**Interstitial Lung Disease**

*adapted notes from talk by B. Drummond*

**Ddx for diffuse interstitial lung infiltrates (*most common):**

- **infxn:** viral, atypical pneumonia, MAI, etc.
- **exposures:**
  - toxins: drugs, radiation
  - hypersensitivity pneumonitis*: occupational, farmer’s lung, etc. (look for exposure to meds, mold, birds)
  - pneumoconiosis* (coal, silica, asbestos, beryllium, etc.)
  - **rheum:**
    - collagen vascular disease: scleroderma, dermatomyositis, SLE, sjogrens, RA
    - vasculitides: Wegener’s, churg-strauss, DAH/pulmonary capillaritis
  - granulomatous disease: sarcoid* (clues: hilar LAD, e-nodosum, hypercalcemia)
  - **other:** eosinophilic pneumonia, LAM, histiocytosis X, pulmonary alveolar proteinosis, BOOP, lipoid pneumonia, Goodpasture’s
  - **idiopathic***: IPF*, NSIP, AIP, RB-ILD, DIP, COP, LIP

***Idiopathic interstitial lung disease is classified by clinicoradio-pathological terms (requiring involvement of pulmonology, radiology, pathology)*

<table>
<thead>
<tr>
<th>clinical</th>
<th>rads</th>
<th>path</th>
<th>tx/px</th>
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</thead>
<tbody>
<tr>
<td><strong>IPF (most common)</strong></td>
<td>50-60 yo, M&gt;F (2:1), indolent course, dry cough</td>
<td>basilar honeycombing</td>
<td>UIP</td>
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<tr>
<td><strong>NSIP (“waste basket”)</strong></td>
<td>variable (?)early IPF</td>
<td>ground glass (“active alveolitis”)</td>
<td>non-specific</td>
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<td><strong>AIP</strong></td>
<td>“Hamman-Rich” = ARDS-like syndrome, very sick, acute-onset, infxn-prodrome, 40-50 yo</td>
<td>ground glass, ARDS</td>
<td>DAD (diffuse alveolar damage)</td>
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<tr>
<td><strong>RB-ILD (respiratory bronchiolitis ILD)</strong></td>
<td>smokers</td>
<td>non-specific</td>
<td>RB-ILD</td>
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<tr>
<td><strong>DIP (desquamative interstitial pneumonia)</strong></td>
<td>smokers</td>
<td>non-specific</td>
<td>DIP (epithelial desquamation of airways)</td>
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<td><strong>cryptogenic organizing pneumonia</strong></td>
<td>50-60 yo; acute prodrome</td>
<td>non-specific</td>
<td>COP</td>
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<td><strong>LIP (lymphocytic interstitial pneumonia)</strong></td>
<td>assoc w/ other systemic illness (e.g. HIV w/ low CD4, myeloma, hepatitis, etc.)</td>
<td>non-specific</td>
<td>sheets of lymphocytes in interstitium</td>
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**Workup**

- H&P to assess for non-idiopathic causes
- screening labs: Cx’s, Ab’s to antigens (for HSP), serologies (ANA, RF, ANCA), lymph proliferation test (if suspect beryllium)
- hi-res chest CT: upper (granulomatous, pneumoconiosis, radiation, drug), lower (IPF, asbestos), peripheral (BOOP, eosinophilic pneumonia)
- PFT’s: restrictive w/ decr diffusion
- Bronch w/ BAL & Bx: assess for infxn & hemorrhage
- VATS: diagnosis, r/o infxn/neoplasm, find treatable processes

Note: After deciding ILD is idiopathic, ask if they have IPF. If convinced that they do, no need for open lung Bx – send for transplant eval.

Ref: ATS consensus statement 2002