# Approach to Internal Medicine

## A Resource Book for Clinical Practice

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## Interstitial Lung Disease

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### DIFFERENTIAL DIAGNOSIS

**PRIMARY (idiopathic)**—usual interstitial pneumonia (UIP), respiratory bronchiolitis-associated interstitial lung disease (RBILD), desquamative interstitial pneumonia (DIP), acute interstitial pneumonia (AIP), non-specific interstitial pneumonia (NSIP), lymphoid interstitial pneumonia (LIP), cryptogenic organizing pneumonia (COP)

**SECONDARY ★DICE★**

- **DRUGS**—chemotherapy (bleomycin), sulfa, penicillin, sulfonylurea, gold, penicillamine, phenytoin, amiodarone, nitrofurantoin
- **INFILTRATIVE**—lymphangitic carcinomatosis, sarcoidosis
- **INFECTIONS**—TB, histoplasmosis, coccidioidomycosis
- **INFLAMMATORY**—rheumatoid arthritis, SLE, scleroderma, ankylosing spondylitis, myositis
- **CONGESTIVE HEART FAILURE**
- **ENVIRONMENT**—**organic dust** (hypersensitivity pneumonitis), **inorganic dust** (asbestos, silica, beryllium, coal worker's pneumoconiosis)
- **EOSINOPHILIA-ASSOCIATED PULMONARY INFILTRATES**—allergic bronchopulmonary aspergillosis (ABPA), parasitic, drugs
- **ETC**—pulmonary histiocytosis X, idiopathic pulmonary hemosiderosis, lymphangioleiomyomatosis, radiation

### CLINICAL FEATURES

**HISTORY**—dyspnea (duration, progression), cough, hemoptysis, wheezes, chest pain, impaired exercise tolerance, occupational history (details of all previous jobs, exposure to gases or chemicals particularly important), environmental exposure (home setting, air-conditioning, pets, hobbies), rash, joint swelling, past medical history (smoking), medications, family history

**PHYSICAL**—vitals (tachypnea, hypoxemia), cyanosis, clubbing (idiopathic pulmonary fibrosis, asbestosis, rheumatoid lung, fibrosing NSIP), decreased chest expansion, crackles (fine), wheezes, cor pulmonale. Note that sarcoidosis and silicosis may have a normal lung examination

**Related Topics**
Allergic Bronchopulmonary Aspergillosis (p. 4)
Restrictive Lung Disease (p. 26)
Rheumatoid Arthritis (p. 297)
Sarcoidosis (p. 483)
Tuberculosis (p. 267)

### INVESTIGATIONS

**BASIC**

- **LABS**—CBC, ANA, RF, anti-CCP antibody, anti-Scl-70, anticentromere antibody, anti-Jo-1 antibody
- **IMAGING**—CXR, CT chest (high resolution), echocardiogram (if suspect pulmonary hypertension)
- **ABG**
- **PFT**

**SPECIAL**

- **BIOPSY**—bronchoscopy (transbronchial biopsy), referral to thoracic surgery for VATS lung biopsy

## DIAGNOSTIC ISSUES

### CHARACTERISTIC CXR PATTERNS FOR INTERSTITIAL LUNG DISEASE

- **UPPER LOBE PREDOMINANCE**—sarcoidosis, hypersensitivity pneumonitis, pneumoconiosis, silicosis, histiocytosis X, PJP, ankylosing spondylitis, ABPA, TB
- **LOWER LOBE PREDOMINANCE**—idiopathic pulmonary fibrosis, asbestosis, rheumatoid arthritis, scleroderma, drugs
- **BILATERAL HILAR/MEDIASTINAL ADENOPATHY WITH INTERSTITIAL INFILTRATES**—sarcoidosis, berylliosis, lymphangitic carcinomatosis, TB, fungal, lymphoma
- **EGGSHELL CALCIFICATION OF HILAR/MEDIASTINAL LYMPH NODES**—silicosis (other pneumoconiosis), TB, fungal
- **CALCIFIED PLEURAL PLAQUES**—asbestos
- **PLEURAL EFFUSIONS WITH INTERSTITIAL INFILTRATES**—HF, lymphangitic carcinomatosis, rheumatoid arthritis, SLE

## MANAGEMENT

**TREAT UNDERLYING CAUSE**—**sarcoidosis** (if stage ≥II or symptomatic, consider glucocorticoids for several months with tapering dose)
**LUNG TRANSPLANT**

## SPECIFIC ENTITIES

### IDIOPATHIC PULMONARY FIBROSIS (IPF)

- **PATHOPHYSIOLOGY**—unknown. Fibrotic rather than inflammatory process; associated with histopathological and/or radiological pattern of usual interstitial pneumonia (UIP)
- **DIAGNOSIS**—high resolution CT chest may show patterns of UIP (honeycombing, interlobular septal thickening, traction bronchiectasis, peripheral, sub-pleural, lack of ground glass pattern), probable UIP, indeterminate for UIP or alternate diagnosis; bronchoscopy (to rule out other causes, mostly infectious); consider open lung biopsy if CT is not consistent with above
- **TREATMENTS**—multidisciplinary discussion for diagnosis and treatment. Referral for lung transplantation should be done early; consider palliative care involvement, pulmonary rehabilitation, vaccinations, supplemental oxygen; consider pirfenidone or nintedanib for mild to moderate disease. Systemic steroids ineffective

## SPECIFIC ENTITIES (CONT'D)

### HYPERSENSITIVITY PNEUMONITIS

- **PATHOPHYSIOLOGY**—inhaled organic antigens → immune response → acute, subacute, or chronic granulomatous pneumonia
- **DIAGNOSIS**—**major criteria** (compatible symptoms, antigen exposure, imaging findings, lavage lymphocytosis, histologic findings [poorly formed granulomas], re-exposure triggers symptoms); **minor criteria** (bilateral crackles, ↓ DLCO, hypoxemia). Combination of major and minor criteria will help raise suspicion of hypersensitivity pneumonitis. Serology may be helpful
- **TREATMENTS**—cessation of exposure, steroids

**ORGANIZING PNEUMONIA** (OP)—previously known as bronchiolitis obliterans organizing pneumonia (BOOP)

- **CAUSES**—**idiopathic** (80%, also known as cryptogenic organizing pneumonia [COP]), **post-infectious** (CMV, influenza, adenovirus, *Chlamydia*), **drugs** (amiodarone, bleomycin, gold, sulfasalazine, cephalosporin, cocaine), **connective tissue disease** (RA, SLE, scleroderma, Sjögren syndrome, dermatomyositis), **immunologic** (essential mixed cryoglobulinemia), **transplantation** (bone marrow, lung, kidney), **malignancy** (MDS, lymphoproliferative diseases, radiation)
- **CLINICAL FEATURES**—about 50% of cases preceded by viral-like respiratory infection. Symptoms include dyspnea on exertion, persistent non-productive cough, and weight loss
- **DIAGNOSIS**—characteristic findings on CXR and CT chest include bilateral, diffuse, ill-defined alveolar opacities distributed peripherally. PFT shows mainly restrictive lung disease pattern
- **TREATMENTS**—*prednisone* 1 mg/kg PO daily for several months with slow taper