

Interstitial Lung Disease (restrictive)

Pathophysiology

- **Inflammatory process** involving the **alveolar wall** (resulting in **widespread fibroelastic proliferation and collagen deposition**).
- Can lead to irreversible fibrosis, distortion of the lung architecture, and impaired gas exchange.
- Prognosis is very variable and depends on diagnosis.

History

- Ask about medication history (esp. chemotherapeutic agents, gold, **amiodarone**, penicillamine, and nitrofurantoin)
- Previous jobs (exposure to asbestos, silicone, beryllium, and coal).

Signs

- **Rales at the bases** are common, **digital clubbing** is common (esp. in idiopathic pulmonary fibrosis), signs of **pulmonary HTN** and cyanosis in advanced disease.

Symptoms

- Dyspnea (at first with exertion then at rest), **cough (nonproductive)**, fatigue, symptoms secondary to another condition.

Interstitial Lung Disease (restrictive) (cont)

Diagnosis

- CXR, CT, PFTs, oxygen desaturation during exercise.
- Bronchoalveolar lavage for culture and cytology yields variable results.
- Tissue biopsy is often required and can be done via fiberoptic bronchoscopy with transbronchial biopsy (limited utility), open lung biopsy, or video-assisted thorascopic lung biopsy (VATS).
- UA can show signs of **glomerular injury in Goodpasture's or Wegener's**.

Causes ILD

Environmental

- Coal worker's pneumoconiosis, silicosis, asbestosis, berylliosis.

Granuloma Associated

- **Sarcoidosis** (along with other organ involvement), Wegener's granulomatosis, Churg-Strauss syndrome.

Alveolar Filling Disease

- Goodpasture's syndrome, Idiopathic pulmonary hemosiderosis, alveolar proteinosis.

Hypersensitivity

- Hypersensitivity pneumonitis, eosinophilic pneumonitis.

Drug Induced

- **Amiodarone**, nitrofurantoin, bleomycin, phenytoin, illicit drugs.

Causes ILD (cont)

Miscellaneous Causes

- Idiopathic pulmonary fibrosis, **bronchiolitis obliertans organizing pneumonia (BOOP)**, ILD associated with connective tissue disorders (RA, scleroderma, SLE, mixed connective tissue disease), ARDS, infection (fungal, TB, viral pneumonia), radiation pneumonitis.

Tests

CXR

- Typical diffuse changes are noted (**reticular, reticulonodular, ground glass, honeycombing**).
- **Honeycombing** refers to a **scarred shrunken lung** and is an end-stage finding with poor prognosis. Air spaces are dilated and there are fibrous scars in the interstitial. Can arise from many causes of ILD.

Pulmonary Function Tests

- A restrictive pattern. **FEV1/FVC is increased >80%**. Both FEV1 and FVC are low. Decreased DLCO. Decreased TLC and RV.

CT Scan

- Shows the extent of fibrosis better than other imaging. **Fibrosis, honeycombing, or traction bronchiectasis**.