Pulmonary Fibrosis

A restrictive lung disease characterised by accumulation of excess fibrous connective tissue in the lung parenchyma which causes progressive shortness of breath and hypoxia due to reduced lung compliance and oxygen diffusion capacity.

Causes

- Upper zone fibrosis = **CARTEx**
  - **C**oal
  - **A**nkylosing spondylitis
  - **R**adiation
  - **T**B
  - **Ex**trinsic allergic alveolitis
- Lower zone fibrosis = **CAID** (more common)
  - **C**onnective tissue disorders (scleroderma, rheumatoid)
  - **A**sbestos
  - **I**diopathic pulmonary fibrosis
  - **D**rugs (nitrofurantoin, amiodarone, methotrexate, cyclophosphamide, gold/sulfasalazine)

Clinical features

- Symptoms
  - Chronic progressive shortness of breath
  - Dry cough
- Signs of pulmonary fibrosis
  - Dyspnoea/oxygen therapy
  - Reduced expansion
  - Fine end-inspiratory crepitations
- Signs of cause
  - Clubbing (idiopathic pulmonary fibrosis)
  - Hand deformity (RA)
  - Telangiectasia, sclerodactaly and microstomia (systemic sclerosis)
  - Butterfly rash (SLE)
  - Lupus pernio (sarcoidosis)
  - Radiation burns
  - Kyphosis (ankylosing spondylitis)

Investigations

- Chest X-ray (reticulonodular shadowing)
- High-resolution CT chest
  - Honeycombing cysts
  - Reticular septal thickening
  - Ground glass changes (indicates inflammatory process which is often steroid responsive)
- Spirometry (restrictive defect with reduced diffusion capacity)
- Blood tests for likely causes (e.g. ANA, RhF, anti-centromere antibody)
- Lung biopsy

Management

- Fibrosis is permanent
- Steroids may help in some cases
- Anti-fibrotics (pirfenidone/nintedanib) may be used in idiopathic pulmonary fibrosis
- Treat/withdraw cause (if possible)
- Long-term oxygen therapy (if $PaO_2 < 7.3kPa$ or $<8kPa$ in presence of pulmonary hypertension or secondary polycythaemia)
- Lung transplant (option for some patients)