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# ILD의 HRCT 진단 – 영상의학적 접근

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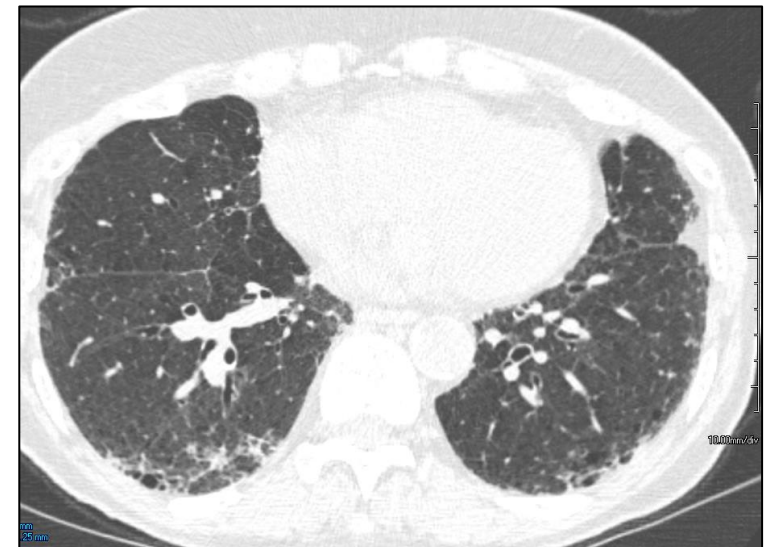
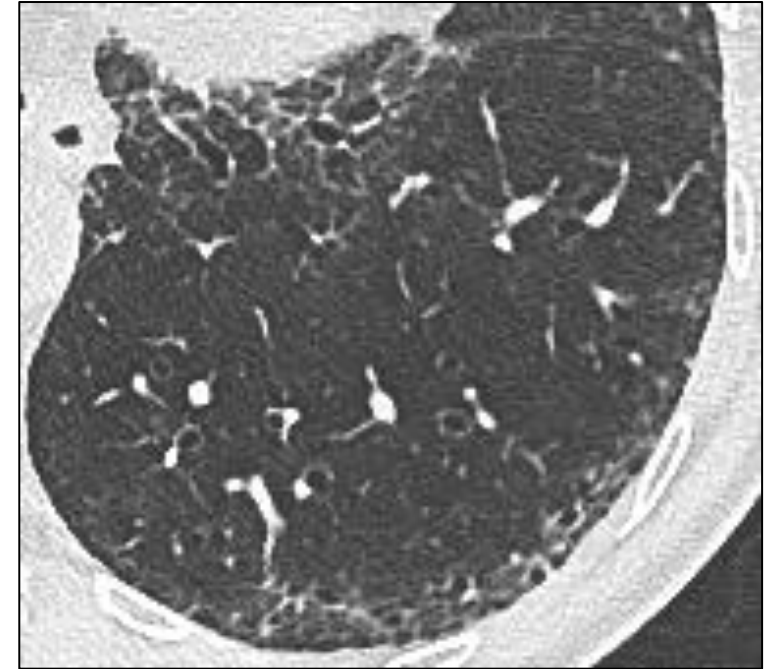
# 차례

- CT Signs of Fibrosis
- HRCT UIP Pattern (2018 & 2022 ATS/ERS/JRS/ALAT Guidelines)
- Other ILD CT Pattern (alternative diagnosis)



## CT signs of fibrosis

- **Reticular opacity**
- **Traction bronchiectasis**
- **Honeycombing cyst**
- **Volume decrease & Lung distortion**
- **Three density sign**
- **Non-emphysematous cysts**

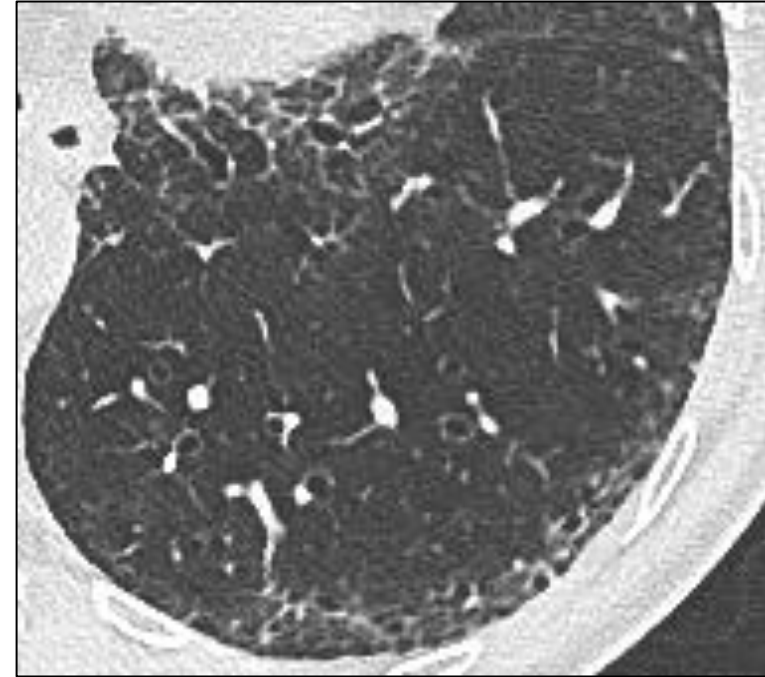
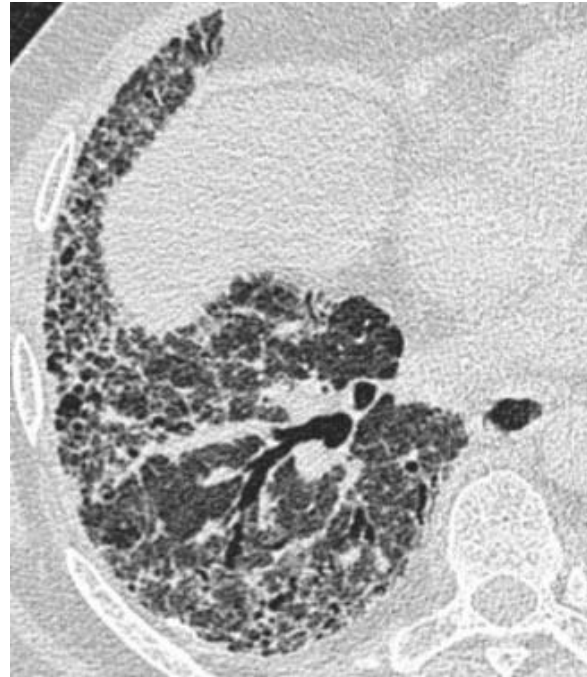


# Reticular opacity

- Innumerable small linear opacities resembling net

– Used in chest radiographs & chest CT

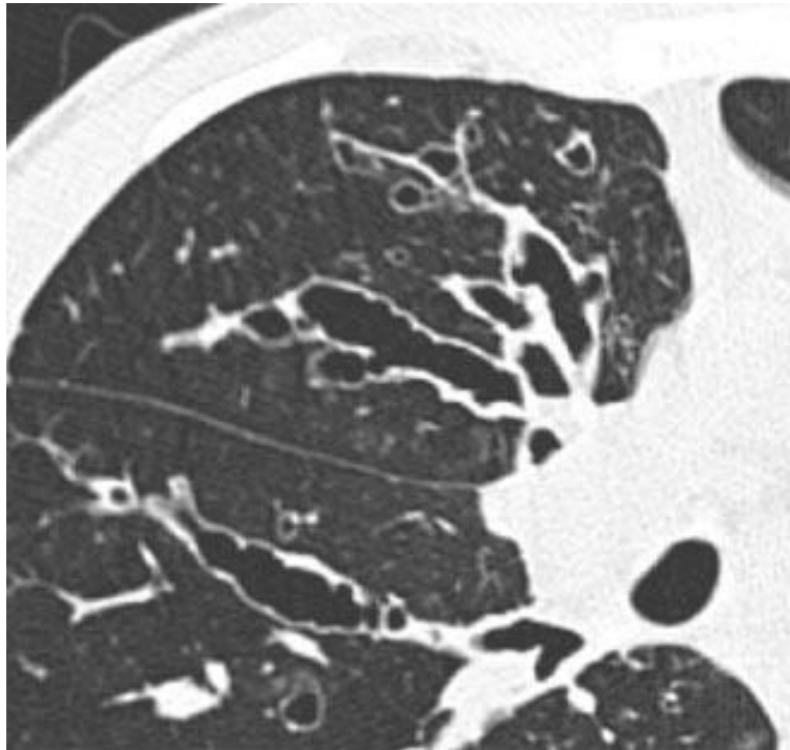
– **Fibrosis**: Interlobular septal thickening, intralobular lines, or cyst walls of honeycombing cysts



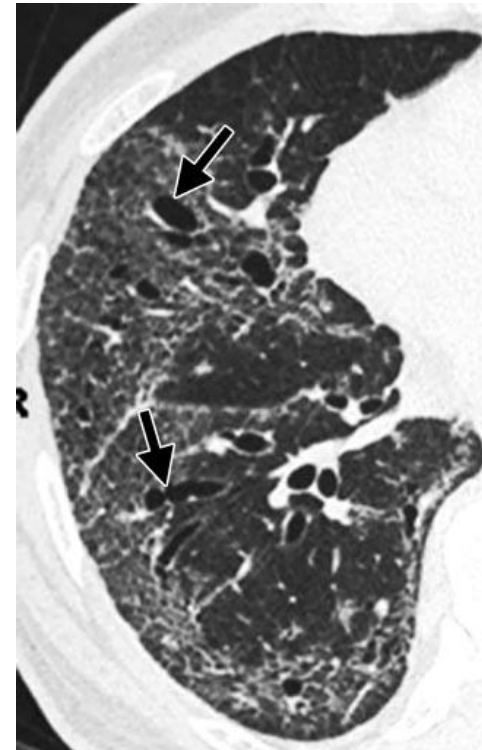
# Traction bronchiectasis

Hansell DM, et al. Radiology 2008

- Irregular bronchial dilatation caused by **surrounding retractile pulmonary fibrosis**
- **Traction bronchiectasis ~ Honeycombing cyst: continuum**



Bronchiectasis



Traction bronchiectasis

# Honeycombing cyst

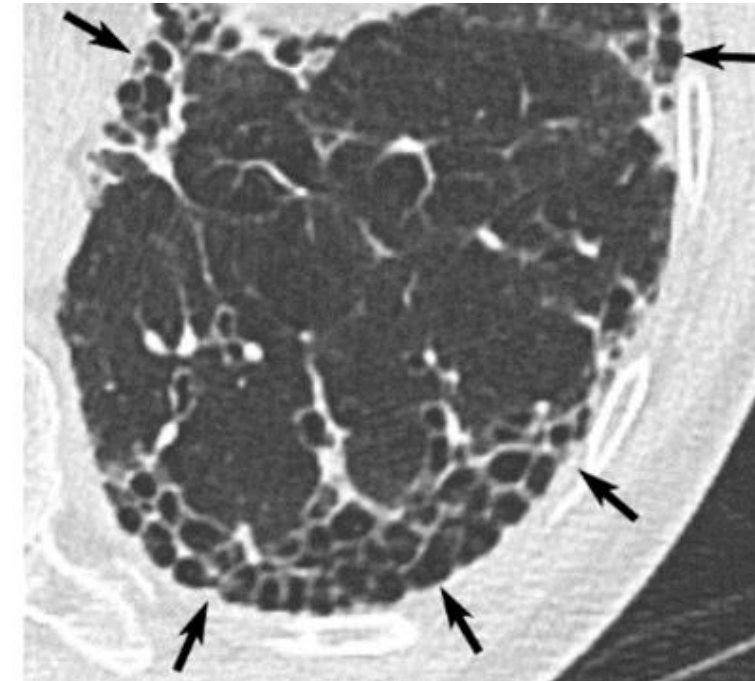
Hansell DM, et al. Radiology 2008  
Travis WD et al. AJRCCM 2013

- **Pathology**

- Destroyed and fibrotic lung tissue containing numerous cystic airspaces with thick fibrous walls

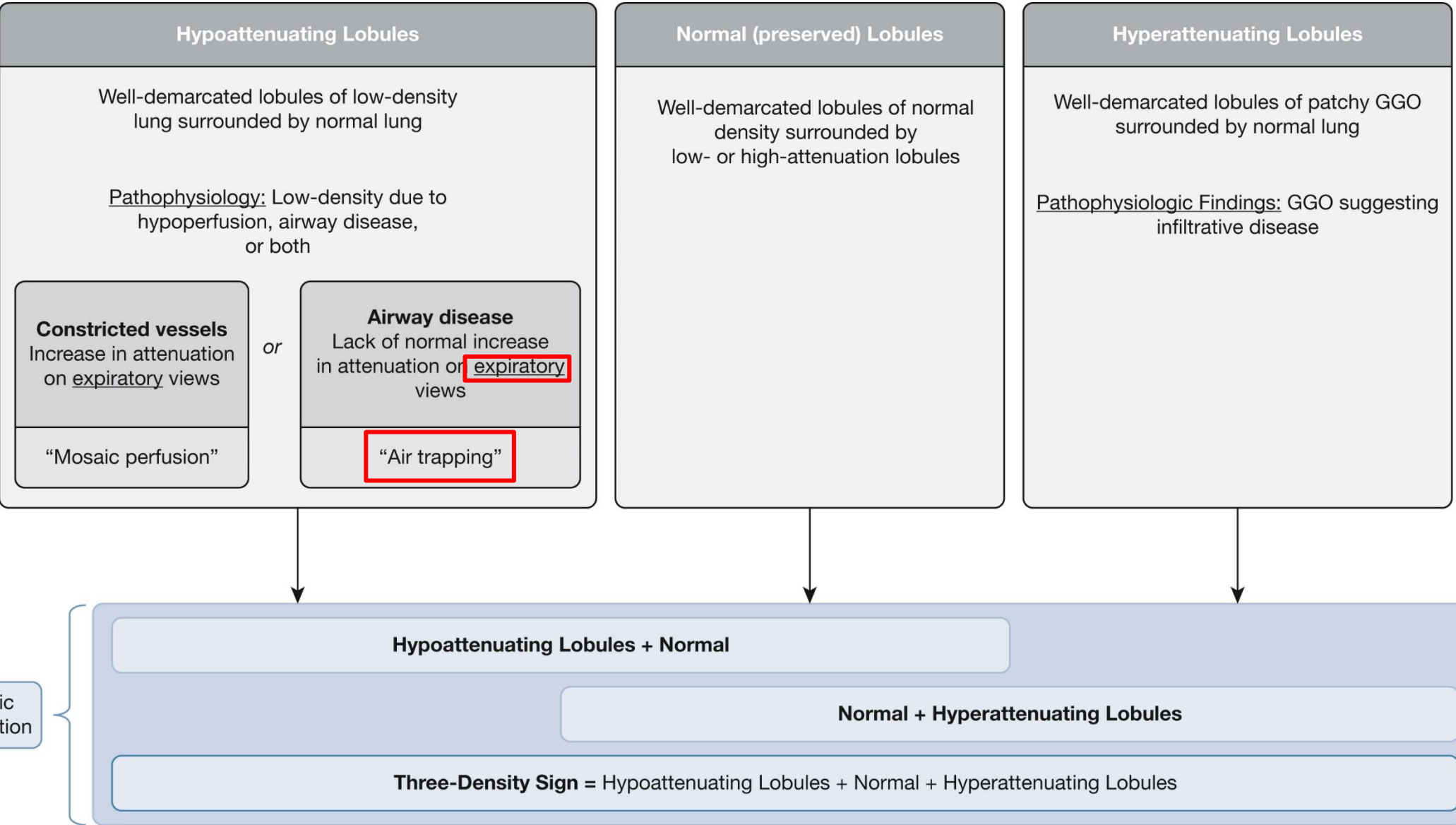
- **CT**

- **Clustered cystic air spaces**, typically 3-10 mm up to 2.5 cm
- Usually **subpleural, characterized by well-defined and shared walls**
- Important **criterion in the diagnosis of UIP**
- Increase in size and number during disease progression
- A single layer can be honeycombing cyst

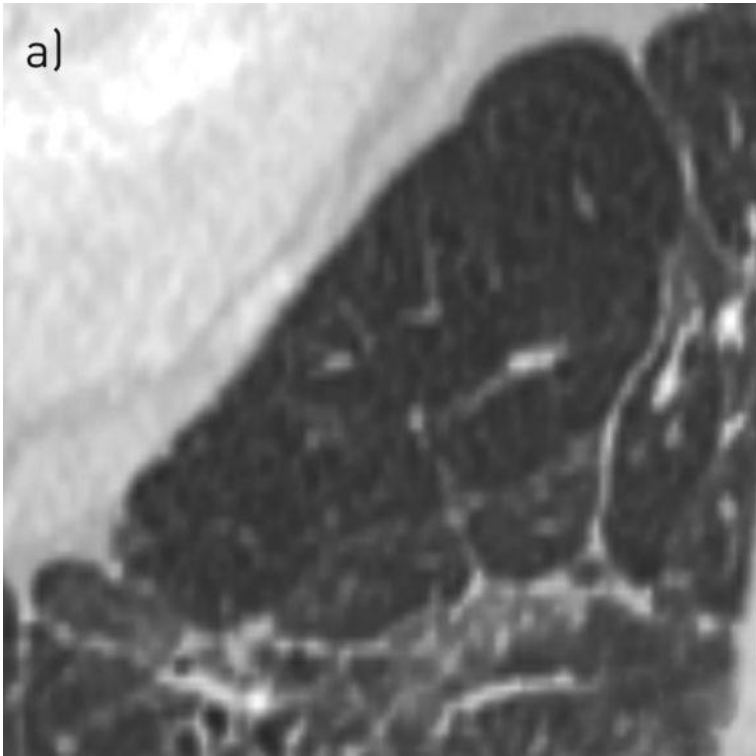




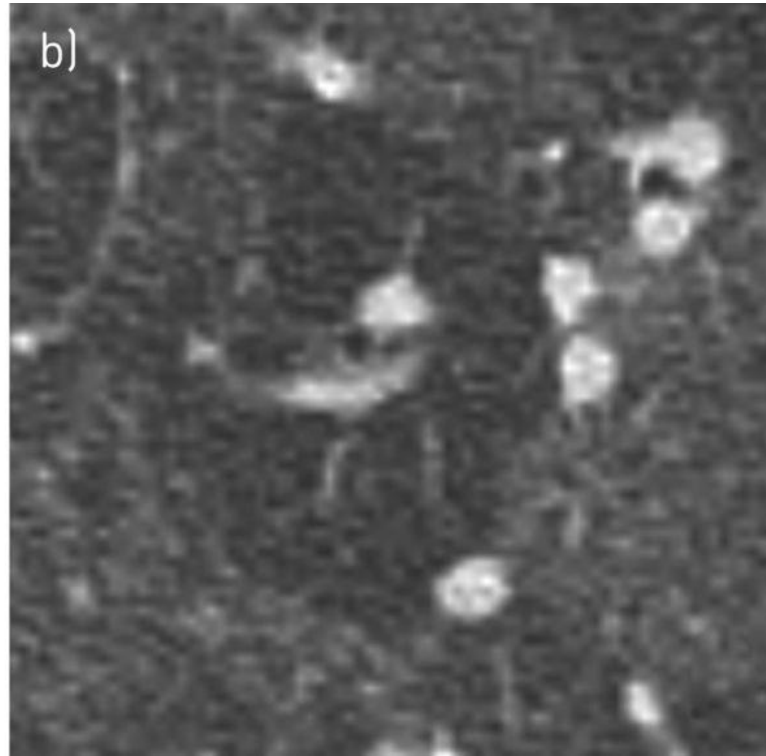
# Mosaic attenuation



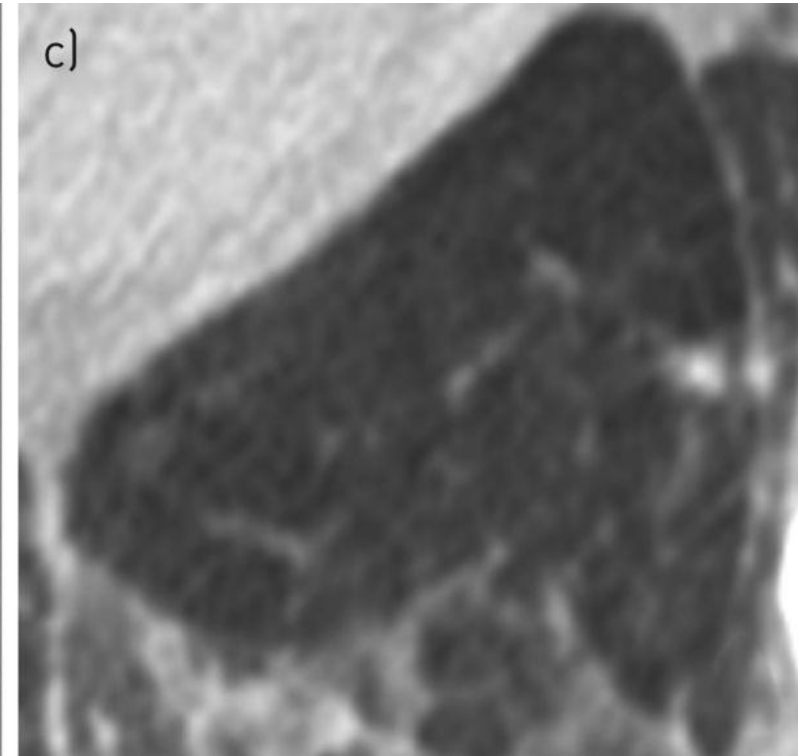
# Mosaic attenuation



Low-density lobules  
Constricted vessels



Preserved lobules  
Increased density of the  
surrounding lung



Expiratory CT  
Air trapping

# Three-density

- Combination

- Normal ap

- High attenu

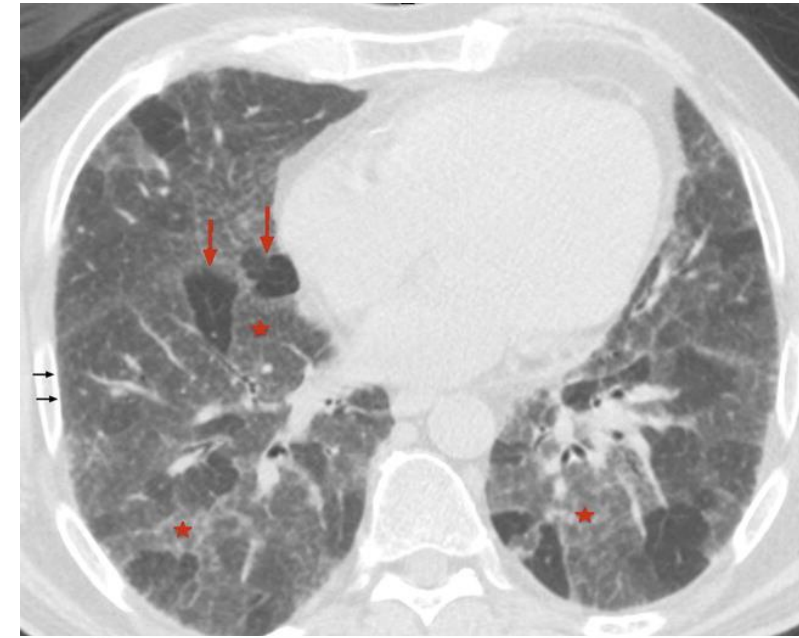
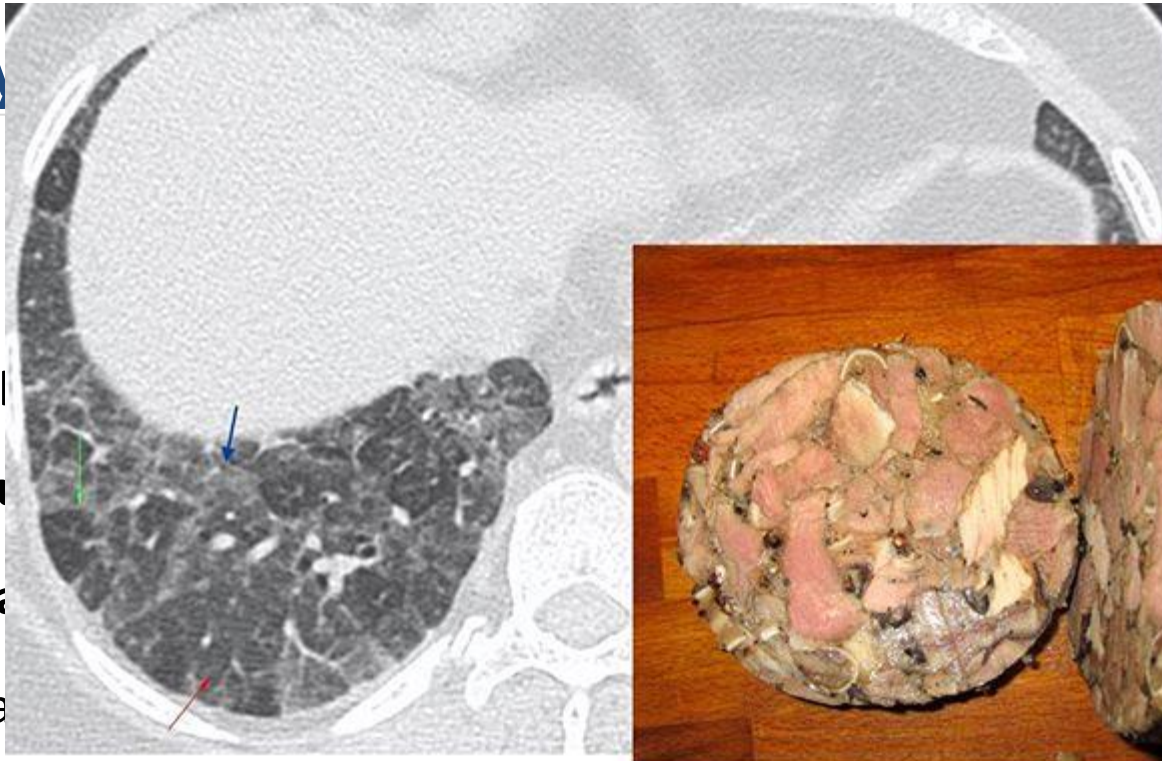
- Lucent area

- Sharply dema

- Headcheese sign

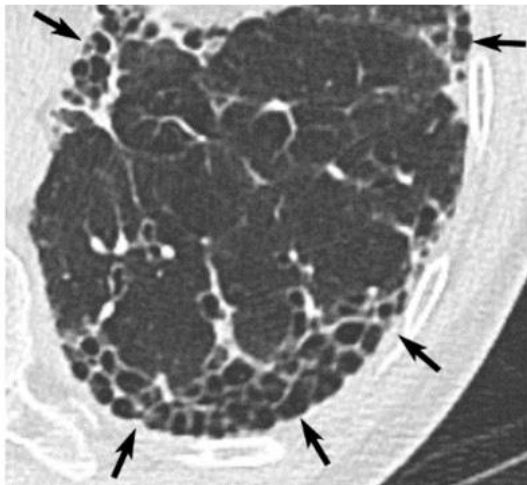
- Mixed obstructive (decreased attenuation and vascularity) and infiltrative (GGO surrounding preserved normal lobules) process

- **Highly specific for fibrotic HP**

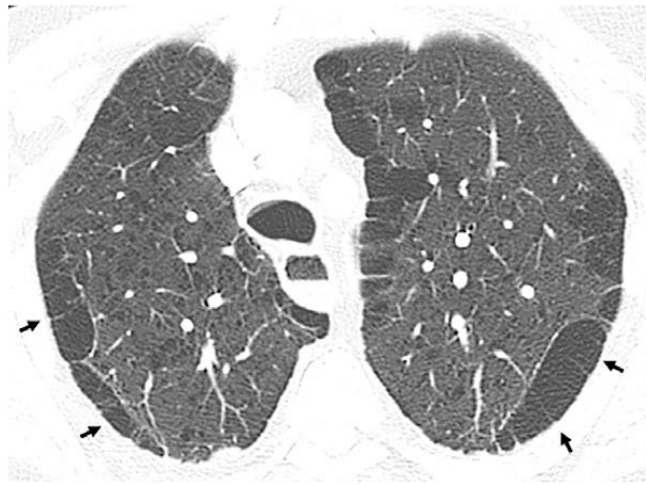


## Non-emphysematous cysts

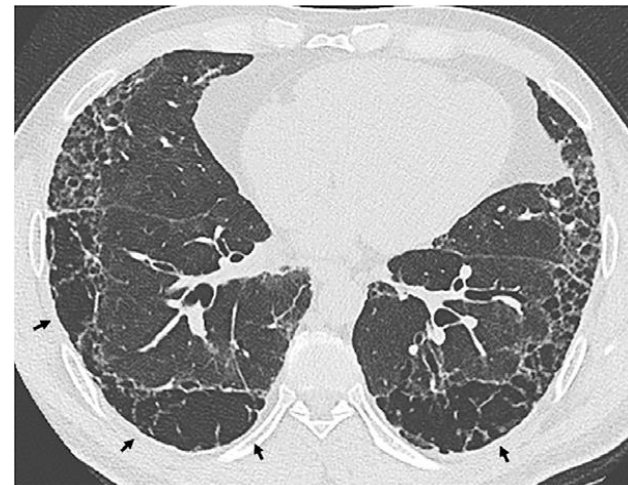
- Lucencies with irregular, well-defined walls
- Cigarette smokers
- DDx from emphysema (: presence of a well-defined wall)
- DDx from honeycombing (: irregular shape, varying size, absence of subpleural predominance, vascular or interlobular core)
- Histology: airspace enlargement with fibrosis, smoking-related interstitial fibrosis



Honeycombing cyst



Emphysema



Non-emphysematous cyst

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# **Idiopathic pulmonary fibrosis (IPF)**

## **HRCT UIP Pattern**

# Usual interstitial pneumonia (UIP) pattern on CT

Ragu G, et al. AJRCCM 2018 and 2022

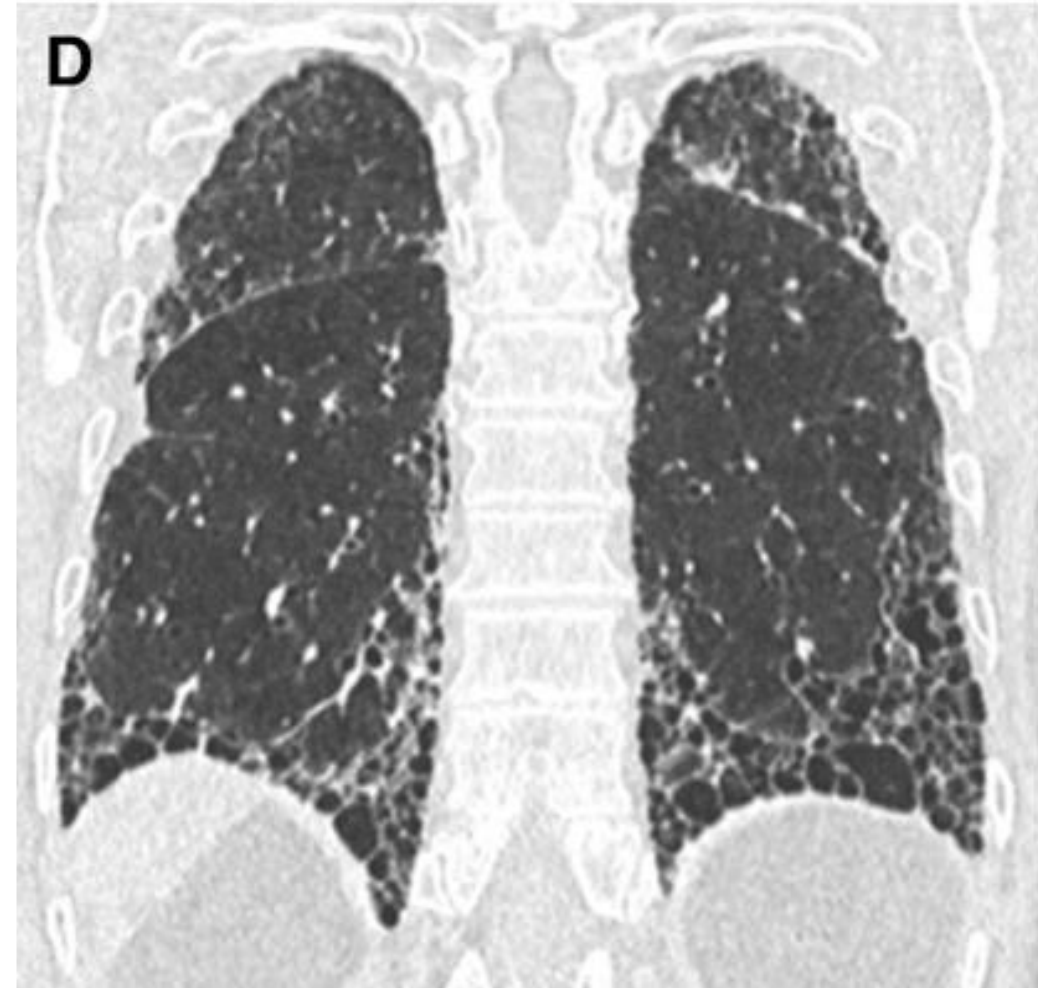
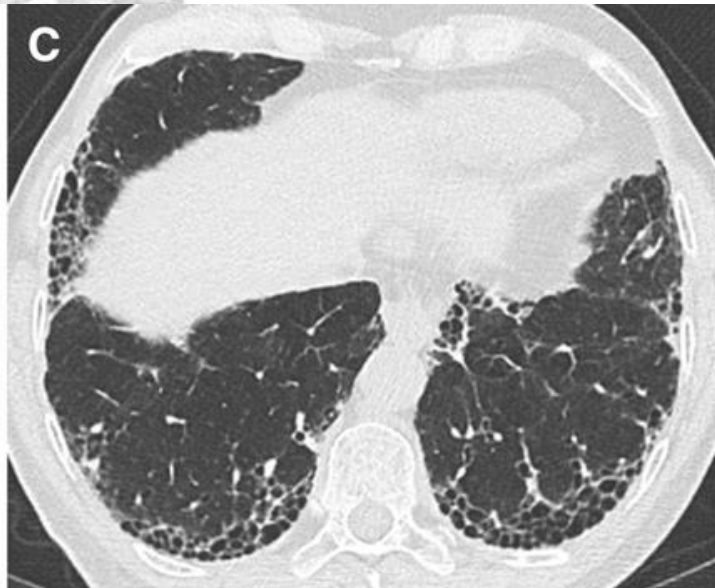
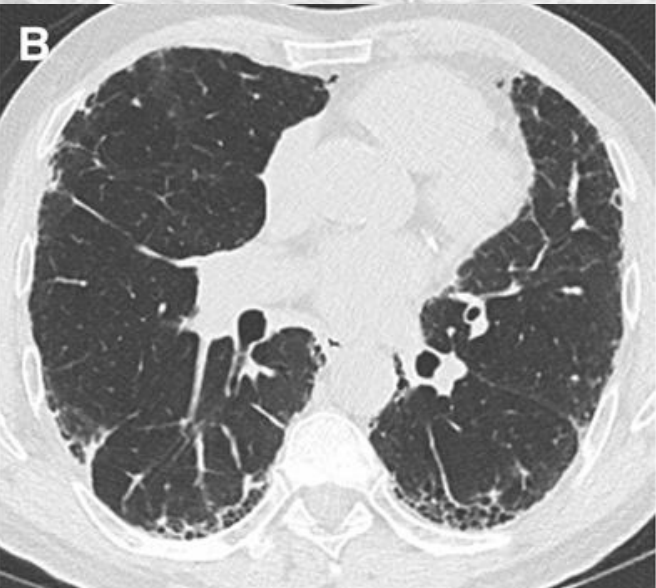
	HRCT Pattern			CT Findings Suggestive of an Alternative Diagnosis
	UIP Pattern	Probable UIP Pattern	Indeterminate for UIP	
Level of confidence for UIP histology	Confident (>90%)	Provisional high confidence (70–89%)	Provisional low confidence (51–69%)	Low to very low confidence (≤50%)
Distribution	<ul style="list-style-type: none"> <li>Subpleural and basal predominant</li> <li>Often heterogeneous (areas of normal lung interspersed with fibrosis)</li> <li>Occasionally diffuse</li> <li>May be asymmetric</li> </ul>	<ul style="list-style-type: none"> <li>Subpleural and basal predominant</li> <li>Often heterogeneous (areas of normal lung interspersed with reticulation and traction bronchiectasis/bronchiolectasis)</li> </ul>	<ul style="list-style-type: none"> <li>Diffuse distribution without subpleural predominance</li> </ul>	<ul style="list-style-type: none"> <li>Peribronchovascular predominant with subpleural sparing (consider NSIP)</li> <li>Perilymphatic distribution (consider sarcoidosis)</li> <li>Upper or mid lung (consider fibrotic HP, CTD-ILD, and sarcoidosis)</li> <li>Subpleural sparing (consider NSIP or smoking-related IP)</li> </ul>
CT features	<ul style="list-style-type: none"> <li>Honeycombing with or without traction bronchiectasis/bronchiolectasis</li> <li>Presence of irregular thickening of interlobular septa</li> <li>Usually superimposed with a reticular pattern, mild GGO</li> <li>May have pulmonary ossification</li> </ul>	<ul style="list-style-type: none"> <li>Reticular pattern with traction bronchiectasis/bronchiolectasis</li> <li>May have mild GGO</li> <li>Absence of subpleural sparing</li> </ul>	<ul style="list-style-type: none"> <li>CT features of lung fibrosis that do not suggest any specific etiology</li> </ul>	<ul style="list-style-type: none"> <li>Lung findings                             <ul style="list-style-type: none"> <li>Cysts (consider LAM, PLCH, LIP, and DIP)</li> <li>Mosaic attenuation or three-density sign (consider HP)</li> <li>Predominant GGO (consider HP, smoking-related disease, drug toxicity, and acute exacerbation of fibrosis)</li> <li>Profuse centrilobular micronodules (consider HP or smoking-related disease)</li> <li>Nodules (consider sarcoidosis)</li> <li>Consolidation (consider organizing pneumonia, etc.)</li> </ul> </li> <li>Mediastinal findings                             <ul style="list-style-type: none"> <li>Pleural plaques (consider asbestosis)</li> <li>Dilated esophagus (consider CTD)</li> </ul> </li> </ul>

# Usual interstitial pneumonia (UIP) pattern CT



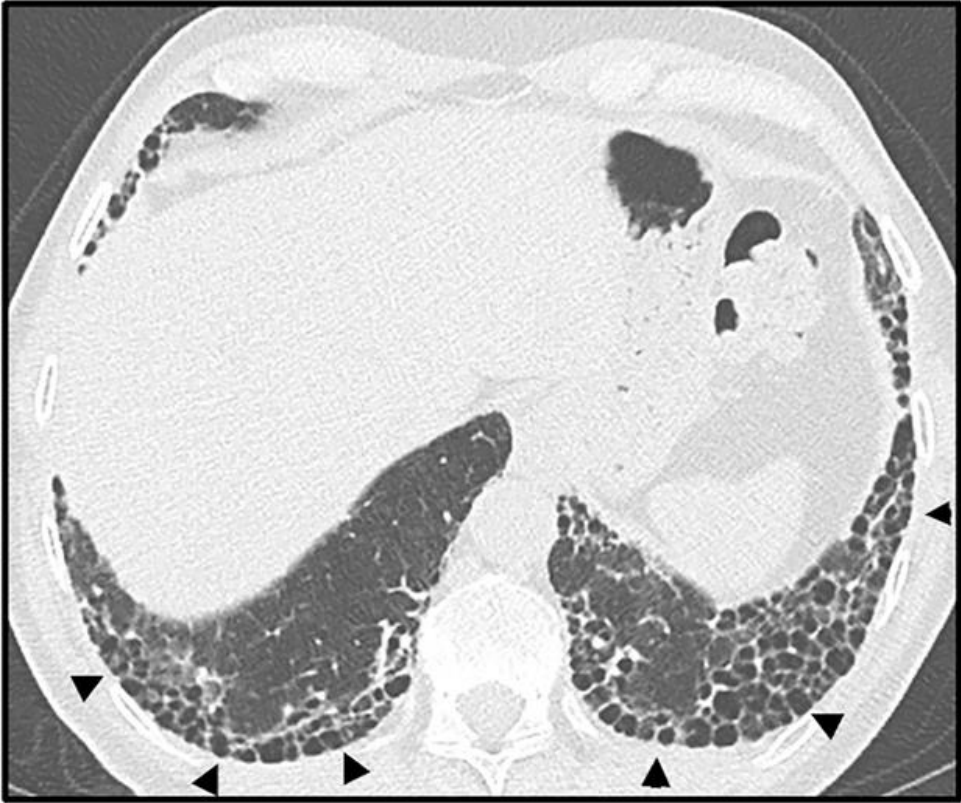
Reticular opacity

Traction  
bronchiectasis +  
Honeycombing



Subpleural and basal predominancy

## Usual interstitial pneumonia (UIP) pattern CT



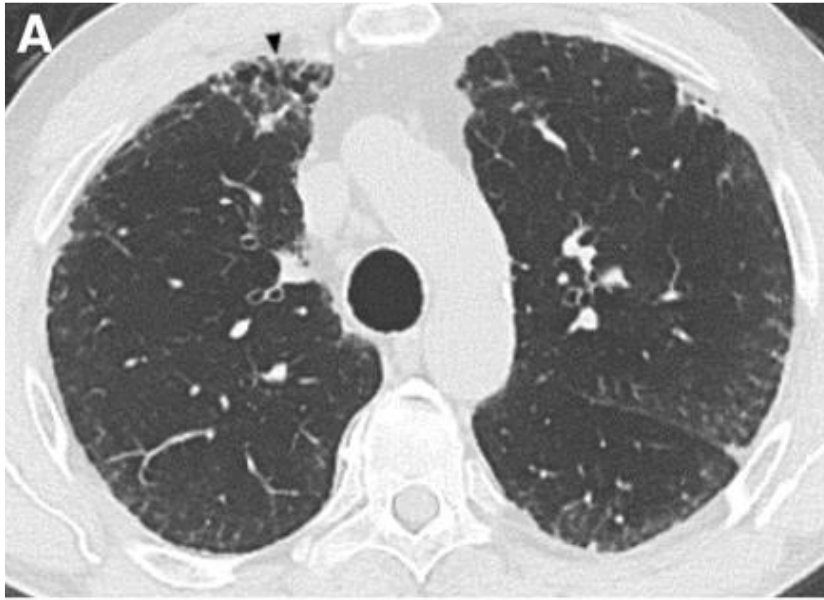
**Traction bronchiectasis + Honeycombing**



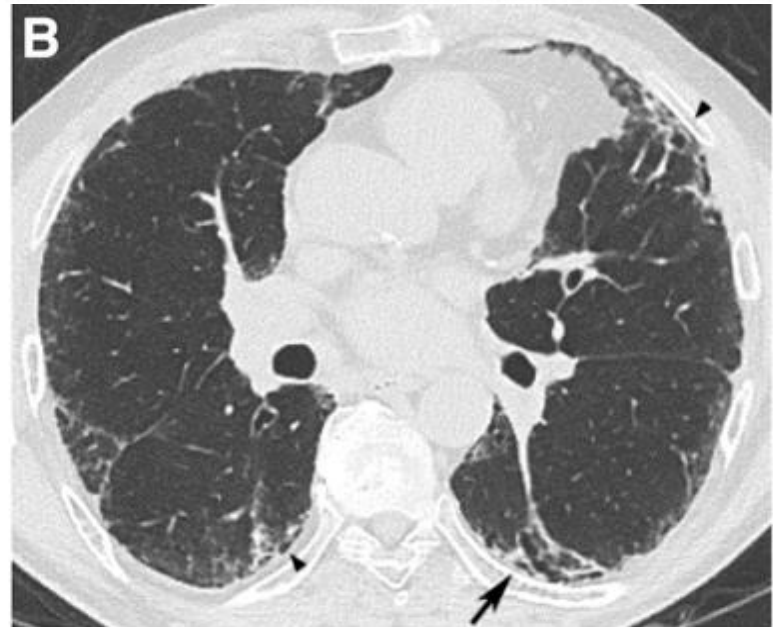
**Subpleural and basal predominance**



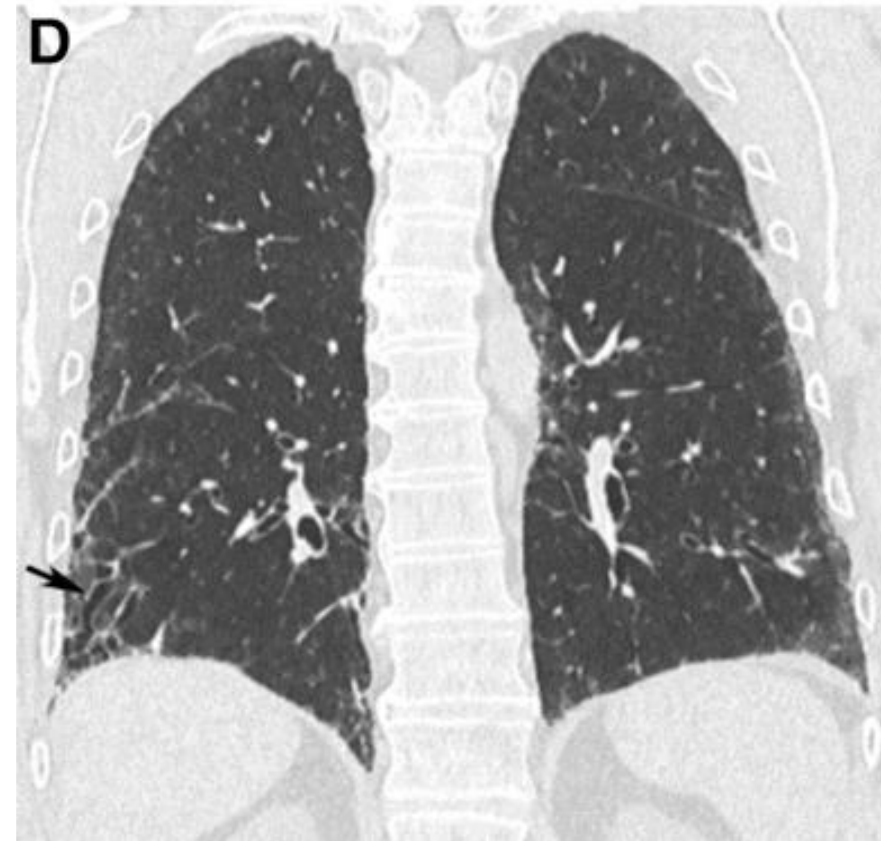
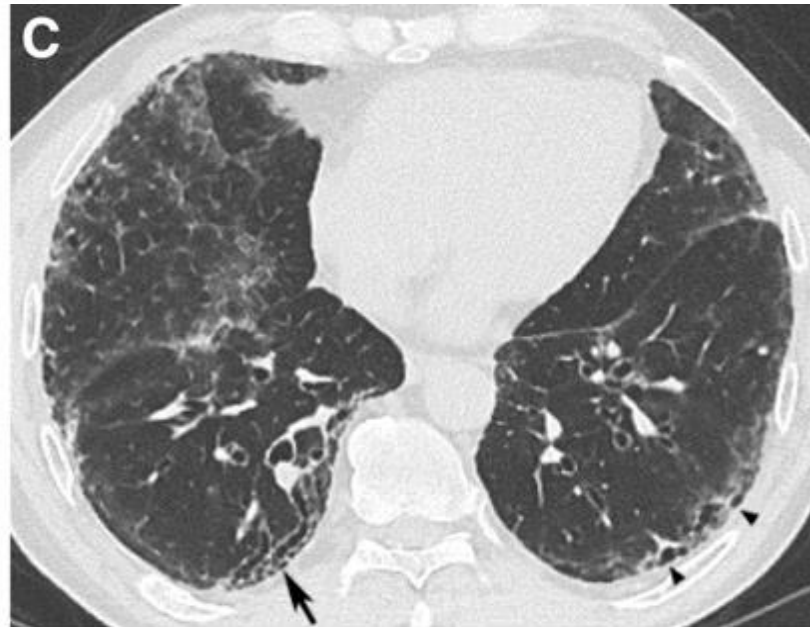
# Probable usual interstitial pneumonia (UIP) pattern CT



**Reticular opacity**



**Traction bronchiectasis + mild GGO**

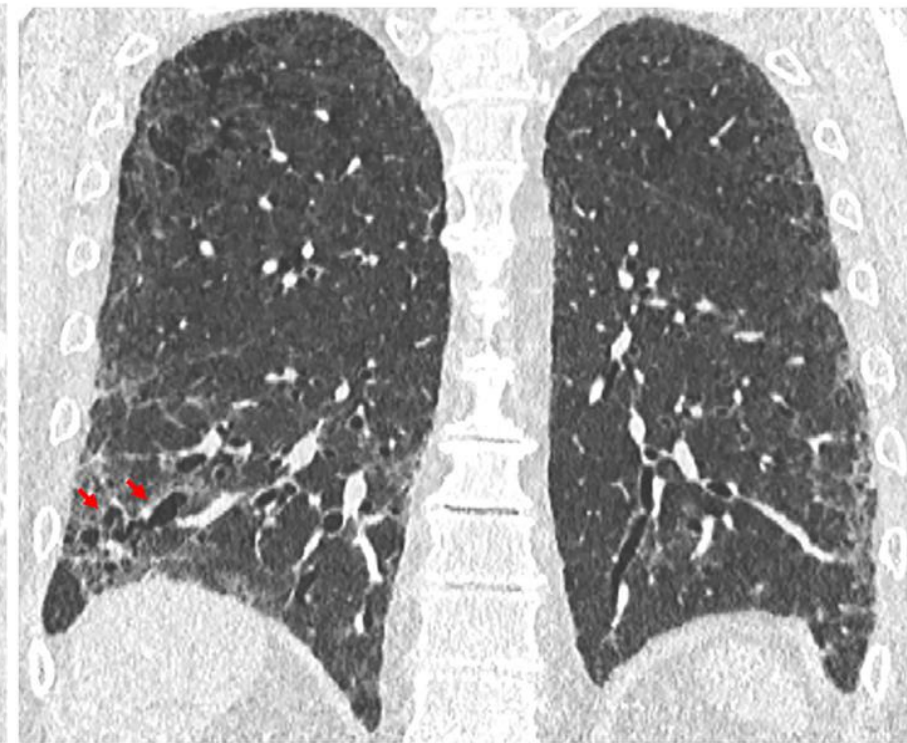
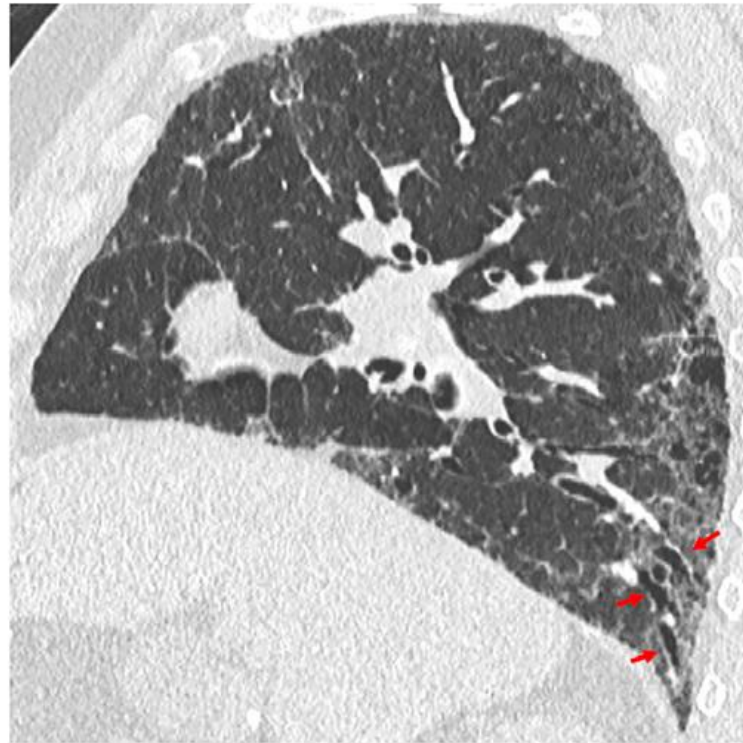


**Subpleural and basal predominance**

## Probable usual interstitial pneumonia (UIP) pattern CT

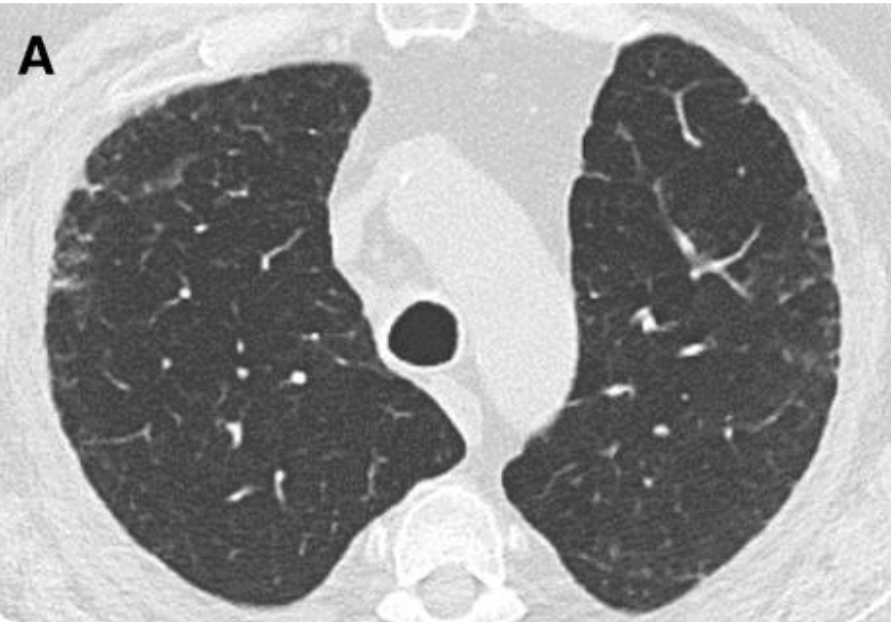


**Reticular opacity + traction bronchiectasis + mild GGO**

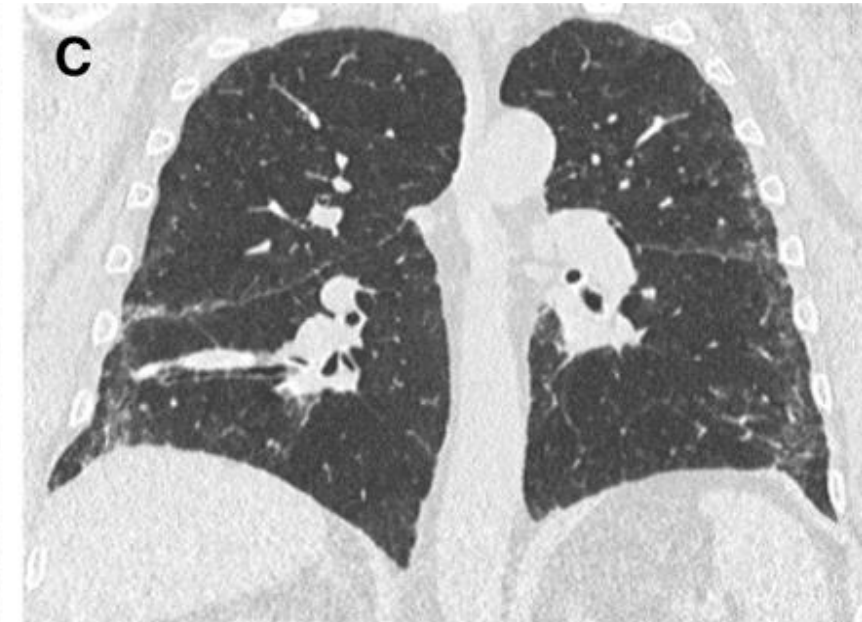
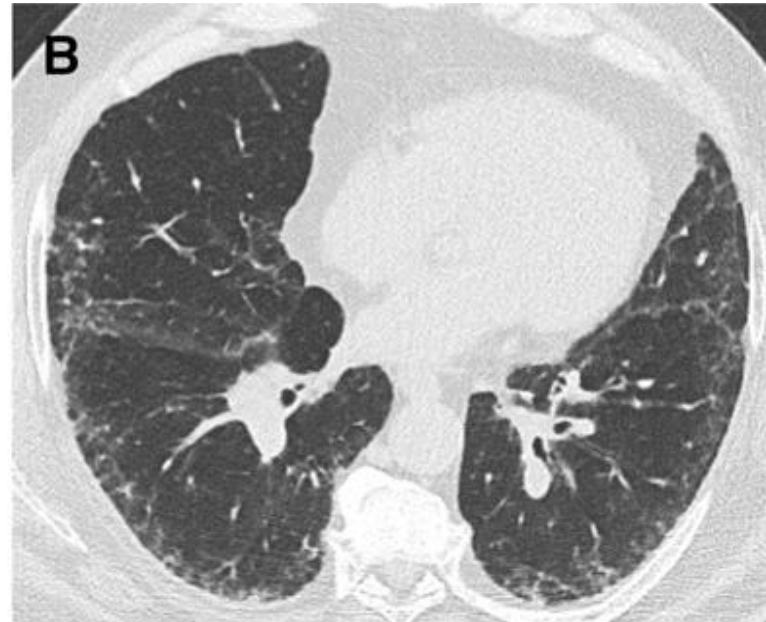


**Subpleural and basal predominance**

## Indeterminate usual interstitial pneumonia (UIP) pattern CT

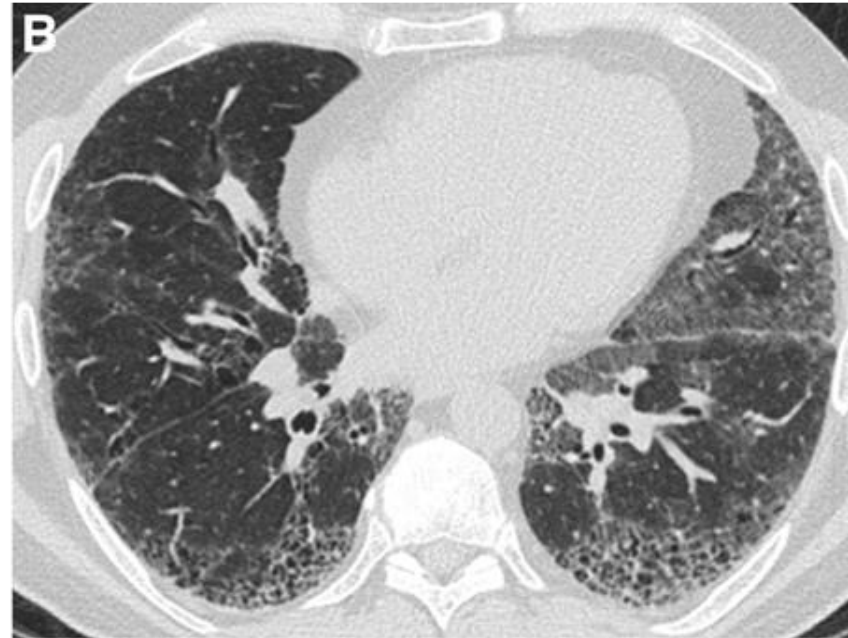


**Reticular opacity + mild GGO**



**Subpleural and basal predominance**

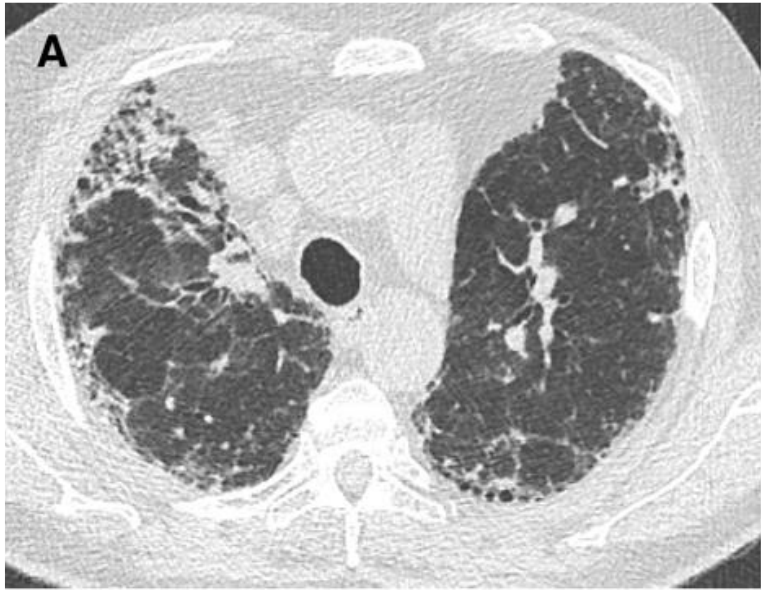
## Indeterminate usual interstitial pneumonia (UIP) pattern CT



**Marked GGO + reticular opacity + traction bronchiectasis**

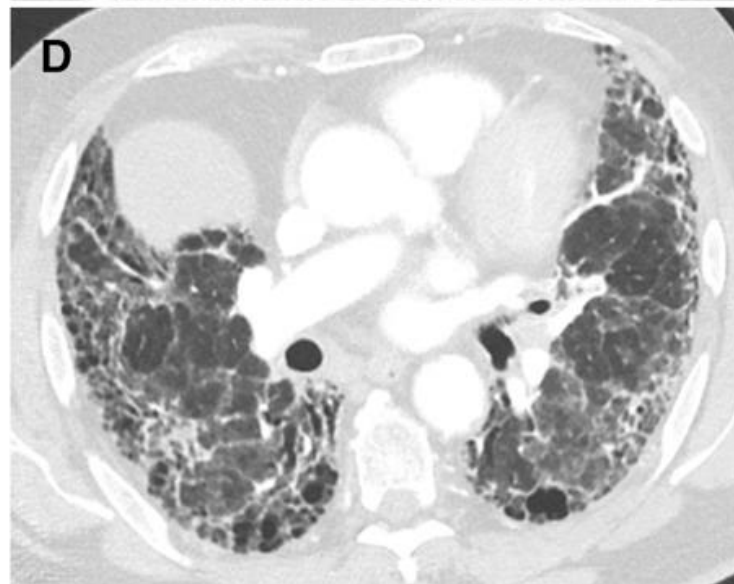
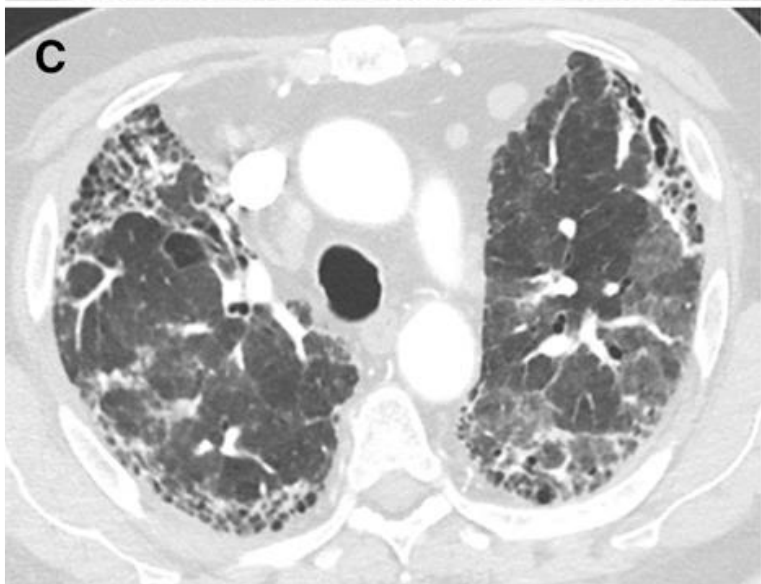
**Subpleural and basal predominance**

## Acute exacerbation of IPF



**Reticular opacity**  
**Traction bronchiectasis**  
**Honeycombing cyst**  
**Subpleural and basal predominancy**

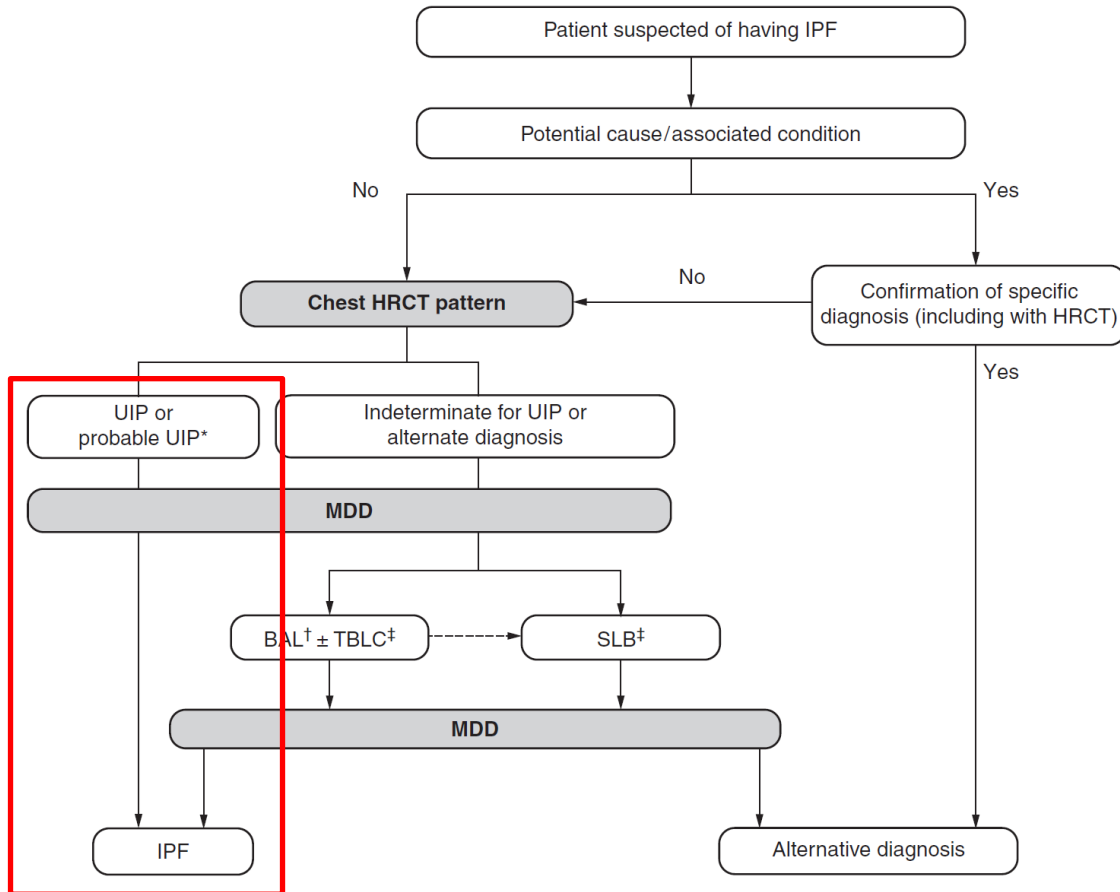
**= UIP pattern**



**New GGO in bilateral lung**  
**= AE-IPF**

**Versus fibrotic NSIP?**

# IPF diagnosis



IPF suspected*		Histopathology pattern <sup>†</sup>			
		UIP	Probable UIP	Indeterminate for UIP or biopsy not performed	Alternative diagnosis
HRCT pattern	UIP	IPF	IPF	IPF	Non-IPF dx
	Probable UIP	IPF	IPF	IPF (Likely) <sup>‡</sup>	Non-IPF dx
	Indeterminate	IPF	IPF (Likely) <sup>‡</sup>	Indeterminate <sup>§</sup>	Non-IPF dx
	Alternative diagnosis	IPF (Likely) <sup>‡</sup>	Indeterminate <sup>§</sup>	Non-IPF dx	Non-IPF dx

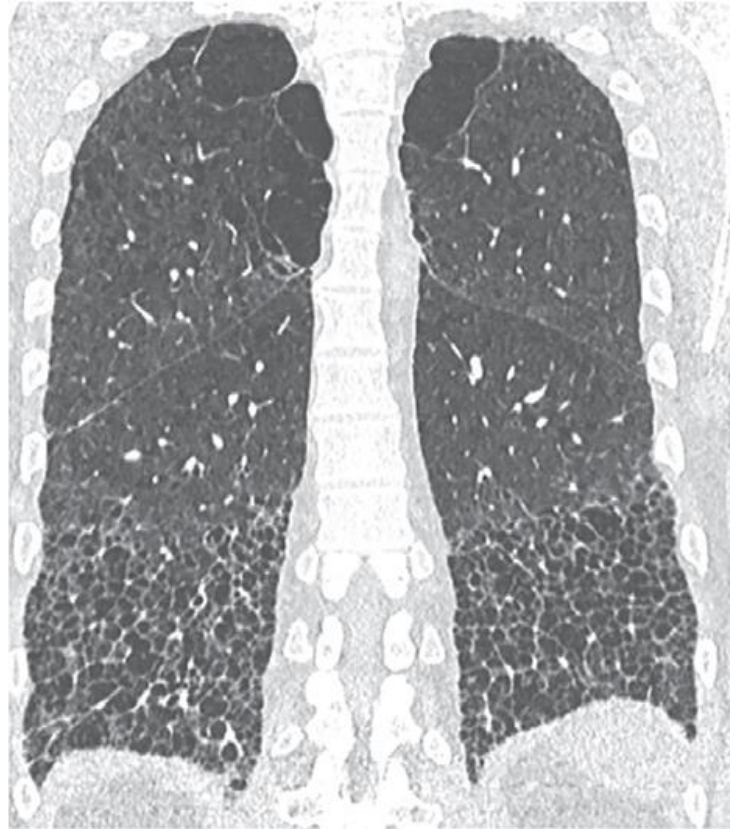
## \*IPF (likely)

- Moderate to severe traction bronchiectasis in a man >50Y or in a woman >60Y old
- Extensive (>30%) reticulation on HRCT and age >70Y
- Increased neutrophils and/or absence of lymphocytosis in BAL
- Multidisciplinary discussion: confident diagnosis of IPF.

\*Appropriate setting: old age, male, smoker

## UIP pattern spectrum

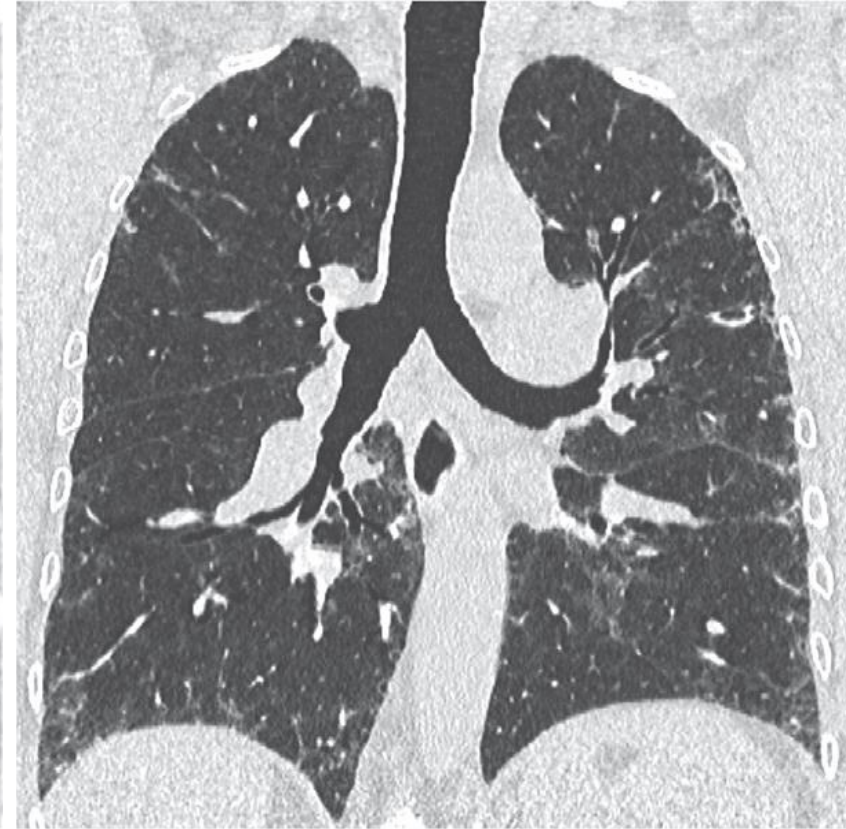
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**UIP pattern**



**Probable UIP pattern**



**Indeterminate UIP pattern**

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## **Other ILD CT Pattern (alternative diagnosis)**

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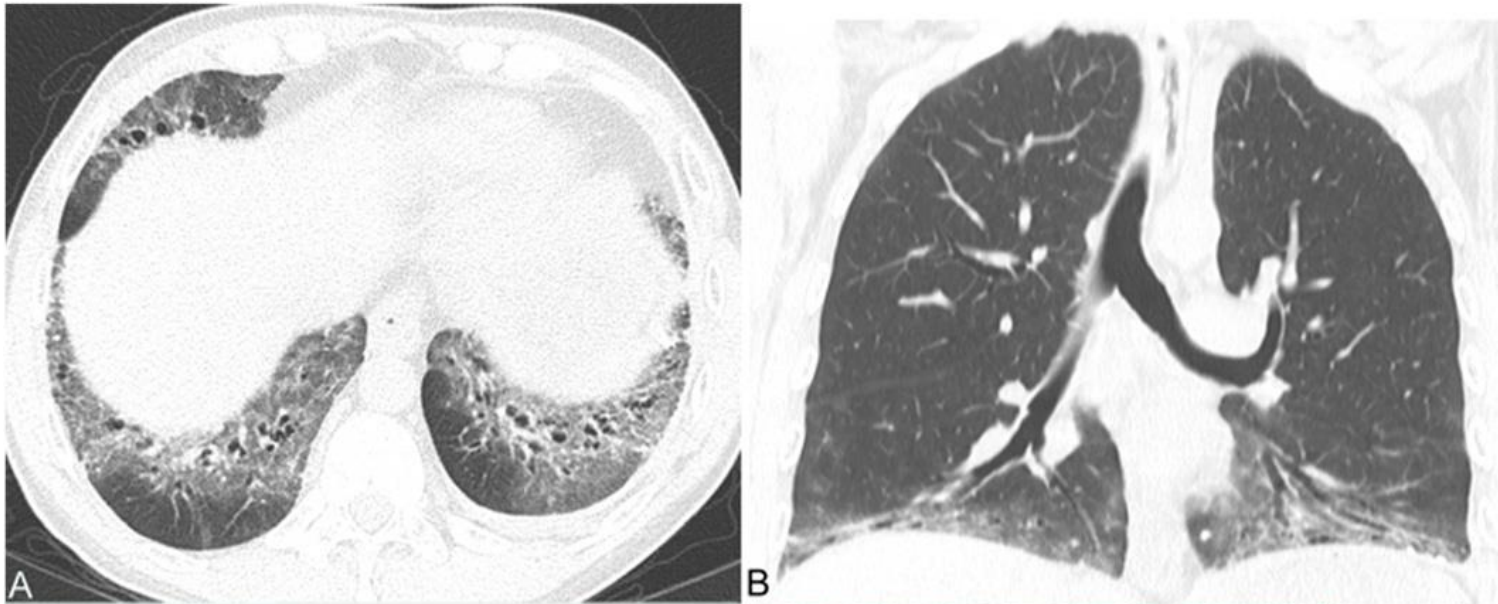
# Usual interstitial pneumonia (UIP) pattern on CT

Ragu G, et al. AJRCCM 2018 and 2022

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Distribution	<ul style="list-style-type: none"> <li>• Subpleural and basal predominant</li> <li>• Often heterogeneous (areas of normal lung interspersed with fibrosis)</li> <li>• Occasionally diffuse</li> <li>• May be asymmetric</li> </ul>	<ul style="list-style-type: none"> <li>• Subpleural and basal predominant</li> <li>• Often heterogeneous (areas of normal lung interspersed with reticulation and traction bronchiectasis/bronchiolectasis)</li> </ul>	<ul style="list-style-type: none"> <li>• Diffuse distribution without subpleural predominance</li> </ul>	<ul style="list-style-type: none"> <li>• Peribronchovascular predominant with subpleural sparing (consider NSIP)</li> <li>• Perilymphatic distribution (consider sarcoidosis)</li> <li>• Upper or mid lung (consider fibrotic HP, CTD-ILD, and sarcoidosis)</li> <li>• Subpleural sparing (consider NSIP or smoking-related IP)</li> </ul>
CT features	<ul style="list-style-type: none"> <li>• Honeycombing with or without traction bronchiectasis/bronchiolectasis</li> <li>• Presence of irregular thickening of interlobular septa</li> <li>• Usually superimposed with a reticular pattern, mild GGO</li> <li>• May have pulmonary ossification</li> </ul>	<ul style="list-style-type: none"> <li>• Reticular pattern with traction bronchiectasis/bronchiolectasis</li> <li>• May have mild GGO</li> <li>• Absence of subpleural sparing</li> </ul>	<ul style="list-style-type: none"> <li>• CT features of lung fibrosis that do not suggest any specific etiology</li> </ul>	<ul style="list-style-type: none"> <li>• Lung findings                             <ul style="list-style-type: none"> <li>○ Cysts (consider LAM, PLCH, LIP, and DIP)</li> <li>○ Mosaic attenuation or three-density sign (consider HP)</li> <li>○ Predominant GGO (consider HP, smoking-related disease, drug toxicity, and acute exacerbation of fibrosis)</li> <li>○ Profuse centrilobular micronodules (consider HP or smoking-related disease)</li> <li>○ Nodules (consider sarcoidosis)</li> <li>○ Consolidation (consider organizing pneumonia, etc.)</li> </ul> </li> <li>• Mediastinal findings                             <ul style="list-style-type: none"> <li>○ Pleural plaques (consider asbestosis)</li> <li>○ Dilated esophagus (consider CTD)</li> </ul> </li> </ul>

# Nonspecific interstitial pneumonia (NSIP)

- Bilateral, symmetric, predominantly lower lung, **bronchovascular bundle distribution**
- **GGO, reticular opacity with traction bronchiectasis**, volume decrease
- Diffuse or subpleural, sometimes **subpleural sparing**
- Commonly associated diseases in NSIP: connective tissue disease, drug toxicity, etc.
- With consolidation: OP pattern, suggesting connective tissue disorder

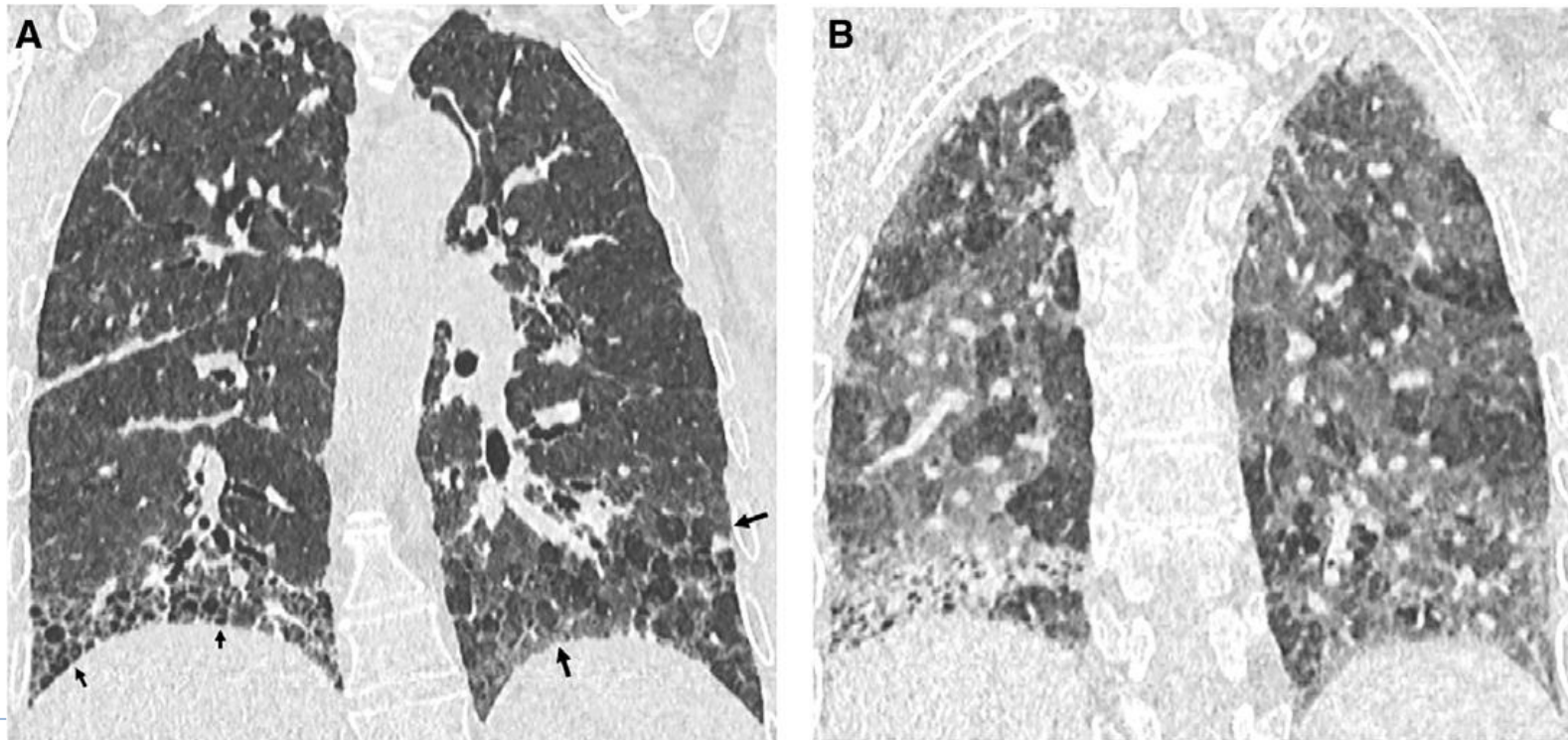


## UIP VS. NSIP

	UIP	NSIP
Median age, Sex	50-70, men	40-50, women
Subpleural sparing	-	+
Reticulation	Most prominent feature (subpleural)	Variable: absent to extensive (subpleural sparing)
GGO	Mild	<b>Most prominent feature</b>
Traction bronchiectasis	Common (subpleural)	Common (bronchovascular bundle)
Honeycombing	<b>Common; subpleural and basal</b>	Uncommon at presentation

# Fibrotic hypersensitivity pneumonitis

- Air-trapping or Three-density sign
- Mid-to-upper lung predominancy
- Centrilobular nodules, central peribronchovascular reticulation and distortion
- Traction bronchiectasis & Honeycombing: may be present but do not predominate

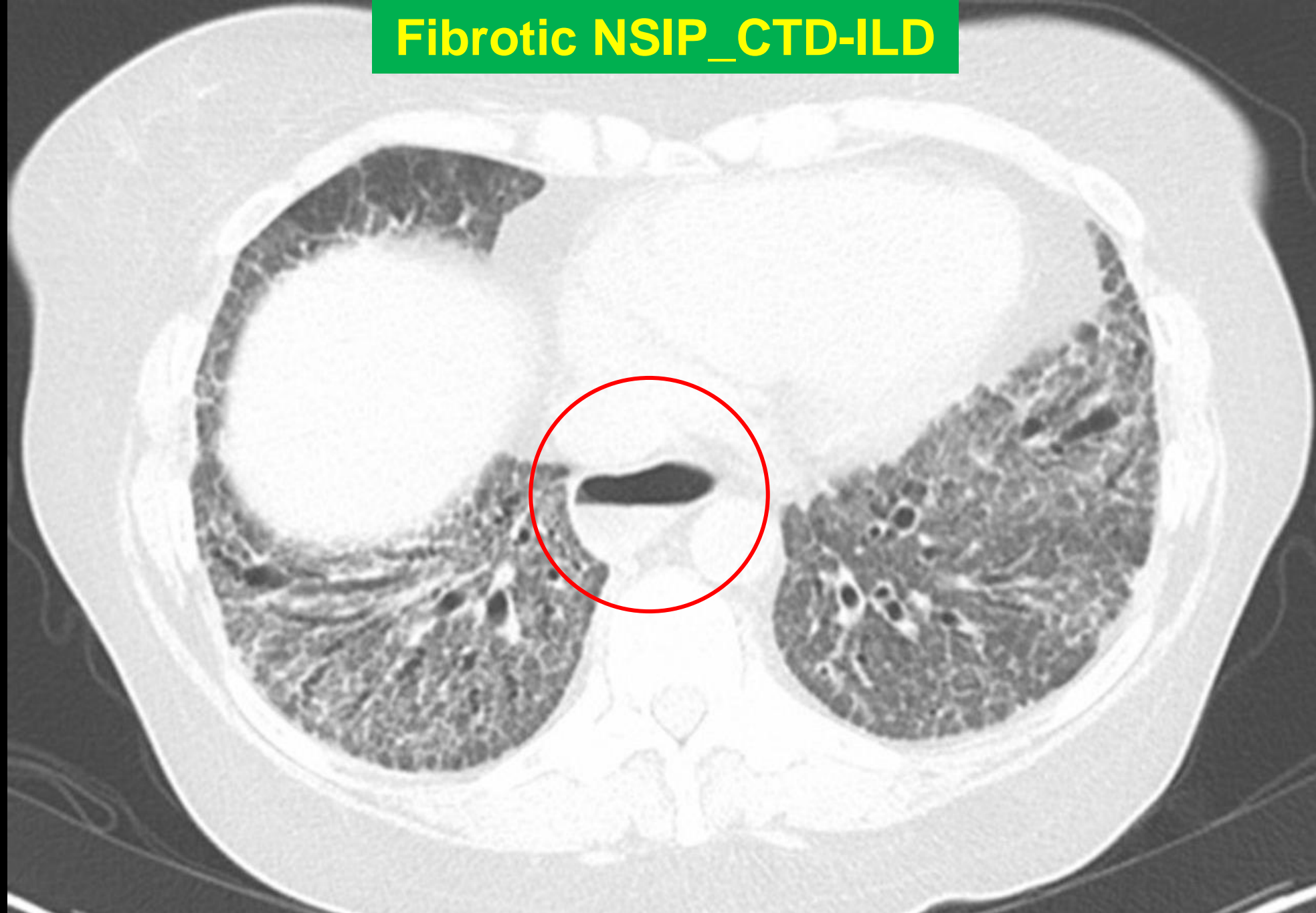


## Connective tissue disease-ILD

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- **Basically indistinguishable from idiopathic interstitial pneumonia**
  - **Tend to cause other thoracic manifestations**
    - Pleural/pericardial effusion or thickening (e.g., SLE)
    - Esophageal dilatation (e.g., scleroderma)
    - Airway involvement: Bronchiectasis, constrictive bronchiolitis
    - Pulmonary artery dilatation
    - Myocarditis
    - Musculoskeletal: soft tissue calcification, joint abnormalities
-

**Fibrotic NSIP\_CTD-ILD**



## Connective tissue disease-ILD



### **Anterior upper lobe sign**

Concentrated fibrosis in the anterior of aspect of the upper lobes while sparing the other aspect of the upper lobes

## Connective tissue disease-ILD

**Exuberant honeycomb sign**  
: >70% of fibrotic portion of lung

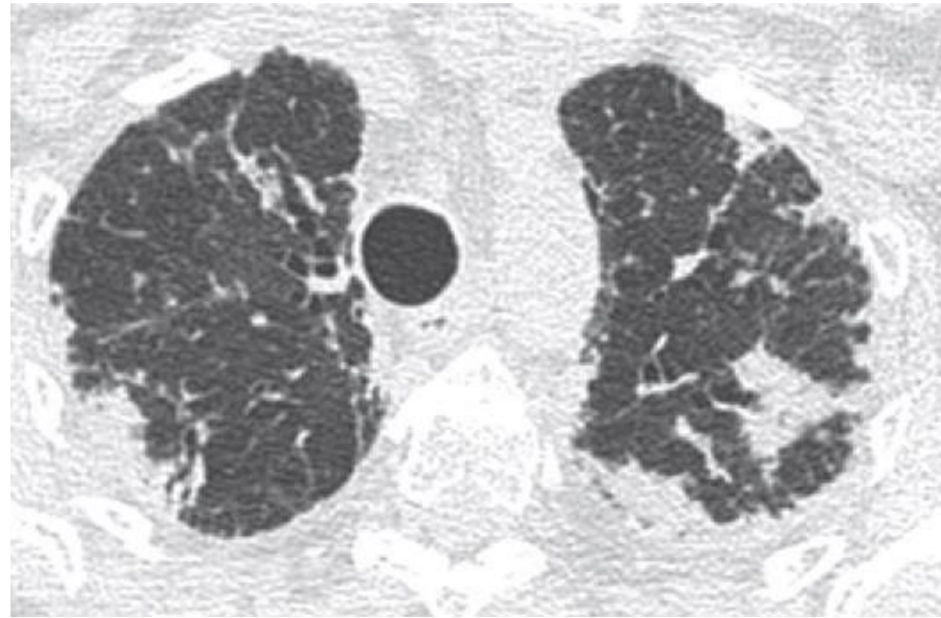


**Straight-edge sign**



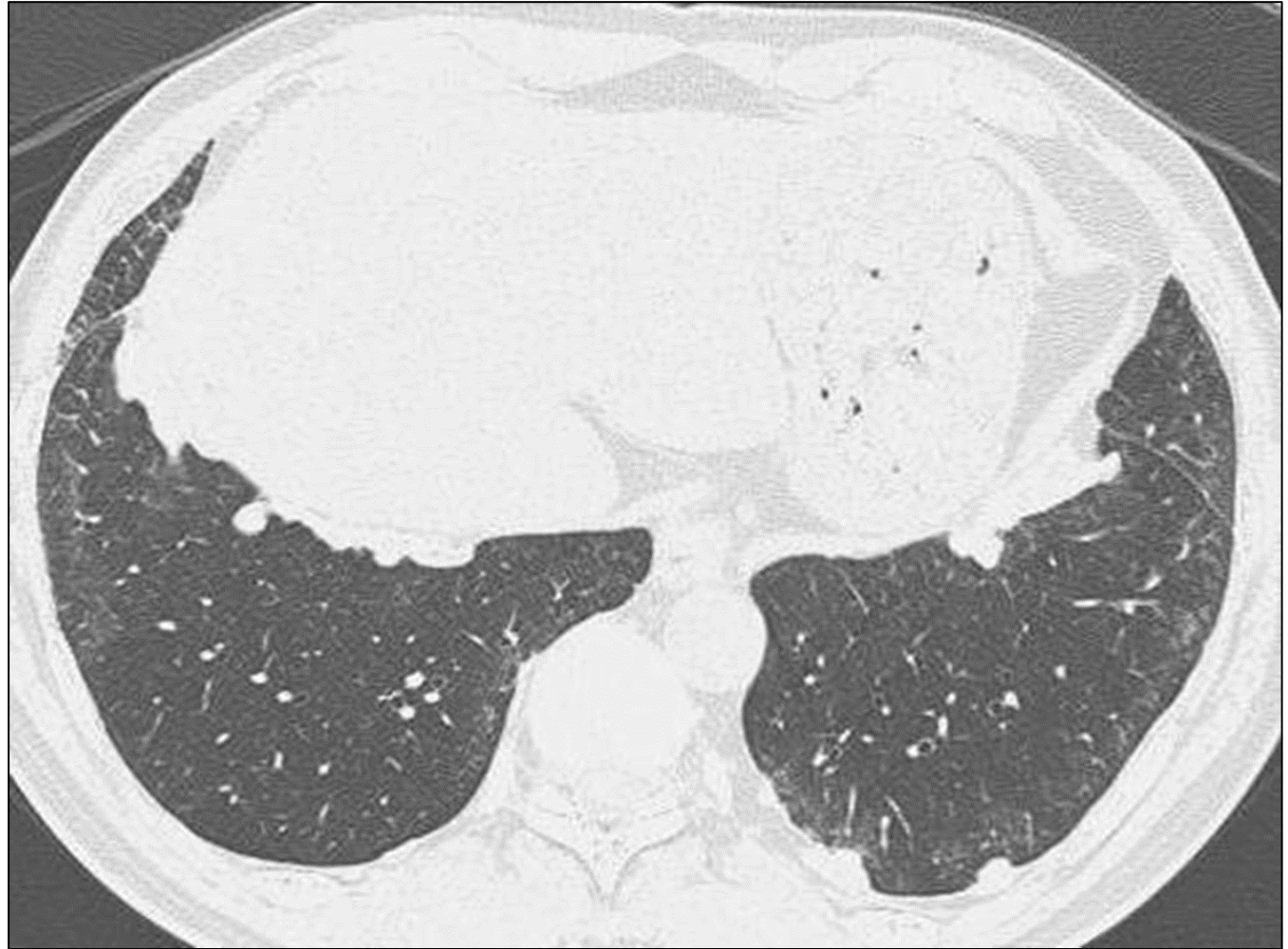
# Pleuroparenchymal fibroelastosis (PPFE)

- A rare interstitial pneumonia
- Irregular (visceral) pleural thickening and “tags” in the upper lungs (triangle)
- Merge with fibrotic changes in the subjacent lung
- Substantial upper lobes volume loss, architectural distortion
- Pneumothorax



# Asbestosis

- Exposure history
- Pleural plaques or diffuse pleural thickening
- Lower lung: subpleural dotlike or branching opacities, less coarse reticular shadow



# End-stage sarcoidosis

- Large cyst, Honeycomb-like cysts: middle and upper lungs
- Fibrotic distortion in peribronchovascular distribution
- Perilymphatic nodules



# Take-home message

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- **Lung fibrosis CT findings**
  - Reticular opacity, traction bronchiectasis, honeycombing cyst, three density sign, non-emphysematous cysts
- **UIP pattern spectrum**
  - UIP, probable UIP, indeterminate UIP pattern definition
- **Alternative diagnosis**
  - NSIP, fibrotic HP, CTD-ILD, PPFE, asbestosis...

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**Thank you for your attentions**

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