Interstitial Lung Disease

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Objectives

- *Define interstitial lung disease*
- *Clinical presentation and history*
- *Physical Exam findings*
- *Diagnostic testing*

Interstitial Lung Diseases

- *Group of diseases classified together because of similar clinical, radiographic, physiologic or pathologic manifestations*
- *Despite the name, the disease also affects the alveoli and airway architecture*

Classification

- *Divided into those with known causes and idiopathic*
  - *Infections*
  - *Exposures*
  - *Idiopathic*

Infections

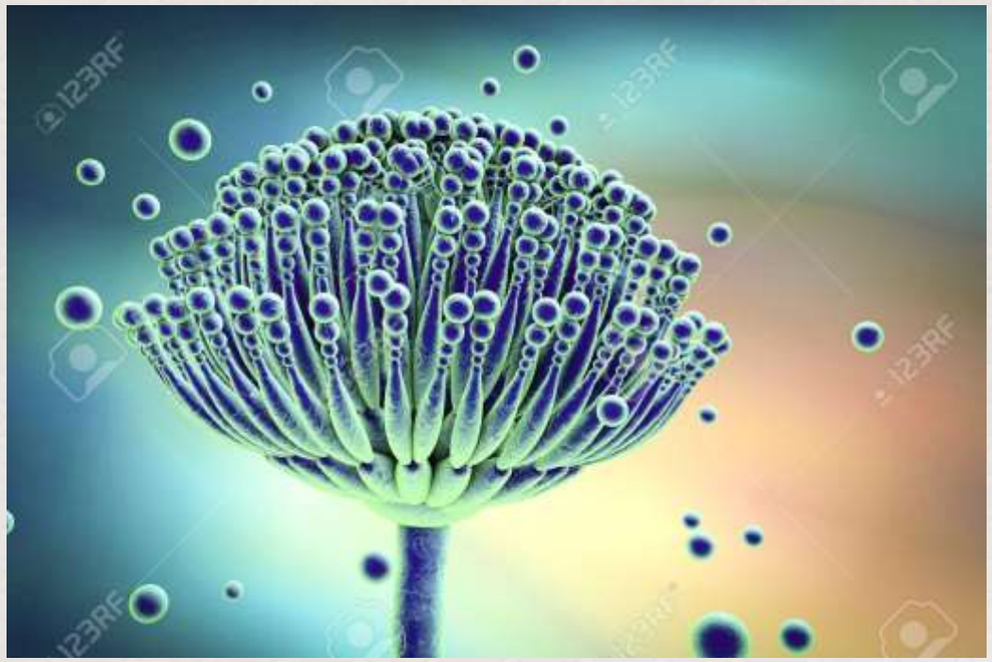
- *Fungal pneumonias*
  - *Coccidioidomycosis*
  - *Cryptococcosis*
  - *Pneumocystis jirovecii*
- *Atypical bacterial pneumonias*
- *Viral pneumonias*

Exposures

- *Occupational and environmental*
  - *Inorganic dusts*
    - *Silicates- asbestos, talc, beryllium*
    - *Carbon- coal dust, graphite*
    - *Metals- tin, aluminum, hard metal dusts*
    - *Others- iron, antimony, hematite, rare earths, etc.*

Exposures

- *Organic dusts*
  - *Thermophilic fungi- farmer's lung, grain handler's lung, air conditioner lung*
  - *Bacteria- humidifier lung*
  - *True fungi- aspergillus*
  - *Animal proteins- bird fancier's disease*



Exposures

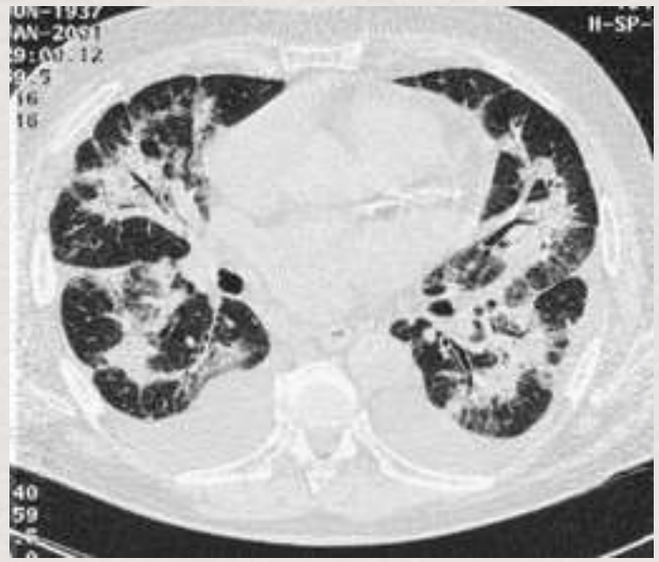
- *Other inhaled agents*
  - *Chemical sources*
    - *Fiber lung, bakelite worker's lung, vinyl chloride, polyvinyl chloride powder*
  - *Gases*
    - *Oxygen, oxides of nitrogen, sulfur dioxide, chlorine gas, methyl isocyanate*
  - *Fumes*
    - *Oxides of zinc, copper, manganese, cadmium, iron, magnesium, nickel, brass*

Exposures

- *Vapors*
  - *Hydrocarbons, thermosetting resins (rubber tire workers), toluene, mercury*
- *Aerosols*
  - *Oils, fats*

Exposures

- *Drug-induced pulmonary toxicity*
  - *Chemotherapy agents, amiodarone*
- *Radiation-induced lung injury*



Idiopathic

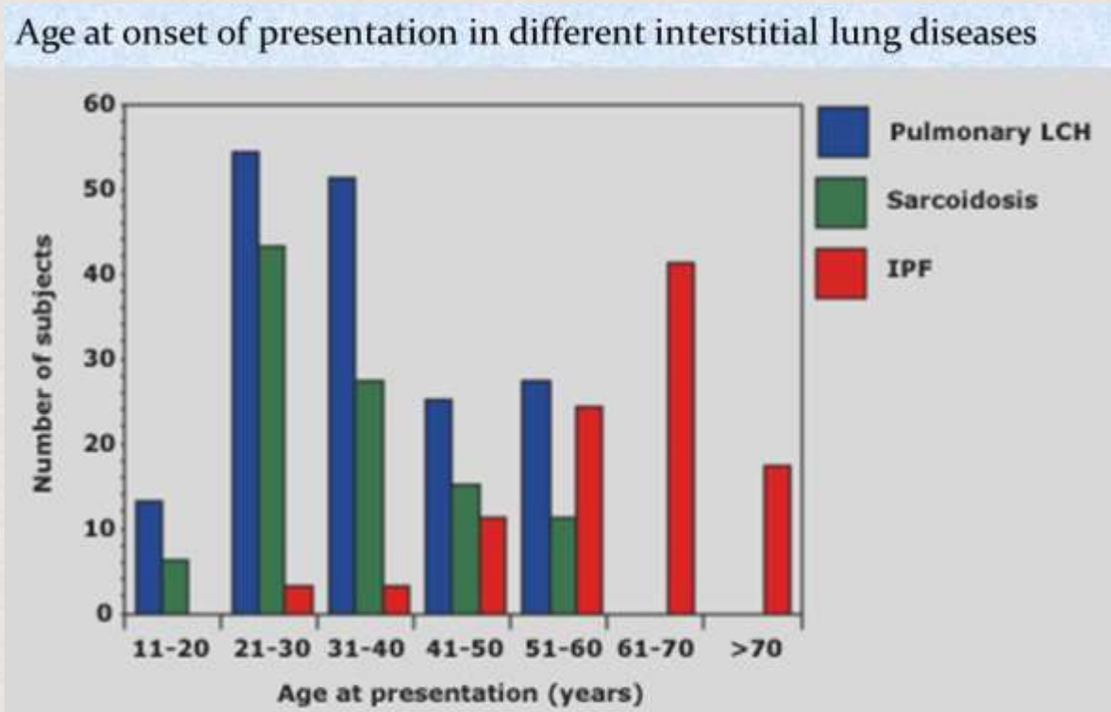
- *Sarcoidosis, cryptogenic organizing pneumonia, idiopathic interstitial pneumonias*

Clinical Presentation

- *Progressive breathlessness with exertion*
- *Persistent nonproductive cough*
- *Pulmonary symptoms associated with another disease*
- *History of occupational exposure*
- *Abnormal chest imaging study*
- *Lung function abnormalities*

History

- *Most important step in initial evaluation*
- *Age and gender*
  - *Age 20-40: sarcoidosis, connective tissue disease*
  - *Female: lymphangioleiomyomatosis, tuberous sclerosis*
- *Onset of symptoms*



Idiopathic pulmonary fibrosis (IPF) has an older age distribution than either pulmonary Langerhans cell histiocytosis (LCH) or sarcoidosis.

History

- *Past medical history*
  - *Connective tissue disease, inflammatory bowel disease, malignancy*
- *Smoking history*
  - *Langerhans cell histiocytosis, respiratory bronchiolitis-interstitial lung disease(RBILD), desquamative interstitial pneumonitis (DIP), idiopathic pulmonary fibrosis(IPF)*
  - *Sarcoidosis, hypersensitivity pneumonitis*
- *Family history*

History

- *Prior medication use and irradiation*
  - *May occur weeks to years after drug has been discontinued*
    - *pneumotox.com*
  - *Radiation injury directly related to the volume of irradiated lung and cumulative dose of radiation*
    - *Acute- 4-12 weeks following treatment*
    - *Late- 6-12 months*
    - *Radiation recall pneumonitis- months to years after radiation when certain antineoplastic agents are administered to patients who have previously received radiation*

History

- *Occupational and environmental exposures*
  - *Don't forget spouse and children*
  - *Duties and known exposures in chronological order*
  - *Hobbies*

Symptoms

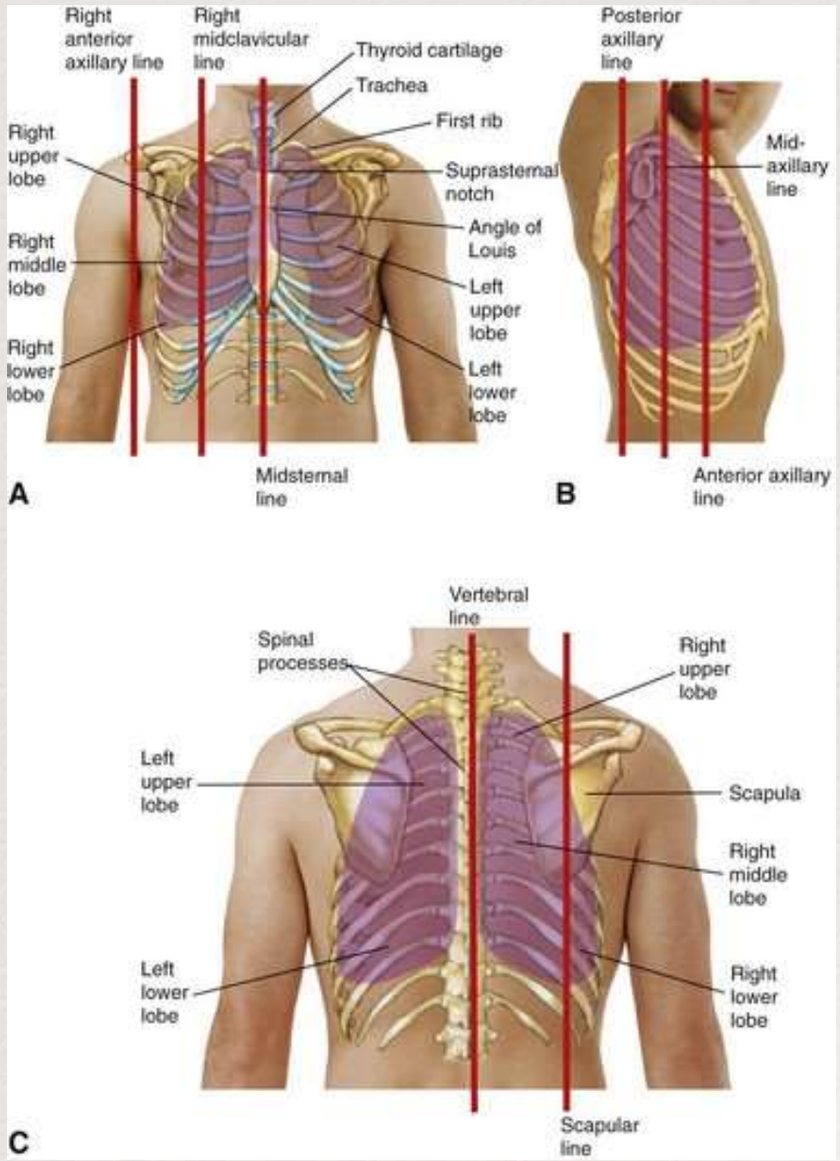
- *Dyspnea*
  - *Sometimes patients may have significant changes on imaging without symptoms*
  - *Sudden worsening may indicate spontaneous pneumothorax*
- *Cough*
  - *Dry, common symptom*

Symptoms

- *Hemoptysis- diffuse alveolar hemorrhage syndromes, lymphangioleiomyomatosis, tuberous sclerosis*
  - *New onset in patient with previous ILD suggests complicating malignancy*
- *Wheezing- uncommon symptom*
- *Chest pain- uncommon symptom, unless associated with underlying connective tissue disorder*
- *Extra pulmonary symptoms*

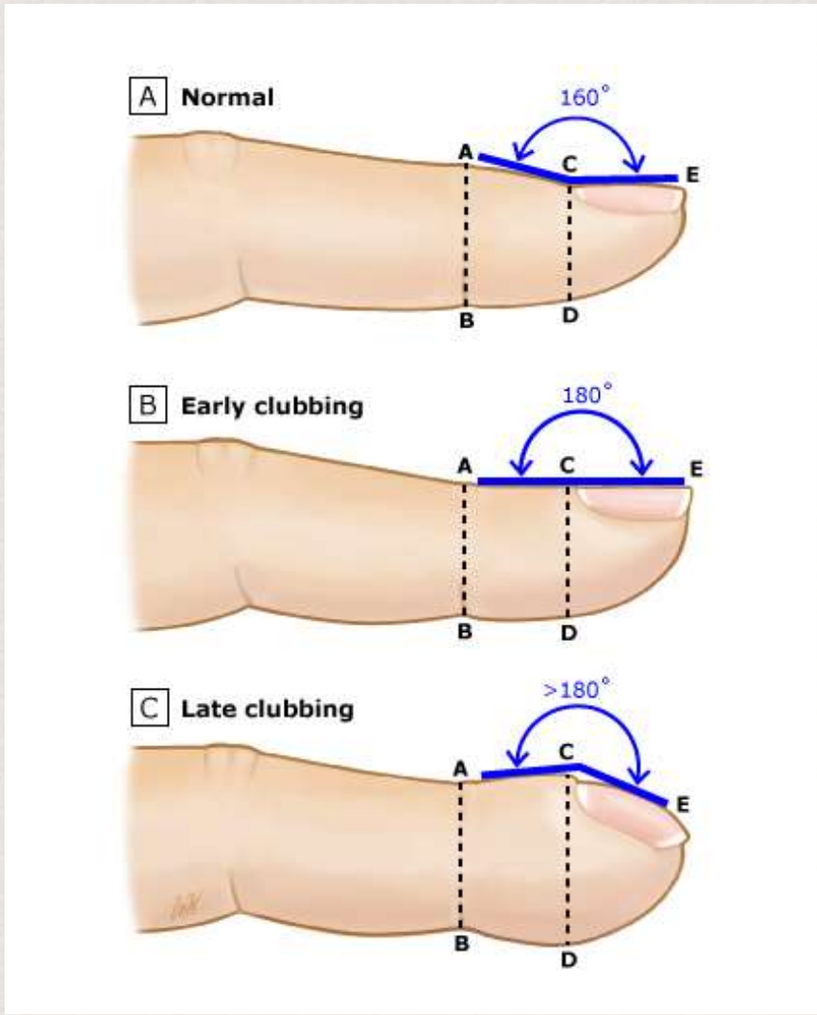
Physical Exam

- *Lung exam*
  - *Frequently abnormal, but nonspecific*
- *Crackles or velcro rales, in lung bases, posterior axillary line in early disease*



Physical Exam

- *Cardiac exam- normal until advanced stages*
  - *Pulmonary hypertension, cor pulmonale*
- *Clubbing*
- *Extrapulmonary findings of systemic disease*



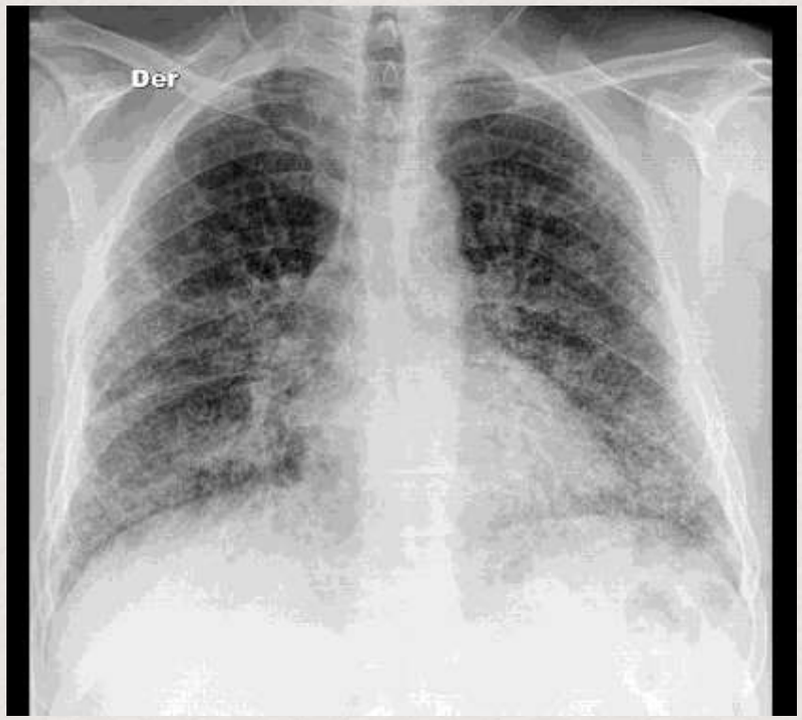
Diagnostic Testing

Laboratory

- *Hepatic and renal function*
- *Hematologic tests to look for anemia, polycythemia, leukocytosis, or eosinophilia*
- *Urinalysis*
- *Serology- ANA, rheumatoid factor*
- *Pulmonary hemorrhage patients- antiGBM antibodies, ANCA, ANA, APL antibodies*
- *Biomarkers- currently for research purposes*

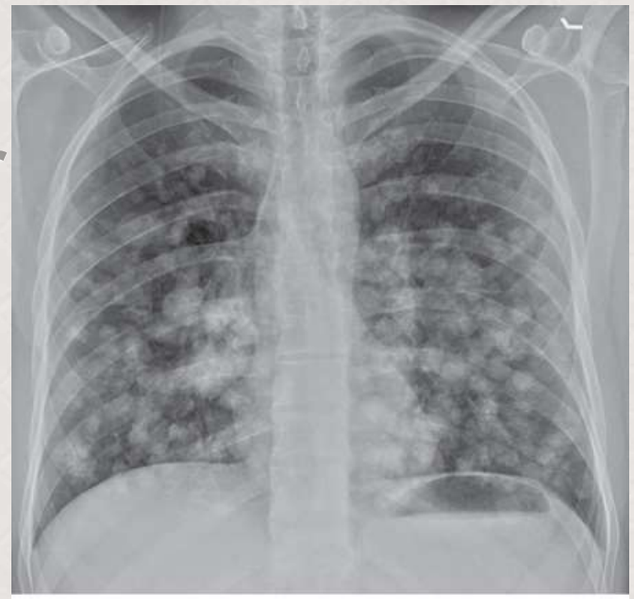
Imaging

- *Chest radiography*
  - *Reticular pattern most common*
    - *Nodular*
    - *Mixed*
- *Pattern poorly correlates to stage of disease*
- *Honeycombing indicates poor prognosis*



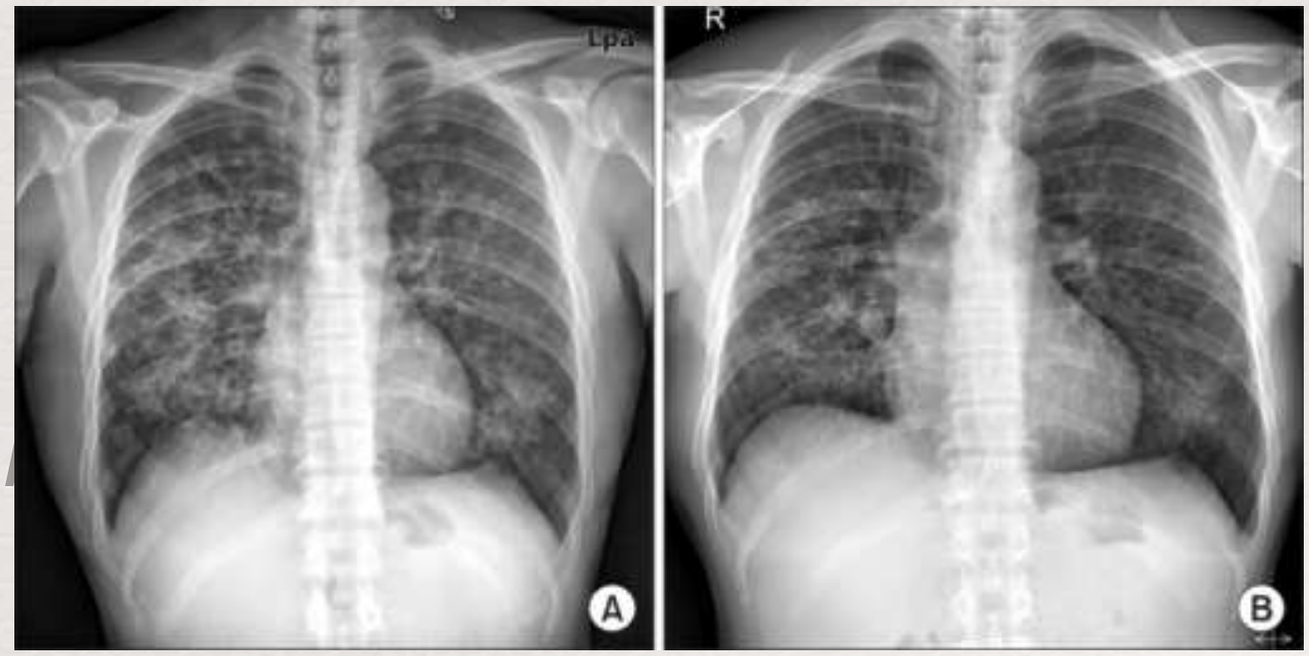
Reticular pattern

Nodular Pattern



Source: Chen MYM, Pope TL, Ott DJ: *Basic Radiology, 2nd Edition*: http://www.accessmedicine.com

Reticulonodular Pattern



CT Scan

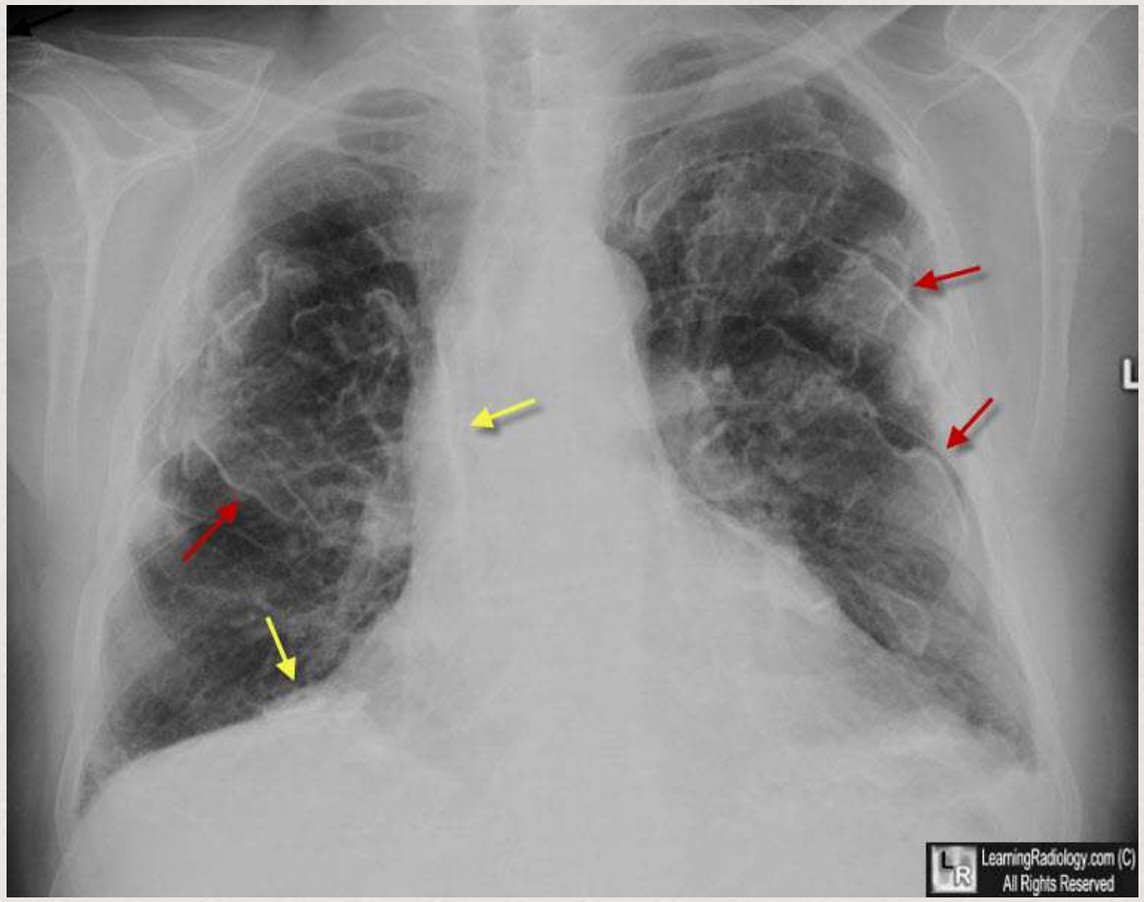
- *High Resolution CT (HRCT) should be obtained in almost all patients with diffuse parenchymal disease*
- *Supine and prone imaging differentiate between dependent atelectasis and interstitial opacities*
- *Inspiratory and expiratory images are helpful when bronchiolitis is suspected*

Patterns on HRCT

- *Septal- interstitial pulmonary edema, lymphatic spread of tumor*
- *Reticular-sarcoidosis, Langerhans cell histiocytosis, lymphangiomyomytosis*
- *Nodular- silicosis, coal worker's pneumoconiosis, sarcoidosis, tuberculosis*
- *Reticulonodular- Langerhans cell histiocytosis, sarcoidosis, lymphatic spread of tumor*
- *Ground glass- NSIP, idiopathic pulmonary hemorrhage*

HRCT Key Findings

- *Bilateral symmetric hilar adenopathy and upper lung zone reticular opacities suggest sarcoidosis or another granulomatous disease*
- *Pleural plaques with linear calcification in association with a basilar predominance of reticular opacities suggest asbestosis*
- *Centrilobular nodules that spare the subpleural space are seen in hypersensitivity pneumonitis, sarcoidosis, Langerhans cell histiocytosis*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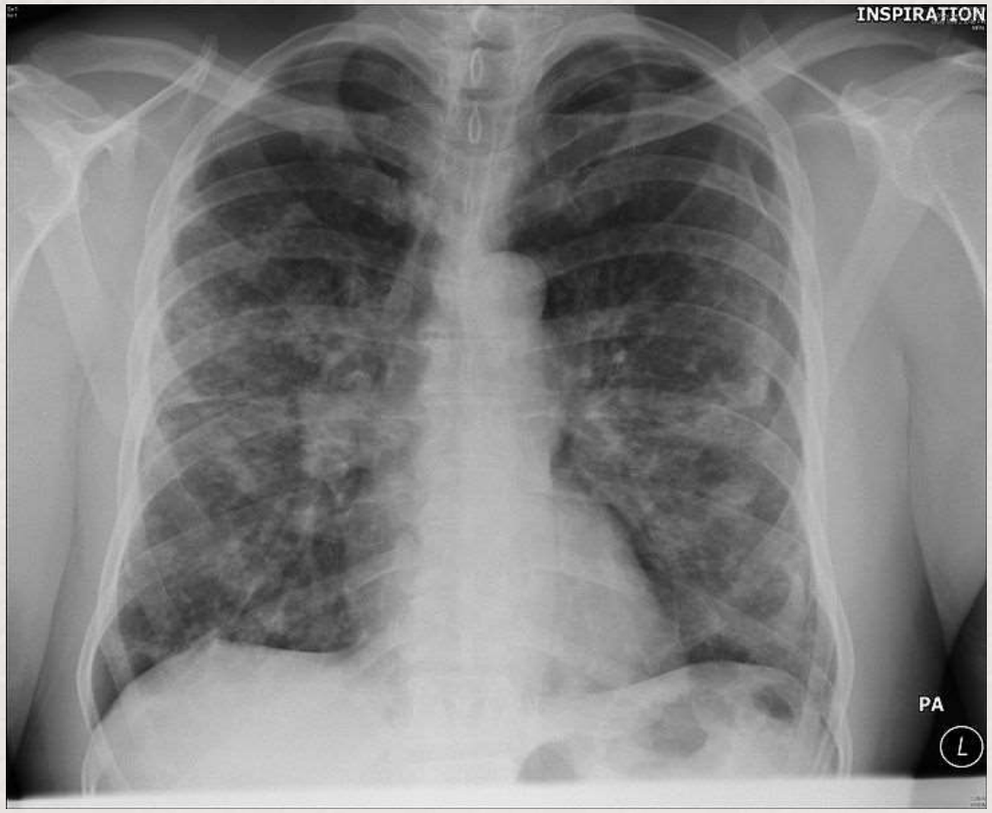
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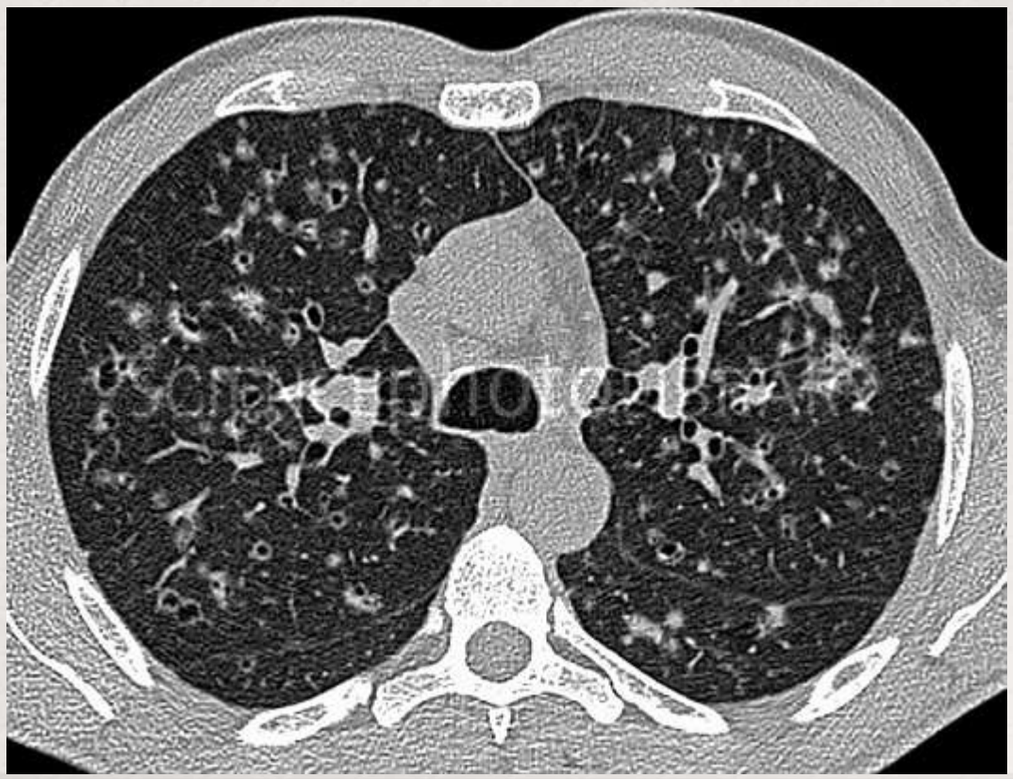
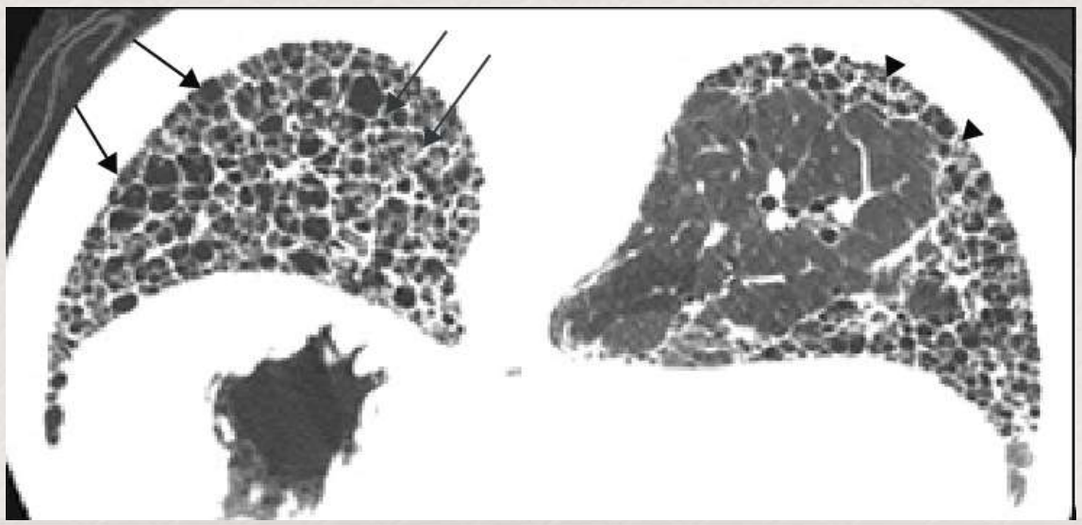


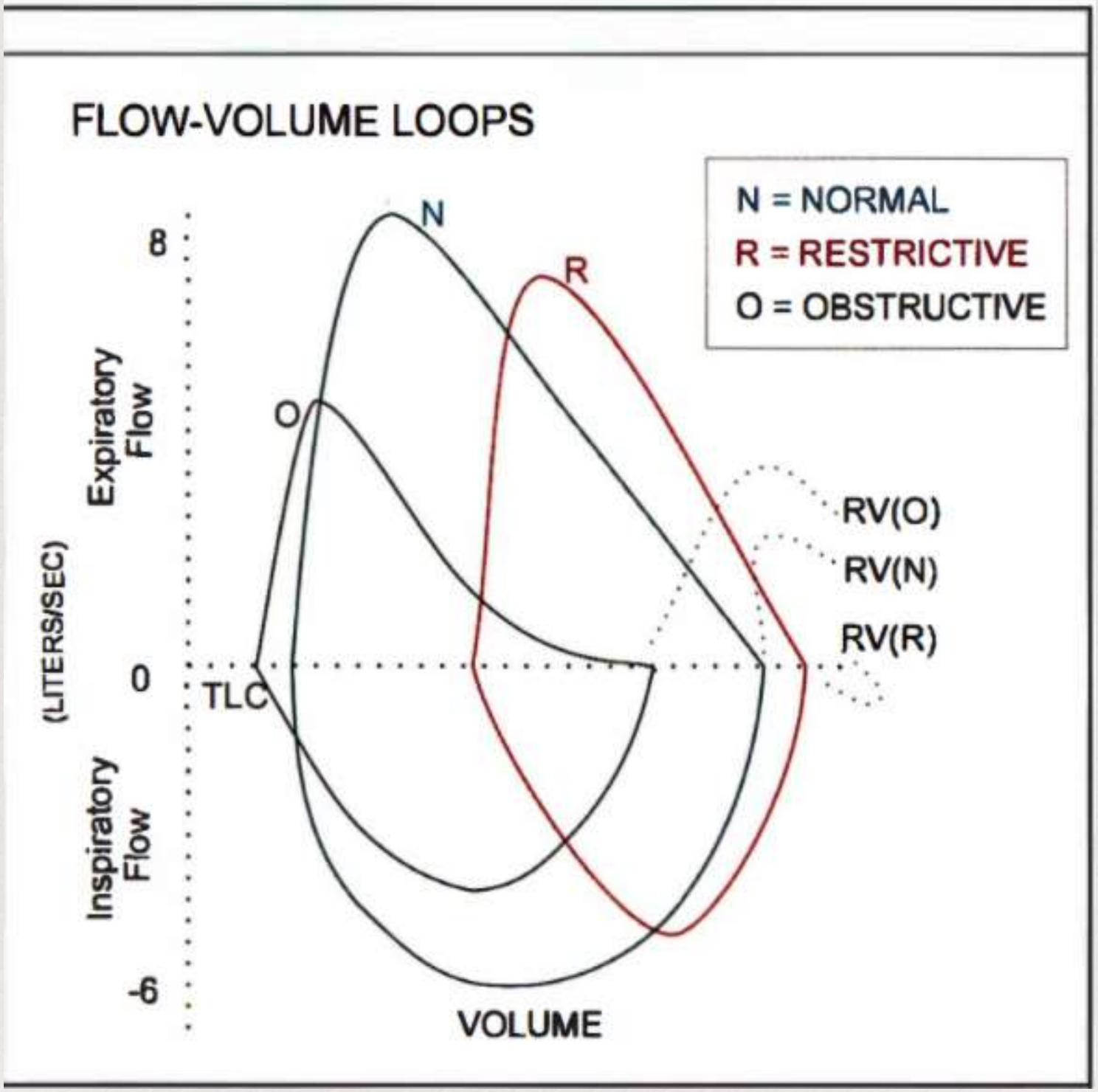
HRCT Key Findings

- *Irregular cysts associated with nodules in the upper and middle lung zones suggest pulmonary Langerhans cell histiocytosis*
- *Basilar and peripheral reticular opacities, traction bronchiectasis, and honeycombing in a subpleural location are classic features associated with usual interstitial pneumonitis (UIP)*



Pulmonary Function Testing

- *Complete lung function tests (spirometry, lung volumes and diffusion capacity) are usually performed on patients suspected of having interstitial lung disease*
  - *Most diseases will have a restrictive pattern with decreases in TLC, FRC and RV*
  - *FEV1/FVC ratio is usually normal or increased*
  - *Lung volumes are further reduced as disease severity progresses*


FLOW-VOLUME LOOPS
N = NORMAL
R = RESTRICTIVE
O = OBSTRUCTIVE
8
Expiratory Flow
(LITERS/SEC)
0
TLC
Inspiratory Flow
-6
N
R
O
RV(O)
RV(N)
RV(R)
VOLUME

Pulmonary Function Tests

- *There is a group of diseases that may present with an obstructive pattern*
  - *Sarcoidosis*
  - *Lymphangioleiomyomatosis*
  - *Hypersensitivity pneumonitis*
  - *Pulmonary Langerhans cell histiocytosis*
  - *Tuberous sclerosis*
  - *Combined COPD and ILD*
  - *Constrictive bronchiolitis*

Diffusion Capacity

- *Reduction is common but nonspecific finding in ILD*
- *Severity does not correlate well with disease prognosis unless <35% predicted*

Gas Exchange Abnormalities

- *Arterial Blood Gases*
  - *May be normal in early ILD*
  - *May reveal hypoxemia and respiratory alkalosis*
- *Cardiopulmonary Exercise Testing*
  - *Oxygen desaturation, high dead space/tidal volume ratio, excessive increase in respiratory rate*

Gas Exchange Abnormalities

- *Pulse Oxygen Saturation*
  - *Serial assessment of resting and exercise gas exchange is one of the methods to follow ILD activity and responsiveness to treatment*
    - *Results of 6MWT have correlated with prognosis in several studies of IPF*

Cardiac Evaluation

- *Important to assess cardiac function in the initial evaluation as heart failure is in the differential diagnosis of ILD*
- *ECG to evaluate for pulmonary hypertension or cardiac disease*
- *BNP if heart failure is suspected*

Cardiac Evaluation

- *No clear guidelines for ECHO*
  - *Consider if abnormal ECG, suspected heart failure, rapid onset of radiologic findings, or moderate to severe reduction in DLCO*
- *Assessment for pulmonary hypertension is important because it may be a clue to underlying etiology of ILD*

Bronchoalveolar Lavage

- *Collect samples of cells and fluid from distal airways and alveoli*
- *Sent for cell counts; cultures for mycobacterial, viral, and fungal pathogens; and cytologic analysis*

Bronchoalveolar Lavage

- *Especially useful in patients with hemoptysis, rapidly progressive disease, or if suspected of having: sarcoidosis, hypersensitivity pneumonitis, pulmonary Langerhans histiocytosis, or infection*
- *Less likely to be helpful if idiopathic pulmonary fibrosis is suspected*
- *No role in assessment of ILD progression or response to therapy*

Lung Biopsy

- *When other testing does not allow for a confident diagnosis, lung biopsy may be necessary*
- *Decision made on a case by case basis*
  - *Atypical or progressive symptoms*
  - *Atypical radiographic features*
  - *Unexplained extrapulmonary manifestations*
  - *Rapid clinical deterioration*
  - *Sudden change in radiographic features*
  - *Conflicting findings on other testing*
  - *Exclude malignancy or infection*

Summary

- *Diffuse parenchymal lung diseases are a group of disorders that are classified together because of similar clinical, radiographic, or pathologic manifestations*
- *Disease is usually recognized following the onset of progressive breathlessness, nonproductive cough, or pulmonary symptoms associated with another disease*
- *Careful documentation of the past medical history is important*
- *Crackles or “velcro rales” are nonspecific and found in most forms of ILD*

Summary

- *Treatment and prognosis varies greatly among ILD, so correct identification is important*
- *High resolution CT should be obtained in most patients as certain findings help narrow the differential diagnosis*
- *ECG can be obtained if there is evidence of pulmonary hypertension or concurrent cardiac disease*
- *Restrictive pattern is generally seen on PFTs*
- *Lung biopsy should be considered if otherwise unable to make a diagnosis*