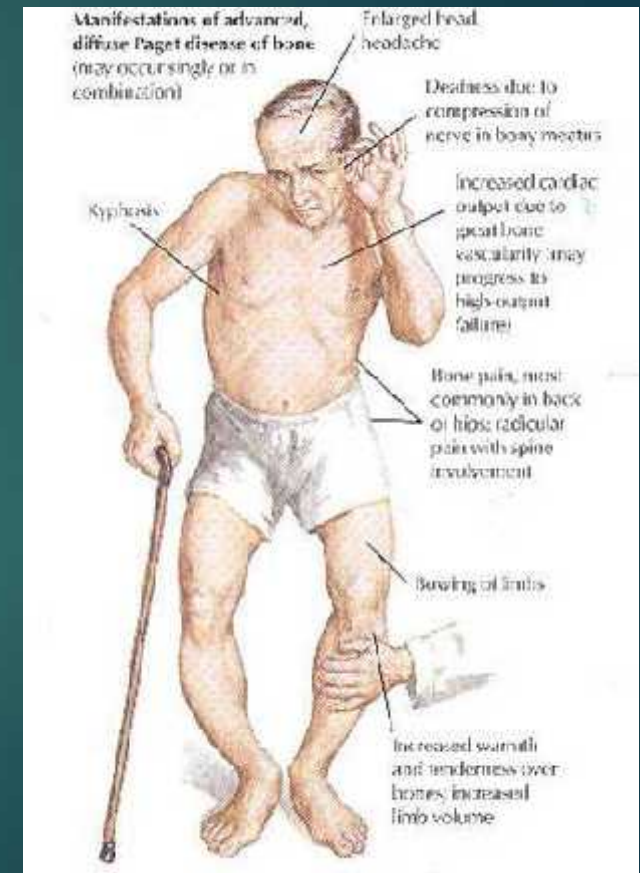
- ✓ 10% – 35%
- ✓ More often in axial skeleton

## Polyostotic Disease

- ✓ 65% - 90%
- ✓ Right sided predominance
- ✓ Usually involves lower extremities

# Clinical Findings - Complications

- ▶ Fractures and bony deformity
- ▶ Secondary osteoarthritis (when around joint)
- ▶ Neurological complications – nerve root compression and cauda equina syndrome
- ▶ Skull involvement
  - Deafness
  - Vertigo
  - Tinnitus
  - Cranial nerve disorders
- ▶ Sarcomatous transformation – osteosarcoma, malignant fibrous histiocytoma, chondrosarcoma
- ▶ Increased bone vascularity – high output heart failure



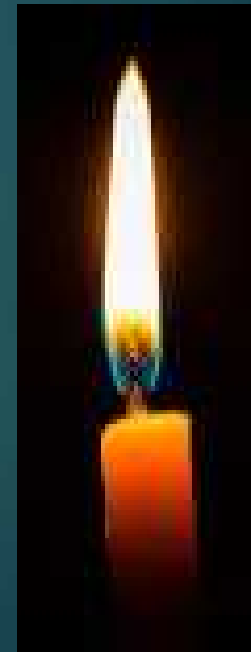
# Imaging

- ▶ Radiography → Pathognomonic Findings
- ▶ Computed tomography
- ▶ Magnetic Resonance Imaging



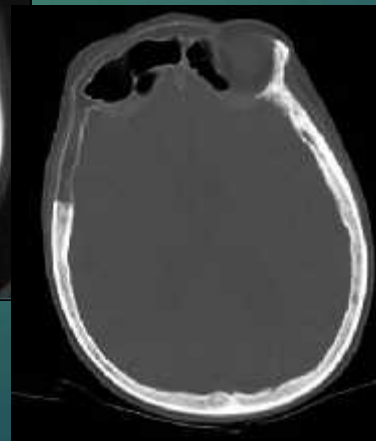
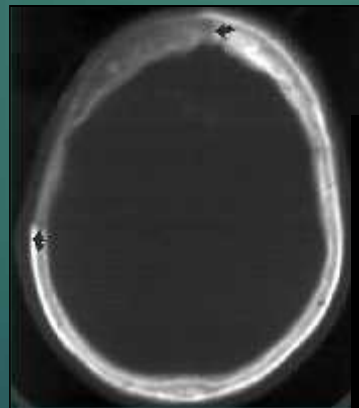
# Lytic Phase

- ▶ Osteoporosis Circumscripta
- ▶ Blade of grass - Flame



# Osteoporosis Circumscripta

- ▶ Skull
- ▶ Well defined large areas of radiolucency
- ▶ Frontal and occipital bones
- ▶ Involves both inner and outer calvarial tables
- ▶ Lack of peripheral sclerosis



# Blade of grass Flame

- ▶ Long bones
- ▶ Begins as a subchondral area of lucency
- ▶ Tip of V-shaped osteolysis
- ▶ Extends towards diaphysis



# Remember!!!

In long bones Paget's disease always starts at the **end** of the bone

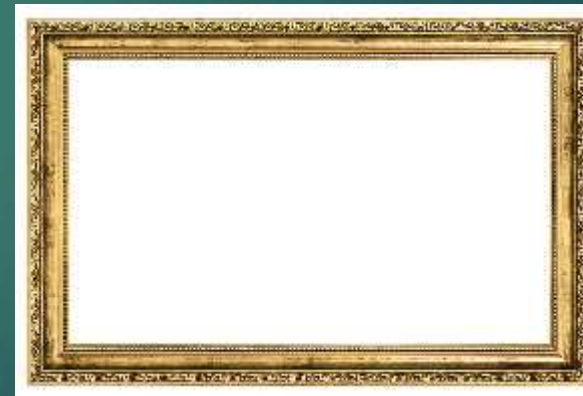
**Exception:** Tibia

→ Lesion in the middle of a long bone with no extension to either end ~~is possible~~



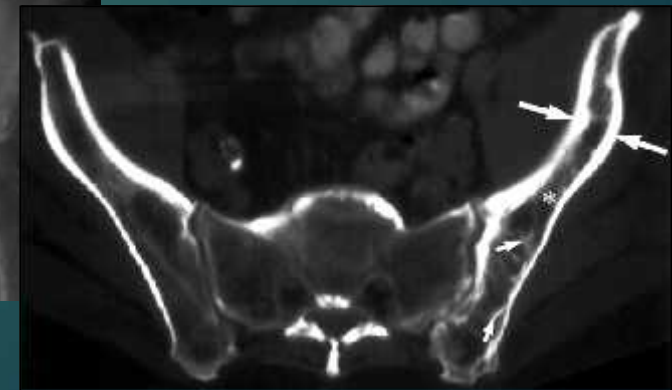
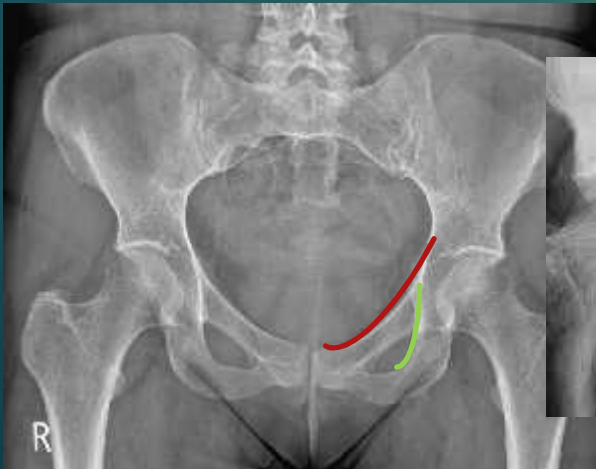
# Mixed Phase

- ▶ Pelvic brim sign
- ▶ Picture frame vertebra



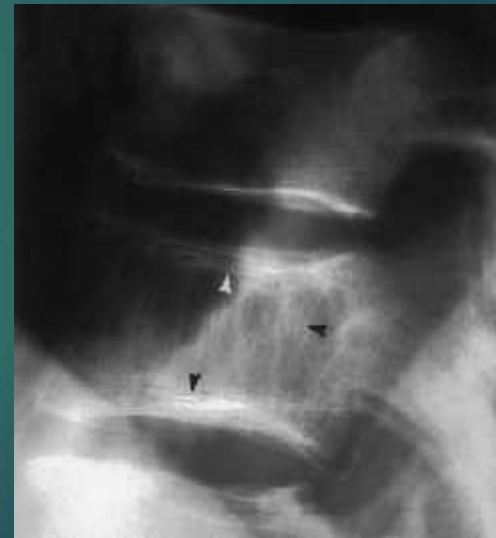
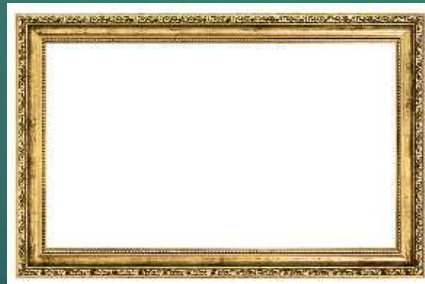
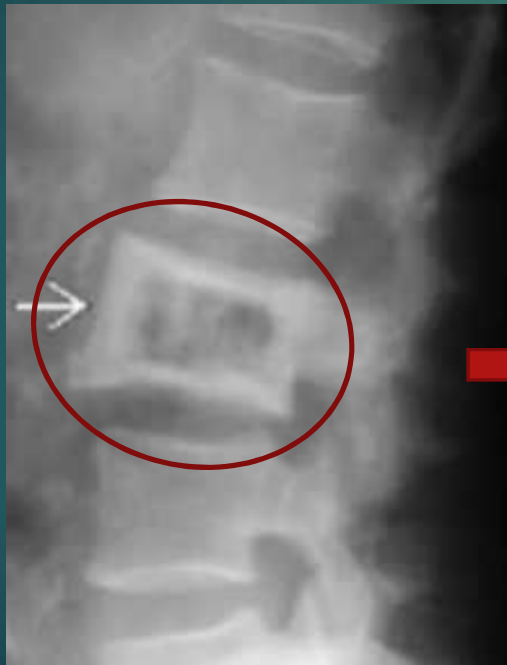
# Pelvic brim sign

- ▶ Cortical thickening of **iliopectineal** and **ilioischial** lines
- ▶ The iliac wing may be involved
- ▶ Often asymmetric
- ▶ Commonly **RIGHT** side

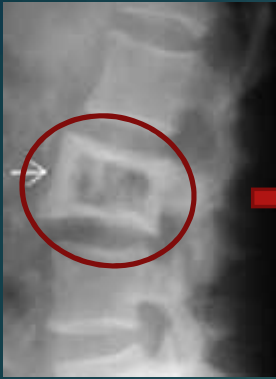


# Picture frame vertebra

- ▶ Thickened cortex of the vertebral bodies
- ▶ Along all 4 borders



# Differential Diagnosis



## Rugger-Jersey spine

Hyperparathyroidism



## Sandwich vertebra

Osteopetrosis



## Hemangioma





# Blastic Phase

- ▶ Tam o'-Shanter skull
- ▶ Cotton wool appearance
- ▶ Ivory vertebra

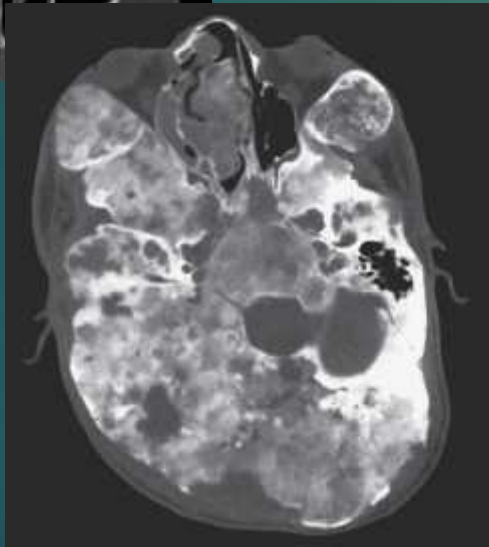


# Tam o' Shanter skull

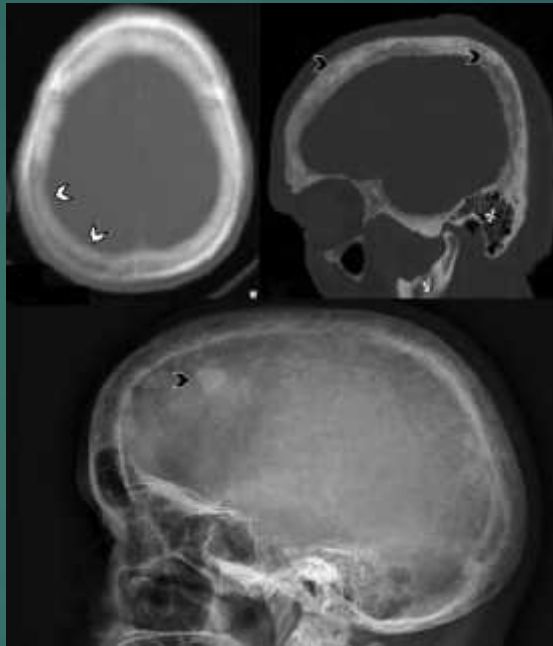
- ▶ Widening of diploic space
- ▶ Enlargement of the cranium
- ▶ Appearance of the skull falling over the facial bones



# Fibrous Dysplasia



**Fibrous Dysplasia**



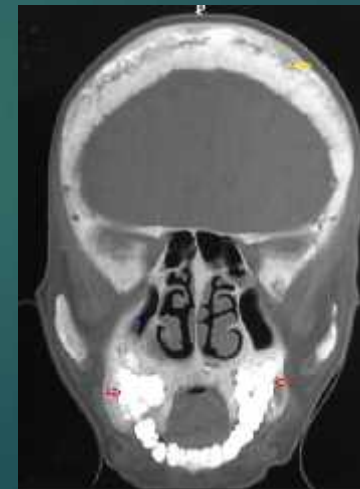
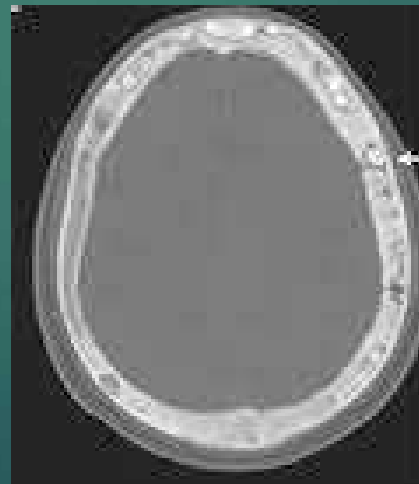
**Paget's Disease**

|                                | Fibrous dysplasia | Paget's disease                         |
|--------------------------------|-------------------|---|
| Age                            | Younger patients  | Older patients                          |
| Location                       |                   |   |
| Symmetry                       | Rather asymmetric | Symmetrical; no hemicranial involvement |
| Orbital involvement            | Common            | Uncommon                                |
| Sphenoid bone involvement      | Common            | Absent                                  |
| Paranasal sinus involvement    | Common            | Absent                                  |
| Computed tomography appearance |                   |   |
| Ground glass                   | Characteristic    | Absent                                  |
| Expanding cortex               | Common            | Common                                  |
| Thin cortical table            | Common            | Absent                                  |
| Associated soft tissue mass    | May occur         | Absent (unless malignant degeneration)  |
| Cyst-like bone lesions         | May occur         | Absent                                  |

Van de Voorde N, et al. Fibrous Dysplasia, Paget's Disease of Bone, and Other Uncommon Sclerotic Bone Lesions of the Craniofacial Bones. Semin Musculoskelet Radiol. 2020 Oct;24(5):570-578.

# Cotton wool appearance

- ▶ Circular areas of sclerosis in previous areas of osteoporosis circumscripta
- ▶ Poorly defined



# Ivory vertebra

- ▶ Diffuse increase in opacity of vertebral body
- ▶ No change in opacity and size of adjacent intervertebral discs



## Differential Diagnosis

Metastasis (prostate)



Lymphoma

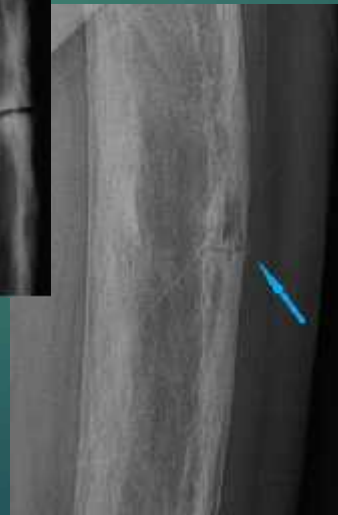


Rare:

- Chordoma
- Unusual infection (TB)
- Myelofibrosis
- Mastocytosis

# Banana Fracture

- ▶ Pathological fracture
- ▶ Orizontally orientated
- ▶ Deformed bones



## Differential Diagnosis



Osteomalacia



# Conclusion

- ▶ Paget's disease → common disease affecting 3 – 4% over 40 years
- ▶ Radiologic patterns related to the phase of disease
  - Lytic
  - Mixed
  - Blastic
- ▶ **Radiography** is sufficient for diagnosis in the majority of cases
- ▶ Complications → sarcomatous transformation



