PACES Hub

V1

Congenital Heart Disease- PDA

Patent Ductus Arteriosus

Clinical features

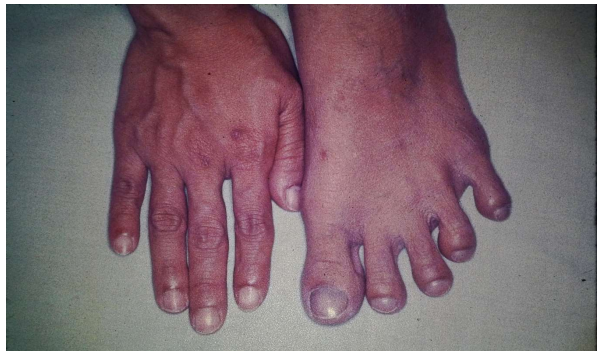
- Continuous machinery-like murmur
- Loudest at left upper sternal border
- If shunt is distal to left subclavian artery → differential clubbing and cyanosis (in lower but NOT upper extremities)

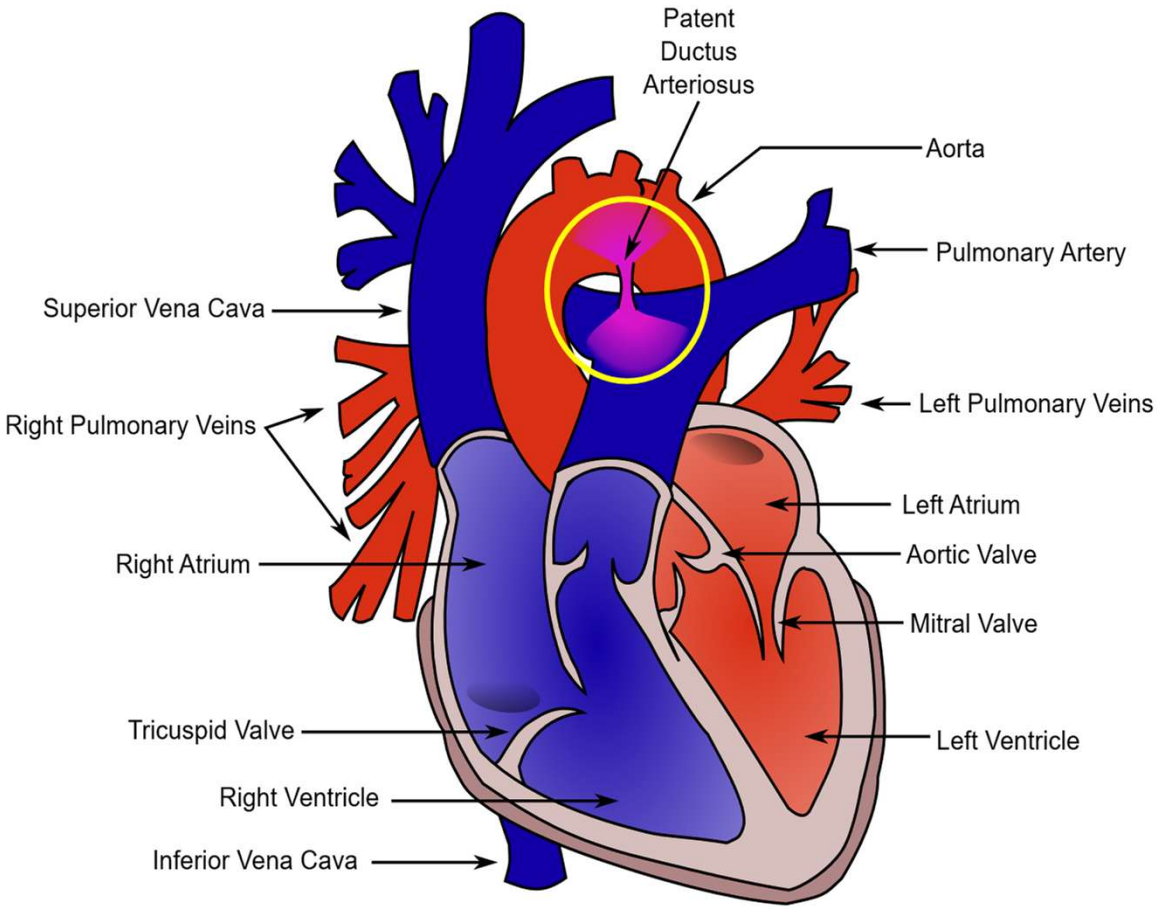
Investigations

- ECG: LVH (in large PDA), RVH (if PH)
- CXR: calcified "trumpet-shaped" duct, enlarged LV and pulmonary arteries
- Echo: continuous main PA flow

Management

- Percutaneous closure if more than trivial shunt





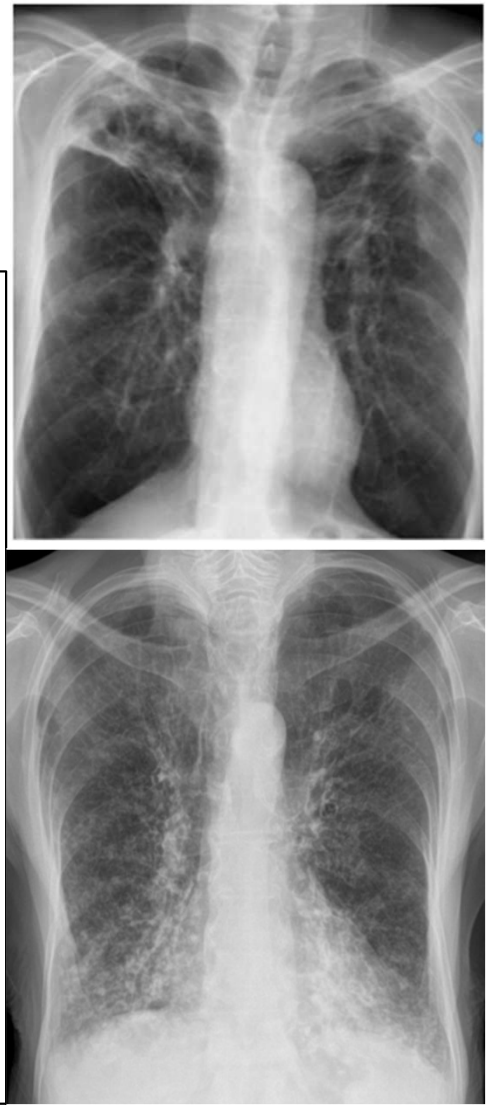
Bronchiectasis

Definition

Permanent pathological bronchial dilation and destruction with impaired mucus drainage which occurs due to repeated infections and/or abnormal antimicrobial host response.

Clinical Features

- Chronically ill appearance, respiratory cachexia
- Moist/loose cough (ask patient to cough)
- Discoloured/mucopurulent +/- bloody sputum (often large volume) in cup
- Clubbing, yellow nail syndrome
- Sinus region tenderness indicative of chronic rhinosinusitis
- +/- hyperinflated chest (if gas trapping), prolonged FET (>5 sec)
- Coarse musical breath sounds:
  - Crackles (usually coarse), wheeze
  - Note distribution (focal vs diffuse, upper vs middle vs lower zone)
- Cardiac:
  - Signs of pulm HTN, RHF/cor pulmonale
  - Do not miss dextrocardia in Kartagener's
- Other extrapulmonary sequelae depending on suspected aetiology: e.g. CF-related diabetes



Aetiology by Distribution

Diffuse:

- Immunodeficiencies (CVID, HIV), primary ciliary dyskinesia (PCD), autoimmune/CTD (e.g. RA, Sjogren's), A1AT deficiency, recurrent infections

Focal:

- Upper zone: cystic fibrosis and variants, TB, allergic bronchopulmonary aspergillosis
- Central/perihilar: ABPA
- Middle/lingular: non-tuberculous mycobacteria e.g. Mycobacterium avium complex (MAC), PCD
- Right lung: aspiration, foreign body
- Lower: idiopathic, immunodeficiencies, recurrent infections or aspiration, fibrotic lung disease, PCD

Psoriatic Arthritis

Clinical Features

Arthritis in up to 25% of patients with psoriasis

5 patterns of joint involvement

1. DIPJ predominant with nail disease (pitting, onycholysis, nail bed hyperkeratosis, nail plate crumbling)

- Nail disease correlates with arthritis activity

2. Asymmetric oligoarthritis (<5 joints involved)
3. Symmetric polyarthritis (RA-like)
4. Axial spondyloarthritis: sacroiliitis, spondylitis
5. Arthritis mutilans (severe, destructive arthropathy)

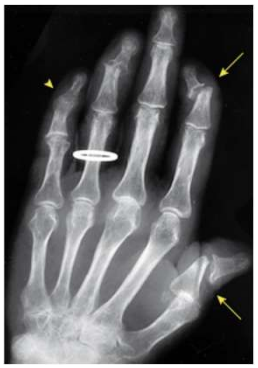
Evidence of psoriatic skin disease

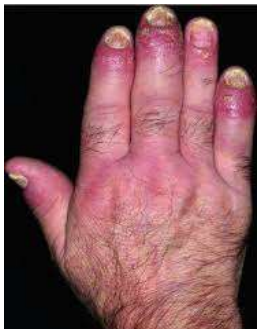
- Scaly, silvery plaques on limb extensor surfaces, scalp, natal cleft, abdomen, behind ears
- Other psoriasis types: palmoplantar pustulosis, guttate
- Note PsA may be seen in absence of ("sine") psoriasis

Dactylitis, enthesitis, extraarticular features as in SpA:

- NB: conjunctivitis more common than uveitis in PsA

Functional limitation





Management

Treat to target approach

Nonpharmacological

- Exercise & physio (esp. for axial disease)
- Orthotics, hand splints for deformity & impaired function
- Photo/UV therapy and emollients for skin psoriasis

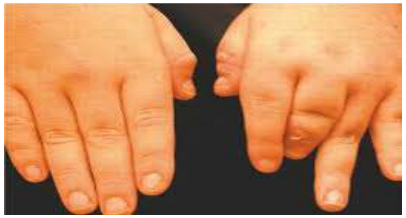
Pharmacological

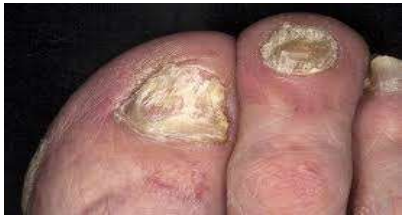
- NSAIDs, local steroid injections for axial + peripheral PsA
- Apremilast (PDE4 inhibitor)), MTX, SSZ, LEF for peripheral
- Avoid systemic steroids as risk of psoriasis flare
- Severe or persistent disease
  - Anti-TNF-α: Infliximab, etanercept, adalimumab
  - Anti-IL-17: Secukinumab
  - Anti-IL12/23: Ustekinumab esp if comorbid IBD
  - Anti-IL23 if no axial disease
  - JAKi as for RA

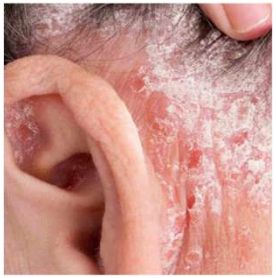
CV and metabolic risk reduction (PsA a/w CVD)

Investigations

- Raised inflammatory markers CRP)
- (Autoantibodies typically negative (RF, ACPA, ANA)
- HLA-B27: positive in ~25% of PsA but not diagnostic
- Inflammatory joint aspirate
- Imaging of affected joints
  - Erosive arthropathy
  - Pencil-in-cup deformity
  - “Rays not rows”: all joints in one digit vs. all DIPs
  - Juxta-articular new bone formation
  - Sacroiliitis







PACES Hub

Parkinsonism

Clinical Features

<u>Motor features</u>

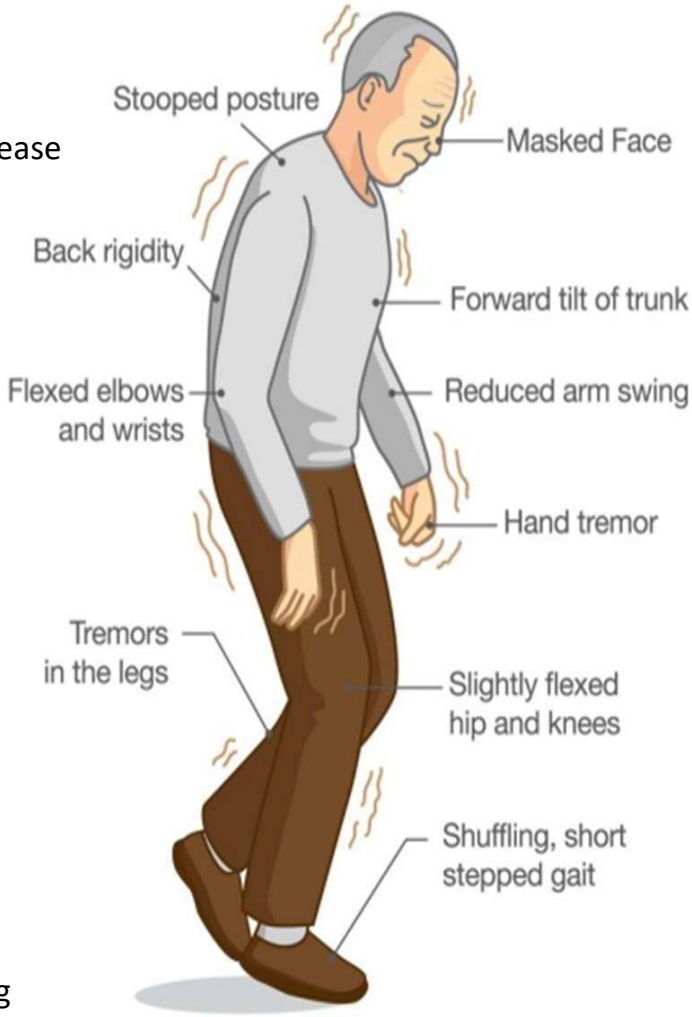
- Decrementing bradykinesia (hallmark)
  - To test → finger to thumb, hand opening, wrist pronation, or toe tapping; look for decrease in amplitude and/or speed; accentuated by cognitive exercises
  - Micrographia, hypomimia, hypophonia
- Rigidity → Upper-limb predominant and asymmetric, "lead-pipe," "cogwheeling"
  - Can unmask by asking patient to forcefully tap contralateral hand or foot
- Postural instability
  - Festinating gait, freezing, reduction in step length, loss of arm swing, loss of heel strike
  - Loss of balance when turning and/or with pull test
- Tremor
  - 4-6 Hz resting tremor most commonly seen ("pill-rolling")
  - Can see kinetic/action tremor and/or postural tremor (accentuated when maintaining position against gravity, i.e. arms raised to 90°)

<u>Non-motor features</u>

- Autonomic dysfunction → orthostatic hypotension, constipation, urinary retention, erectile dysfunction
- Anosmia
- REM sleep disorder
- Neuropsychiatric manifestations & cognitive dysfunction (frontal executive dysfunction)
- Seborrheic dermatitis

<u>Primitive reflexes</u> → glabellar tap, rooting/sucking reflex, grasp/utilization behaviour

<u>Other</u> → Restriction in upward gaze / complex ophthalmoplegia in PSP; olfactory nerve testing



Cushing's syndrome

Clinical Features

Somatic
- Adiposity
  - Increased visceral / intraabdominal fat
  - Increased facial width (moon face)
  - Buffalo hump / dorsal fat pad
  - Decreased subcutaneous / limb fat
- Abdominal striae
- Thin skin
- Bruising
- Proximal myopathy
- Skin pigmentation → secondary cause

Metabolic
- Insulin resistance / diabetes mellitus
- Systemic arterial hypertension
- Hypogonadism
- Osteoporosis

Neuropsychiatric
- Cognitive dysfunction
- Psychosis

Other
- Hypercoagulable, increased VTE risk
- Propensity for infections
- Increased cardiovascular disease risk

Aetiology

Primary (adrenal)
- Adenoma : carcinoma = 2:1
- Bilateral macronodular adrenal hyperplasia
- Primary pigmented nodular adrenocortical disease

Secondary (pituitary, "Cushing's disease")
- ACTH-secreting pituitary adenoma

Tertiary (hypothalamic)
- CRH-secreting lesion (rare)

Ectopic (usually paraneoplastic)
- Small-cell lung cancer
- Pancreatic cancer
- Thyroid cancer

Exogenous (iatrogenic or factitious)
- Long-term or high-dose steroids
- Most common cause overall