Interstitial Lung Disease

Known causes
- Hypersensitivity pneumonitis
- Asbestosis
- Drug reactions
- Connective Tissue Disease

Idiopathic
- UIP  Usual interstitial pneumonia
- NSIP  Nonspecific interstitial pneumonia
- COP  Cryptogenic organizing pneumonia
- AIP  Acute interstitial pneumonia
- RBILD  Respiratory bronchiolitis–associated interstitial lung disease
- DIP  Desquamative interstitial pneumonia
- LIP  Lymphocytic interstitial pneumonia
- PPFE  Pleuroparenchymal fibroelastosis
Factors which increase a person's risk for pulmonary fibrosis

Idiopathic pulmonary fibrosis (IPF) 101

- Idiopathic pulmonary fibrosis (IPF) most common and deadly type of pulmonary fibrosis
- Similar survival as compared to Non-Small Cell Lung Cancer
- Usual interstitial pneumonitis (UIP) is the imaging pattern and histology in IPF
- Conversely, UIP almost always IPF (>95%)
- In 2014, FDA approved new therapies for the treatment of patients with IPF

Secondary Pulmonary Lobule

- Interlobular septa
- Bronchus
- Artery
- Alveoli

ATS Guidelines for UIP

- Subpleural basilar predominant fibrosis
- Reticulations
- Honeycombing
- Absence of features that would suggest and alternative diagnosis
CANNOT B UIP

- Consolidation
- Air trapping
- Nodules
- Non-solid or ground glass
- O is for holes or cysts
- T is for top
- B is for bronchovascular

Honeycombing vs bronchiectasis
ATS Guidelines for Possible UIP

- Subpleural basilar predominant fibrosis
- Reticulations
- Honeycombing
- Absence of features that would suggest and alternative diagnosis

Possible UIP

Subpleural and basilar predominant
Possible UIP

Reticulations

Pre honeycombing to honeycombing

Reticulations

Honeycombing
ATS guidelines for inconsistent with UIP

- Inconsistent with UIP Pattern (Any of the Seven Features)
  - Upper or mid-lung predominance
  - Peribronchovascular predominance
  - Extensive ground glass abnormality (extent > reticular abnormality)
  - Consolidation in bronchopulmonary segment(s)/lobe(s)
  - Profuse micronodules (bilateral, predominantly upper lobes)
  - Discrete cysts (multiple, bilateral, away from areas of honeycombing)
  - Diffuse mosaic attenuation/air-trapping (bilateral, in three or more lobes)


Upper lobe fibrosis

- Upper lobe fibrosis with volume loss
- Relative sparing of the lower lobes
- Centrilobular nodules
**Ground-glass opacity**

- Ground glass opacity and traction bronchiectasis in NSIP

**Mimickers of Fibrosis**

- Fibrosis caused by osteophyte
- Reversible atelectasis
**NSIP**

- Lower lobe fibrosis
- Follows bronchovascular distribution
- Homogeneous
- Ground glass opacity
- Dilated esophagus

### NSIP
- Lower lobe fibrosis
- Follows bronchovascular distribution
- Homogeneous
- Ground glass opacity
- Dilated esophagus

---

### Chronic Hypersensitivity Pneumonitis
- Upper lobe predominant
- Bronchovascular distribution
- Air trapping

Chronic Hypersensitivity Pneumonitis

Centrilobular nodules and early fibrosis

Stage 4 Sarcoidosis

- Upper lobe predominant fibrosis
- Bronchovascular distribution

**Stage 4 Sarcoidosis**

Upper lobe predominant fibrosis that follows bronchovascular distribution

**Hiatal Hernia**

Hiatal hernia and gastro-esophageal reflux is associated with fibrosis of the lung
**Pulmonary Artery**

- Pulmonary artery enlargement is associated with pulmonary hypertension and can be seen in patients with fibrosis.

**Enlarged Lymph Nodes**

- Mediastinal lymphadenopathy can occur in patients with pulmonary fibrosis.

Combined pulmonary fibrosis and emphysema

Honeycombing

Emphysema

Non-radiological features that point toward IPF diagnosis

- Older age
- Male sex
- History of smoking
- Basilar crackles
- Decreased DLCO, FVC
- Decreased performance on 6 minute walk test


Radiological features that point toward IPF diagnosis

- Lower lobe predominant fibrosis
- Sub-pleural fibrosis
- Reticulations and traction bronchiectasis
- Honeycombing
- Volume loss
- Absence of consolidation, air trapping, nodules


Case 1: 55 year old female with dyspnea.

What is the most likely diagnosis?
Case 1: 55 year old female with dyspnea.

- NSIP
  - Lower lobe predominant fibrosis
  - Bronchovascular distribution
  - Absence of honeycombing
  - Ground glass opacity

Case 2: 73 year old female with long standing dyspnea

What is the most likely diagnosis?
Case 2: 73 year old female with long standing dyspnea

• CHP
  – Upper lobe fibrosis
  – Bronchovascular
  – Air trapping

Case 3: 37 year old with chronic cough

What is the most likely diagnosis?
Case 3: 37 year old with chronic cough

- Stage 4 Sarcoid
  - Upper lobe fibrosis
  - Bronchovascular distribution
  - Cystic changes

Case 4: 45 year old with chronic cough

What is the most likely diagnosis?
Case 4: 45 year old with chronic cough

- Organizing pneumonia
  - Consolidation
  - Bronchovascular distribution

Case 5: 68 year old female with increasing shortness of breath and cough

What is the most likely diagnosis?
Case 5: 68 year old female with increasing shortness of breath and cough

• Possible UIP
  – Subpleural basilar predominant fibrosis
  – Absence of Honeycombing
  – Reticulations and traction bronchiectasis

Case 6: 78 year old man with shortness of breath

What is the most likely diagnosis?
Case 6: 78 year old man with shortness of breath

- UIP
  - Subpleural basilar predominant fibrosis
  - Honeycombing
  - Reticulations and traction bronchiectasis

Radiology and Pulmonary communication

- Radiology report should provide supporting words for correct diagnosis
- Ancillary findings in report include pulmonary artery size, hiatal hernia and liver cirrhosis
- Discussion with pulmonologist for first CT to review clinical information
- Multidisciplinary conference is gold standard