What is Pulmonary Fibrosis?

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Interstitial Lung Disease
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Pulmonary Fibrosis

Normal Lung Histology
• Inflammation
Pulmonary Fibrosis
Interstitial Lung Disease
Pulmonary Fibrosis

Interstitial Lung Disease

- Fibrosis
Pulmonary Fibrosis  Interstitial Lung Disease

Interstitial Lung Disease
Pulmonary Fibrosis Foundation

Normal Lung Explant
Pulmonary Fibrosis

IPF Lung Explant
Interstitial Lung Disease

Interstitial Lung Disease

Pulmonary Fibrosis

Idiopathic Pulmonary Fibrosis

IIP
Pulmonary Fibrosis

- Pulmonary Fibrosis Mortality
  - 55.1/1,000,000 for females
  - 61.9/1,000,000 for males

- Compared to analyses from 1970, the mortality increased 28.4% in men and 41.3% in women.

### Table 2. Predicted Mortality Rates per 1,000,000 in Men and Women with Pulmonary Fibrosis in 2008

<table>
<thead>
<tr>
<th>Age Strata (yr)</th>
<th>Men Predicted Mortality Rate per 1,000,000</th>
<th>95% Confidence Interval</th>
<th>Women Predicted Mortality Rate per 1,000,000</th>
<th>95% Confidence Interval</th>
</tr>
</thead>
<tbody>
<tr>
<td>45–54</td>
<td>18</td>
<td>16–19</td>
<td>17*</td>
<td>15–18</td>
</tr>
<tr>
<td>55–64</td>
<td>71</td>
<td>67–75</td>
<td>52*</td>
<td>48–56</td>
</tr>
<tr>
<td>65–74</td>
<td>306*</td>
<td>295–317</td>
<td>185*</td>
<td>177–193</td>
</tr>
<tr>
<td>75–84</td>
<td>827*</td>
<td>802–852</td>
<td>494*</td>
<td>478–510</td>
</tr>
<tr>
<td>&gt; 85</td>
<td>1,380*</td>
<td>1,320–1,443</td>
<td>942*</td>
<td>908–977</td>
</tr>
</tbody>
</table>

* p < 0.05 for comparison of predicted 2008 mortality rates and actual 2003 mortality rates.
• 64 yo female presents with 3 months of cough and dyspnea
• Notices some increase in cough with dressing in changing room
• Exam notable for mid-late inspiratory crackles and squeaks.
Case 1
Pulmonary Fibrosis

Case 1
Pulmonary Fibrosis

Case 1
67 yo male presents with 2-3 yrs of cough and progressive dyspnea

Exam unremarkable except for mid-late inspiratory crackles and clubbing
Case 2
Pulmonary Fibrosis

Case 2
Pulmonary Fibrosis

Case 2
Distinct chronic fibrosing interstitial pneumonia of unknown cause, primarily occurring in older adults, is limited to the lungs, and has typical imaging and pathologic findings of UIP.
• 72 yo male presents with 2 months of cough and dyspnea
• Recovered from “pneumonia” and played tennis with his granddaughter 5 days ago
• Exam notable for mid-late inspiratory crackles.
Case 3
Pulmonary Fibrosis

Case 3
Pulmonary Fibrosis Foundation

Aren’t All ILDs the Same?

Bjoraker et al. Am J Respir Crit Care Med
1998; 157:199-203
What is the natural history of patients with IPF?


Interstitial Lung Disease

- Drug-Induced Diseases
- Genetic / Inherited
- Systemic Diseases
  - Collagen Vascular Diseases
  - Common Variable Immunodeficiency Disorder
- Idiopathic
  - Idiopathic Interstitial Pneumonias
  - Sarcoidosis
  - Primary Disorders
  - LAM

Exposures
- Avocational
- Environmental
- Occupational
Clinical Classification of ILD: Drug and Treatment Induced

<table>
<thead>
<tr>
<th>Antibiotics</th>
<th>Neurotropic and psychotropic</th>
<th>Chemotherapeutic agents</th>
<th>Nitrosoureas</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nitrofurantoin</td>
<td>Dilantin</td>
<td></td>
<td>Carmustine (BCNU)</td>
</tr>
<tr>
<td>Sulfasalazine</td>
<td>Fluoxetine</td>
<td></td>
<td>Lomustine (CCNU)</td>
</tr>
<tr>
<td>Cephalosporin</td>
<td>Carbamazepine</td>
<td></td>
<td>Others</td>
</tr>
<tr>
<td>Minocycline</td>
<td>Chemotherapeutic agents</td>
<td></td>
<td>Procarbazine</td>
</tr>
<tr>
<td>Ethambutol</td>
<td>Antibiotics</td>
<td></td>
<td>Nilutemide</td>
</tr>
</tbody>
</table>

| Antiarrrhythmic             |                             |                         |                             |
| Amiodarone                  |                             |                         |                             |
| ACE-Inhibitors              |                             |                         |                             |
| Tocainide                   |                             |                         |                             |
| Beta-blocking agents        |                             |                         |                             |

| Anti-inflammatory           |                             |                         |                             |
| Gold                        |                             |                         |                             |
| Penicillamine               |                             |                         |                             |
| Nonsteroidal anti-         |                             |                         |                             |
| inflammatory agents        |                             |                         |                             |

| Antimetabolites             | Methotrexate                |                         |                             |
|                             | Azathioprine                |                         |                             |
|                             | Cytosine arabinoside        |                         |                             |

Schwarz MI, King TE. Interstitial lung disease. 3rd Ed. Malden, MA 1998
Scleroderma
Polymyositis - dermatomyositis
Systemic lupus erythematosus
Rheumatoid arthritis
Ankylosing spondylitis
Mixed connective tissue disease
Primary Sjögren’s syndrome
Behcet's syndrome
Clinical Classification of ILD: Primary Disorders

Sarcoidosis  
Eosinophilic granuloma  
Amyloidosis  
Lymphangioleiomyomatosis  
Tuberous sclerosis  
Neurofibromatosis  
Lymphangitic carcinomatosis  
Gaucher’s disease  
Niemann-Pick disease  
Hermansky-Pudlak syndrome  
Adult respiratory distress syndrome  
Bone marrow transplantation  
Acquired immune deficiency syndrome (AIDS)  
Post-infection  
Pulmonary vasculitis  

Respiratory bronchiolitis  
Interstitial cardiogenic pulmonary edema  
Familial hemophagocytic lymphohistocytosis  
Diabetes mellitus  
Lysinuric protein deficiency  
Alveolar filling diseases  
Alveolar proteinosis  
Diffuse alveolar hemorrhage syndromes  
Lipoid pneumonia  
Bronchioloalveolar carcinoma  
Pulmonary lymphoma  
Chronic aspiration  
Eosinophilic pneumonia  
Alveolar microlithiasis  
Alveolar sarcoidosis  
Cryptogenic organizing pneumonia

Schwarz MI, King TE. Interstitial lung disease. 3rd Ed. Malden, MA 1998
Inorganic

Silicosis
Asbestosis
Talc pneumoconiosis
Kaolin pneumoconiosis
Diatomaceous earth pneumoconiosis
Aluminum oxide fibrosis
Berylliosis
Hard metal fibrosis
Coal worker’s pneumoconiosis
Baritosis (barium)

Schwarz MI, King TE. Interstitial lung disease. 3rd Ed. Malden, MA 1998
Clinical Classification of ILD: Occupational and environmental exposure

Organic (hypersensitivity pneumonitis)
- Bagassosis (sugar cane)
- Bird breeder’s lung (pigeons, parakeets, etc.)
- Dove handler’s disease
- Farmer’s lung
- Coffee worker’s lung
- Tobacco grower’s lung
- Coptic disease (mummy wrappings)
- Cheese worker’s lung
- Fishmeal worker’s lung
- Furrier’s lung
- Meat worker’s lung
- Mushroom worker’s lung
- Paprika splitter’s lung
- Miller’s lung (wheat flour)
- Wood worker’s disease
- Sequoiosis

Malt worker’s lung
- Tea grower’s lung Suberosis (cork)
- Lycoperdonosis (Lycoperdon puffballs)
- Compost lung
- Humidifier lung
- Sauna taker’s lung
- Woodman’s disease (oak and maple)
- Pauli’s hypersensitivity pneumonitis (reagent)
- Pituitary snuff disease
- Detergent worker’s lung (isocyanates)
- Japanese summer-type hypersensitivity
- Thatched roof lung
- Familial hypersensitivity pneumonitis (wood dust)
- Vineyard sprayer’s lung
- Laboratory worker’s lung (rat urine)

Schwarz MI, King TE. Interstitial lung disease. 3rd Ed. Malden, MA 1998
Pulmonary Fibrosis

Hitchcock’s “The Birds”

Clinical Classification of ILD:
Idiopathic and Autoimmune disorders

Acute interstitial pneumonia (Hamman-Rich syndrome)
Idiopathic pulmonary fibrosis
Familial idiopathic pulmonary fibrosis
Lymphocytic interstitial pneumonitis
Bronchiolitis obliterans organizing pneumonia
Nonspecific interstitial pneumonitis
Desquamative interstitial pneumonitis

Autoimmune hemolytic anemia
Idiopathic thrombocytopenic purpura
Cryoglobulinemia
Inflammatory bowel disease
Celiac disease
Whipple’s disease
Primary biliary cirrhosis
Chronic active hepatitis
Cryptogenic cirrhosis

Schwarz MI, King TE. Interstitial lung disease. 3rd Ed. Malden, MA 1998
Interstitial Lung Disease

Drug-Induced Diseases

Exposures
- Avocational
- Environmental
- Occupational

Genetic / Inherited

Systemic Diseases
- Collagen Vascular Diseases
- Common Variable Immunodeficiency Disorder

Idiopathic
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