Restrictive Lung Diseases

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Restrictive lung diseases

- Chest wall
- Lung parenchyma
- Neuromuscular disorders
- Typical PFT pattern:
  - Small lung volumes
  - Reduction of flow throughout but in proportion, so FEV_1/FVC is often well preserved
  - Normal vs low D_LCO
    - parenchymal abnormalities
    - atelectasis (in advance cases of chest wall/neuromuscular disorders)

Chest Wall

- Kyphoscoliosis
- Obesity
- Limited expansion of the chest wall during inspiration, resulting in lower lung volumes and a restrictive lung syndrome picture
- Tidal breathing nearer closing volume, resulting in hypoxia from shunting through the bases of the lungs (very important factor for hypoxia post-op)

Neuromuscular disorders

- Respiratory muscle weakness
  - ALS, Muscular dystrophies
  - Guillain-Barre Syndrome
  - Myasthenia Gravis
- Reduce PEF
- Smaller lung volumes
- Reduction of flow throughout
- Often well preserved FEV_1/FVC
- In severe expiratory muscle weakness, the effectiveness of cough might be impaired
- Increased risk of aspiration (lower and posterior lobar segments)

Lung parenchyma

- Diffuse parenchymal lung diseases, often collectively referred to as the interstitial lung diseases (ILDs)
- The term *interstitial* is misleading since most of these disorders are also associated with extensive alterations of alveolar and airway architecture

Clinical presentation

- Dyspnea on exertion
- Persistent nonproductive cough
- Abnormal chest x-ray
  - Reticular, reticulo-nodular patterns
  - Distribution (bases, periphery)
  - Honeycombing
  - Ground-glass pattern (HRCT criteria, not on CXR)
- Pulmonary symptoms associated with another disease, such as a connective tissue disease
- Lung function abnormalities

Diagnosis

- History
  - Age (sarcoidosis, CT-ass. ILDs, LAM, Histiocytosis, inherited forms of IPF present btw age 20 to 40)
  - Gender (LAM limited to female, RA-ass ILDs and pneumoconiosis more common in men)
  - Smoking (Langerhans cell histiocytosis, DIP, IPF, respiratory bronchiolitis, Goodpasture's syndrome (worsening of disease)) or non-smoking (sarcoidosis, hypersensitivity pneumonitis)
  - Medication use (amiodarone, furantoin, chemotherapy agents, oils, radiotherapy)

Diagnosis (cont'd)

- History of…
  - Occupational exposure (dusts such as in asbestosis, berylliosis and coal worker's lung; gases, chemicals)
  - Environmental (farmer's lung, pigeon breeder's disease, extrinsic allergic alveolitis)
- P/E, chest x-ray, HRCT, ABG, spirometry, lung volume measures, D_LCO, blood tests
- Gallium scan
- Lung biopsy (BAL, BBx, TBBx, OLBx)

Clinical Classification of ILDs

- Idiopathic fibrotic disorders
- Primary diseases
- Occupational and environmental diseases
- Drug-induced
- Associated with connective tissue diseases

Not based on histology

Idiopathic fibrotic disorders

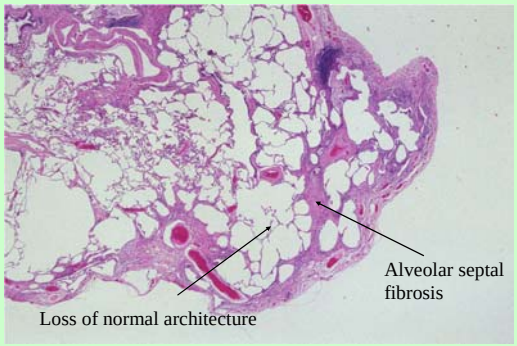
- Acute interstitial pneumonitis (Hamman-Rich syndrome)
- Idiopathic pulmonary fibrosis (IPF)
- Familial IPF
- Desquamative interstitial pneumonitis (DIP)
- Bronchiolitis obliterans organizing pneumonia (BOOP)
- Non-specific interstitial pneumonitis (NSIP)
- Lymphocytic interstitial pneumonitis (LIP) (Sjogren's, CTD, AIDS)
- Autoimmune pulmonary fibrosis (IBD, PBC, ITP, AHA)

Not for exam purposes, do not worry

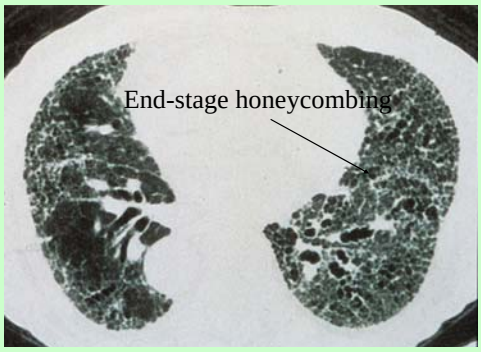
Idiopathic Pulmonary Fibrosis

- Insidious onset of dyspnea in patients between 50 and 70 associated with dry cough
- Late inspiratory crackles at the bases (velcro-like)
- Clubbing in 40-75% of cases
- Chest x-ray
  - Reticular pattern
  - Linear densities
  - Honeycombing
  - Pleural involvement uncommon (suggests another diagnosis)

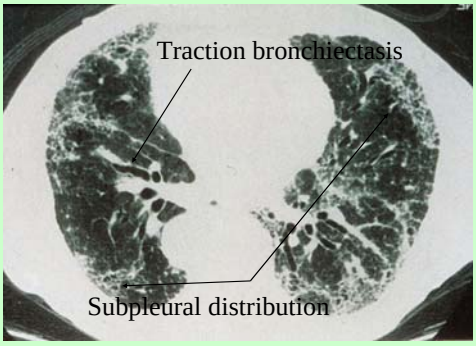
Histology of IPF



HRCT of IPF



HRCT of IPF

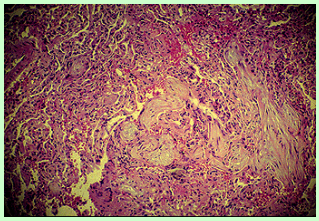


Treatment

- Corticosteroids trials
  - 1/3 of patients will respond
  - Suppress chronic alveolitis
- Cyclophosphamide
- Azathioprine
- Colchicine
  - Alternative treatment
- Oxygen, annual influenza vaccination
- Lung transplant

BOOP

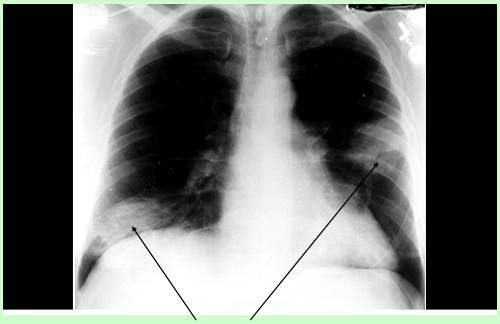
- Formerly called Cryptogenic organizing pneumonia
- Pneumonia-like illness with an excessive proliferation of granulation tissue within the small airways/alveolar ducts, associated with chronic inflammation of the surrounding alveoli



BOOP

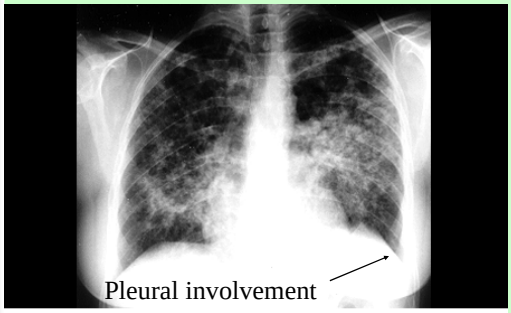
- Flu-like illness of acute onset with cough, fever, malaise, weight loss
- Chest x-ray
  - Peripheral distribution
  - Fleeting infiltrates, often wedge-shaped
  - Pleural involvement common

Chest x-ray of BOOP

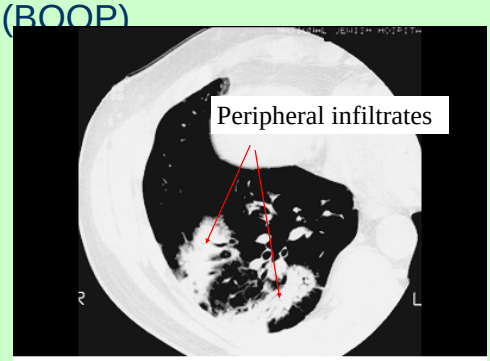


Peripheral, wedge-shaped infiltrates

Chest x-ray of BOOP



CTscan of the CHEST (BOOP)



Treatment of BOOP

- Complete recovery in 2/3 of patients with corticosteroids, frequent relapses upon tapering
- Spontaneous improvement over 3 to 6 months in some patients
- Can sometimes progress to an ARDS-like picture

Primary (unclassified) diseases

- Sarcoidosis
- Langerhans cells histiocytosis
- Lymphangioleiomyomatosis (LAM)
- Amyloidosis
- Pulmonary vasculitis
- ARDS
- AIDS
- Diffuse alveolar hemorrhage
- Alveolar microlithiasis

Not for exam purposes, do not worry

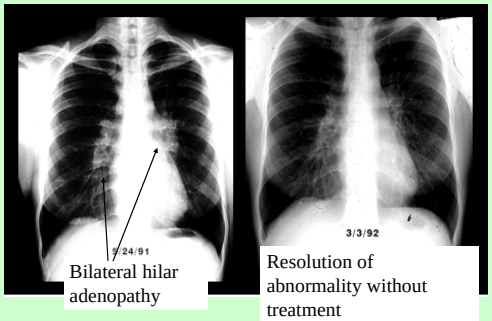
Sarcoidosis

- Onset of disease most commonly between 20 and 40 years of age
- Typically asymptomatic patients with an abnormal chest x-ray
- Löffgren's syndrome
  - Erythema nodosum
  - Bilateral hilar adenopathy
  - Arthralgias
- Heerfordt's syndrome (uveoparotid fever)
  - Facial nerve palsy
  - Parotitis

Stage of Sarcoidosis

- Based on the chest x-ray appearance
- Stage 0: Normal chest x-ray
- Stage 1: Bilateral hilar adenopathy
- Stage 2: Bilateral hilar adenopathy with parenchymal infiltration
- Stage 3: Parenchymal infiltration without hilar adenopathy
- Stage 4: Advanced fibrosis

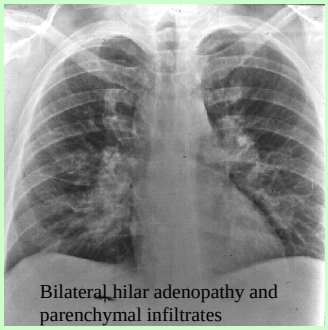
Sarcoidosis stage 1



Bilateral hilar adenopathy

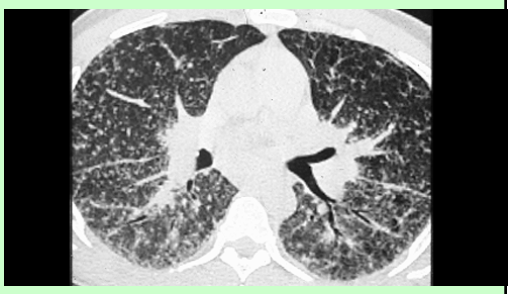
Resolution of abnormality without treatment

Sarcoidosis stage 2

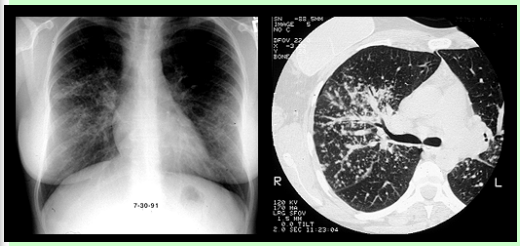


Bilateral hilar adenopathy and parenchymal infiltrates

Sarcoidosis stage 2, HRCT



Sarcoidosis stage 3



Lung infiltrates with absence of hilar adenopathy
Stage 4: end-stage lung fibrosis

Natural history & treatment

- 30-50% of cases will remit spontaneously in a period of up to 3 years
- 30% will show progression over the next 5 to 10 years
- 30% will remain stable
- Follow clinical symptoms and PFTs
- Corticosteroids, MTX, azathioprine, hydroquinolone

Occupational/Environmental

- Silicosis
- Asbestosis
- Coal worker's pneumoconiosis
- Berylliosis
- Talc pneumoconiosis
- Bird breeder's lung
- Farmer's lung

Not for exam purposes, do not worry

Drug-induced ILDs

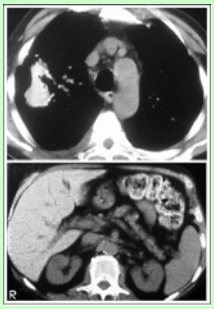
- Furantoin, sulfazalazine
- Amiodarone, propanolol
- Gold, penicillamine
- Dilantin
- Bleomycin, busulfan, cyclophosphamide, methotrexate, azathioprine
- Radiation
- Oxygen toxicity
- Narcotics

Not for exam purposes, do not worry

Drug-induced diseases

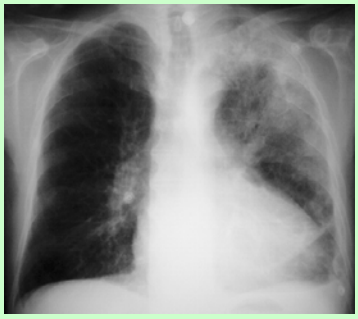
- Amiodarone
  - 4-6% incidence, usually after at least 1 month of use, most while receiving at least 400mg/d
  - Insidious dyspnea, non-productive cough, often associated with a low-grade fever
  - Interstitial or alveolar pattern
  - Pleural effusion uncommon
  - Can progress even if the drug is discontinued
  - Lesions are *sometimes* denser than surrounding tissues and have a high attenuation because of the iodine deposition

Amiodarone-induced lung disease, CTscan



High attenuation of the lung consolidation and the liver parenchyma due to iodine deposition

Amiodarone-induced lung disease



Drug-induced diseases (cont'd)

- Bleomycin
  - Most common chemotherapeutic drug to produce lung toxicity
  - Up to 20% of treated patients will develop clinical pulmonary disease (1% mortality)
  - Frequent monitoring of D_LCO may help predict subsequent clinical disease
  - Definite correlation between prior or concurrent thoracic XRT
  - Synergic toxicity with O_2 (careful intra-op and post-op)

Connective tissue diseases

- Scleroderma
- Polymyositis-Dermatomyositis
- SLE
- RA
- Mixed CTDs
- Ankylosing spondylitis

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Conclusion

- Multiple etiologies are the cause of the restrictive lung diseases but some diseases are more common than others
- History and P/E are key to diagnosis
- Careful interpretation of the PFTs
- Serial review of the chest x-rays and HRCT
- Question: Why do obesity cause hypoxemia when the patient assumes a recumbent position?