Progressive Fibrotic Interstitial Lung Disease (PF-ILD)

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ILD Clinical Presentation Topics

1. ILD Definition and Patient Population
2. Clinical Information on the PF-ILD Concept
3. Rationale for New Code
ILD Definition and Patient Population

• **Encompasses a large group of pulmonary disorders**
  – Affects the interstitium (tissue and space around the alveoli)

• **General consensus is that some form of injury of the alveolar epithelial cells initiates an inflammatory response coupled with repair mechanisms**\(^1\)
  – Reflected pathologically as inflammation, fibrosis, or a combination of both
  – Alteration of the interstitial space leads to clinical symptoms consistent with restrictive ventilatory deficit and poor gas exchange

• **Some patients with different types of ILD can develop a distinct, progressive fibrosing phenotype**
  – Similar to Idiopathic Pulmonary Fibrosis (IPF) with worsening of respiratory symptoms, lung function, quality of life, functional status, and early mortality\(^2, 3, 4\)

• **High unmet need for further characterization and treatment options**\(^5\)
  – Two interventional studies are evaluating treatment benefits of two anti-fibrotic therapies in non-IPF progressing fibrosing ILD\(^6, 7\)

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PF-ILD Concept

• **Historically:** Likely grouped many of the ILDs into a general category of pulmonary fibrosis
  – Lots of heterogeneity
  – Difficult to study
Improved survival

- Women, younger age, good steroid response

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M Turner-Warwick et al., Cryptogenic fibrosing avleolitis: response to corticosteroid treatment and its effect on survival, 35 THORAX 593-9 (1980).
Response to Steroid Therapy in UIP and NSIP

P<0.05 for difference in distribution of responses between patients with UIP and NSIP.

K Flaherty et al., Clinical significance of histological classification of idiopathic interstitial pneumonia, 42 EUR. RESPIR. J. 275-83 (2002).
PF-ILD Concept

• **Historically:** Likely grouped many of the ILDs into a general category of pulmonary fibrosis
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• **Currently:** Emphasis on diagnosis
  – Helps determine therapy and prognosis
  – Likely helped success in IPF clinical trials
  – Fails to account for subsequent disease behavior
Baseline DLco and change in FVC are associated with poor prognosis in patients with fibrotic interstitial pneumonia

<table>
<thead>
<tr>
<th>Factors</th>
<th>Hazard Ratio</th>
<th>95% CI</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male</td>
<td>2.724</td>
<td>1.277–5.813</td>
<td>0.010</td>
</tr>
<tr>
<td>Initial DLco % predicted</td>
<td>0.972</td>
<td>0.949–0.996</td>
<td>0.022</td>
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<tr>
<td>6-month change in FVC</td>
<td>0.925</td>
<td>0.893–0.958</td>
<td>&lt;0.001</td>
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<tr>
<td>Age</td>
<td>1.027</td>
<td>0.992–1.064</td>
<td>0.134</td>
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<tr>
<td>NSIP</td>
<td>0.854</td>
<td>0.349–2.093</td>
<td>0.730</td>
</tr>
<tr>
<td>Resting PaO₂</td>
<td>0.995</td>
<td>0.961–1.031</td>
<td>0.798</td>
</tr>
<tr>
<td>Initial FVC % predicted</td>
<td>0.987</td>
<td>0.964–1.010</td>
<td>0.262</td>
</tr>
</tbody>
</table>

Initial Diagnosis No Longer Prognostic after Accounting for Change in Lung Function
PF-ILD Concept

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- **Future**: Combine diagnosis and behavior (phenotype)
  - Although no FDA-approved treatments exist for patients with fibrotic ILD other than IPF, several clinical trials are in progress

PF-ILD Concept

UC – unclassifiable
HP – hypersensitivity pneumonitis
Rationale for New Code

• Enable more specific identification of patients with ILD with a progressive fibrotic phenotype

• Facilitate research and characterization of this patient population

• Enhance understanding of the diseases to aid in diagnosis, disease management, and treatment

• Facilitate more responsive monitoring and tracking of ILDs, while enhancing providers’ ability to accurately respond to the type of ILD diagnosed

• Streamline claims processing and enable clearer diagnoses in patient records, particularly as new treatments become available