# Approach to Interstitial Lung Disease


King, Talmadge. “Approach to the adult with interstitial lung disease.” *UpToDate* 2012.

King, Talmadge. “Overview of sarcoidosis” and “Treatment of pulmonary sarcoidosis with corticosteroids.” *UpToDate* 2012.

**MKSAP 12.**

### Key Points:
- Suspect interstitial lung diseases with insidious dyspnea, even with minimal or no radiographic findings
- A thorough history with exposures and systemic ROS is key
- Firm diagnosis for IPF requires ruling out treatable causes and considering biopsy for atypical presentations

### Clinical presentation
- Symptoms: dyspnea (insidious onset vs. acute) and cough (minimal production) – timing of onset is key
- **Hx:**
  - Age and gender
  - Tobacco use
  - Medications
  - Occupational and environmental exposures (including those of household contacts)
  - Family history
  - HIV risk factors
  - Rheumatology ROS
- **Physical exam (no findings sensitive or specific):**
  - O2 saturation (with exercise)
  - Bilateral inspiratory crackles
  - Clubbing: IPF, asbestos, malignancy
  - Extrapulmonary systemic findings (skin, joints, etc.)
- Complications: pneumothorax, hemorrhage

### Tests
- Consider: infection (TB, fungal, PCP, etc), neoplasm, systemic disease
- **CXR:** reticular / reticulonodular (not sensitive or specific)
- **High resolution CT (see UpToDate for exhaustive radiological ddx) – get prone and supine images for specificity**
  - Reticular:
    - Upper: granulomatous, pneumoconiosis, radiation, drug
    - Lower: IPF, asbestosis
    - Peripheral: BOOP, eosinophilic pneumonia
- **PFT:** restrictive with decreased diffusion
  - FVC, TLC, FRC, RV low
  - DLco low
  - FEV1/FVC normal – high
  - Obstructive spirometry with sarcoid, COPD (underlying), LAM
- Special: antibodies to antigens (for HSP), serologies (ANA, RF, ANCA), lymphocyte proliferation test (beryllium)
- **Biopsy**
  - Bronchoscopy with lavage and transbronchial biopsy: good to r/o infection or hemorrhage
  - VATS: preferred method, for the purposes of:
    - Diagnosis (particularly when potential treatments harmful or not responding)
    - R/O infection or neoplasm
    - Find treatable process
Treatment
- Steroids: variable responsiveness (sarcoid, beryllium, HSP, BOOP, NSIP, DIP, RB-ILD
  - Prophylaxis for PCP, osteoporosis, and other side effects
- Supportive Care
- Lung transplantation: for survival and functional status

Epidemiology and diseases: sarcoid, IPF, HSP, and pneumoconiosis most common causes of ILD
- Sarcoïdosis
  - Stages and spontaneous remission rates:
    - I: hilar adenopathy, 60-80%
    - II: hilar adenopathy with parenchymal opacities, 50-70%
    - III: parenchymal opacities without hilar, <30%
    - IV: fibrosis
  - Poor prognostic factors: onset >40 years, symptoms >6 months, 3 or more organs, skin lesions, bone cysts, arthritis, African descent, increasing infiltrates
  - Rx: for symptoms or advanced disease (given rates of spontaneous remission without steroids)
    - Steroids and steroid-sparing agents (antimalarials, methotrexate, azathioprine)
- IPF
  - Onset: late middle age, slowly progressive
  - Honeycombing with less ground glass, basilar predominance
  - Pathology: usual interstitial pneumonia (UIP) – can diagnose based on HRCT / clinical scenario without bx
  - Rx: poor response to steroids; interferon gamma under investigation
  - Transplantation
- Hypersensitivity pneumonitis (HSP)
  - Acute, subacute, or chronic
  - +/- known trigger
  - BAL: lymphocytic lavage
  - Steroid responsive and need to remove exposure
- Pneumoconiosis
  - Coal, silica, asbestosis, beryllium
  - Can pre-dispose to TB superinfection (silicosis)
- Other:
  - Bronchiolitis obliterans organizing pneumonia
    - 80% idioapathic, although associated with systemic diseases
    - Diffuse patchy infiltrates
    - Resolution without treatment, steroids for progressive disease
  - Vasculitis: Wegener’s, Churg-Strauss, pulmonary capillaritis with diffuse alveolar hemorrhage
  - Eosinophilic pneumonia
  - Pulmonary alveolar proteinosis

Categories of diffuse parenchymal lung diseases (from ATS/ERS consensus)
- Known cause (drugs, CVD)
- Idiopathic interstitial pneumonias
  - Idiopathic pulmonary fibrosis
  - Other interstitial pneumonias: NSIP, DIP, RB-ILD, AIP, COP, LIP
- Granulomatous (sarcoid)
- Other forms (lymphangioleiomyomatosis, histiocytosis, eosinophilic pneumonia)