Idiopathic Interstitial Pneumonias
David A. Lynch, MB

Idiopathic interstitial pneumonias: 2012 update
David A Lynch, MB

Learning objectives
• Recognize CT features of idiopathic interstitial pneumonias, primarily fibrosing interstitial pneumonias
• Recognize importance of multidisciplinary diagnosis

2002 ATS-ERS Classification of idiopathic interstitial pneumonia

<table>
<thead>
<tr>
<th>MORPHOLOGIC PATTERN</th>
<th>IDIOPATHIC CLINICAL SYNDROME</th>
</tr>
</thead>
<tbody>
<tr>
<td>Usual IP</td>
<td>Idiopathic pulmonary fibrosis</td>
</tr>
<tr>
<td>Nonspecific IP pattern</td>
<td>Idiopathic NSIP</td>
</tr>
<tr>
<td>Desquamative IP pattern</td>
<td>Desquamative IP</td>
</tr>
<tr>
<td>Respiratory bronchiolitis</td>
<td>Respiratory bronchiolitis ILD</td>
</tr>
<tr>
<td>Organizing pneumonia</td>
<td>Cryptogenic organizing pneumonia</td>
</tr>
<tr>
<td>Diffuse alveolar damage</td>
<td>Acute IP</td>
</tr>
<tr>
<td>Lymphoid IP pattern</td>
<td>Idiopathic LIP</td>
</tr>
</tbody>
</table>

IIP: relative frequency

<table>
<thead>
<tr>
<th>PATTERN</th>
<th>FREQUENCY</th>
</tr>
</thead>
<tbody>
<tr>
<td>Usual IP</td>
<td>40%</td>
</tr>
<tr>
<td>Nonspecific IP pattern</td>
<td>20%</td>
</tr>
<tr>
<td>Desquamative IP pattern</td>
<td>20%</td>
</tr>
<tr>
<td>Respiratory bronchiolitis</td>
<td>20%</td>
</tr>
<tr>
<td>Organizing pneumonia</td>
<td>10%</td>
</tr>
<tr>
<td>Diffuse alveolar damage</td>
<td>10%</td>
</tr>
<tr>
<td>Lymphoid IP pattern</td>
<td>Rare</td>
</tr>
</tbody>
</table>

Prognosis

Mortality from pulmonary fibrosis increased in the United States from 1992 to 2003.

**Diagnostic criteria for UIP**

**UIP pattern**
- Subpleural, basal predominance
- Reticular abnormality
- Honeycombing with or without traction bronchiectasis
- Absence of features inconsistent with UIP pattern

**Possible UIP pattern**
- Subpleural, basal predominance
- Reticular abnormality
- Absence of features inconsistent with UIP pattern

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**Honeycombing in UIP**

- Present in 70-80% of cases of UIP
- Strongest indicator of UIP on CT
- Median survival
  - UIP with honeycombing: 2.1 years
  - UIP without honeycombing: 5.8 years

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**Subpleural, Reticular Absence**

ATS/ERS/JRS/ALAT Guidelines for Diagnosis and Management.

**UIP**

ATS/ERS/JRS/ALAT Guidelines for Diagnosis and Management.

**Idiopathic Pulmonary Fibrosis:**
- honeycombing
- bronchiectasis

**Statement:**
- Basal features with or without traction bronchiectasis

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**Diagnostic criteria for UIP**


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**Diagnostic criteria for UIP**

Diagnostic criteria for UIP

Inconsistent with UIP pattern
- Upper or midlung predominance
- Peribronchovascular predominance
- Extensive ground glass abnormality (reticular)
- Profuse micronodules (bilateral, predominantly upper lobes)
- Discrete cysts (multiple, bilateral, away from areas of honeycombing)
- Diffuse mottled attenuation (bilateral, in three or more lobes)
- Consolidation in segments or lobes


Predictive value of CT diagnosis of UIP

<table>
<thead>
<tr>
<th>Study</th>
<th>Correctness of first choice diagnosis of UIP</th>
<th>Correctness of confident first choice diagnosis</th>
<th>% cases of UIP without confident CT diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mathieson</td>
<td>88%</td>
<td>95%</td>
<td>72%</td>
</tr>
<tr>
<td>Hunninghake</td>
<td>85%</td>
<td>96%</td>
<td>52%</td>
</tr>
<tr>
<td>Flaherty</td>
<td>100%</td>
<td>100%</td>
<td>63%</td>
</tr>
<tr>
<td>Tsubamoto</td>
<td>100%</td>
<td>91%</td>
<td>9%</td>
</tr>
<tr>
<td>Elliot</td>
<td>88%</td>
<td>88%</td>
<td>50%</td>
</tr>
<tr>
<td>Silva</td>
<td>84%</td>
<td>100%</td>
<td>67%</td>
</tr>
</tbody>
</table>

Sverzellati et al. Radiology 2010: 254;957-964

UIP: atypical features

- 55 subjects with biopsy-proven UIP
- 20 had high prob CT diagnosis of UIP
- Differential Dx in 34 low-prob CT cases:
  - NSIP (53%)
  - Chronic HP (12%)
  - Sarcoid (9%)

Accelerated deterioration in IPF

Baseline

5 months later

Causes of UIP pattern

- Collagen vascular disease
- Asbestosis
- Hypersensitivity pneumonitis

UIP: lung cancer

- Prevalence ranges from 5-15%
- Often peripheral, lower lobe
- May be multifocal
- Nodules, lobulation, air bronchograms

Kishi et al. JCAT, 2006;30:95-9
NSIP

- Disease that does not correspond to one of the existing categories of IIP
- Good prognosis
- Often an underlying cause


NSIP

- Usually presents with fibrosing interstitial pneumonia
- Substantially better survival than UIP
- ± Associated collagen vascular disease or HP

NSIP: HRCT

- Basal predominance
- Peribronchovascular/subpleural sparing
- Confluent pattern
- Volume loss
- Ground glass
- Reticular
- Traction bronchiectasis
- Consolidation +/-
- Honeycombing rare

Other entities mimicking radiologic pattern of NSIP

- UIP
- NSIP
- HP
- LIP
- DIP/RA-ILD

Underlying diseases in NSIP
- Collagen vascular disease
- Environmental exposures
- Drug toxicity
- Cigarette smoking?

NSIP: natural history

- 2000
- 2002
- 2004
- 2010

Serial evaluation of NSIP
- 23 subjects followed for median 61 months
- Decrease in extent of ground glass
- Increase in reticular pattern
- Increase in extent of honeycombing
- Increase in subpleural distribution
- Increase in number of subjects with CT suggestive of IPF (from 3 to 6)

<table>
<thead>
<tr>
<th>Imaging features of CHP, UIP, NSIP</th>
<th>CHP</th>
<th>UIP</th>
<th>NSIP</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ground-glass opacities</td>
<td>36</td>
<td>46</td>
<td>50</td>
</tr>
<tr>
<td>Reticulation</td>
<td>36</td>
<td>46</td>
<td>50</td>
</tr>
<tr>
<td>Honeycombing</td>
<td>23</td>
<td>31</td>
<td>4</td>
</tr>
<tr>
<td>Lobular decreased attenuation</td>
<td>29</td>
<td>20</td>
<td>17</td>
</tr>
<tr>
<td>Centrilobular nodules</td>
<td>20</td>
<td>7</td>
<td>7</td>
</tr>
<tr>
<td>Subpleural sparing</td>
<td>4</td>
<td>2</td>
<td>32</td>
</tr>
<tr>
<td>Lower lung predominance</td>
<td>11</td>
<td>38</td>
<td>47</td>
</tr>
</tbody>
</table>

Silva et al. Radiology 2008;246:288-97

<table>
<thead>
<tr>
<th>Smoking-related lung diseases</th>
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<tbody>
<tr>
<td>RB</td>
</tr>
<tr>
<td>RB-ILD</td>
</tr>
<tr>
<td>DIP (IPF)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Respiratory bronchiolitis: CT</th>
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</thead>
<tbody>
<tr>
<td>• Centrilobular nodules</td>
</tr>
<tr>
<td>• Ground glass</td>
</tr>
<tr>
<td>• Emphysema ±</td>
</tr>
</tbody>
</table>

Smoking-related lung diseases

Respiratory bronchiolitis: CT
**Respiratory bronchiolitis interstitial lung disease**

- Ground glass
- Centrilobular nodules
- Emphysema
- Air trapping

**Desquamative interstitial pneumonia: HRCT**

- Basal, peripheral prominence
- Ground glass
- ± Reticular
- ± Cysts

**Continuum of smoking-related lung disease**

<table>
<thead>
<tr>
<th>Path</th>
<th>CT</th>
</tr>
</thead>
<tbody>
<tr>
<td>RB</td>
<td>Centrilobular</td>
</tr>
<tr>
<td>RB-ILD</td>
<td>Centrilobular + ground glass</td>
</tr>
<tr>
<td>DIP</td>
<td>Ground glass +/- cysts, centrilobular</td>
</tr>
</tbody>
</table>

**Continuum of smoking-related lung disease**

<table>
<thead>
<tr>
<th>Symptoms/physiologic impairment</th>
</tr>
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<tbody>
<tr>
<td>RB</td>
</tr>
<tr>
<td>RB-ILD</td>
</tr>
<tr>
<td>DIP</td>
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Heyneman AUR1999;173: 1617-22

**Interstitial lung abnormalities in cigarette smokers**

- ILA found in 194/2416 subjects (8%)
  - Predominantly ground glass abnormality
- Associated with:
  - current smoking
  - Pack years of smoking
- Decreased TLC
- Lower likelihood of meeting COPD criteria
- Decreased quantitative % emphysema


**Organizing pneumonia: HRCT**

- Subpleural/ peribronchial
- Consolidation
- Ground glass
Organizing pneumonia: perilobular thickening

Organizing pneumonia: reverse halo sign

Causes of OP pattern

- Cryptogenic
- Collagen vascular disease (polymyositis/dermatomyositis)
- Drug toxicity
- Aspiration

LIP: HRCT

- Ground glass attenuation
- Cysts
- Reticular abnormality
- Usually underlying disease (esp. Sjogren syndrome)

Multi-disciplinary approach to IIP diagnosis

- 58 cases with suspected IIP
- Independent review followed by clinical-radiology-pathology consensus
- Consensus review caused alteration of:
  - 53% of radiologist diagnoses
  - 34% of clinician diagnoses
  - 19% of pathologist diagnoses

Summary

- Idiopathic interstitial pneumonias comprise fibrosing and non-fibrosing entities
- Honeycombing is the most important discriminator between UIP and non-UIP
- Multidisciplinary diagnosis makes a difference:
  - Sometimes the radiologist is right!

Flaherty et al. Am J Respir Crit Care Med. 2004; 170:904-10