Metabolic and Endocrine Bone Disease Imaging

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Tumoral Calcinosi
Osteoporosis

Osteoporosis is the most common metabolic bone disorder. It has been defined by the National Institutes of Health as an age-related disorder characterized by decreased bone mass and increased susceptibility to fractures in the absence of other recognizable causes of bone loss.
Osteoporosis

- **Type 1. Involutional osteoporosis** affects mainly trabecular bone, occurs in women during the 15-20 years after the menopause, and is related to a lack of estrogen. This is thought to account for wrist and vertebral crush fractures, which occur through areas of principally trabecular bone.

- **Type 2. Senile involutional osteoporosis.** The fractures of old age seen at the hip, proximal humerus, pelvis and asymptomatic vertebral wedge fractures. This affects both trabecular and cortical bone and represents progressive loss of bone mass from the peak around the age of 18-35 years.

- **Secondary osteoporosis** is due to an underlying medical condition, such as renal disease, malabsorption, or hormonal imbalance, or to medical treatment such as steroids or certain anticonvulsants.
Osteoporosis Measurement

- Plain film,
  - Subjective
  - Radiogrammetry
  - Radioabsoptiometry
- SPA
- DPA
- DEXA
- QCT
- US
- MRI
DEXA

DEXA has very high

accuracy

(the difference in the measurement from a known standard)

and

precision

(observed deviation of serial measurements with time),

both short and long term, to within 1% at the hip and spine
Bone Densitometry

WHO uses T scores

- **Normal**
  - > -1 SD below young adult

- **Osteopenia**
  - -1 - 2.49 SD

- **Osteoporosis**
  - <= -2.5 SD

- **Established (Manifest) Osteoporosis**
  - + Fxs, usually spine, hip, proximal humerus, wrist, rib
T score is compared to reference population, 20-45 years, same sex, any race, any weight.

Z score is matched for age, sex, weight and ethnicity.
# Osteoporosis - OGI

<table>
<thead>
<tr>
<th>REGION</th>
<th>BMD (^1) g/cm(^2)</th>
<th>Young Adult (^2) %</th>
<th>Age Matched (^3) %</th>
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<tr>
<td>NECK</td>
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<td>WARDS</td>
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<td>80</td>
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<td>TROCH</td>
<td>0.598</td>
<td>64</td>
<td>65</td>
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<td>L1</td>
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<td>48</td>
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<td>L3</td>
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<td>L4</td>
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<td>L1-L4</td>
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Bone Densitometry
FRAX – Fracture risk assessment tool

- Age
- Sex
- Weight / Height - BMI
- Spontaneous previous adult fracture
- Parent fractured hip
- Current smoking
- Glucocorticoids
- Rheumatoid arthritis
- Secondary osteoporosis
- Alcohol
- Bone mineral density
Bone Densitometry

Risk Factors

Fracture probability

>20% High
10-20% Middle
<10% Low

Treat
BMD
No action
Osteoporosis - Secondary

- Regional
  - Disuse

- General
  - Hormonal
  - Malabsorption
  - Drugs
    - Steroids
    - Anticonvulsants
Disuse Osteoporosis
Disuse Metaphyseal Lucency

Acute

1m

2m
Disuse Osteoporosis of Sesamoid

[Images of radiographs showing acute and 6 weeks later stages]

Acute

6w later
“Hawkins” Scaphoid
Disuse Osteoporosis

Sag PDFS

Sag T1FS Gd

Patchy Enhancement
52F injured left tibia after fall with persistent pain exacerbated with prolonged standing
Cushing’s Osteoporosis

- Syndrome
  - ↑ cortisol

- Pituitary 80% Cushing’s disease
  - 90% adenoma
  - 20% visible radiographically

- Adrenal 20%
  - Adenoma
  - Carcinoma

- Ectopic ACTH (Ca bronchus)
Cushing’s Osteoporosis

- 50% of Cushings syndrome adults are osteoporotic

- 30-50% pathologic fractures (trabecular-spine)

- Children also growth retardation

- Cortisol multifactorial effect on bone
  - Growth hormone
  - Hypogonadism
  - Calcium absorption
  - Renal calcium excretion
Cushing’s Osteoporosis

[Image of X-ray showing bone density and possible osteoporosis markers]
Cushing’s Osteoporosis

Codfish vertebrae 85F
Vitamin D

- 1,25 dihydroxycholecalciferol

- 1αOH

- Kidney

- PTH

- 1,25 (OH₂)D₃

- Bone

- Growth plate

- Stimulates osteoid mineralisation

- Intestine

- 25- OH-D3

- 25- OH-D3

- PO₄

- PO₄

- Ca

- Ca

- Diet

- Absorption

- Liver disease

- Anticonvulsants

- Renal disease

- Vit D res

- Parathyroid dis

- Bone

- PO₄

- Intestine

- Ca
Rickets

- Increased uncalcified osteoid in the immature skeleton

- Lack of Vitamin D
  - Dietary
  - Malabsorption
  - Renal tubular disease
Rickets – Growth Plate changes

- Widened growth plate
- Metaphysis
  - Fraying
  - Splaying
  - Cupping
  - Spurs
- Diaphysis
  - Indistinct cortex
- Rickety rosary
- Looser’s zones
Rickets – Changes of Soft Bones

- Bowing
- Triradiate pelvis
- Harrison’s sulcus
  - Soft ribs
- Scoliosis
- Biconcave vertebrae
- Basilar invagination
- Craniotabes
Rickets – Follow up

- Dietary Rickets
  - Full recovery on Rx
Rickets – Follow up

6 year old with bowed legs
X-linked Hypophosphatemic Rickets

- Vitamin D resistant Rickets
- Familial Hypophosphatemic Rickets
- X-linked
  - Phosphate levels equally low, M=F
  - $1^\alpha$ hydroxylation reduced in males

- Imaging
  - Identical to dietary Rickets
X-linked Hypophosphatemic Rickets

- 1,25 dihydroxycholecalciferol

\[ \downarrow \text{PO}_4 \]

\[ \downarrow \text{Ca} \]

\[ \text{PTH} \]

Kidney

1αOH

Bone

\[ \uparrow \text{PO}_4 \]

1,25 (OH\textsubscript{2})D\textsubscript{3}

Growth plate

Stimulates osteoid mineralisation

Intestine

\[ \uparrow \text{Ca} \]

Liver disease

Vit D res

Parathyroid dis

Diet

Absorption

Renal disease

Anticonvulsants
X-linked Hypophosphatemic Rickets
Hypophosphatasia

- Tissue nonspecific alkaline phosphatase
  - TNSALP
- Causes defective mineralisation of bone
- Low serum alkaline phosphatase
- High serum phosphoethanolamine
Hypophosphatasia

- Perinatal – Fatal
- Infantile – 50% fatality
- Childhood – Rickets
- Adult – Poorly healing stress fractures
- Odontohypophosphatasia – Loss of teeth
Hypophosphatasia - Neonatal

- Profoundly deficient mineralization
  - Knees
  - Wrists
  - Costochondral
- Fractures
Hypophosphatasaia - Infantile

- Physes
  - Widened

- Metaphyses
  - Cupped
  - Frayed

- Demineralised epiphyses.

- Widened cranial sutures

- Craniostenosis brachycephaly
Hypophosphatasia - Childhood

- Physis
  - Widened

- Metaphysis
  - Cupped
  - Frayed

- Demineralised epiphyses

- Widened cranial sutures

- Craniostenosis brachycephaly
Hypophosphatasia - Adult

- Osteomalacia
- With ↓ bone density
Hypophosphatasia - Adult

27F multiple fractures, some incomplete, poorly healing, over 3 yrs
Osteomalacia

- ↑ uncollcified osteoid in the mature skeleton
- ↓ bone density
- Looser’s zones
  - Scapula
  - Femoral neck
  - Femoral shaft.
  - Pubic rami
  - Ribs
- Coarsened ill defined trabeculae
- Bone softening
  - Protrusio
  - Bowing
  - Biconcave vertebrae
  - Basilar invagination
Hyperparathyroidism

• Primary
  • Parathyroid adenoma 90%
    • 2% are multiple
  • Hyperplasia of all four glands 5%
    • Familial
  • Carcinoma
  • Ectopic
  • MEN type 1 (hyperplasia or adenoma)
    • Pituitary adenoma and pancreatic tumor
Hyperparathyroidism

- **Secondary**
  - Failure to excrete phosphate in renal failure
  - Phosphate binds with Calcium
  - Due to ↓ serum Calcium
Hyperparathyroidism - Imaging

- Primary and secondary HPT have similar findings now that patients with renal failure have increased life expectancy.

- Previously thought that Brown tumors were more common in Primary HPT
Renal Osteodystrophy

- Renal glomerular disease
  - Bilateral reflux nephropathy
  - Pyelonephritis
  - Chronic glomerulonephritis
- Osteomalacia or Rickets
  - Failure to hydroxylate
- Secondary hyperparathyroidism
  - Failure to excrete phosphate
- Osteosclerosis
- Calcification more prominent in adults

Primary oxalosis
Hyperparathyroidism – Imaging - Bones

- Osteopenia
  - Ground-glass

- Resorption
  - Subperiosteal
    - Fingers
    - Proximal tibia
    - Lateral clavicle
    - Symphysis pubis
    - Ischial tuberosity
    - Medial femoral neck

- Cortical
  - Cortical tunneling
  - Pepper pot skull

- Osteosclerosis
  - Rugger jersey spine

- Brown tumors
  - Solitary sign in 3%

- Bone softening
Renal Osteodystrophy and Brown Tumors

Rib brown tumors 30F
Hyperparathyroidism – Imaging – Soft Tissues

- ST Cal
  - Arteries
  - Periarticular
    - Capsule
    - Tendon
    - Tumoral

ROD Tumoral calcinosis
Hyperparathyroidism – Imaging - Joints

- Marginal erosions
  - DIPJ
  - Ulnar side base of 5th MC
  - Hamate
  - No JSN

- Subchondral collapse

- Chondrocalcinosis
  - CPPD
  - Gout
Dialysis Spondylosis

Beta2 microglobulin destructive spondyloarthropathy
Primary and secondary HPT

Primary oxalosis
Primary oxalosis
secondary hyperparathyroidism

- Primary
  - Hereditary hyperoxaluria
  - AR, enzyme deficiency - carboligase
  - Diffuse calcium oxalate deposits

- Secondary
  - Disturbance of bile acid metabolism
  - Usually diseases of terminal ileum
Primary oxalosis
secondary hyperparathyroidism
HypoParathyroidism

- Hormone deficient
  - Surgery, Idiopathic

- Imaging
  - Osteosclerosis
  - DISH like ossification
  - Thickened calvarium
  - Sutural diastasis
  - Basal ganglia calcification
Pseudohypoparathyroidism

- Hormone resistant
  - End organ unresponsiveness to PTH
    - Usually bone and renal

- X – linked dominant F>M

- Clinical – Albright’s hereditary osteodystrophy
  - Short stature, thickset features
  - ↓ Calcium, ↑Phosphate, normal or ↑ PTH

- Imaging
  - Short 4\textsuperscript{th} > 5\textsuperscript{th} > 1\textsuperscript{st} metacarpals/tarsals
  - Exostoses
  - Basal ganglia calcification
  - Soft tissue calcification
PseudoPseudohypoparathyroidism

• Similar phenotypically to pseudohypoparathyroidism, but with normal plasma calcium
38 year old male with foot and heel pain with history of congenital deformity and obesity
Albright’s hereditary osteodystrophy

• PseudohypoPTH (PHP) and Pseudo-pseudohypoPTH (PPHP)

• X linked dominant; females > males

• PHP
  • Low Ca2+, high Ph

• PPHP (normocalcemic form of PHP)
  • Normal Ca2+ and Ph
PHP and PPHP

- Clinical features
  - Short stature, Obesity, Round face, Brachydactyly

- Typical radiographic findings
  - ST calcification and ossification – plaquelike, assymetric, parallel skin surface
  - Basal ganglion Ca+ and Calvarial thickening
  - Short MC, MT, phalanges- especially 1st, 4th MC
  - Premature physeal fusion
  - Exostoses- centrally located with right angle to bone
  - Cone epiphyses
  - Wide bones
Thyroid Disorders

- Hyperthyroidism
- Thyroid Acropachy
- Hypothyroidism
Thyroid Acropachy

- 0.5% of thyrotoxicosis
- After Rx
- Exophthalmos
- Painless STS of fingers
- Pretibial myxoedema
- Finger clubbing
Thyroid Acropachy
Finger clubbing

- Periosteal new bone MC and proximal phalanges
- Radial aspect of bone
- Dense and solid
- STS
Thyroid Acropachy
Hypothyroidism Cretinism

- Appendicular skeleton
  - Delayed appearance of ossification centers
  - Delayed epiphyseal closure
  - Short slender long bones
  - Endosteal thickening
  - Dense metaphyseal bands.
  - Coxa vara with short femoral neck

Chronological Age = 22 years
Hypothyroidism Cretinism

- Skull
  - Brachycephaly
  - Wormian bones
  - Delayed sutural closure
  - Poorly developed sinuses and mastoids.
Hypothyroidism Cretinism

- **Axial skeleton**
  - Flattened vertebrae
  - Wide discs
- Thoracolumbar kyphosis.
- Hypoplastic bullet L1 or L2
- Segmental sternum

Chronological Age = 22 years
Pituitary Disorders

- Acromegaly
- Cushing’s disease
- Hypopituitarism
Acromegaly

- Excessive growth hormone on mature skeleton
  
- Skull
  - Thickened vault
  
- Thorax and spine
  - ↑ sagittal diameter of chest with kyphosis
  - Enlarged vertebrae

- Appendicular skeleton
  - ↑ width of bones with normal cortical thickness
  - Terminal phalangeal tufting
  - Prominent entheses
  - Widened joint spaces
  - Premature OA
  - Osteoporosis
  - ↑ heel pad thickness
Acromegaly

- Excessive growth hormone on mature skeleton

- Skull
  - Thickened vault

- Thorax and spine
  - ↑ sagittal diameter of chest with kyphosis
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  - Widened joint spaces
  - Premature OA
  - Osteoporosis
  - ↑ heel pad thickness
Hypopituitarism

• Pituitary Dwarfism
  • Damage to anterior lobe of pituitary in childhood

• Delay in appearance and growth of ossification centers

• Delay in closure
Paget’s Disease

- Excessive abnormal remodeling of bone
- Middle age 3%
- Old age 10%
- Spine 75%
- Skull 65%
- Pelvis 40%
- Proximal femur 75%
Paget’s Disease

- Middle aged and elderly
- Excessive and abnormal remodeling of bone
- Initial osteolytic phase
- Subsequent osteosclerotic phase
- Enlarged bone with increased density and coarse trabecular

Paget’s cortex
Paget’s Disease

- Radiographic findings
- Active Osteolytic phase
  - Osteoporosis circumscripta
  - Advancing wedge of lucency
Paget’s Disease

- Radiographic findings
- Mixed phase
  - Skull
    - Osteoporosis circumscripta with sclerosis
  - Pelvis
    - Mixed osteolytic and osteosclerotic
  - Long bones
    - Diaphyseal lucency
    - Epi/Meta sclerosis
Paget’s Disease

- Radiographic findings

- Osteosclerotic phase
  - Skull
    - Thickened vault
  - Spine
    - Enlarged vertebrae
    - Coarse trabeculae
  - Pelvis
    - Often asymmetric
  - Long bones
    - Cortical thickening
    - Medullary encroachment
Paget’s Disease

- Complications
  - Osseous deformity
  - Fractures
  - Nerve entrapment
  - Neoplasms
  - Osteomyelitis
  - Extramedullary hematopoiesis
  - Gout
  - Degenerative joint disease

Pathological fractures in Paget’s
Alkaptonuria / Ochronosis

- Absence of homogentisic acid oxidase
- Pigmentation
- Arthropathy
- Osteoporotic with dense disc calcification
- Larger joints show DJD
Idiopathic Tumoral Calcinosis

• 10-30Yrs, black > white, M=F

• Elevated phosphate, normal calcium
  • Renal tubular phosphate resorption

• Single or multiple, firm, tumor like, painless

• Hips, Shoulders, Elbows, Ankles

• Dense, Flocculent, Amorphous,

• 1-20cm, fluid levels

• Recur if resected

• Dx of exclusion Periarticular calcified masses
Tumoral Calcinosis

- DDX
  - Idiopathic
  - Renal osteodystrophy
  - Gout
  - Hyperparathyroidism
  - Collagen vascular Disease
  - Hypervitaminosis D
  - HADD
Homocystinuria – Metacarpal index

Average 2-5 abnormal if >8.4 male, >9.2 female

Homocystinuria 40F Marfanoid
Osteopetrosis