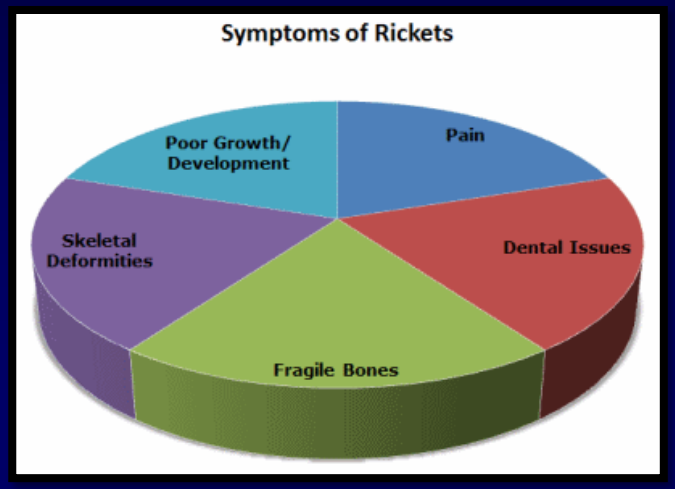
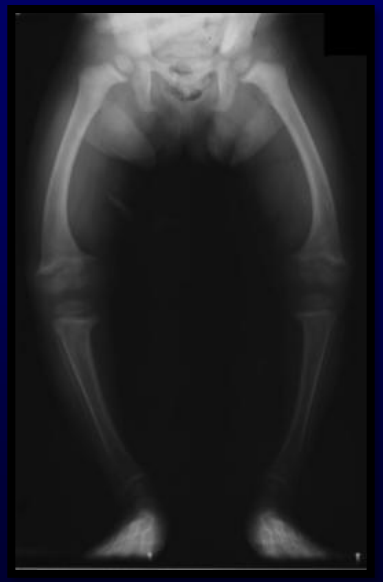
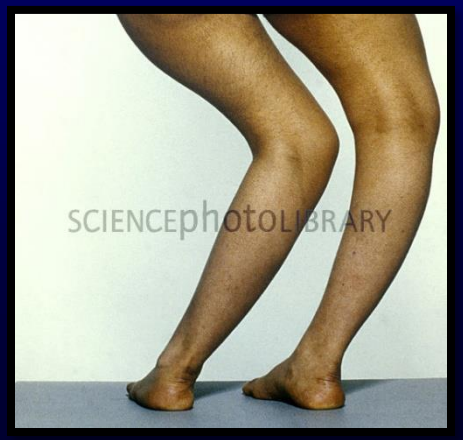
Lecture 3

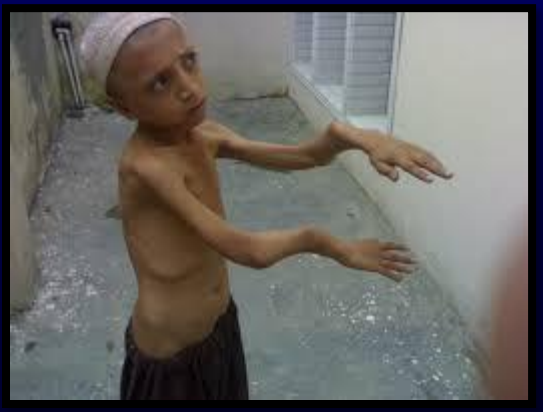
RICKETS & OSTEOMALACIA



- Vitamin D deficiency or abnormal metabolism of vitamin D.
- Children: Rickets
- Adults: osteomalacia
- Decreased mineralization of bone, unmineralized matrix
- Increase risk of fractures







HYPERPARATHYROIDISM (HPT)

Hyperparathyroidism classification

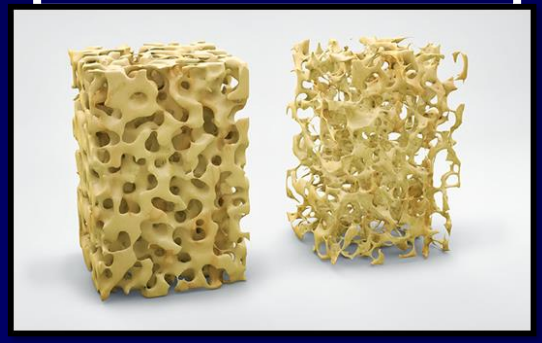
Different causes and features of hyperparathyroidism - raised parathormone (PTH).

	primary	secondary	tertiary
pathology	Hyperfunction of parathyroid cells due to hyperplasia, adenoma or carcinoma.	Physiological stimulation of parathyroid in response to hypocalcaemia.	Following long term physiological stimulation leading to hyperplasia.
associations	May be associated with multiple endocrine neoplasia.	Usually due to chronic renal failure or other causes of Vitamin D deficiency.	Seen in chronic renal failure.
serum calcium	high	low / normal	high
serum phosphate	low / normal	high	high
management	Usually surgery if symptomatic. Cincacalcet can be considered in those not fit for surgery.	Treatment of underlying cause.	Usually cinacalcet or surgery in those that don't respond.

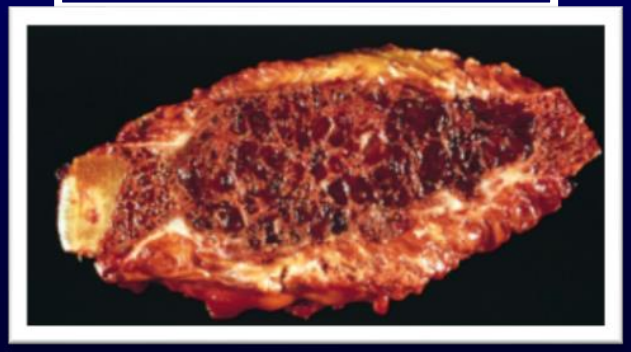
NICE have issued guidance for the use of cinacalcet in what they call refractory secondary hyperparathyroidism which is classified as tertiary hyperparathyroidism in this tblable. http://www.nice.org.uk/TA117

HPT CLINICALLY

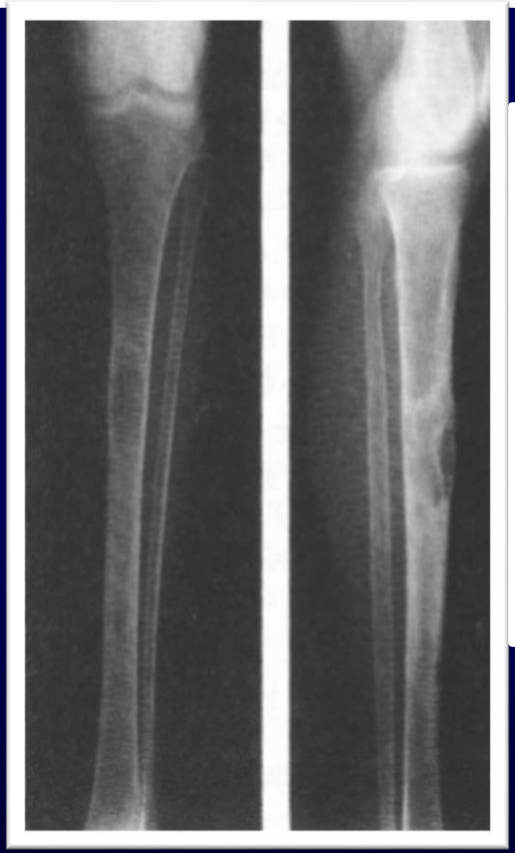
OSTEOPOROSIS



BROWN TUMOR



OSTEITIS FIBROSA CYSTICA



Abbreviated OFC, also known as osteitis fibrosa, osteodystrophia fibrosa, and von Recklinghausen's disease of bone (not to be confused with von Recklinghausen's disease, neurofibromatosis type I)

Summary

Metabolic Disorders of Bone

- **Osteopenia** and **osteoporosis** represent histologically normal bone that is decreased in quantity. In osteoporosis the bone loss is sufficiently severe to significantly increase the risk of fracture. The disease is very common, with marked morbidity and mortality from fractures. Multiple factors including peak bone mass, age, activity, genetics, nutrition, and hormonal influences contribute to its pathogenesis.
- **Osteomalacia** is characterized by bone that is insufficiently mineralized. In the developing skeleton, the manifestations are characterized by a condition known as **rickets.**
- **Hyperparathyroidism** arises from either autonomous or compensatory hypersecretion of PTH and can lead to **osteoporosis**, **brown tumors,** and **osteitis fibrosa cystica**. However, in developed countries, where early diagnosis is the norm, these manifestations are rarely seen.

PAGET DISEASE OF BONE (OSTEITIS DEFORMANS)

- Increased badly formed bone structure.
- 3 phases (lytic, mixed, sclerotic)
- 1% in USA; geographic variation
- Genetic and environmental factors
- 50% of familial Paget and 10% of sporadic have SQSTM1 gene mutations (+RANK & -OPG)
- Viruses (measles and RNA viruses)??

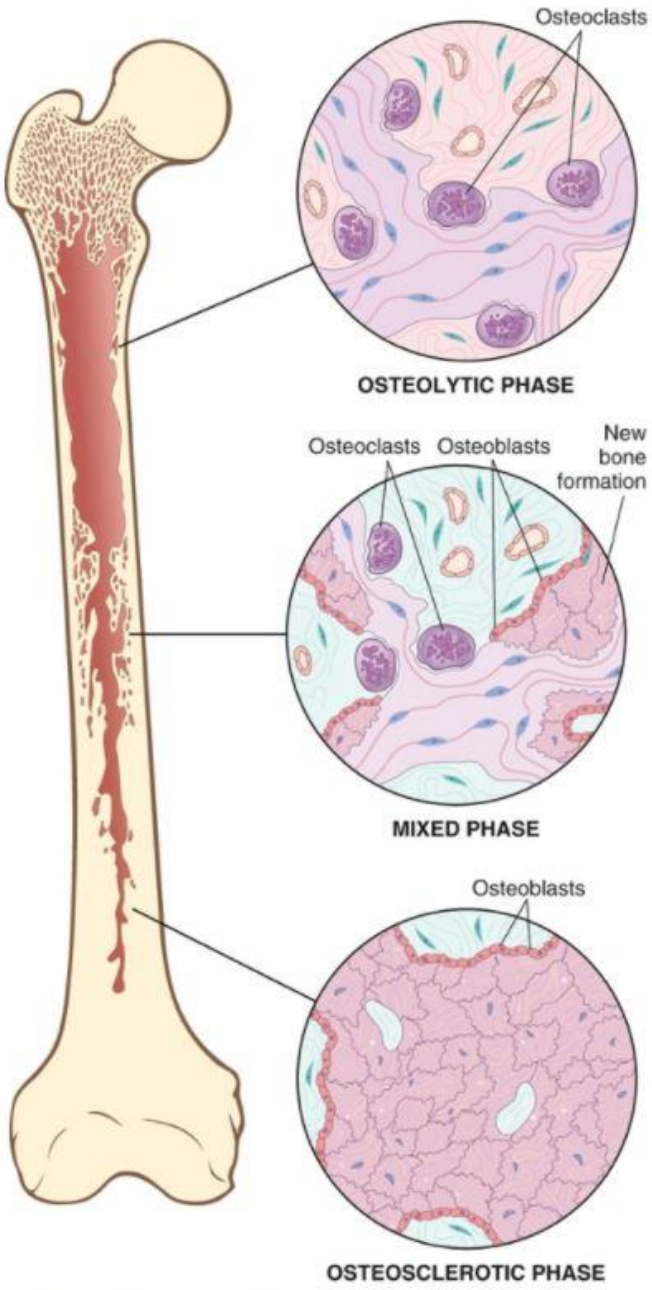
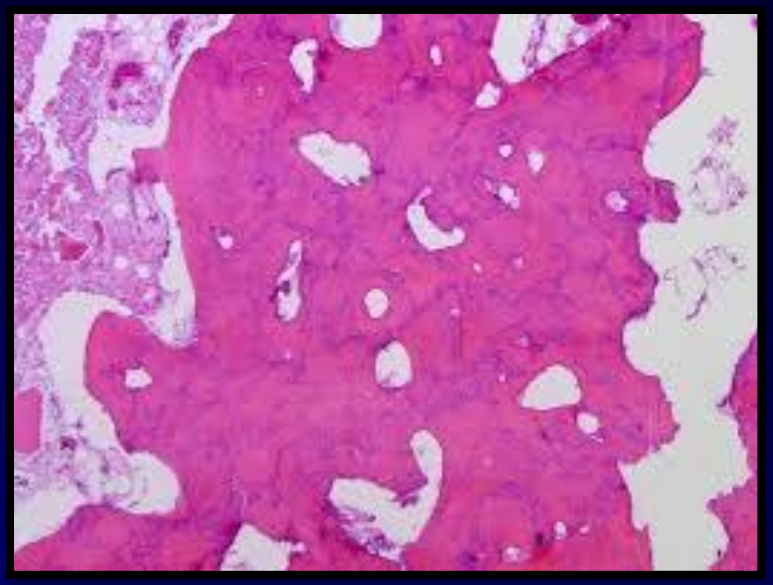
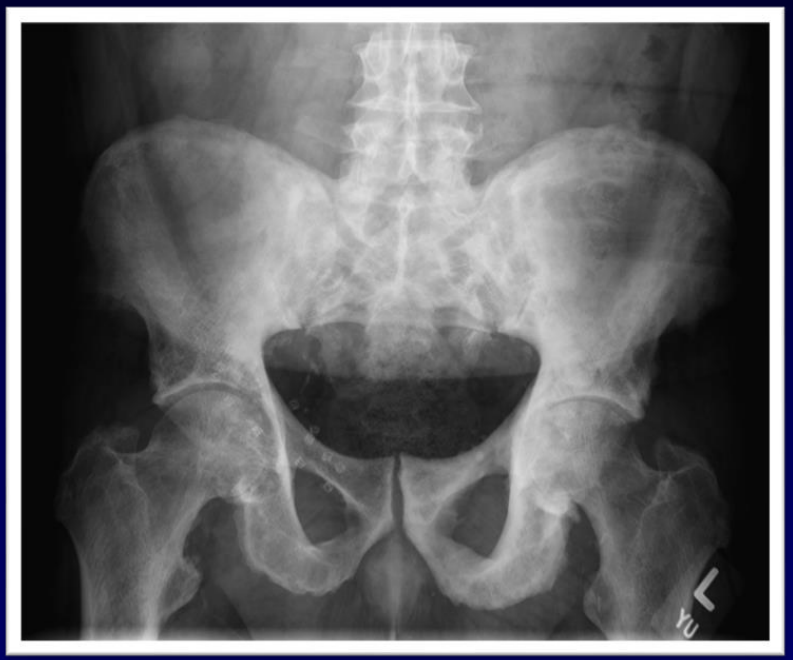
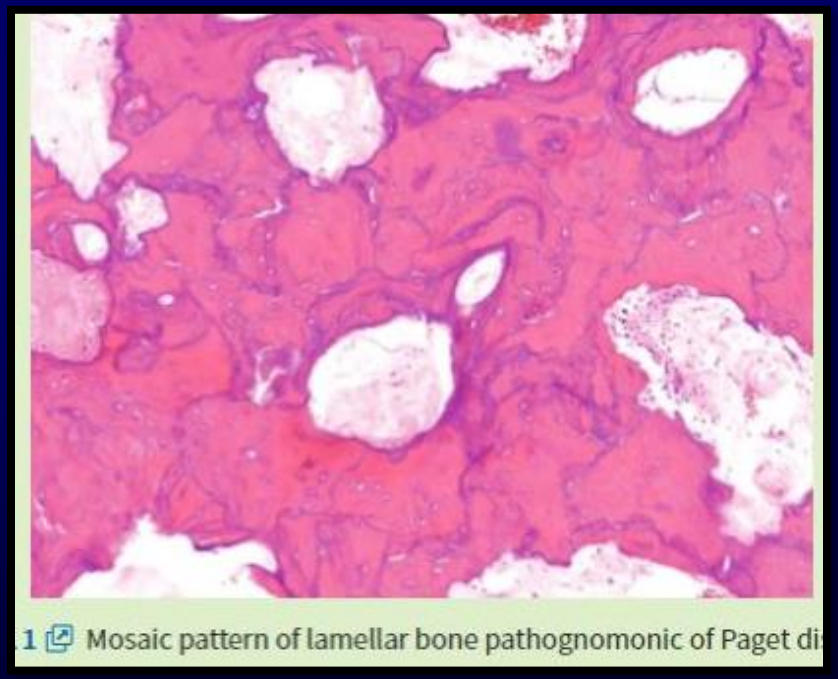


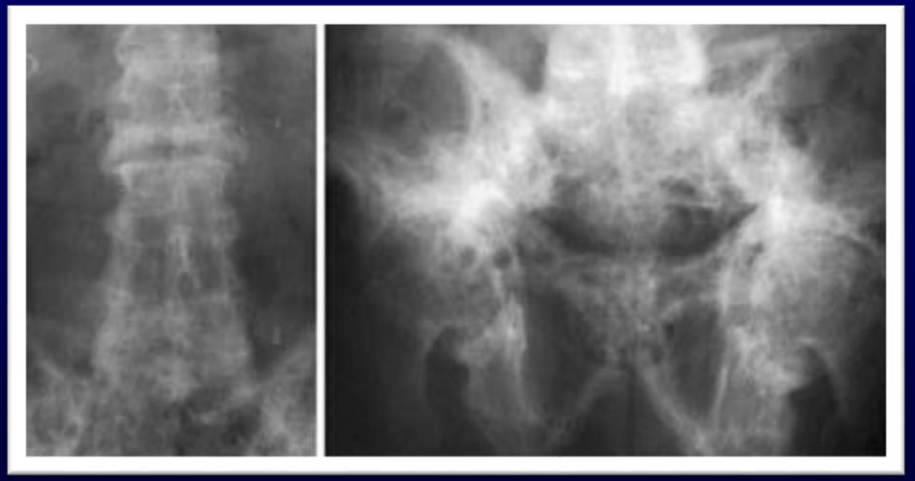
FIG. 21.10 Diagrammatic representation of Paget disease of bone demonstrating the t...







1 Mosaic pattern of lamellar bone pathognomonic of Paget di



PAGET CLINICALLY:

- **85% polystotic; 15% monostotic**
- **Axial skeleton more affected (prox. Femur)**
- **Most are mild and asymptomatic (pain)**
- **Pain: microfractures or nerve compression**
- ***Leontiasis ossea* (lion face); *platybasia* (invagination of skull base); secondary osteoarthritis; fractures; osteosarcoma (1%)**
- **DX: x-ray; ↑ serum Alk P, Normal Ca and PO4**

Leontiasis ossea (lion face); platybasia