RESTRICTIVE LUNG DISEASE

reduced expandability.
decreased compliance.

Chest wall abnormalities
- fibrothorax
- Kyphoscoliosis
- morbid obesity

Respiratory muscle weakness
- polio
- myasthenia gravis

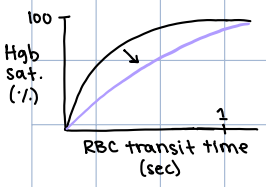
Abnormal lung parenchyma
- sarcoidosis
- idiopathic pulmonary fibrosis (IPF)

PFT → ↓FVC and ↓FEV1 BUT ↑FEV1/FVC (or normal)

↓IRV, ↓TV, ↓ERV, ↑RV, ↓VC, ↓FRC, ↓IC, ↓TLC

INTERSTITIAL/DIFFUSE PARENCHYMAL LUNG DISEASE

Pathophysiology: diffusion impairment



- transit time = 1 sec at normal HR

If HR↑ → RBCs move through capillary faster → ↓ transit time → hgb not fully saturated → hgb desaturation

Diagnosis: ABG - ↑A-a gradient

High resolution chest CT - detects sub-radiographic diseases, distinguish btwn active inflammation (ground glass) and fibrosis, suggest diagnosis

granulomatous | inhalation | Connective tissue disease | idiopathic interstitial pneumonias | drug-induced | Other disease related | Other

SARCOIDOSIS

growth of tiny collections of inflammatory cells - non-caseating granulomas

More common in black pts

Stage 1: lymphadenopathy ONLY
- observation - may resolve on its own
- trial of steroids after infection ruled out
- lymph node / skin lesion biopsy. Chest radiograph

Stage 2: Parenchymal involvement plus adenopathy

Stage 3: Parenchymal involvement ONLY

Stage 4: Fibrosis

Diagnosis: ① + granulomas ② no other cause

Treatment: none or corticosteroids prednisone (5-40mg)

Lofren's Syndrome
acute form of sarcoidosis
- fever
- hilar adenopathy
- erythema nodosum

UNKNOWN ETIOLOGIES

IDIOPATHIC PULMONARY FIBROSIS (IPF)

Most common interstitial pneumonia
- older, male, smoker, SOB x year

reticular (fibrosis), honeycombing

Diagnosis of exclusion

non-IPF idiopathic interstitial pneumonia
- non-specific - no pattern
  - ground glass
- respiratory bronchiolitis
  - ground glass
  - bronchial wall thickening
- desquamative
  - ground glass
- Cryptogenic - patchy, peripheral consolidation
- acute lymphocytic

Evaluation of Interstitial Lung Disease

history and physical exam
↓
CBC + diff, electrolytes, BUN, creatinine
↓
PFT
↓
CXR and high res. CT
↓
Clinical diagnosis → certain → diagnosis
Clinical diagnosis → not obtainable by broncoscopy → biopsy (VAT or open lung) → diagnosis
Clinical diagnosis → broncoscopy → diagnosis
broncoscopy → not diagnostic → biopsy (VAT or open lung) → diagnosis

KNOWN ETIOLOGIES

PNEUMOCONIOSES
diffuse parenchymal lung diseases caused by inhalation of inorganic dusts

- dust particle needs to be 0.5 to 5 microns. Deposited first in respiratory bronchioles.

Silicosis
exposure to silica (mining, quarrying, tunneling, stone cutters, sandblasting, glass manufacturing, faundry work, enameling, quartz crystal, rubber industry)

Pathophysiology: inhalation of silica particles → deposition in bronchioles → phagocytosis by macrophages → macrophage dies → release toxic silica particles → reingestion → inflammation and fibroblast migration/proliferation

- nodules develop to contain particles → nodules coalesce → Late-stage silicosis (PMF)

Chronic	Accelerated	PMF	Acute
10-20 yr development	occurs faster higher level of exposure	Small nodules coalesce	Triggers hypersensitivity
CXR - profusion of small, rounded densities within upper lobe zones (early). • Progression - nodules ↑ in number and coalesce. Middle/lower lobe involved Eggshell calcifications also coal workers and Hodgkins UPPER LOBE		Pts w/ chronic silicosis < pts w/ accelerated Nodules coalesce into conglomerate masses CXR - hyperinflation, eggshell calcification Silicotuberculosis	MASSIVE exposure • acute onset (weeks) CXR - alveolar filling process in lower zones

Coal workers Pneumoconiosis
black lung disease due to coal exposure

Rate and quantity of dust accumulation most important factor in pathogenesis

Clinical Presentation: Similar to silicosis - simple, chronic, PMF

Diagnosis: CXR - Same as silicosis UPPER LOBE

Black color of nodules distinguishes from silicotic nodules

Coal macule → nodule lesions run in random directions v. concentric lesions in Silicotic

Caplan Syndrome is swelling and scarring of lungs due to pneumoconiosis + RA

Talc
paint, pharmaceutical, rubber, and cosmetics industries

Talcosilicosis: Caused by talc mined w/ high silica content

Talcoasbestosis: Crystalline talc contaminated by asbestos fibers

Talcosis: inhalation of pure talc → bronchitis

IV talc injection - cutting heroin w/ talc → granulomas → pHTN

Diagnosis: need exposure hx, ↑scan, Clinical presentation UPPER LOBE

Berylliosis
aerospace, automotive, computer, ceramics, nuclear industries

- looks like Sarcoidosis but less common. Hilar lymphadenopathy, non-necrotizing granulomas. Non-infectious.

UPPER LOBE

Asbestos LOWER LOBE

Occupations - plumbers, pipefitters, electricians, insulation, carpenter, boilermakers, welders, cutters

Industries - construction, shipbuilder, manufacturing, railways, textile mills

Risk factors - fiber, dose, intensity, ± ventilation, dust control measures

Pathophysiology: direct toxic effect, release of inflammatory mediators, fibroblast recruitment

Diagnosis: photomicrograph dumbbell-shaped asbestos body

PLEURAL PLAQUES

- >20 yrs after exposure
- 50% people exposed to asbestos
- dense → lower lobe on diaphragm
- ± calcifications on CXR and CT

BAPE

- first 20 yrs after exposure
- pleuritic chest pain and fever
- exudative fluid - eosinophil predominant
- ± diffuse pleural thickening

MESOTHELIOMA - Cancer

of mesothelial cells of pleura

- 20-40yrs after exposure but RARE
- not very treatable pts die quickly

Asbestosis - Scarring of lungs

- >30 years after exposure
- long term, heavy exposure

Diagnosis - exposure, dyspnea, crackles in 2+ locations, ↓lung volume, honeycombing
- honeycombing indicates scarring

LOOKS like IPF - history + plaques

Clinical Presentation SOB → dyspnea

HYPERSENSITIVITY PNEUMONITIS

Pathophysiology: immune mediated response to inhaled organic antigen
- derived from microorganisms or proteins
- occupational exposure

Antigen exposure → sensitization of T-cells over time
Antigen exposure → lymphocytic inflammation and macrophage activation → granulomatous inflammation

Clinical Presentation: acute (sudden onset, heavy exposure)
subacute (3-6 months)
chronic (causes scarring to lung)

Diagnosis: exposure (history or IgG antibodies)
clinical presentation (radiographic, PFT abnormalities)
lymphocytosis (bronchoalveolar lavage) - lots of lymphs on branch
histopathology (loose formed granulomas and lymphocytic inflammation)
- peribronchiolar inflammation and fibrosis

Treatment: avoid irritants. Steroids.