

Idiopathic Pulmonary Fibrosis in Images

Interpreting HRCT and
Identifying the UIP Pattern

A CME-certified Program

Jointly provided by The Potomac Center for Medical Education and Rockpointe



This activity is supported by an independent educational grant from
Boehringer Ingelheim Pharmaceuticals, Inc.

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Interstitial Lung Disease

Known causes

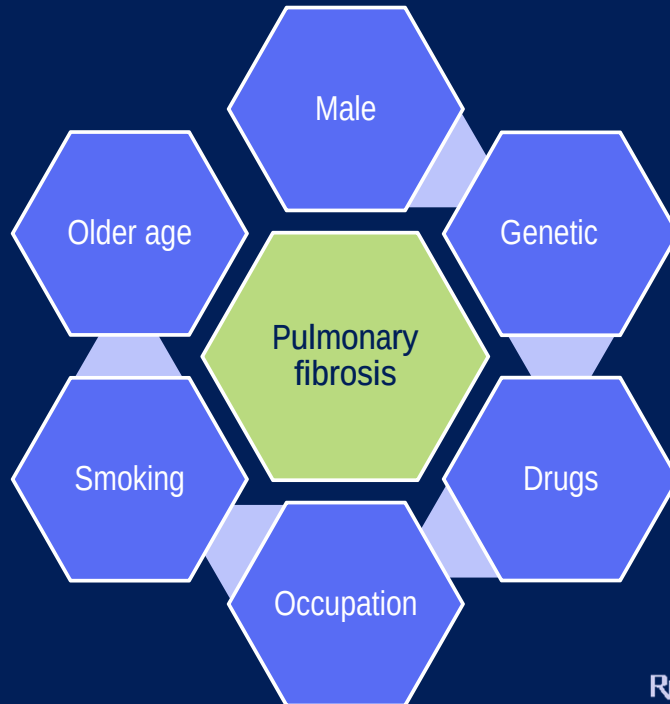
- Hypersensitivity pneumonitis
- Asbestosis
- Drug reactions
- Connective Tissue Disease

Idiopathic

UIP Usual interstitial pneumonia	NSIP Nonspecific interstitial pneumonia
COP Cryptogenic Organizing Pneumonia	AIP Acute interstitial pneumonitis
RBILD Respiratory bronchiolitis-associated interstitial lung disease	DIP Desquamative interstitial pneumonia
LIP Lymphocytic Interstitial Pneumonia	PPFE Pleuroparenchymal fibroelastosis

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Factors which increase a persons risk for pulmonary fibrosis



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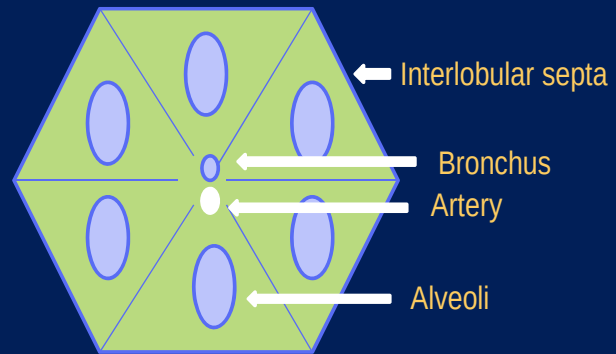
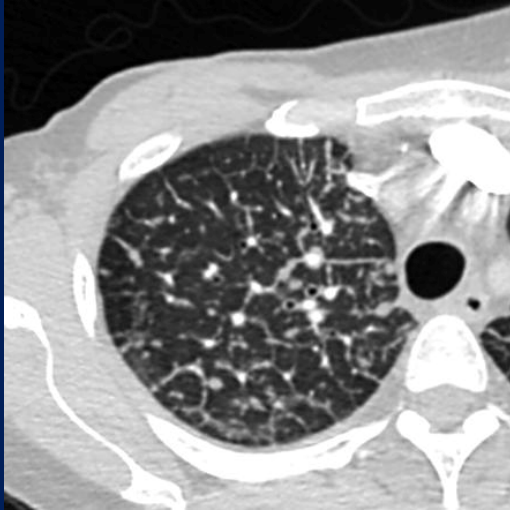
Idiopathic pulmonary fibrosis (IPF) 101

- 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

Xia, Meng, et al. "Treatment Destiny In Idiopathic Pulmonary Fibrosis Patients Following Approval Of Two Drug Therapies." A103. IPF: MORE ABOUT THERAPY AND OUTCOMES. American Thoracic Society, 2016. A2697-A2697.

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Secondary Pulmonary Lobule



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ATS Guidelines for UIP



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

Reghu G, et al. ATS/ERS/JRS/ALAT Committee on Idiopathic Pulmonary Fibrosis. An official ATS/ERS/JRS/ALAT statement: Idiopathic pulmonary fibrosis: evidence-based guidelines for diagnosis and management. Am J Respir Crit Care Med. 2011 Mar 15;183(6):788-824.

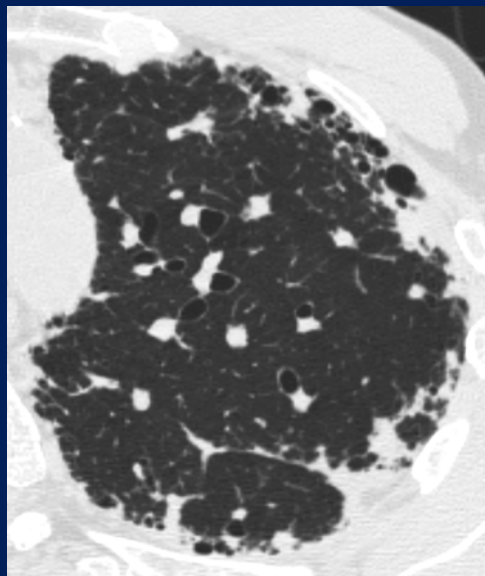
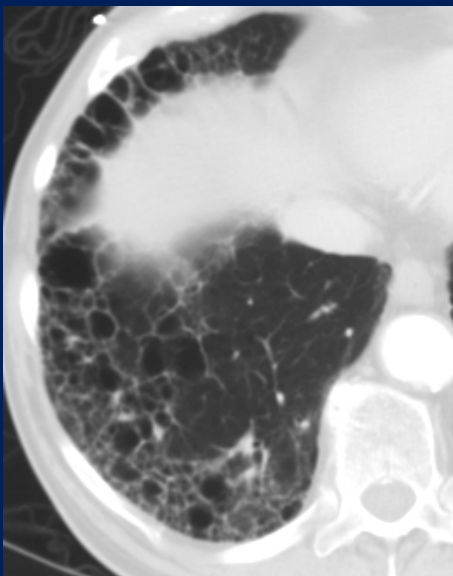
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CANNOT B UIP

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

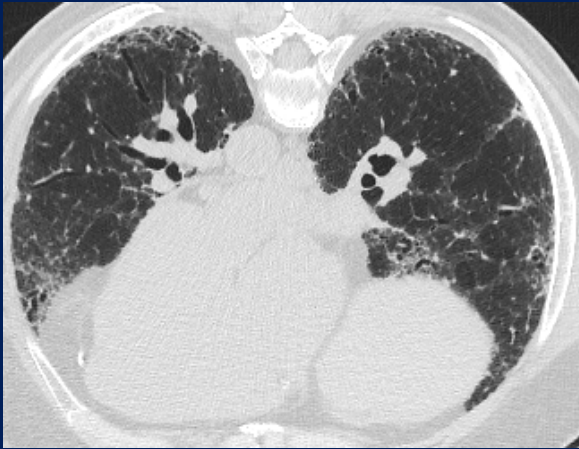
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Honeycombing vs bronchiectasis



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ATS Guidelines for Possible UIP



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

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Possible UIP

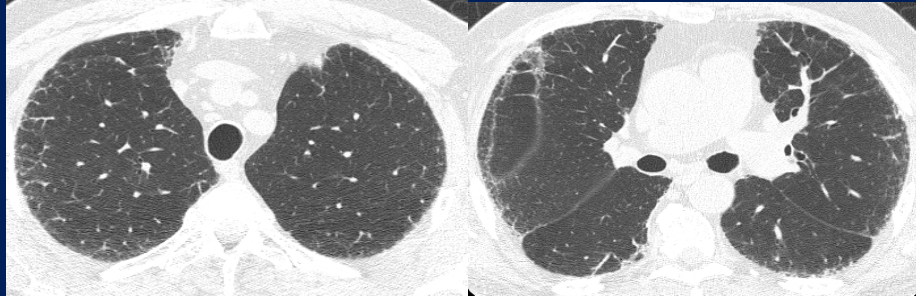


Subpleural and basilar
predominant

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Possible UIP

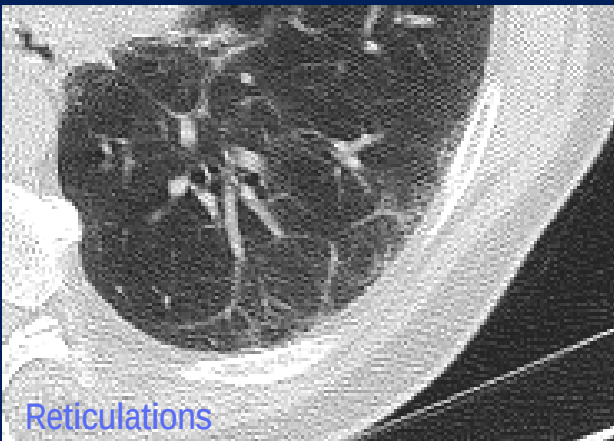
Reticulations



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Pre honeycombing to honeycombing

Reticulations



Honeycombing



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ATS guidelines for inconsistent with UIP

- Inconsistent with UIP Pattern (Any of the Seven Features)

- Upper or mid-lung predominance

- Peribronchovascular predominance

Distribution

- Extensive ground glass abnormality (extent > reticular abnormality)

- Consolidation in bronchopulmonary segment(s)/lobe(s)

- Profuse micronodules (bilateral, predominantly upper lobes)

Increased density

- Discrete cysts (multiple, bilateral, away from areas of honeycombing)

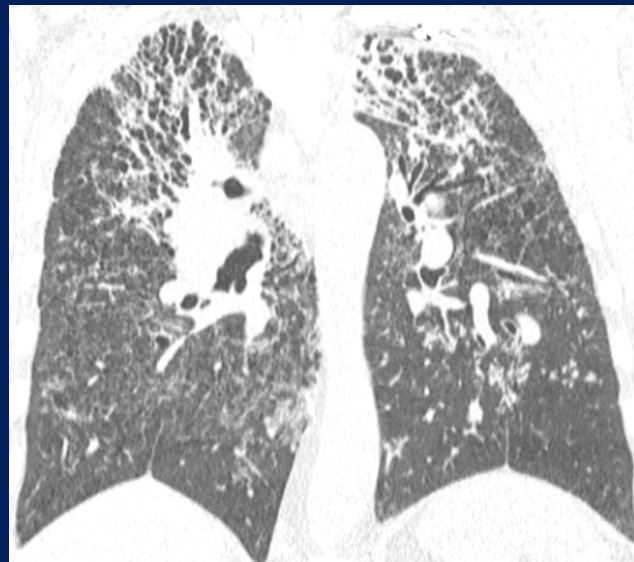
- Diffuse mosaic attenuation/air-trapping (bilateral, in three or more lobes)

Decreased density

Raghu G, et al. ATS/ERS/JRS/ALAT Committee on Idiopathic Pulmonary Fibrosis. An official ATS/ERS/JRS/ALAT statement: idiopathic pulmonary fibrosis: evidence-based guidelines for diagnosis and management. Am J Respir Crit Care Med. 2011 Mar 15;183(6):788-824.

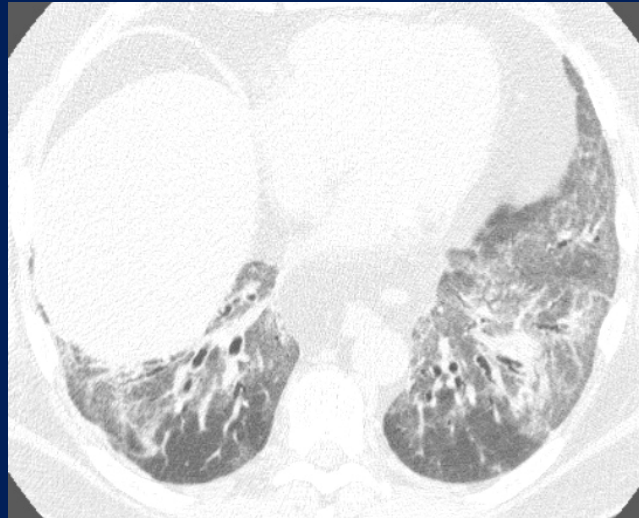
Upper lobe fibrosis

- Upper lobe fibrosis with volume loss
- Relative sparing of the lower lobes
- Centrilobular nodules



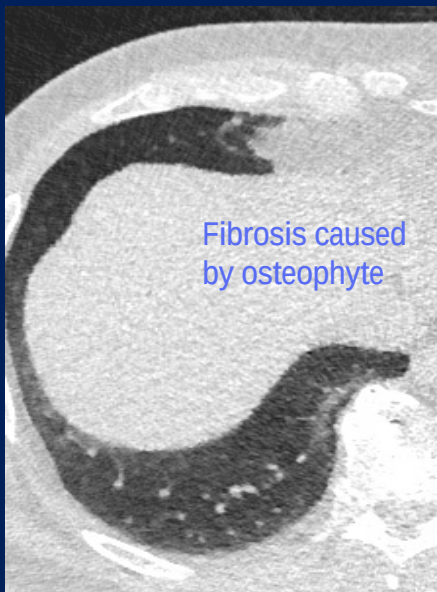
Ground-glass opacity

- Ground glass opacity and traction bronchiectasis in NSIP



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Mimickers of Fibrosis



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CHP

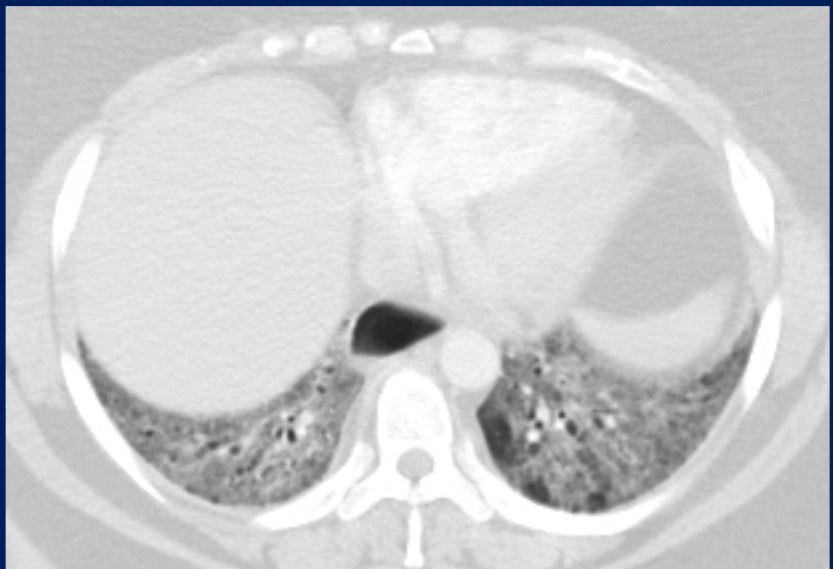
Sarcoid

UIP

NSIP

NSIP

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

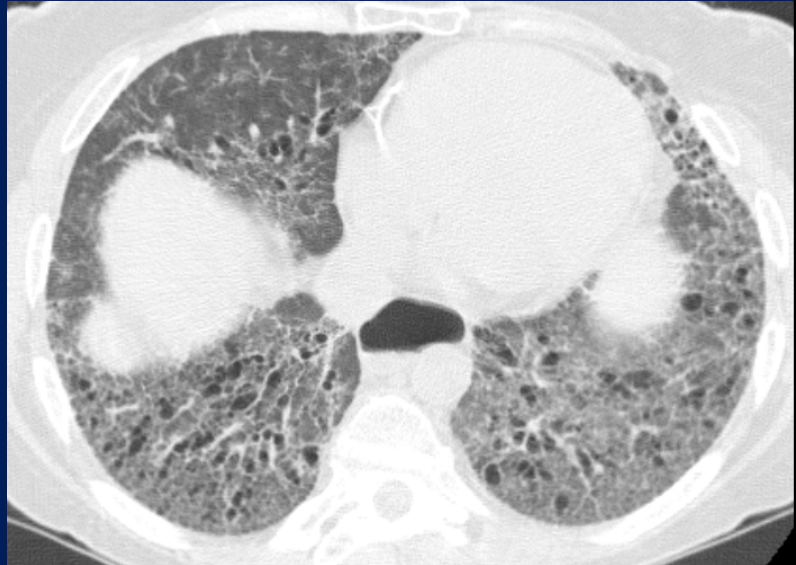


Sasaki, Shinichi, et al. "Diagnostic Significance Of Biomarkers For Chronic Fibrosing Idiopathic Interstitial Pneumonias (Usual Interstitial Pneumonia Vs Non-Specific Interstitial Pneumonia)." C39. IPF: MORE ON DIAGNOSIS AND THERAPY. American Thoracic Society, 2016. A4999-A4999.

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NSIP

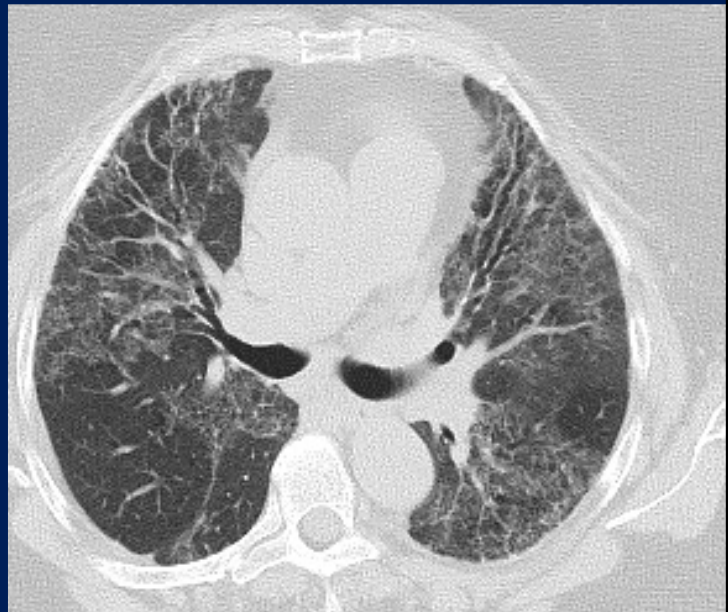
- Lower lobe fibrosis
- Follows bronchovascular distribution
- Homogeneous
- Ground glass opacity
- Dilated esophagus



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Chronic Hypersensitivity Pneumonitis

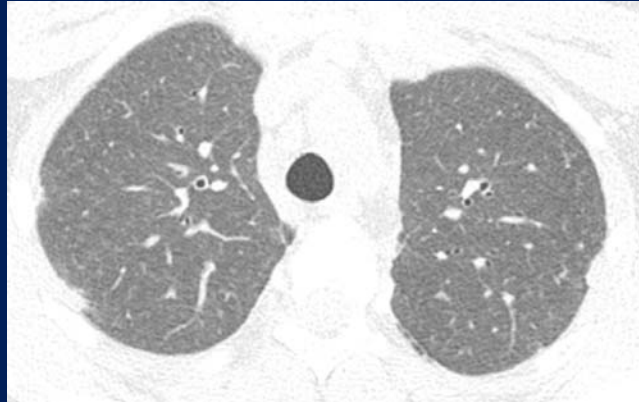
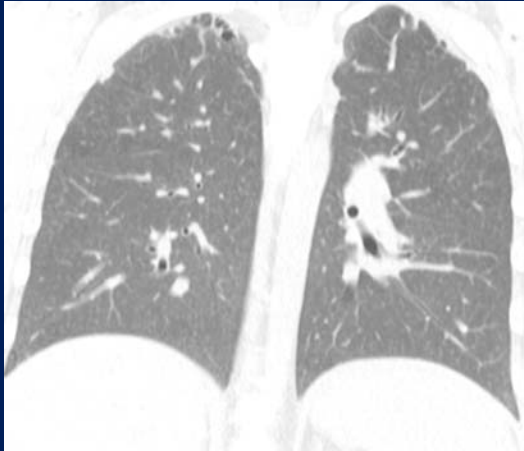
- Upper lobe predominant
- Bronchovascular distribution
- Air trapping



Silva, C. Isabela S., et al. "Chronic Hypersensitivity Pneumonitis: Differentiation from Idiopathic Pulmonary Fibrosis and Nonspecific Interstitial Pneumonia by Using Thin-Section CT 1." *Radiology* 246.1 (2008): 288-297.

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Chronic Hypersensitivity Pneumonitis

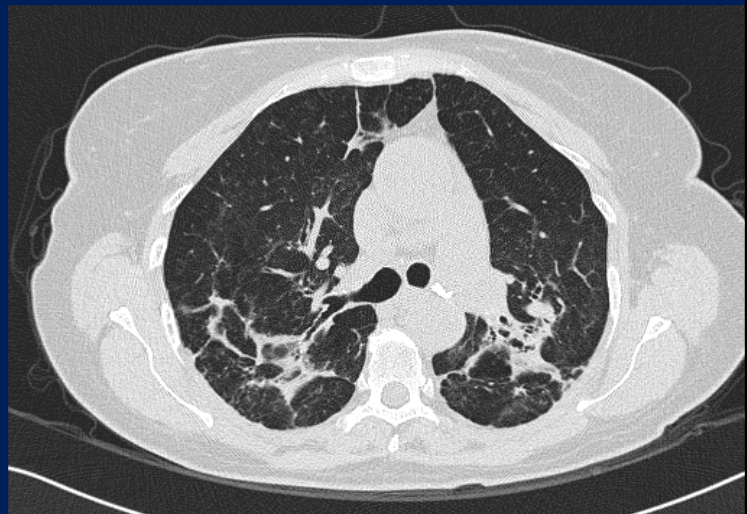


Centrilobular nodules and early fibrosis

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Stage 4 Sarcoidosis

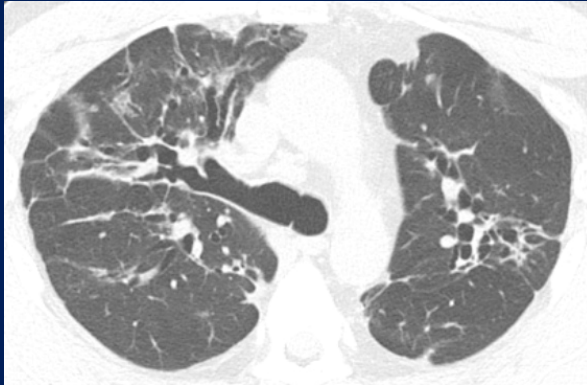
- Upper lobe predominant fibrosis
- Bronchovascular distribution



Brauner, M. W., et al. "Pulmonary sarcoidosis: evaluation with high-resolution CT." *Radiology* 172.2 (1989): 467-471.

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Stage 4 Sarcoidosis

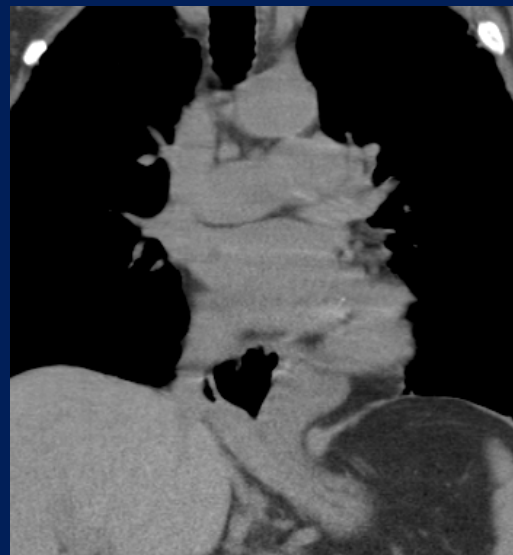


Upper lobe predominant fibrosis that follows bronchovascular distribution



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Hiatal Hernia

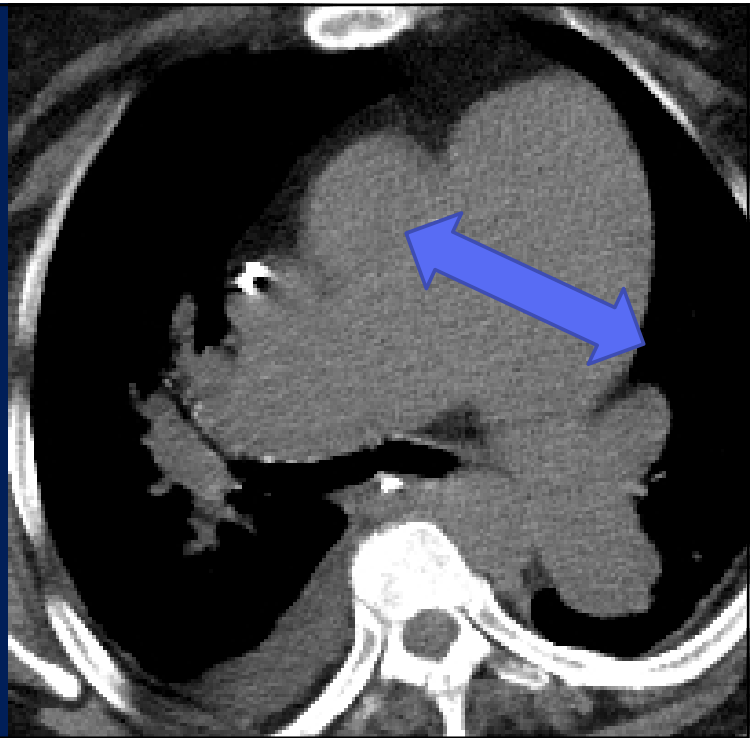


Hiatal hernia and gastro-esophageal reflux is associated with fibrosis of the lung

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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



Souza, Carolina Althoff, et al. "Idiopathic interstitial pneumonias: prevalence of mediastinal lymph node enlargement in 206 patients." *American Journal of Roentgenology* 186.4 (2006): 995-999.

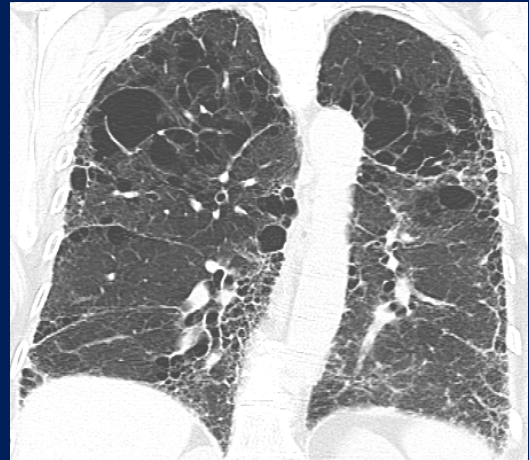
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Combined pulmonary fibrosis and emphysema



Honeycombing

Emphysema



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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

Raghu G, et al. ATS/ERS/JRS/ALAT Committee on Idiopathic Pulmonary Fibrosis. An official ATS/ERS/JRS/ALAT statement: idiopathic pulmonary fibrosis: evidence-based guidelines for diagnosis and management. Am J Respir Crit Care Med. 2011 Mar 15;183(6):788-824.
Rosewarne, D., et al. "The idiopathic interstitial pneumonias—a survival guide." *Imaging* (2014).

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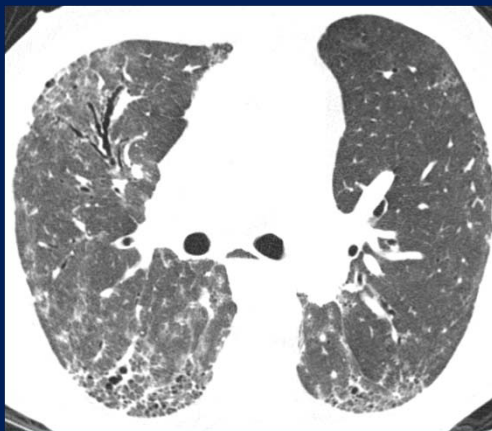
Radiological features that point toward IPF diagnosis

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

Raghu G, et al. ATS/ERS/JRS/ALAT Committee on Idiopathic Pulmonary Fibrosis. An official ATS/ERS/JRS/ALAT statement: idiopathic pulmonary fibrosis: evidence-based guidelines for diagnosis and management. *Am J Respir Crit Care Med*. 2011 Mar 15;183(6):788-824.
Rosewarne, D., et al. "The idiopathic interstitial pneumonias—a survival guide." *Imaging* (2014).

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Case 1: 55 year old female with dyspnea.



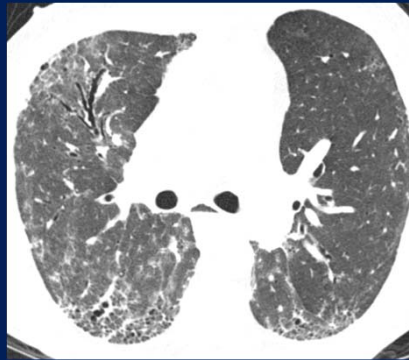
What is the most likely diagnosis?

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Case 1: 55 year old female with dyspnea.

- NSIP

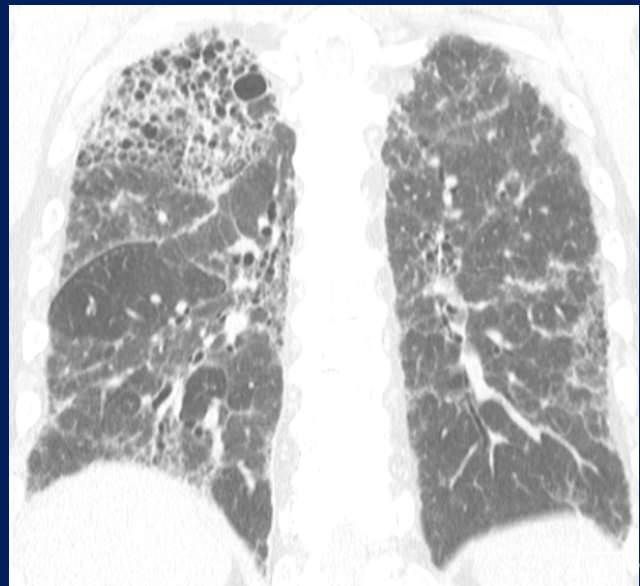
- Lower lobe predominant fibrosis
- Bronchovascular distribution
- Absence of honeycombing
- Ground glass opacity



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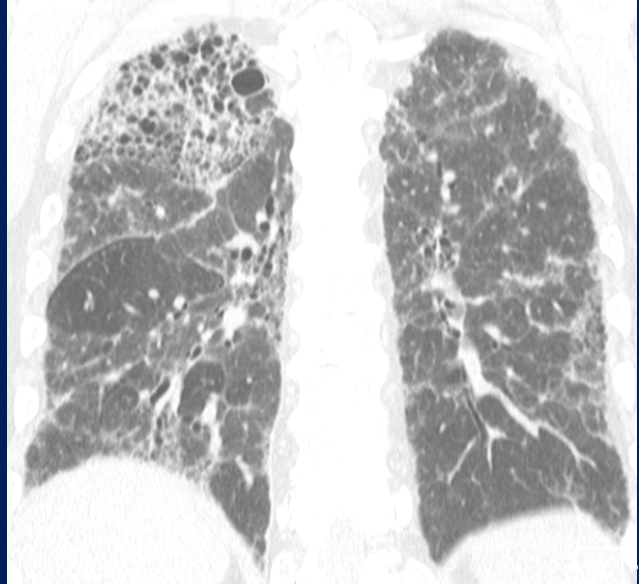
Case 2: 73 year old female with long standing dyspnea

What is the most likely diagnosis?



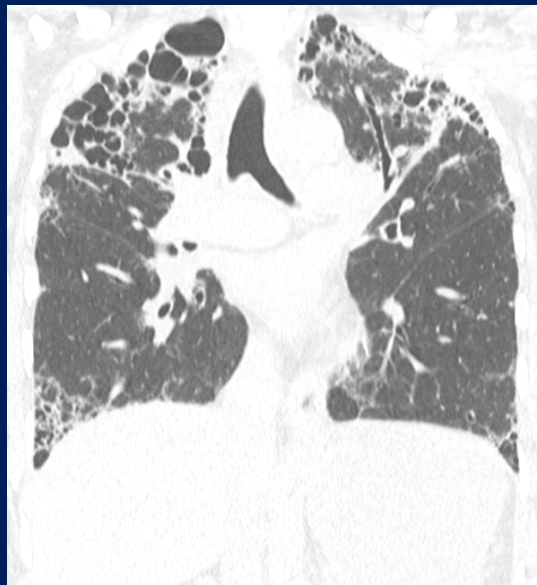
Case 2: 73 year old female with long standing dyspnea

- CHP
 - Upper lobe fibrosis
 - Bronchovascular
 - Air trapping



Case 3: 37 year old with chronic cough

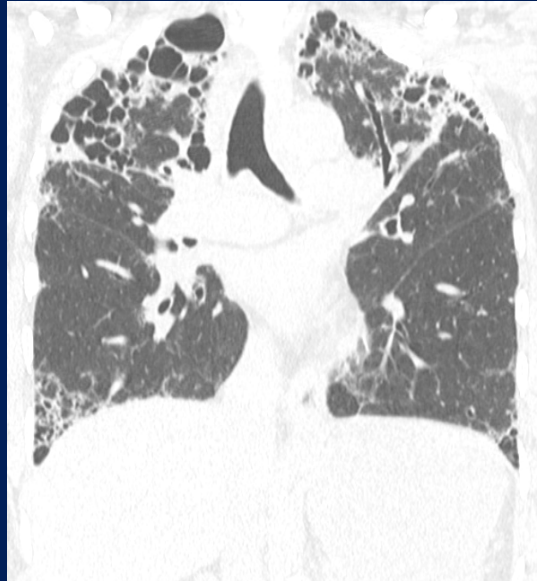
What is the most likely diagnosis?



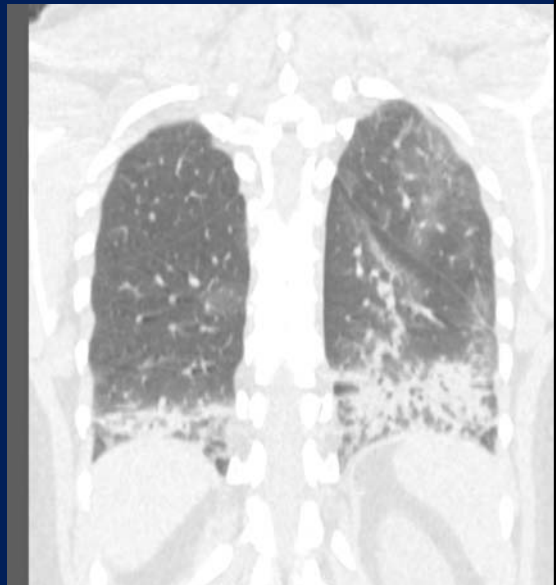
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Case 3: 37 year old with chronic cough

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



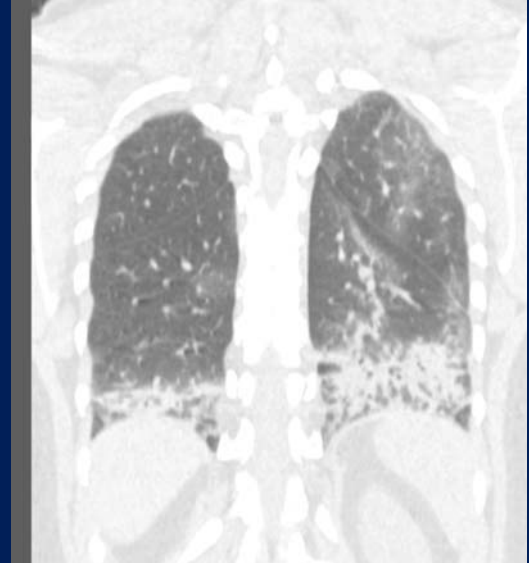
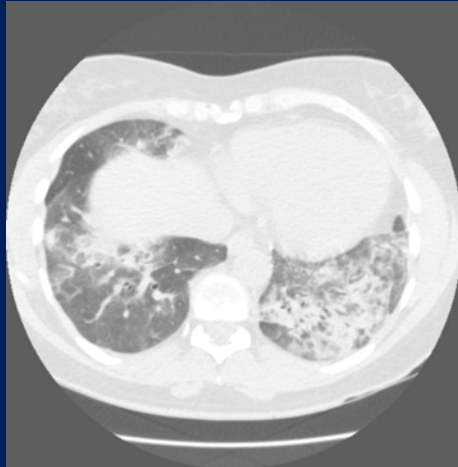
Case 4: 45 year old with chronic cough



What is the most likely diagnosis?

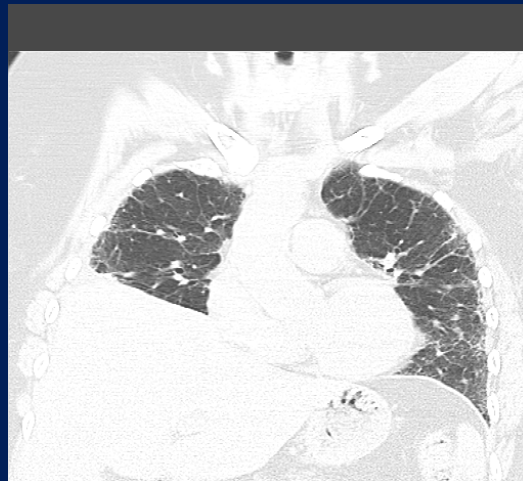
Case 4: 45 year old with chronic cough

- Organizing pneumonia
 - Consolidation
 - Bronchovascular distribution



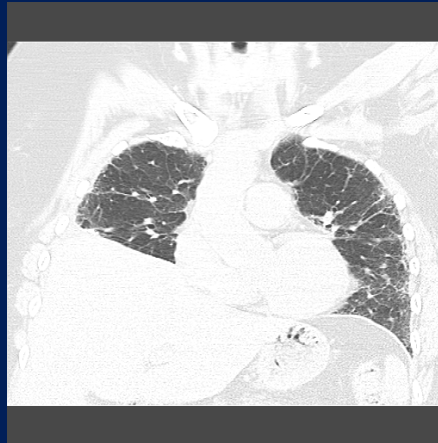
Case 5: 68 year old female with increasing shortness of breath and cough

What is the most likely diagnosis?



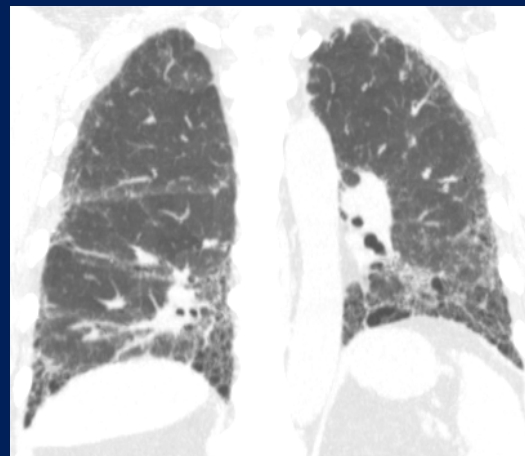
Case 5: 68 year old female with increasing shortness of breath and cough

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



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Case 6: 78 year old man with shortness of breath

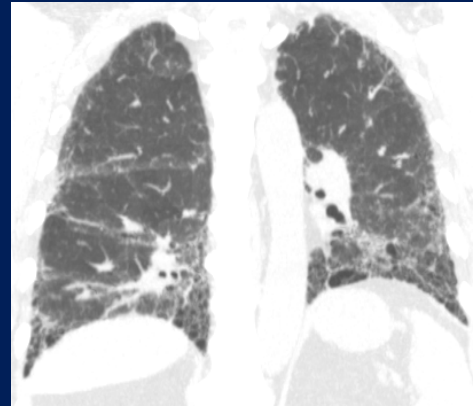


What is the most likely diagnosis?

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Case 6: 78 year old man with shortness of breath

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



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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

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