
HRCT in ILD

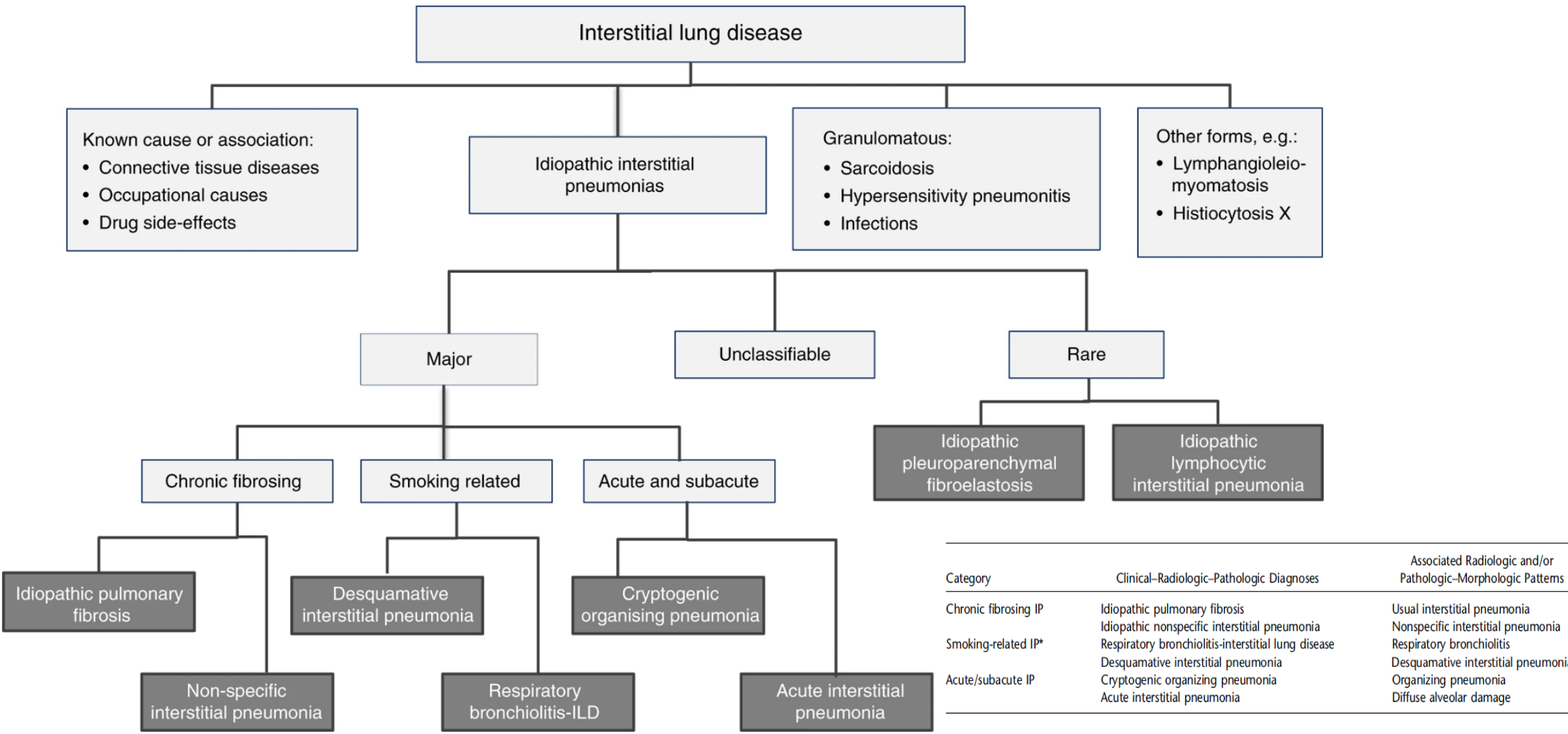
Recognizing Key Diagnostic Patterns

서울대학교병원 영상의학과 이종혁

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

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2013 ATS/ERS classification of idiopathic interstitial pneumonias

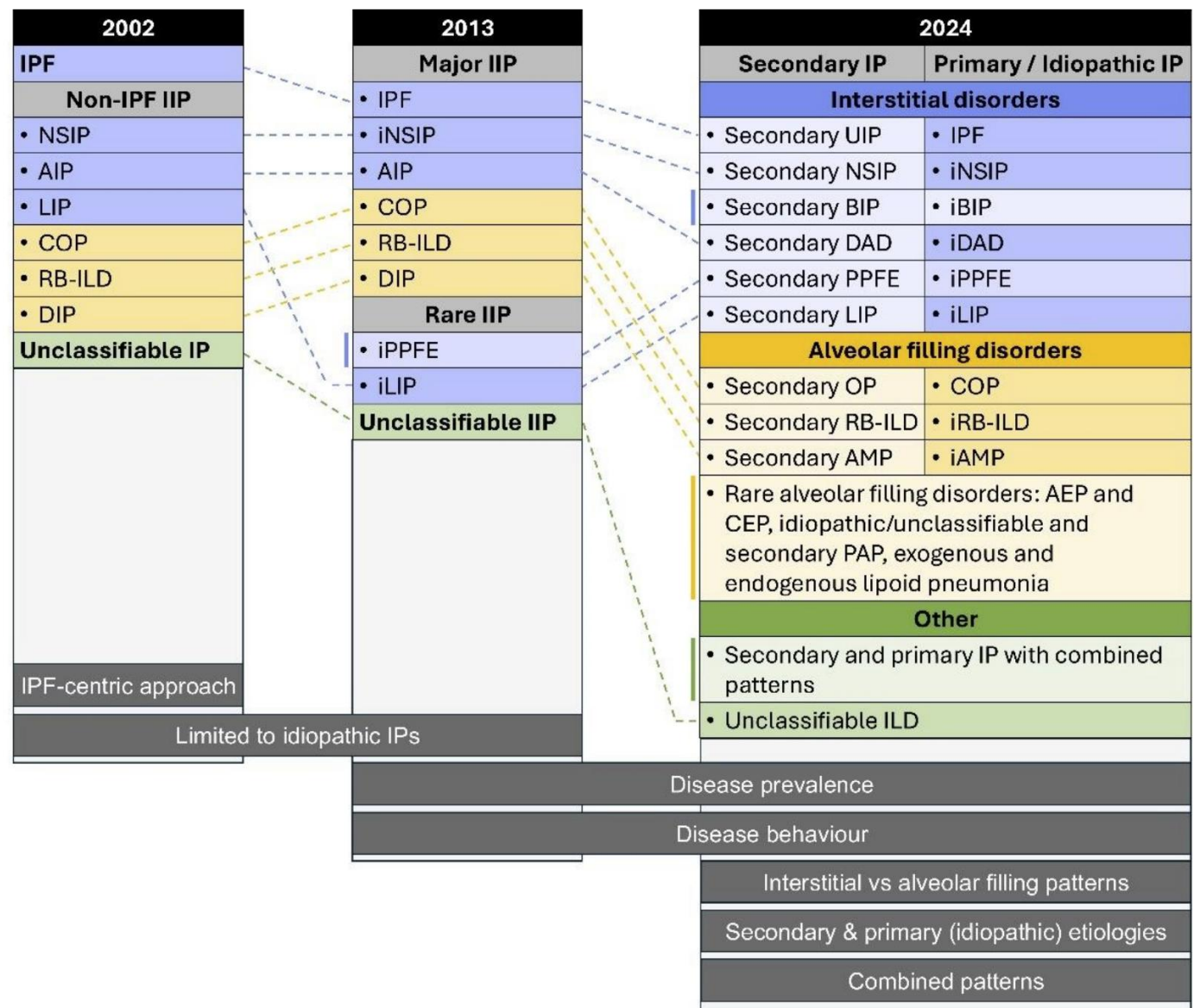


Category	Clinical–Radiologic–Pathologic Diagnoses	Associated Radiologic and/or Pathologic–Morphologic Patterns
Chronic fibrosing IP	Idiopathic pulmonary fibrosis Idiopathic nonspecific interstitial pneumonia	Usual interstitial pneumonia Nonspecific interstitial pneumonia
Smoking-related IP*	Respiratory bronchiolitis-interstitial lung disease Desquamative interstitial pneumonia	Respiratory bronchiolitis Desquamative interstitial pneumonia
Acute/subacute IP	Cryptogenic organizing pneumonia Acute interstitial pneumonia	Organizing pneumonia Diffuse alveolar damage

2024 ERS/ATS Multidisciplinary Classification of interstitial pneumonias

Morphologic patterns by Pathology and Imaging		Major clinical-radiologic-pathologic diagnoses	
		Secondary	Primary / Idiopathic
Interstitial patterns	Usual interstitial pneumonia (UIP)	Secondary UIP (e.g., CTD, HP, medications)	Idiopathic pulmonary fibrosis (Idiopathic UIP)
	Nonspecific interstitial pneumonia (NSIP)	Secondary NSIP (e.g., CTD >>> HP, medications)	Idiopathic NSIP
	Bronchiolocentric interstitial pneumonia (BIP)*	Secondary BIP (e.g., HP >>> CTD, aspiration, inhalational exposures, medications)	Idiopathic BIP (provisional diagnosis*)
	Diffuse alveolar damage (DAD)	Secondary DAD (multiple causes)	Idiopathic DAD (acute interstitial pneumonia)
	Pleuroparenchymal fibroelastosis (PPFE)	Secondary PPFE (e.g., IPF, CTD, HP, medications, radiation, transplant [restrictive allograft syndrome, pulmonary infection [post-tuberculosis], occupational)	Idiopathic PPFE
	Lymphoid interstitial pneumonia (LIP)	Secondary LIP (e.g., CTD, immune deficiency)	Idiopathic LIP
Alveolar filling patterns	Organising pneumonia (OP)	Secondary OP (e.g., CTD, post-infectious, medications, aspiration)	Cryptogenic OP (Idiopathic OP)
	Respiratory bronchiolitis-ILD (RB-ILD)	Secondary RB-ILD (e.g., smoking >>> CTD, medications, aspiration, hereditary)	Idiopathic RB-ILD
	Alveolar macrophage pneumonia [†] (AMP)	Secondary AMP (e.g., smoking >>> CTD, medications, aspiration, hereditary)	Idiopathic AMP
	Rare alveolar filling disorders	e.g., Acute and chronic eosinophilic pneumonia, pulmonary alveolar proteinosis, lipid pneumonia (See Supplemental Table 1)	
Other	Combined pattern	Multiple combinations (e.g., NSIP + OP, UIP + PPFE)	
	Unclassifiable pattern	Unclassifiable ILD (multiple undefined patterns)	

2024 ERS/ATS Multidisciplinary Classification of interstitial pneumonias



Interstitial lung abnormality

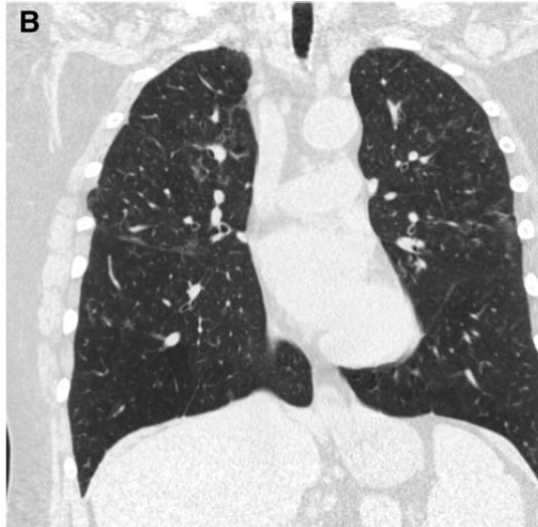
Chest CT showing bilateral and nondependent ground-glass opacities, reticular abnormalities, lung distortion, traction bronchiectasis, and/or honeycombing involving $\geq 5\%$ of a lung zone*

Definition of interstitial lung disease for those with ILAs

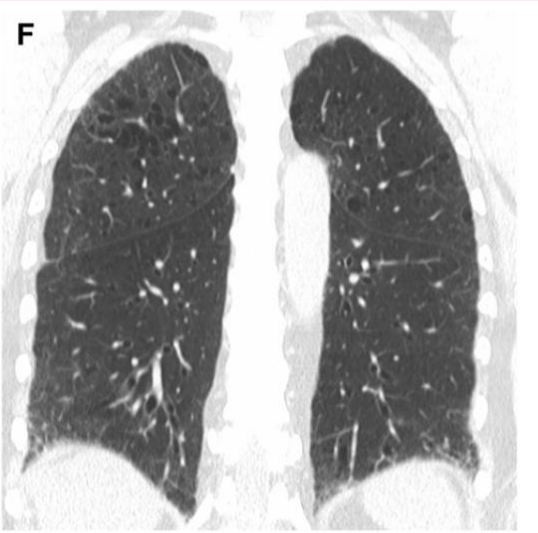
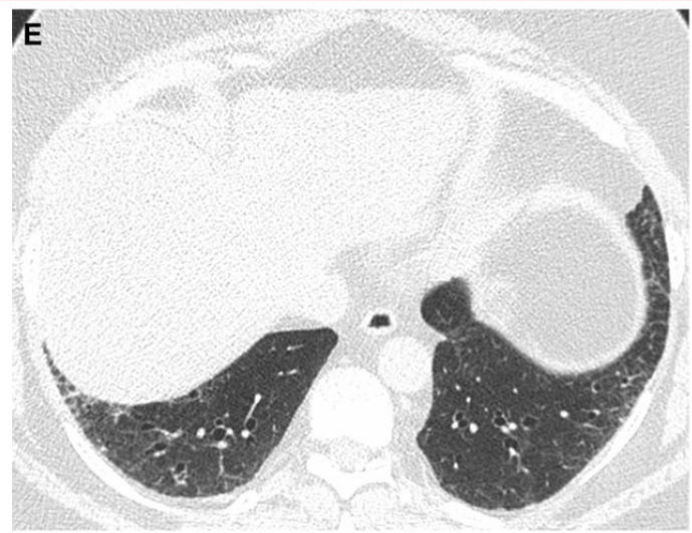
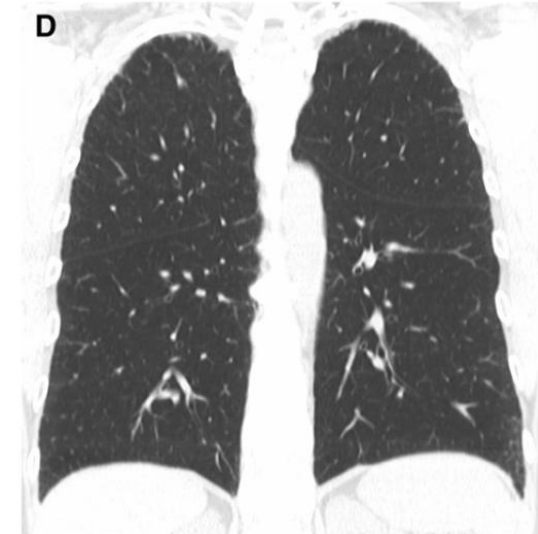
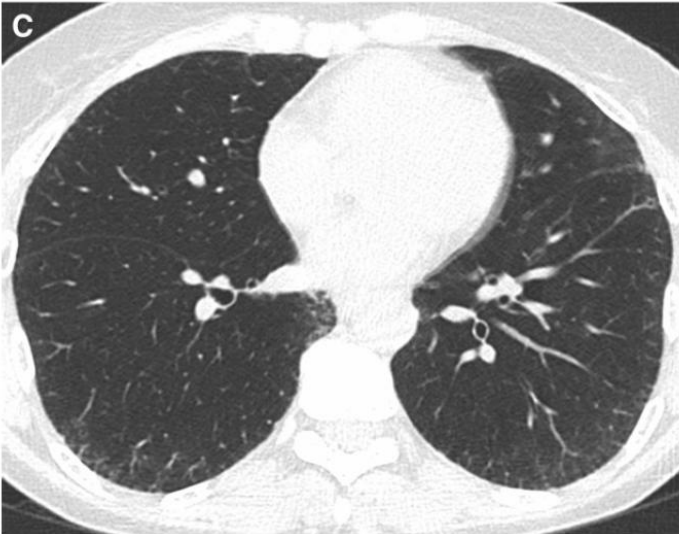
In a person with CT features of ILAs, at least one of the following criteria must be present to define ILD*

- **Symptoms:** Any amount of dyspnea and/or cough that a clinician attributes to ILD
- **Physiology** (any of)
 - Any abnormality in FVC, TLC, or DL_{CO} that a clinician attributes to ILD (defined as a value or z-score below the lower limit of normal)
 - Satisfies physiologic criteria for progressive pulmonary fibrosis that a clinician attributes to ILD (9)
- **Imaging** (any of the following on chest CT)
 - Fibrotic abnormalities (honeycombing and/or reticulation with traction bronchiectasis) involving $\geq 5\%$ of total lung volume by visual estimate
 - Progressive fibrotic abnormality on serial chest CT
 - Presence of a major fibrotic ILD pattern on chest CT (i.e., UIP/probable UIP, fibrotic HP, or fibrotic NSIP)
- **Pathology:** Presence of a major fibrotic ILD pattern (i.e., UIP/probable UIP, fibrotic HP, or fibrotic NSIP)

Interstitial lung abnormality



Nonsubpleural ILA



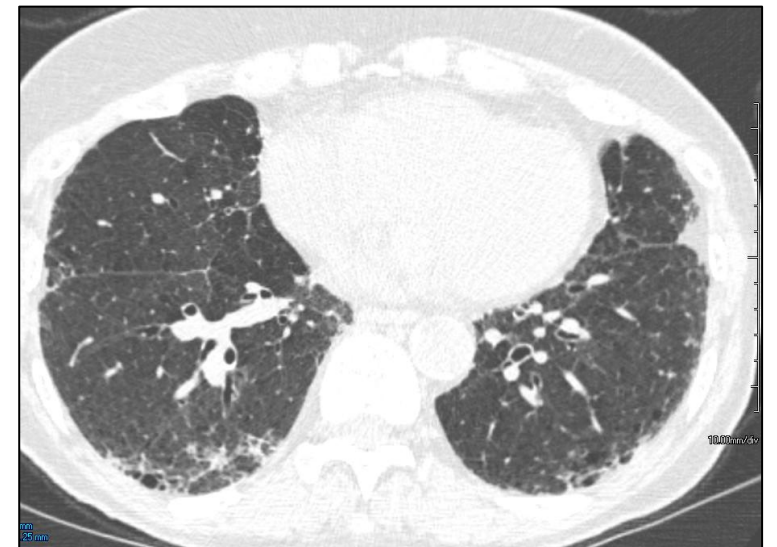
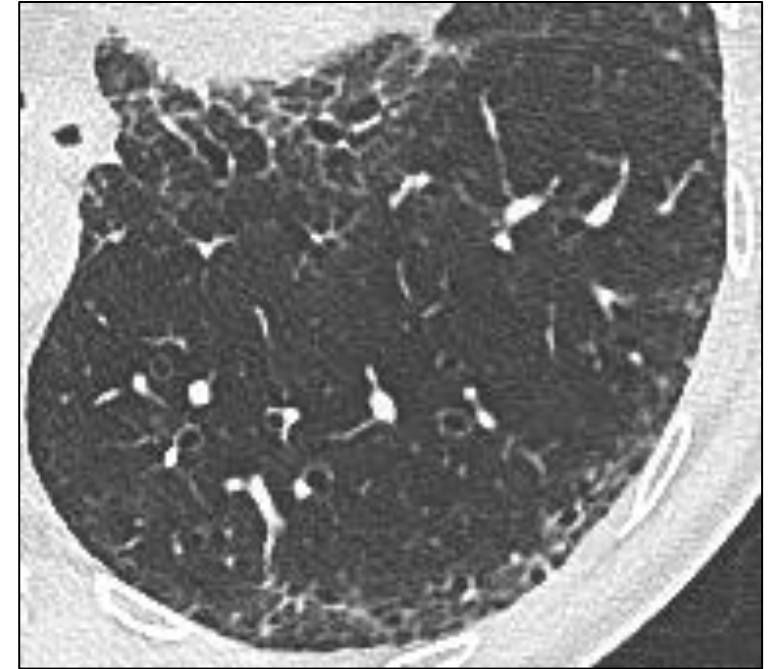
Subpleural fibrotic ILA

Subpleural nonfibrotic ILA

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

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



Reticular opacity

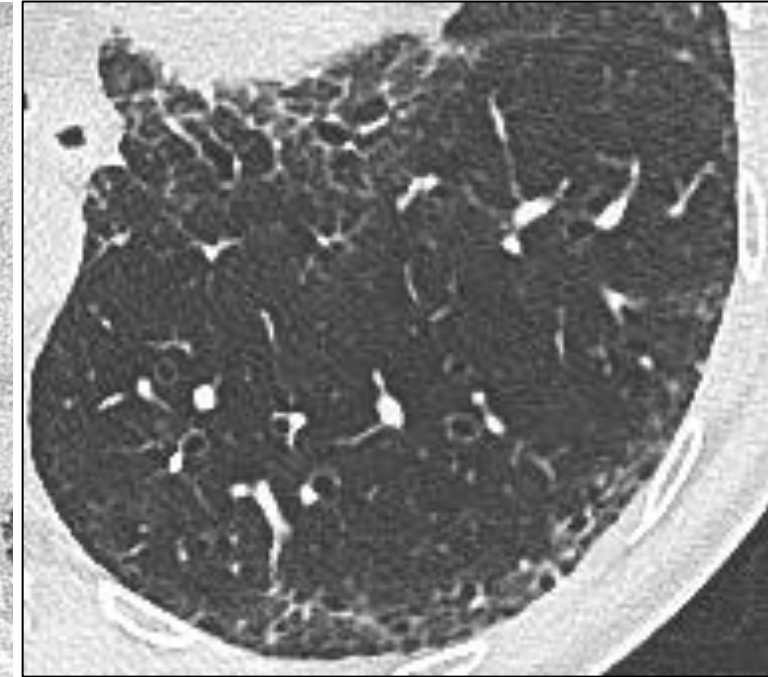
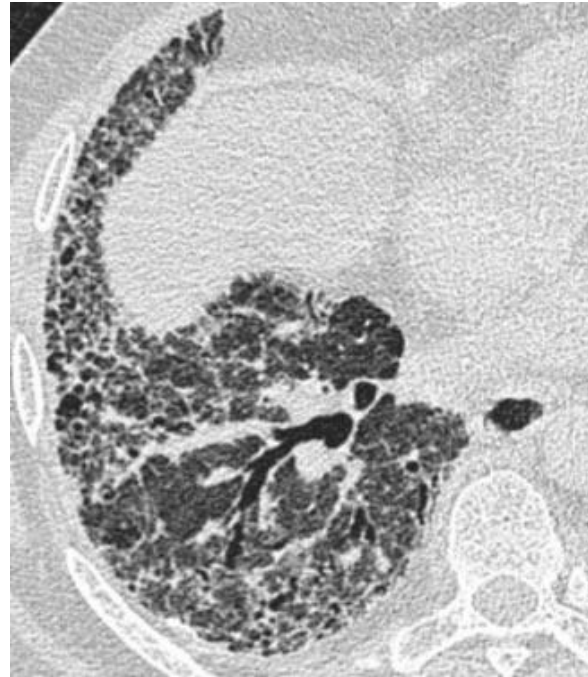
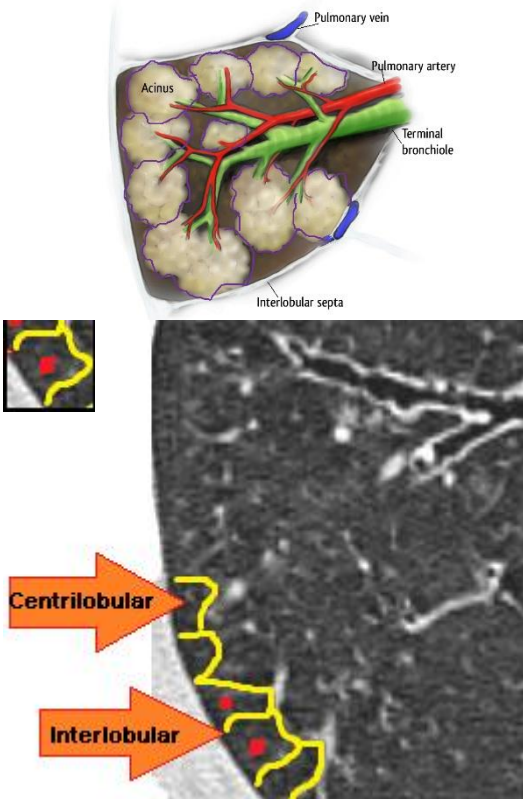
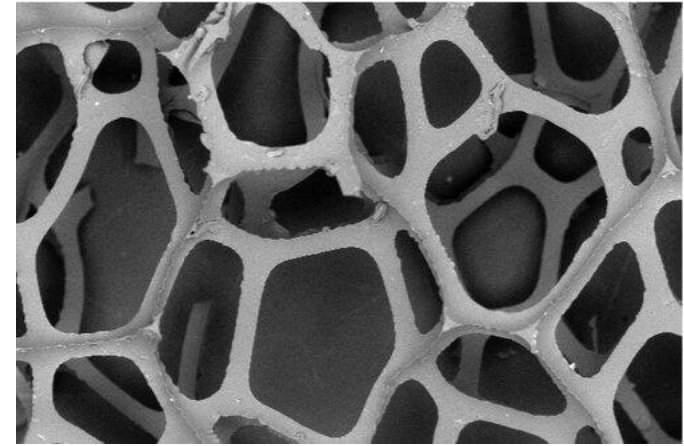
reticulation +

1. 명사 그물코, 망상 조직, 그물 세공(network)
2. 명사 사진 그물 모양의 주름 ((감광 유제(乳劑)에 생기는))
3. 명사 수도관 망

- Innumerable small linear opacities resembling net

– Used in chest radiographs & chest CT

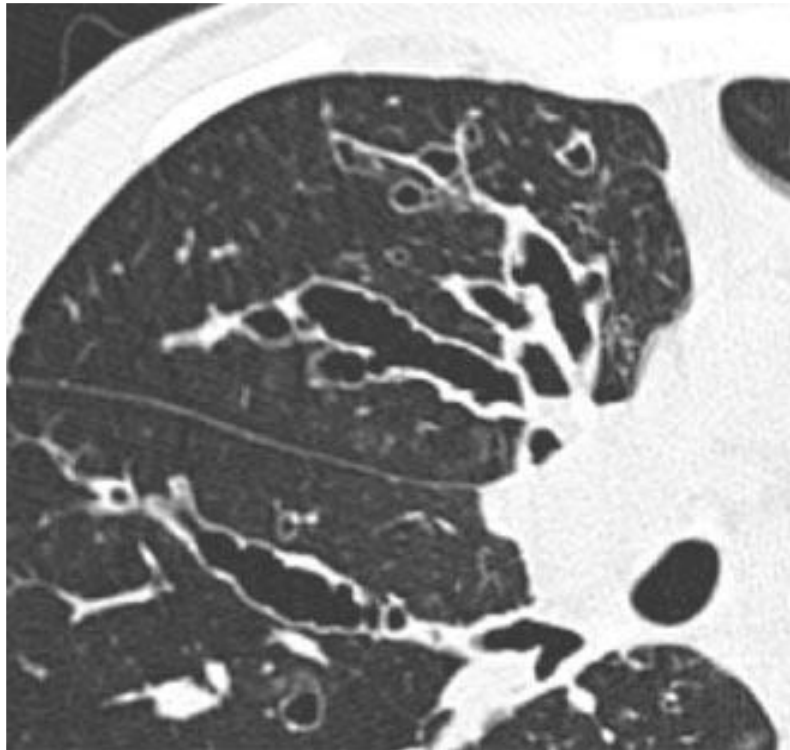
– **Fibrosis**: interlobular septal thickening or cyst walls of honeycombing cysts



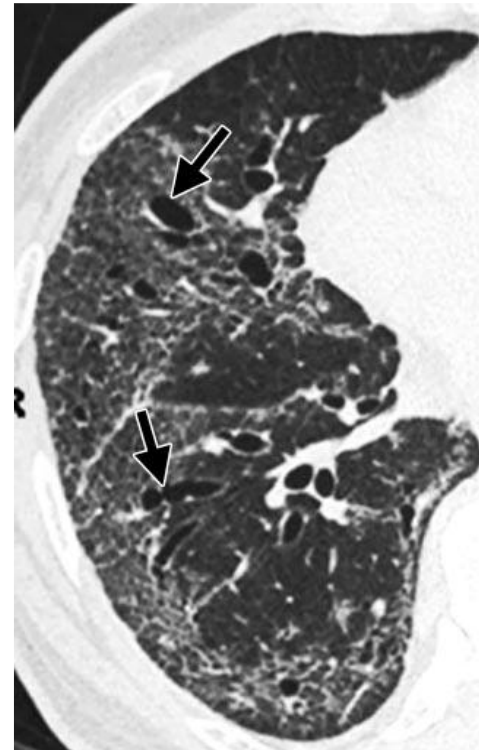
Traction bronchiectasis

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

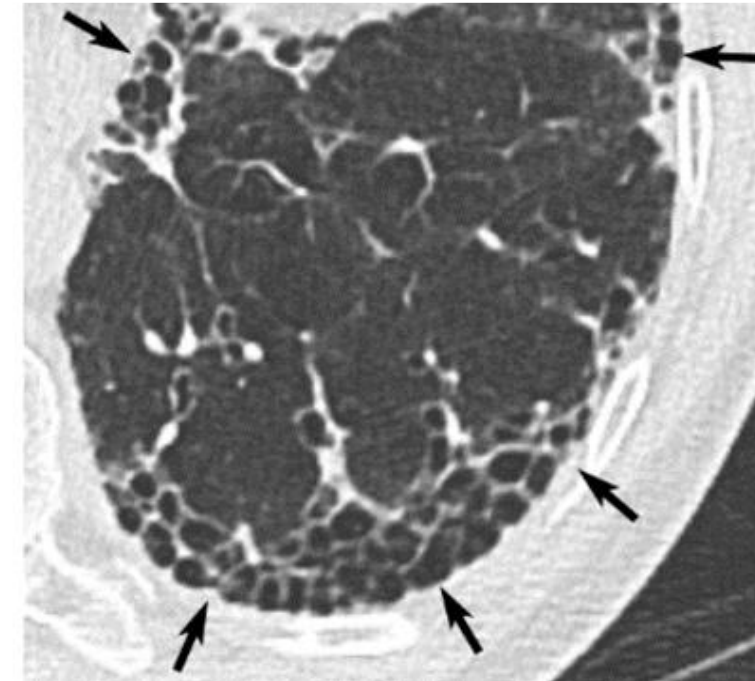
Hansell DM, et al. Radiology 2008
Travis WD et al. AJRCCM 2013

- **Pathology**

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

- **CT**

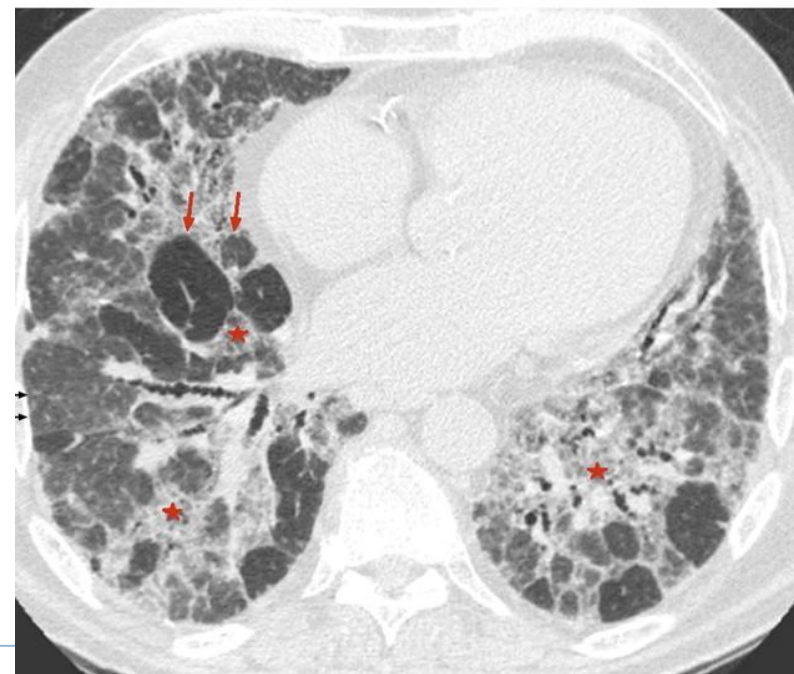
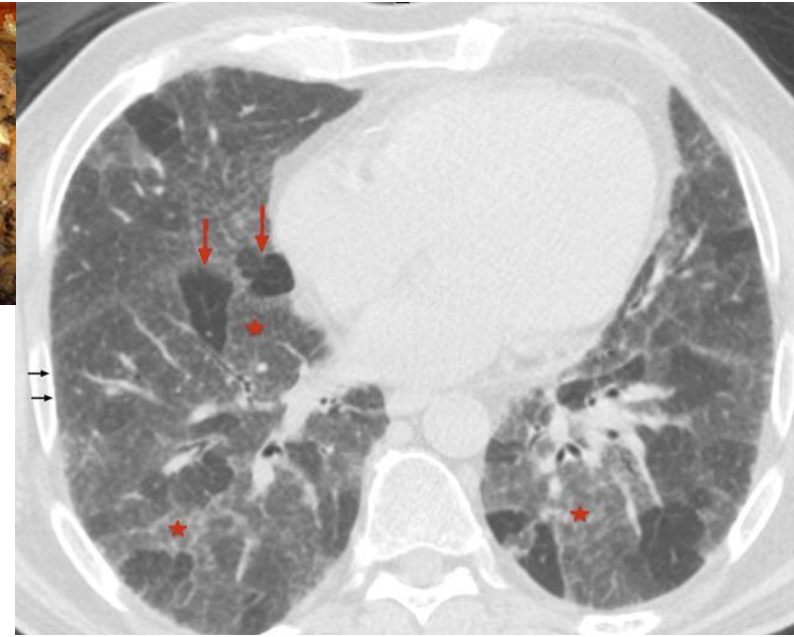
- **Clustered cystic air spaces**, typically 3-10 mm up to 2.5 cm
- Usually **subpleural, characterized by well-defined and shared walls**
- Important **criterion in the diagnosis of UIP**
- Increase in size and number during disease progression
- A single layer can be honeycombing cyst





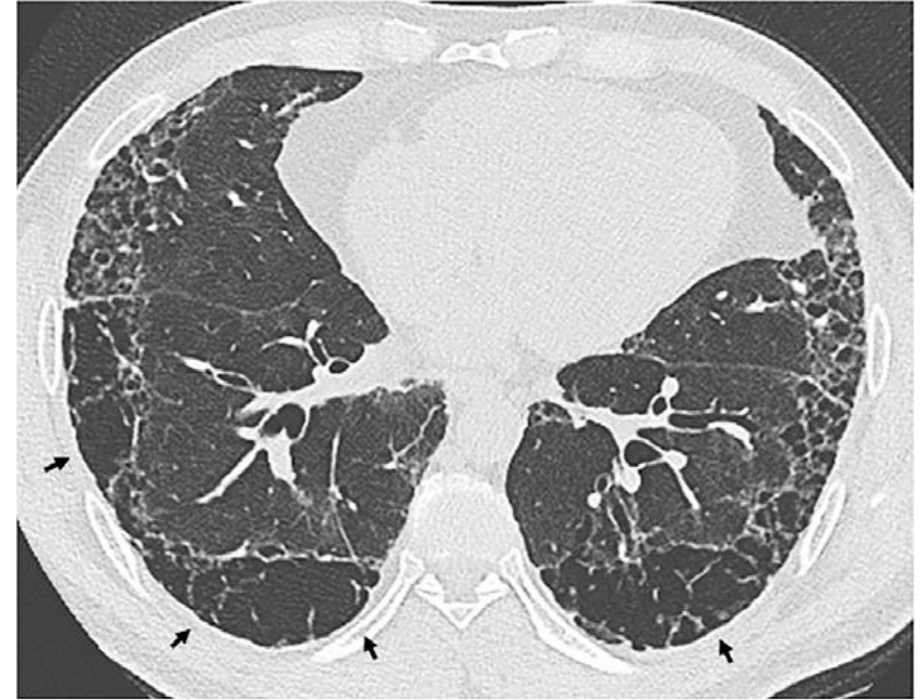
Three-density pattern

- **Combination of three attenuation**
 - Normal appearing lung
 - High attenuation (GGO)
 - Hypoattenuating area (air-trapping & associated decreased vascularity)
- Sharply demarcated from each other
- Headcheese sign
- **Highly specific for fibrotic HP**



Non-emphysematous cysts

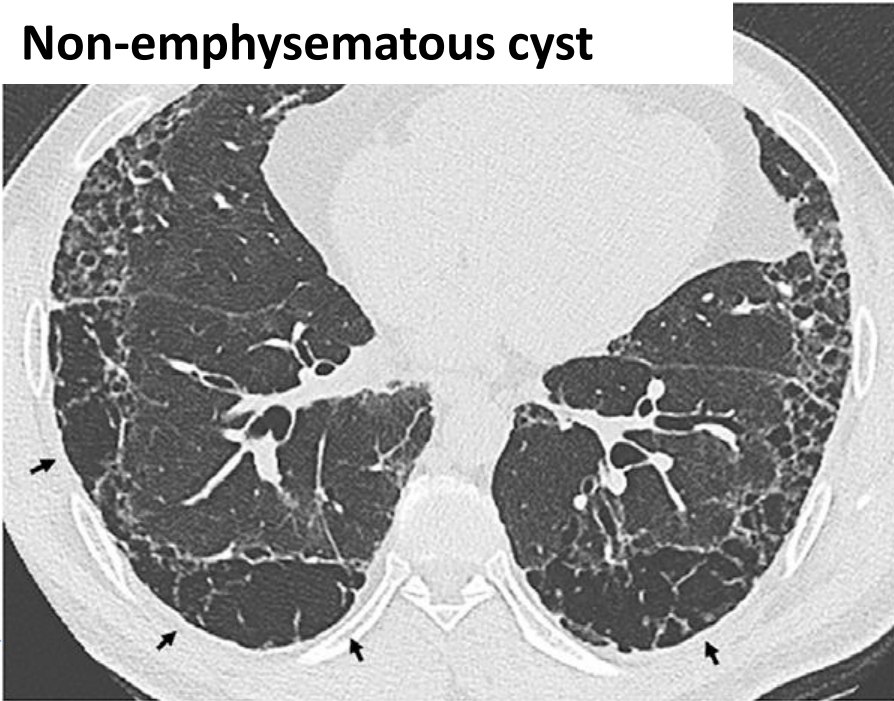
- Non-emphysematous cysts
 - SRIF (smoking related interstitial fibrosis)
 - Airspace enlargement with fibrosis
 - RB-ILD with fibrosis
 - Respiratory bronchiolitis with fibrosis
- Pathology: fibrotic alveolar septal thickening with hyalinized collagen bundles and hyperplastic smooth muscle
- **CT: Cysts with irregular, well-defined (=1mm) walls; juxtapleural sparing (upper lateral and lower dorsal)**



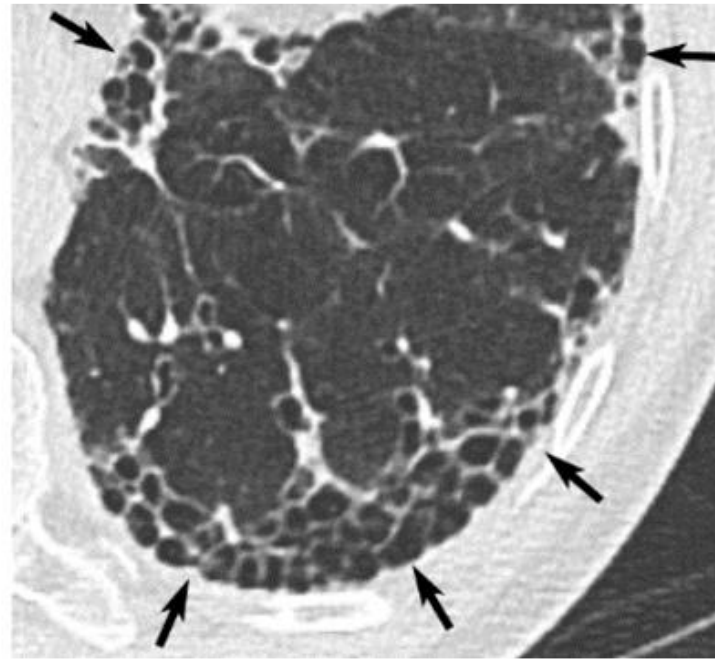
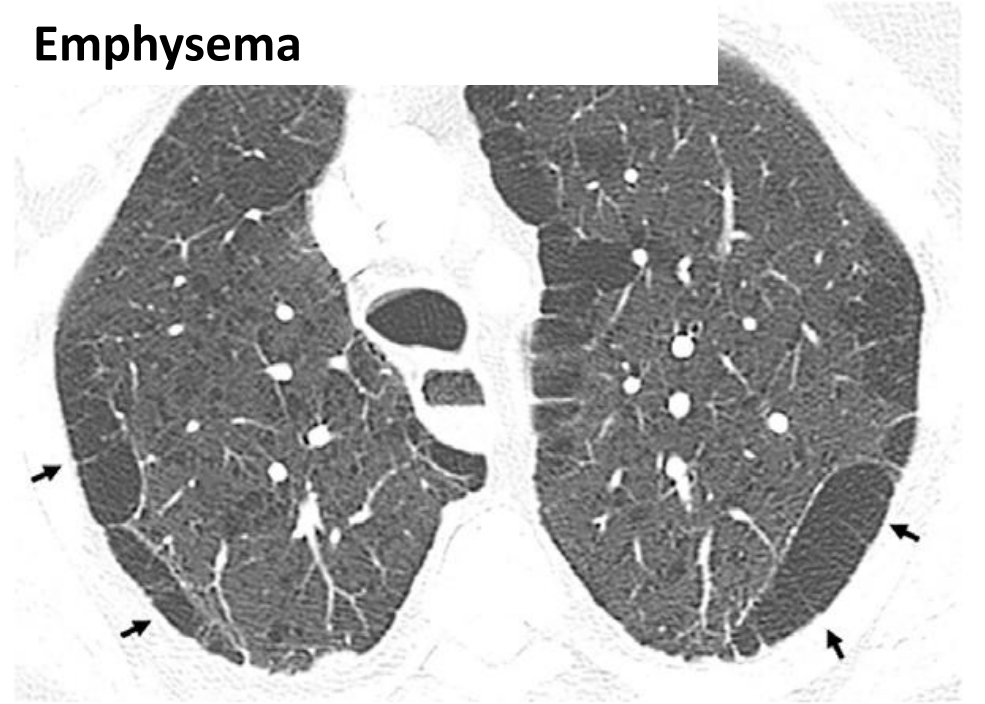
Non-emphysematous cysts

- DDx from emphysema: thin or indiscernible wall; absence of GGO, reticular opacity
- DDx from honeycombing: thick wall, uniform cyst; subpleural predominance, no interrupted septa

Non-emphysematous cyst



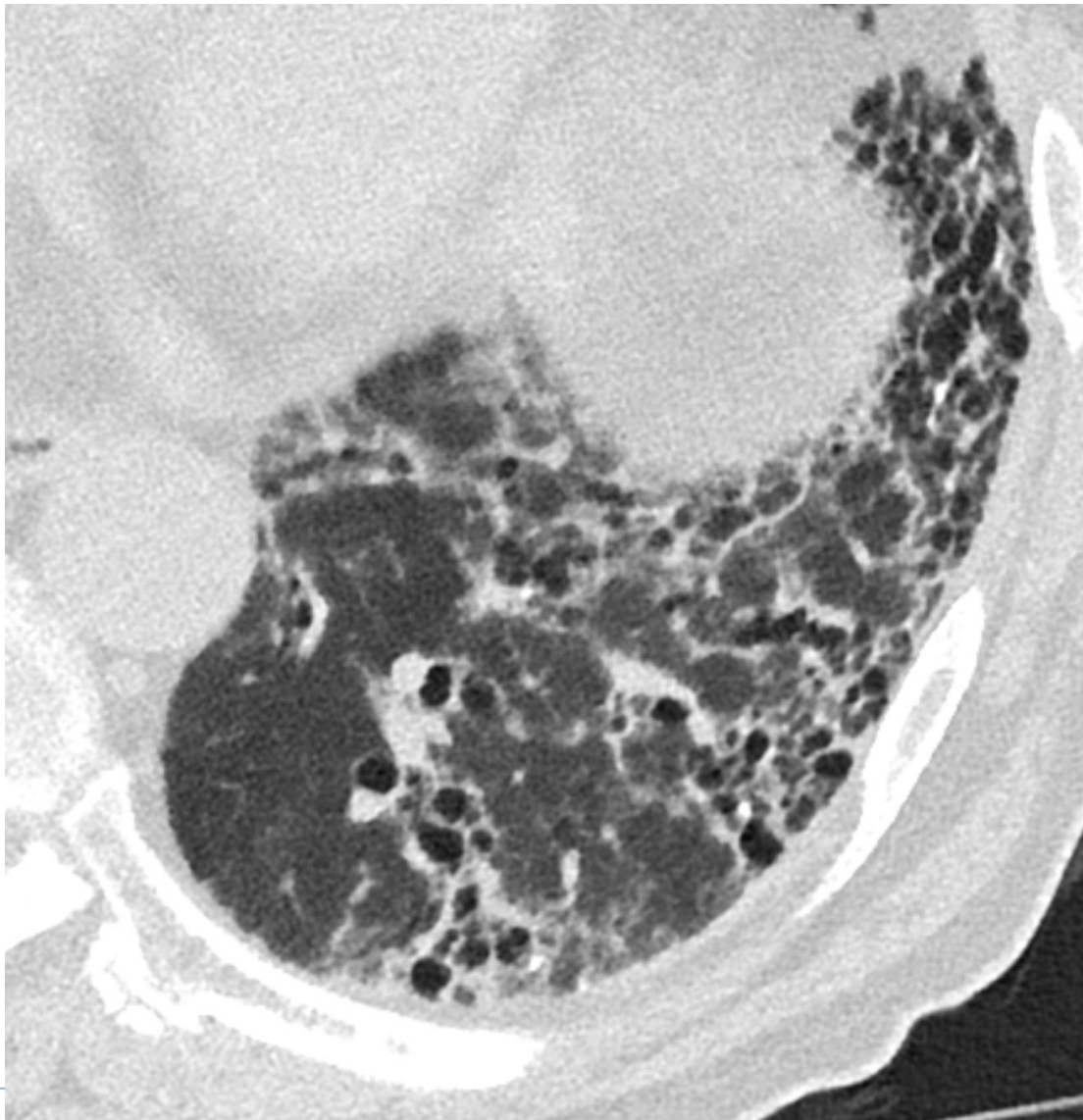
Emphysema



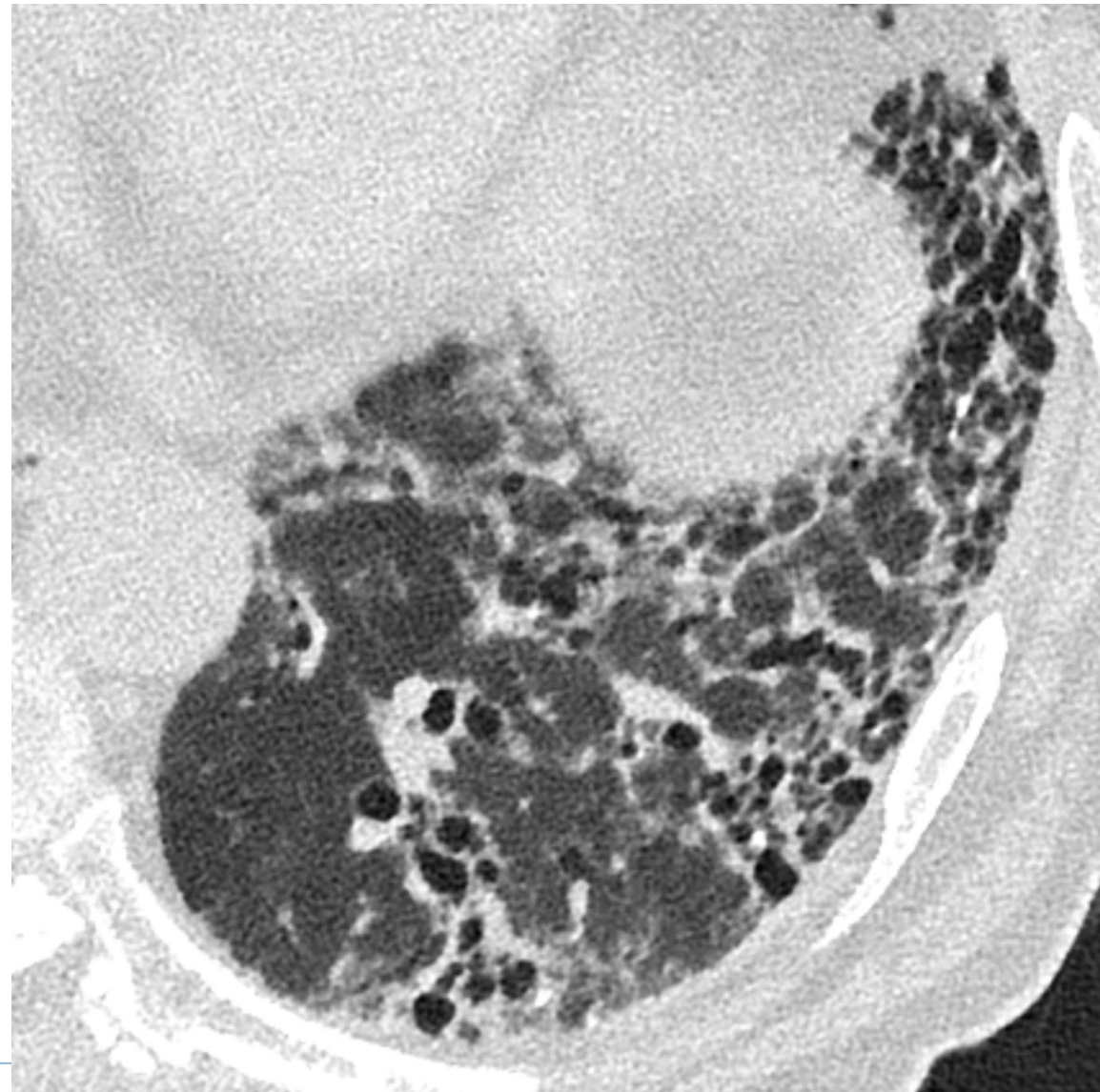
Honeycombing
cyst

Photon-counting CT for ILD

70keV, Br64, QIR 3, 1mm, 1024x

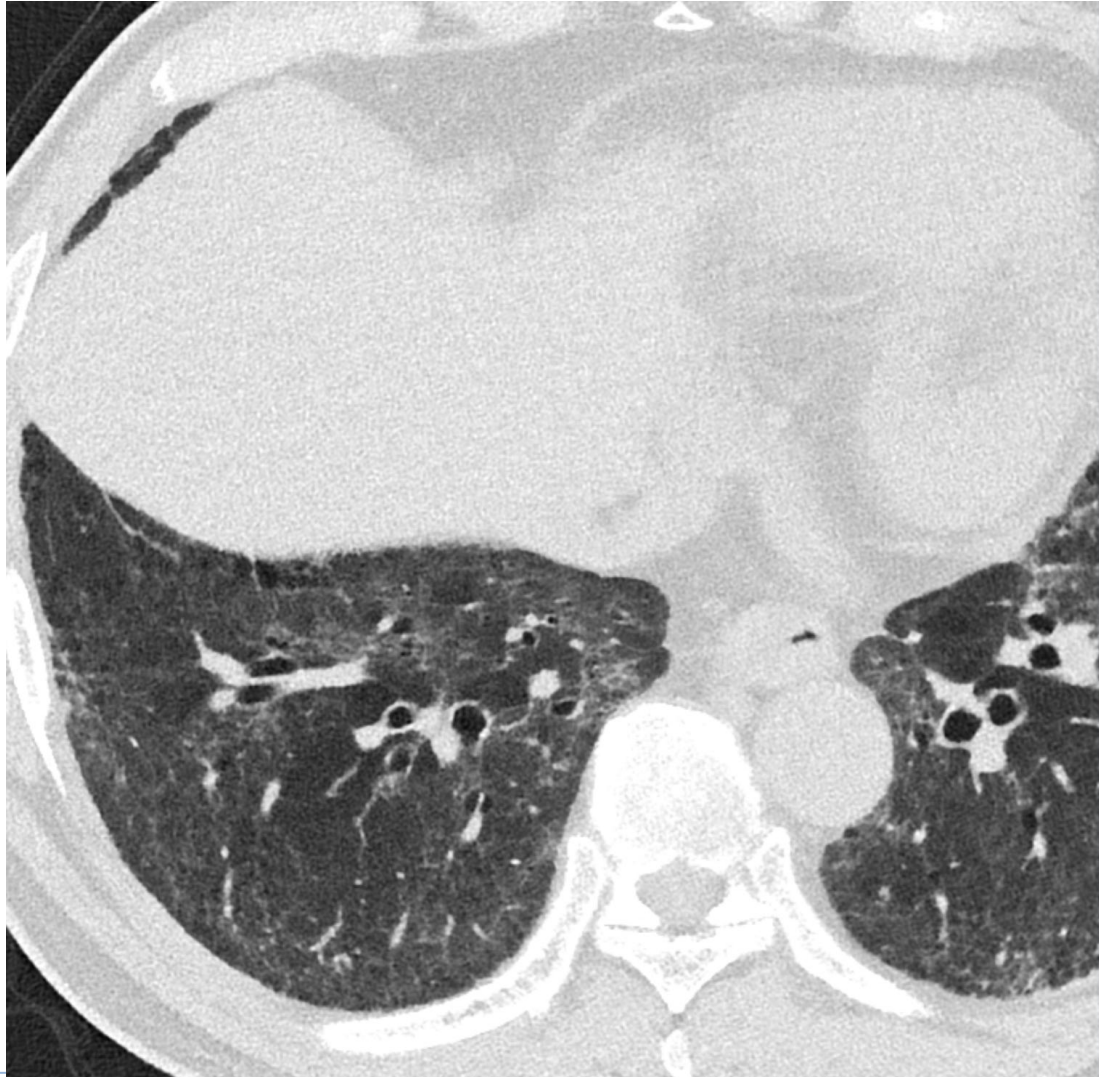


70keV, Br64, QIR 3, 0.4mm, 1024x



Photon-counting CT for ILD

70keV, Br64, QIR 3, 1mm, 1024x

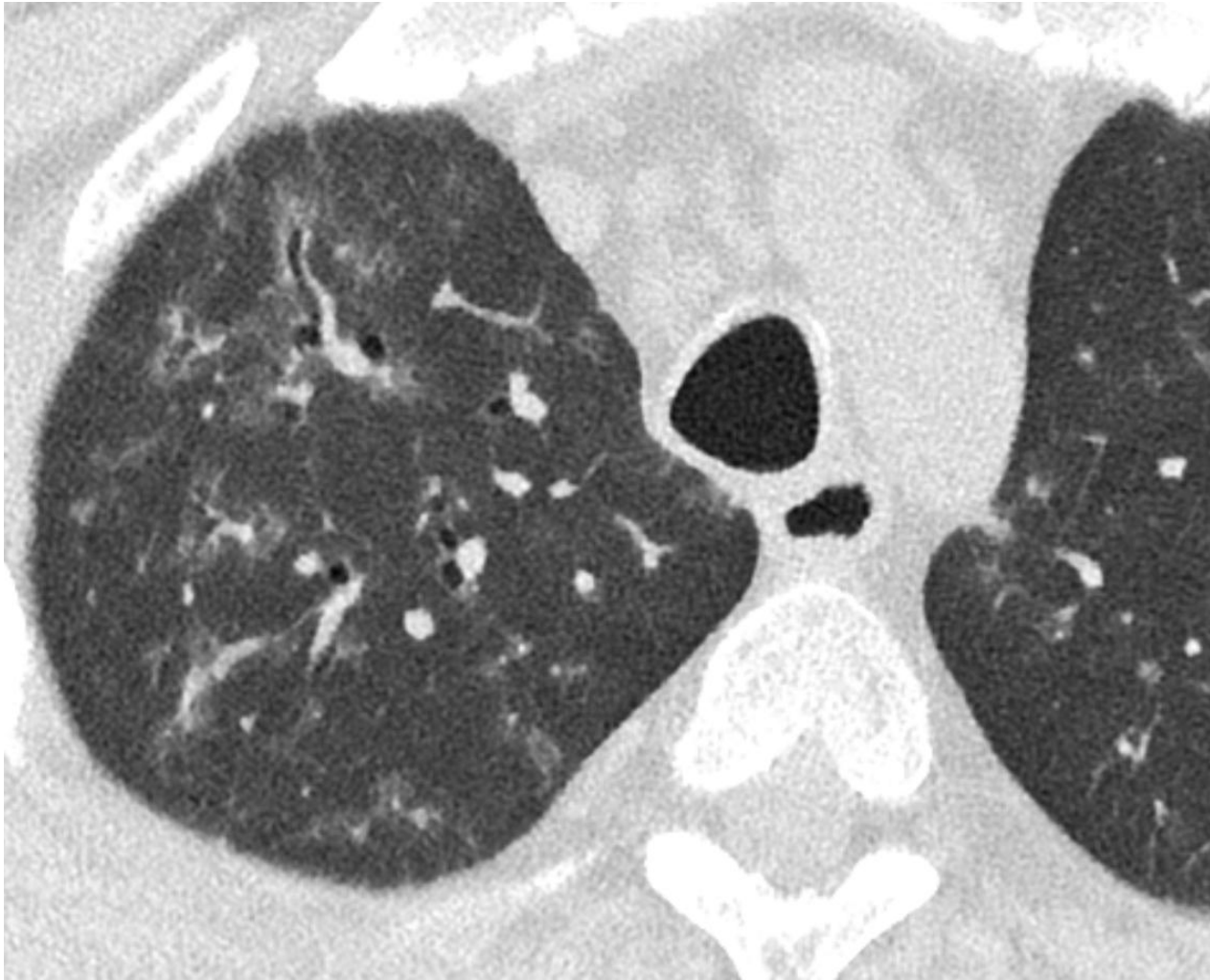


120kV, YC, iDose(4), 1mm, 512x

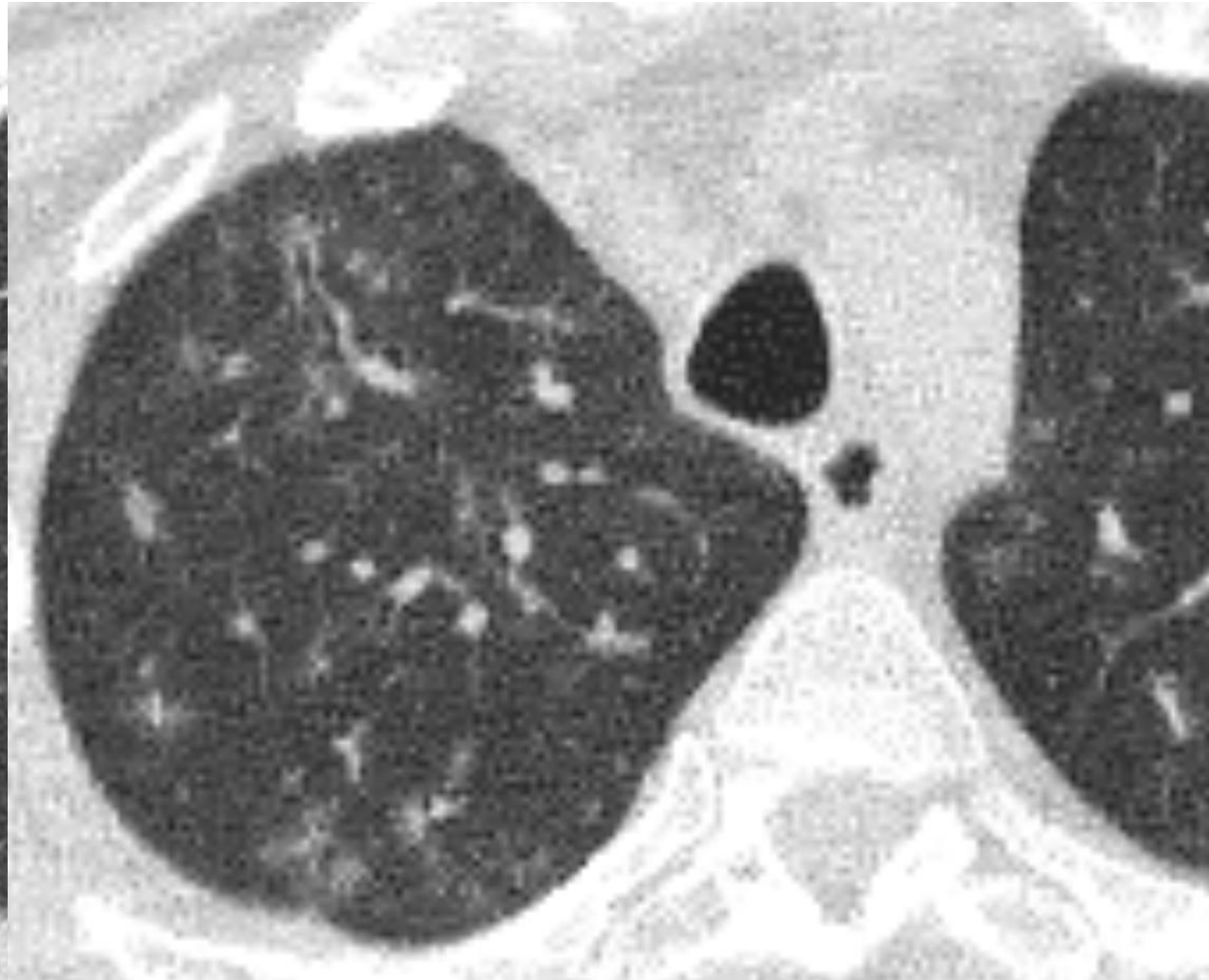


Photon-counting CT for ILD

70keV, Br64, QIR 3, 1mm, 1024x



120kV, Bone, AR50%, 1.25mm, 512x

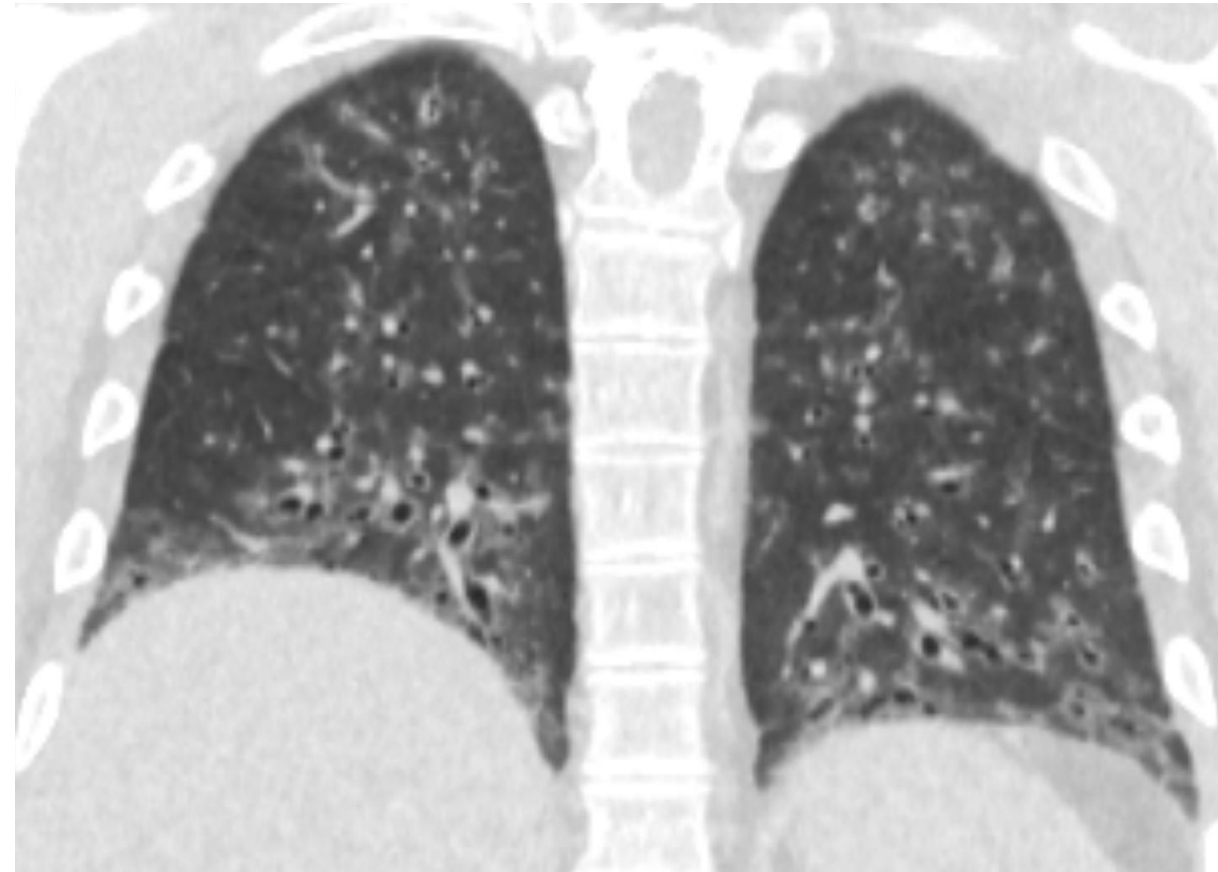


Photon-counting CT for ILD

70keV, Br64, QIR 3, 1mm, 1024x



120kV, Bone, AR50%, 1.25mm, 512x



차례

- ILD Classification & ILA
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Usual interstitial pneumonia (UIP) pattern on CT

Ragu G, et al. AJRCCM 2018 and 2022

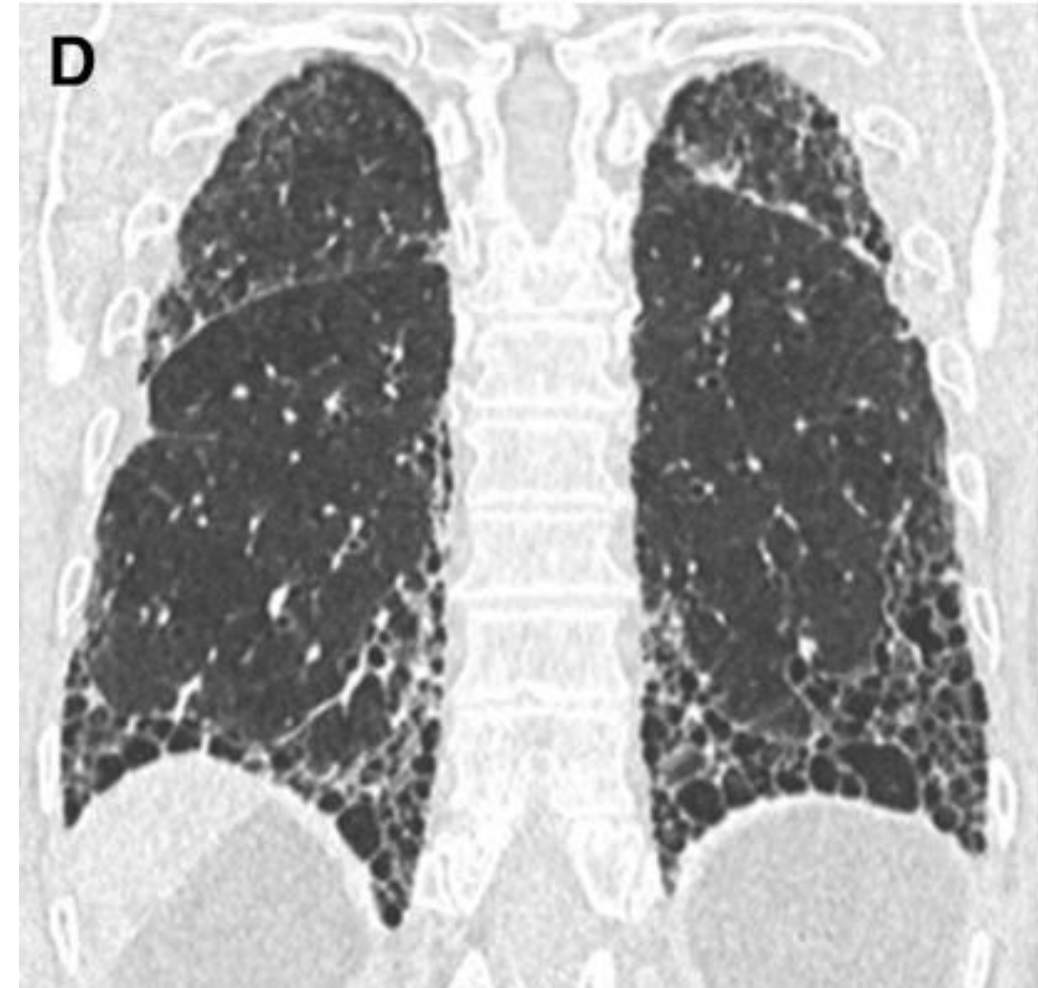
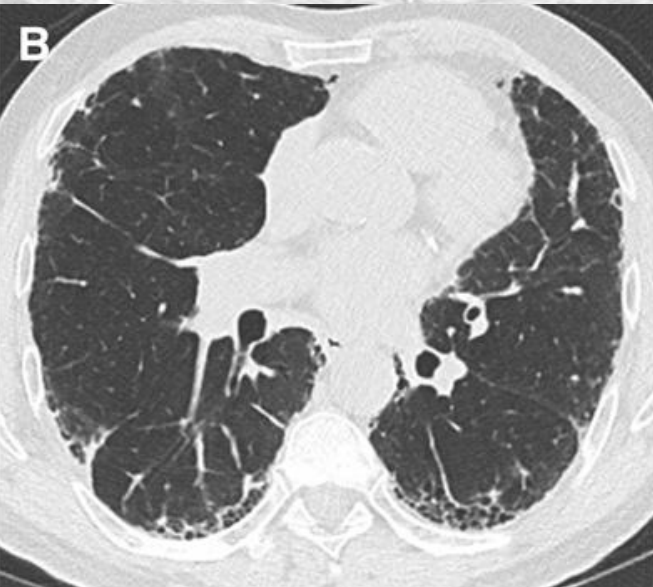
	HRCT Pattern			CT Findings Suggestive of an Alternative Diagnosis
	UIP Pattern	Probable UIP Pattern	Indeterminate for UIP	
Level of confidence for UIP histology	Confident (>90%)	Provisional high confidence (70–89%)	Provisional low confidence (51–69%)	Low to very low confidence (≤50%)
Distribution	<ul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

Usual interstitial pneumonia (UIP) pattern CT



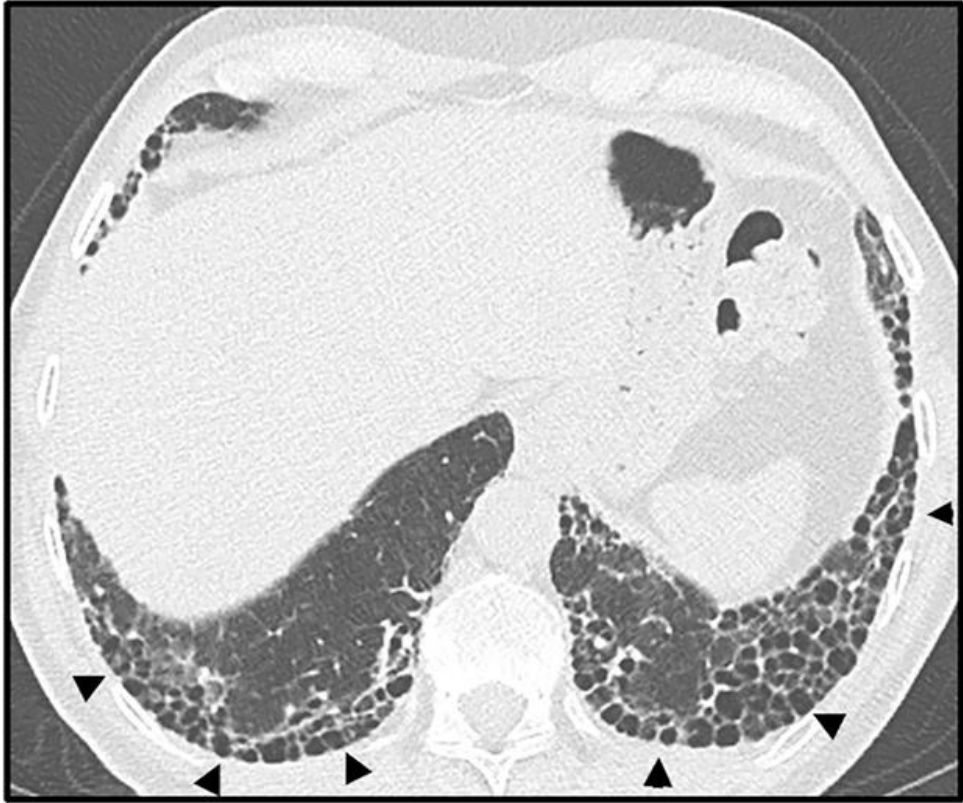
Reticular opacity

Traction
bronchiectasis +
Honeycombing



Subpleural and basal predominance

Usual interstitial pneumonia (UIP) pattern CT



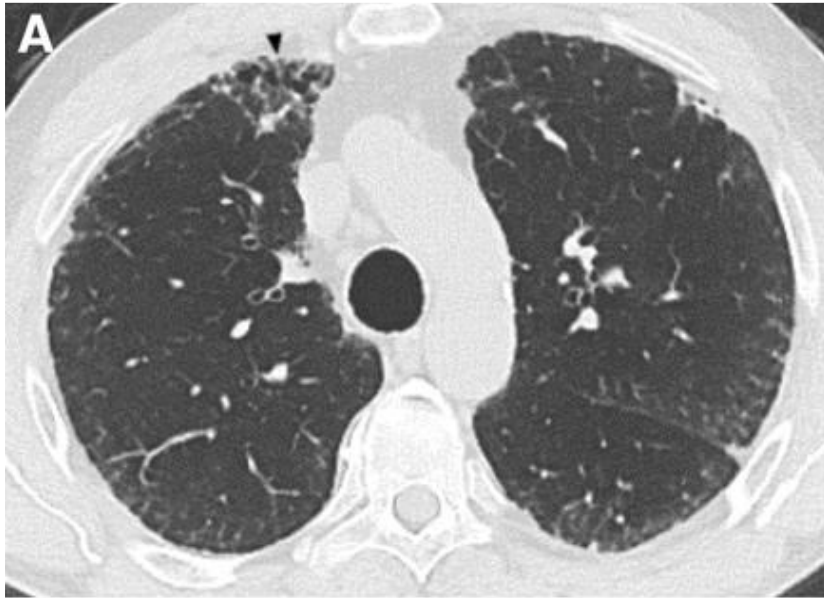
Traction bronchiectasis + Honeycombing



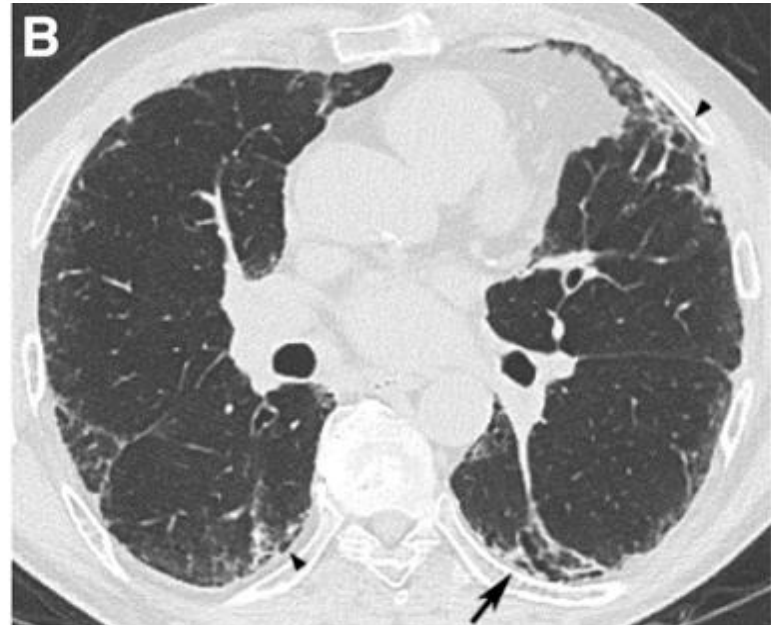
Subpleural and basal predominance



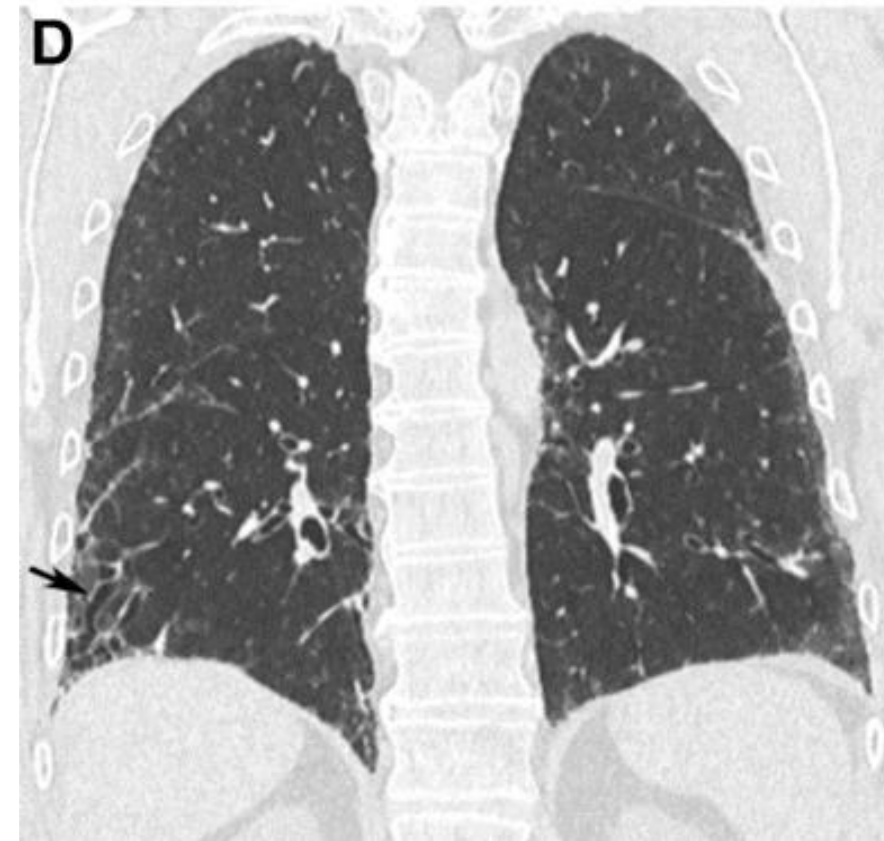
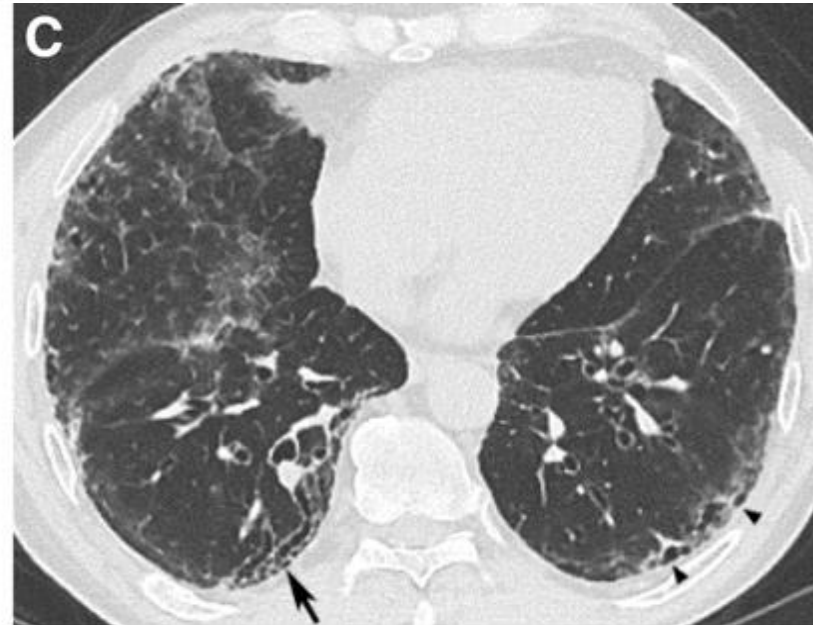
Probable usual interstitial pneumonia (UIP) pattern CT



Reticular opacity



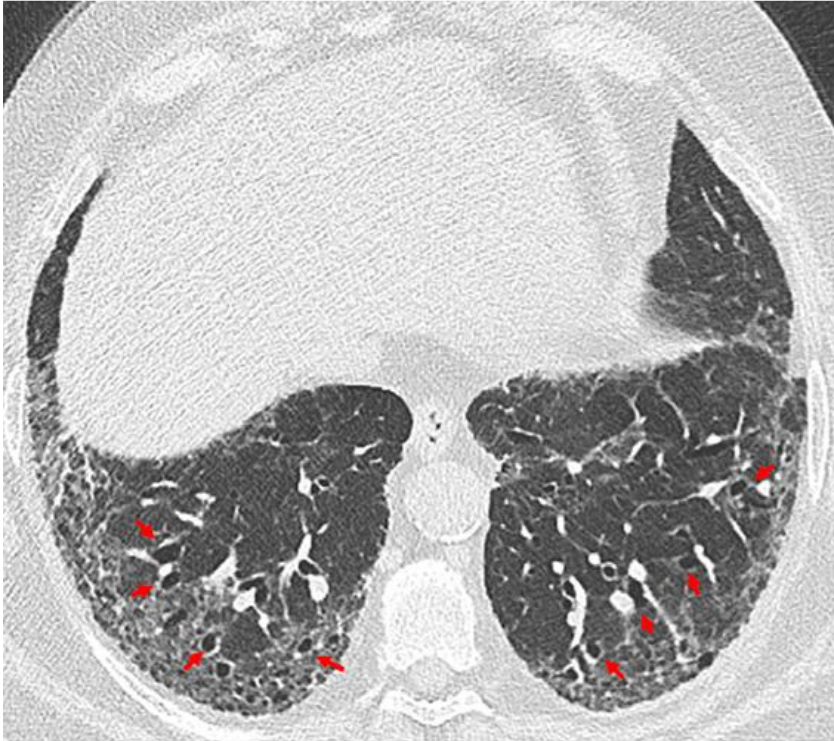
Traction bronchiectasis + mild GGO



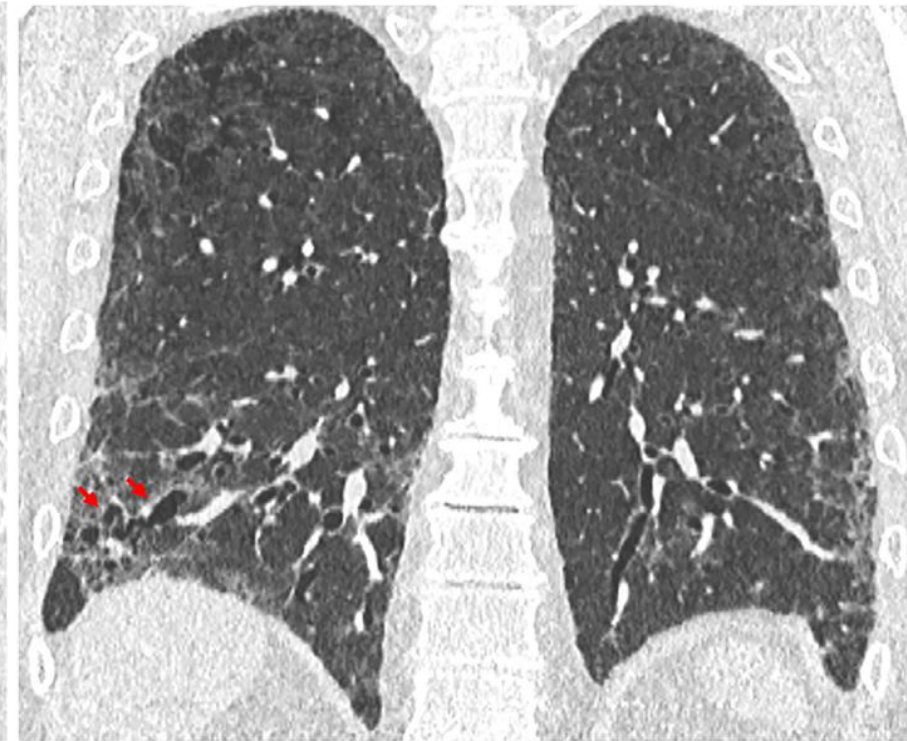
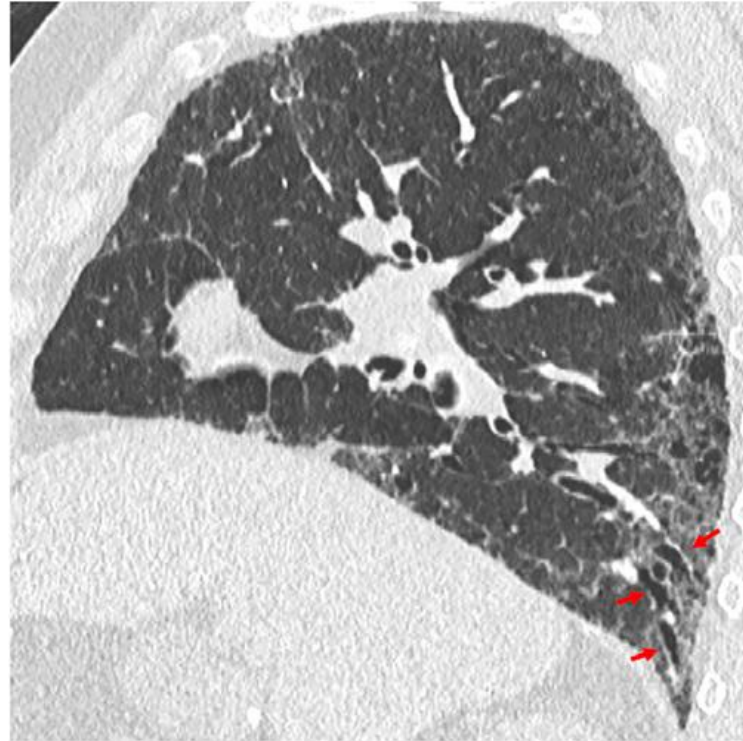
Subpleural and basal predominance

Probable usual interstitial pneumonia (UIP) pattern CT

Raghu G, et al. AJRCCM 2018 and 2022



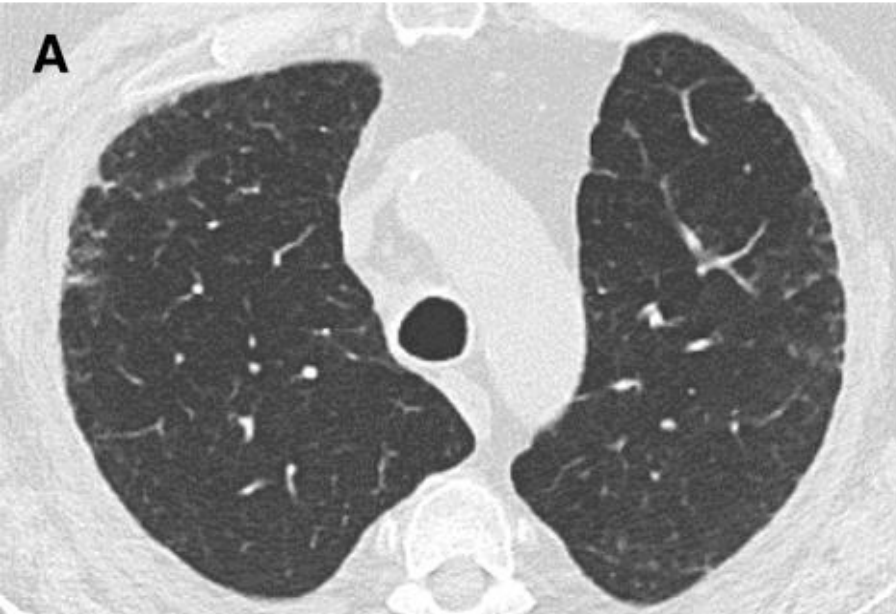
Reticular opacity + traction bronchiectasis + mild GGO



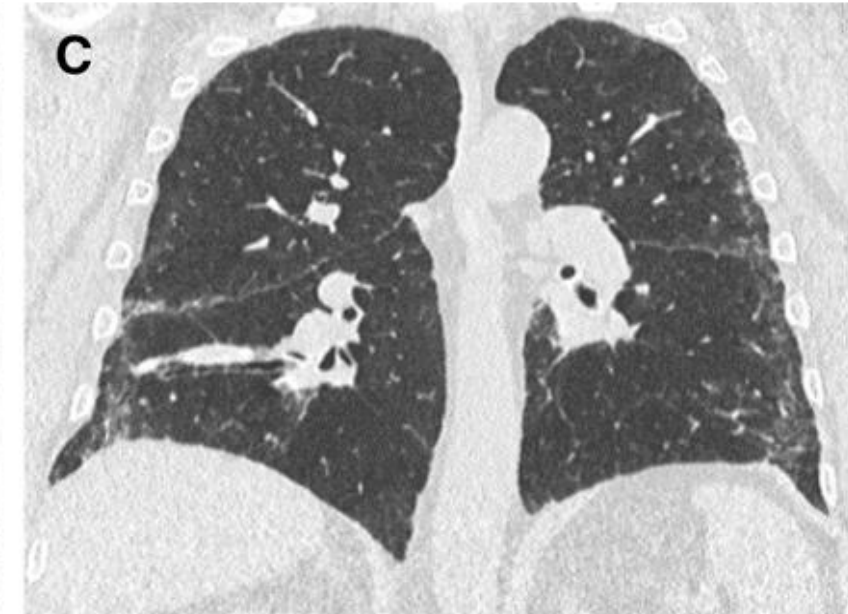
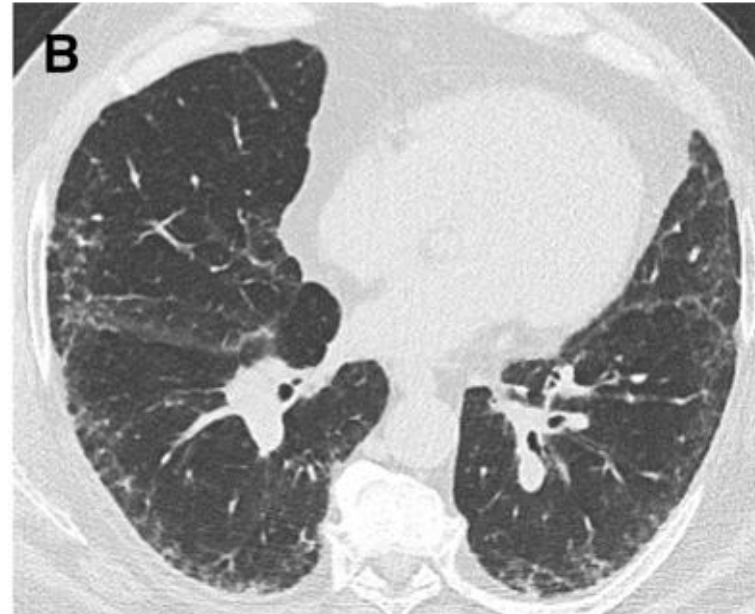
Subpleural and basal predominance

Indeterminate usual interstitial pneumonia (UIP) pattern CT

Raghu G, et al. AJRCCM 2018 and 2022



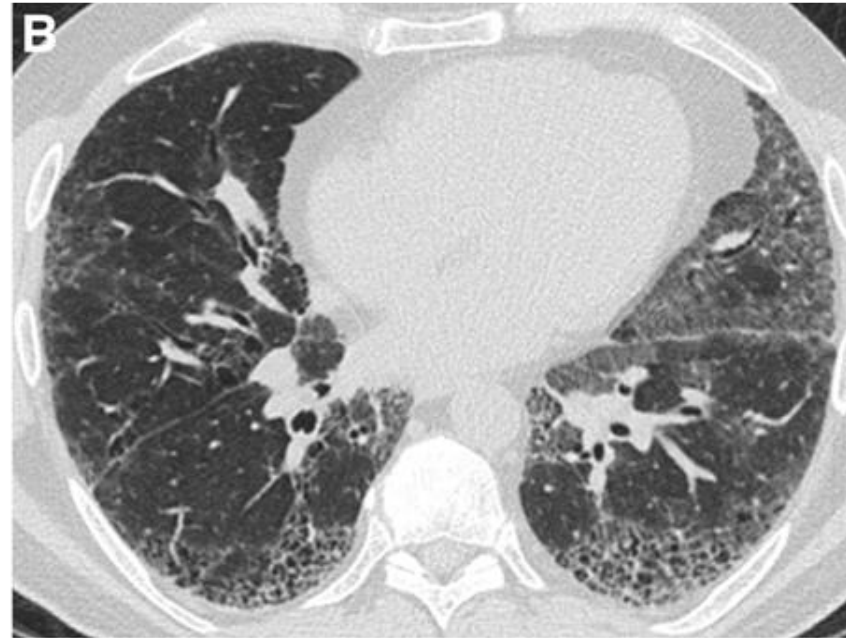
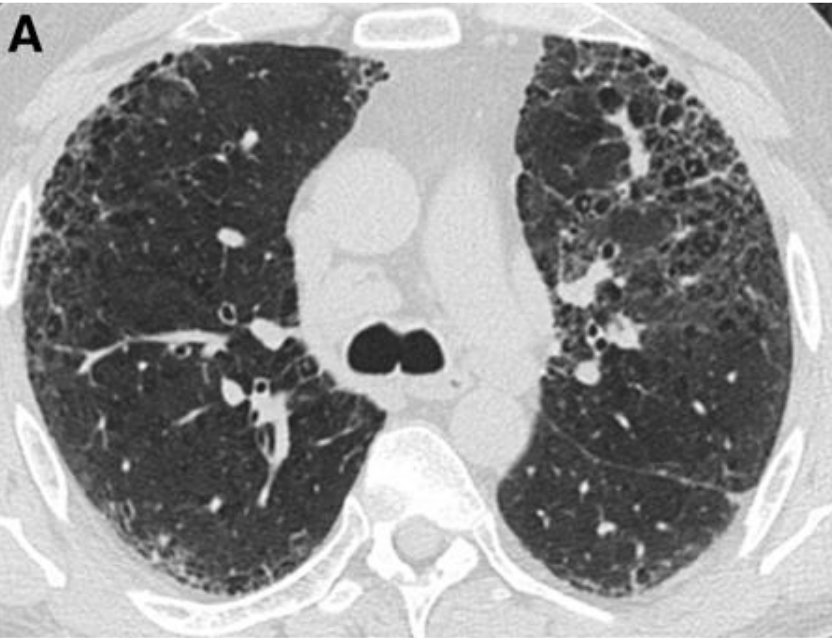
Reticular opacity + mild GGO



Subpleural and basal predominance

Indeterminate usual interstitial pneumonia (UIP) pattern CT

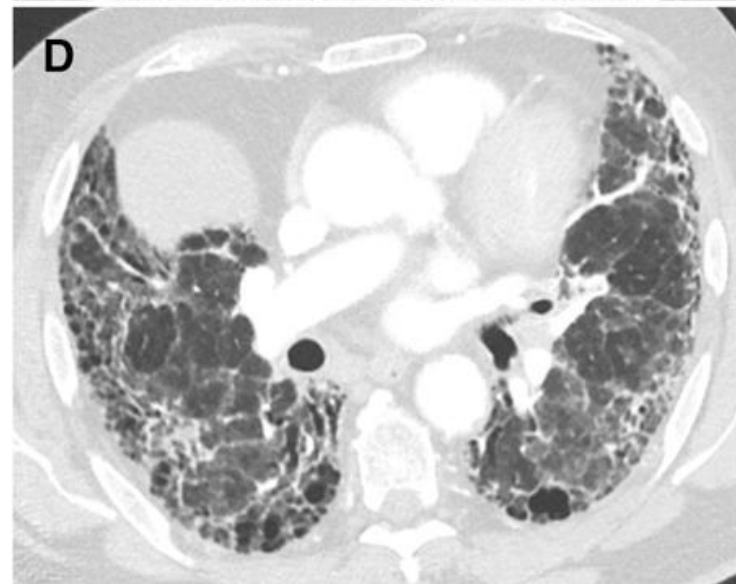
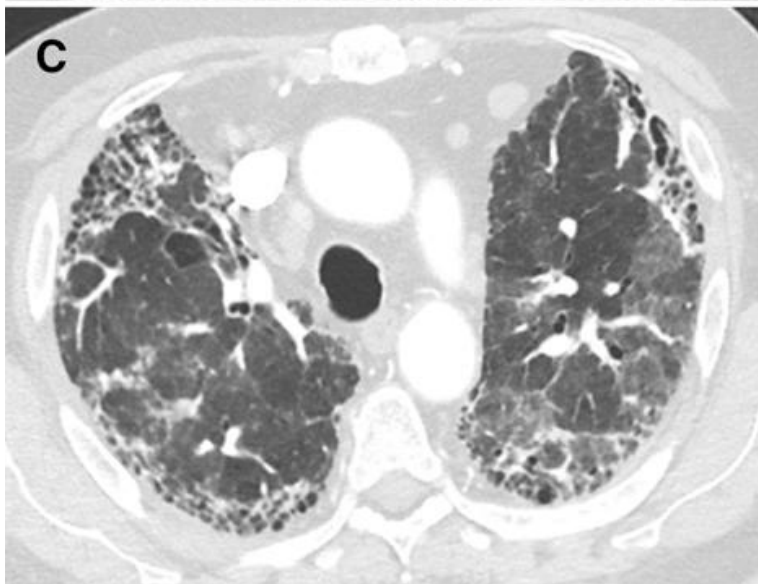
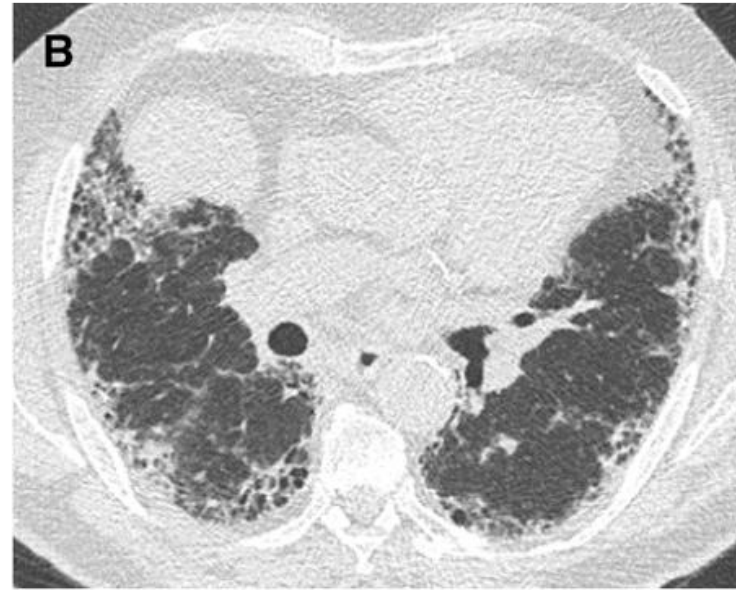
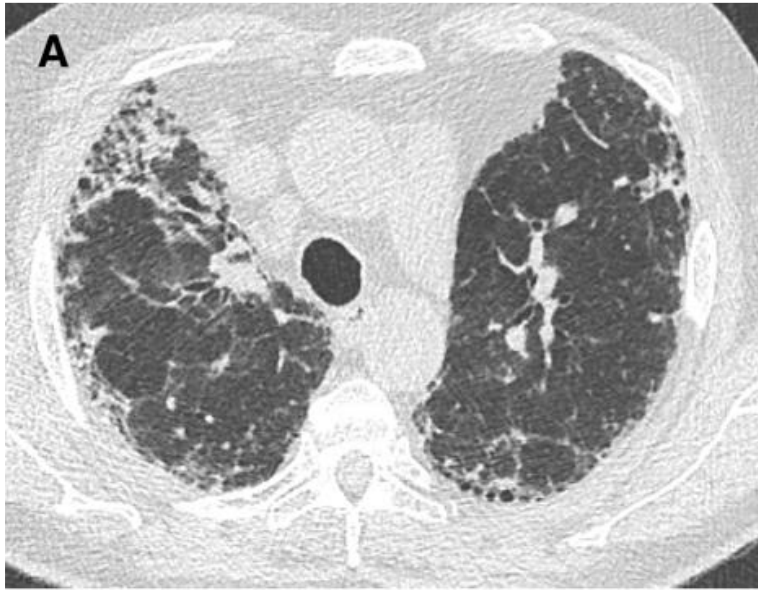
Raghu G, et al. AJRCCM 2018 and 2022



Marked GGO + reticular opacity + traction bronchiectasis

Subpleural and basal predominance

Acute exacerbation of IPF

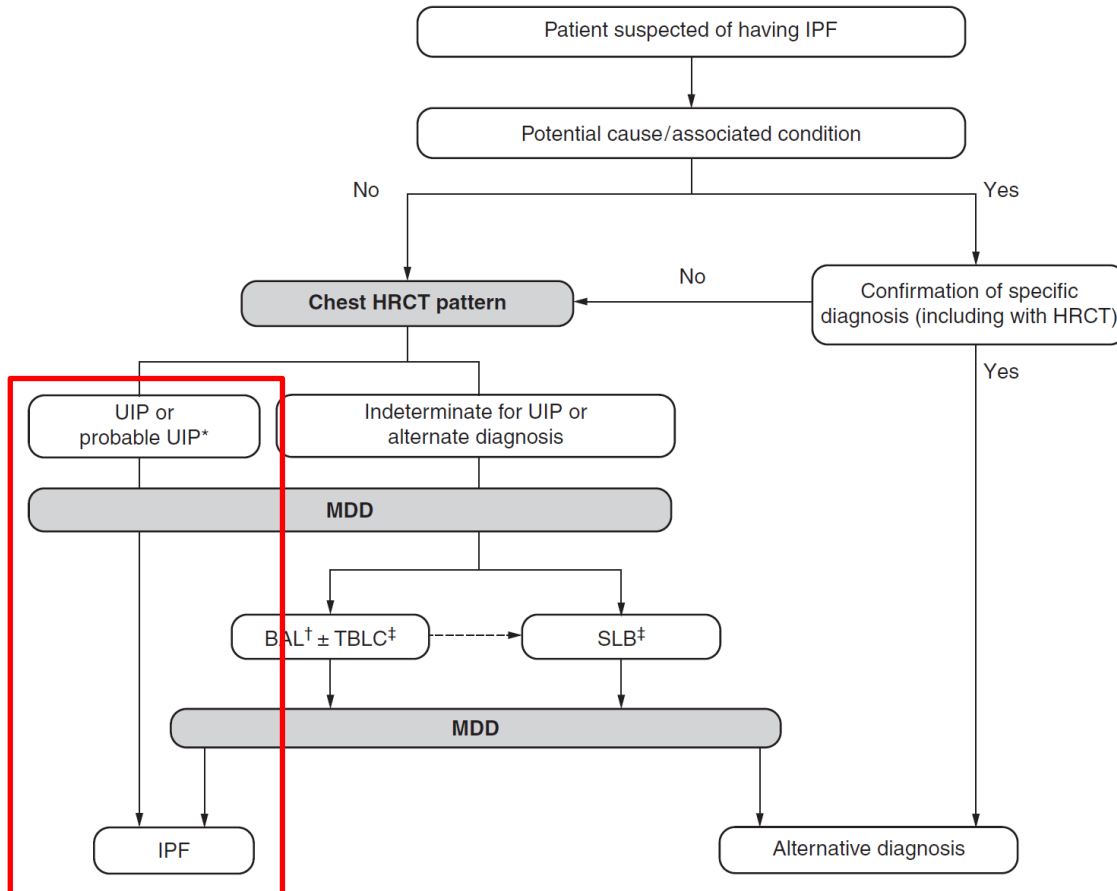


Reticular opacity
Traction bronchiectasis
Honeycombing cyst
Subpleural and basal predominance

= UIP pattern

New GGO in bilateral lung
= AE-IPF

Versus fibrotic NSIP?



IPF suspected*		Histopathology pattern [†]			
		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) [‡]	Non-IPF dx
	Indeterminate	IPF	IPF (Likely) [‡]	Indeterminate [§]	Non-IPF dx
	Alternative diagnosis	IPF (Likely) [‡]	Indeterminate [§]	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

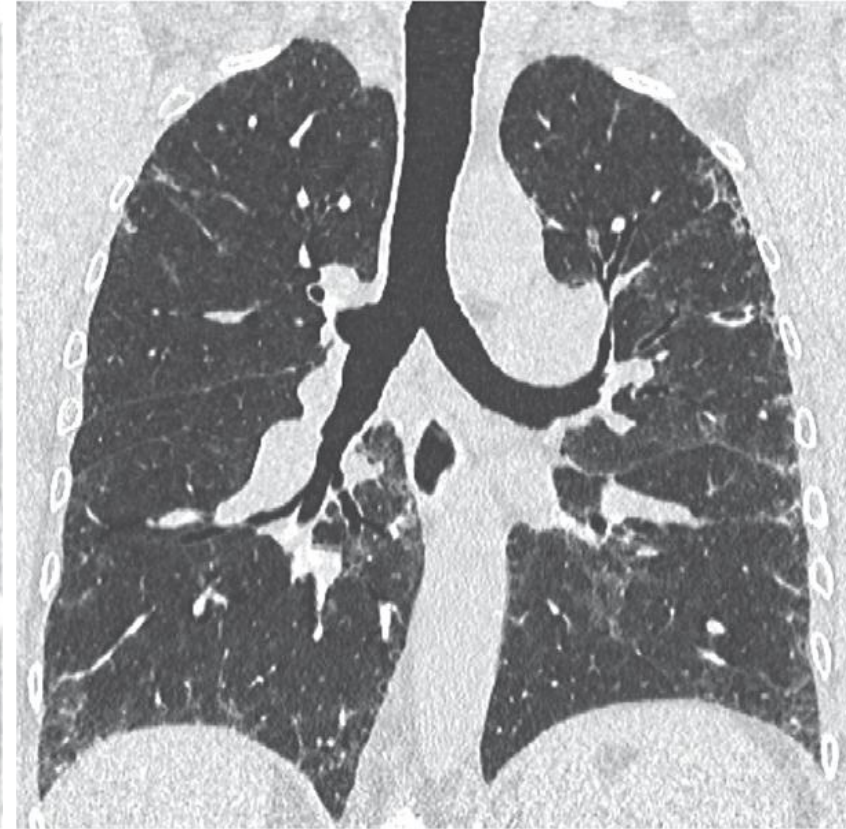
UIP pattern spectrum



UIP pattern



Probable UIP pattern



Indeterminate UIP pattern

차례

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 - **Other ILD CT Pattern (alternative diagnosis)**
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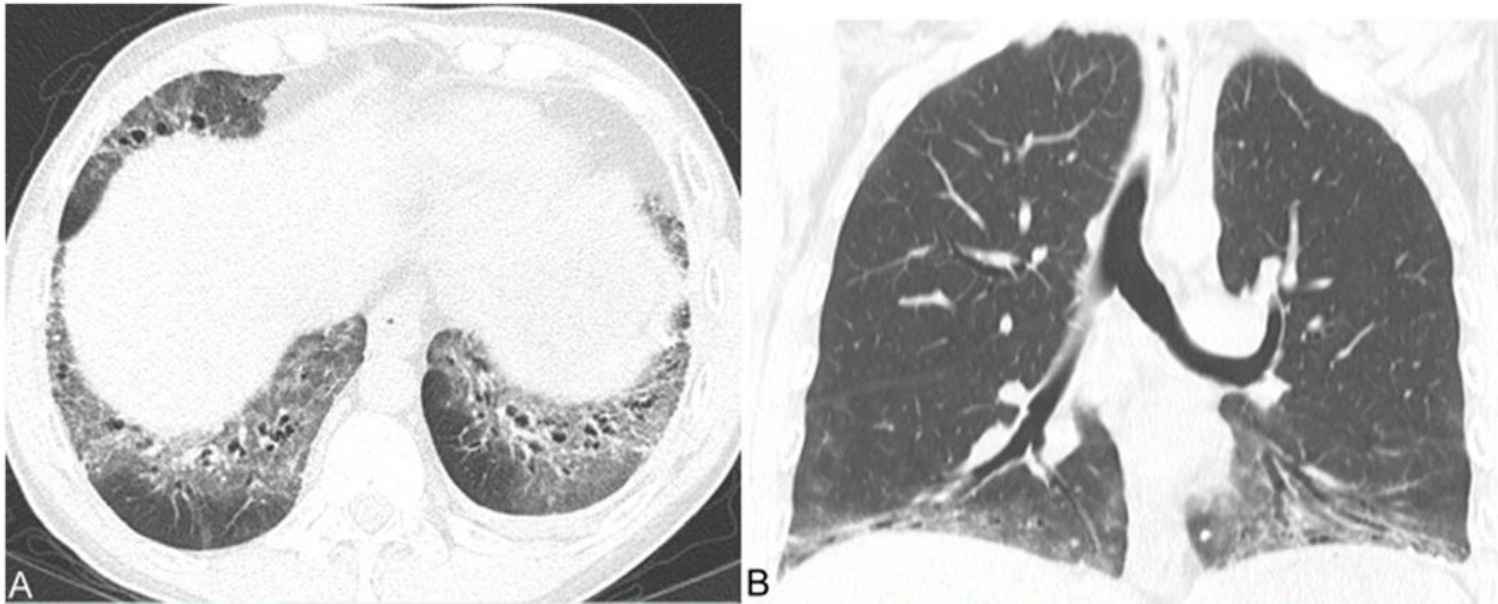
Alternative diagnosis

Ragu G, et al. AJRCCM 2018 and 2022

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

Nonspecific interstitial pneumonia (NSIP)

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

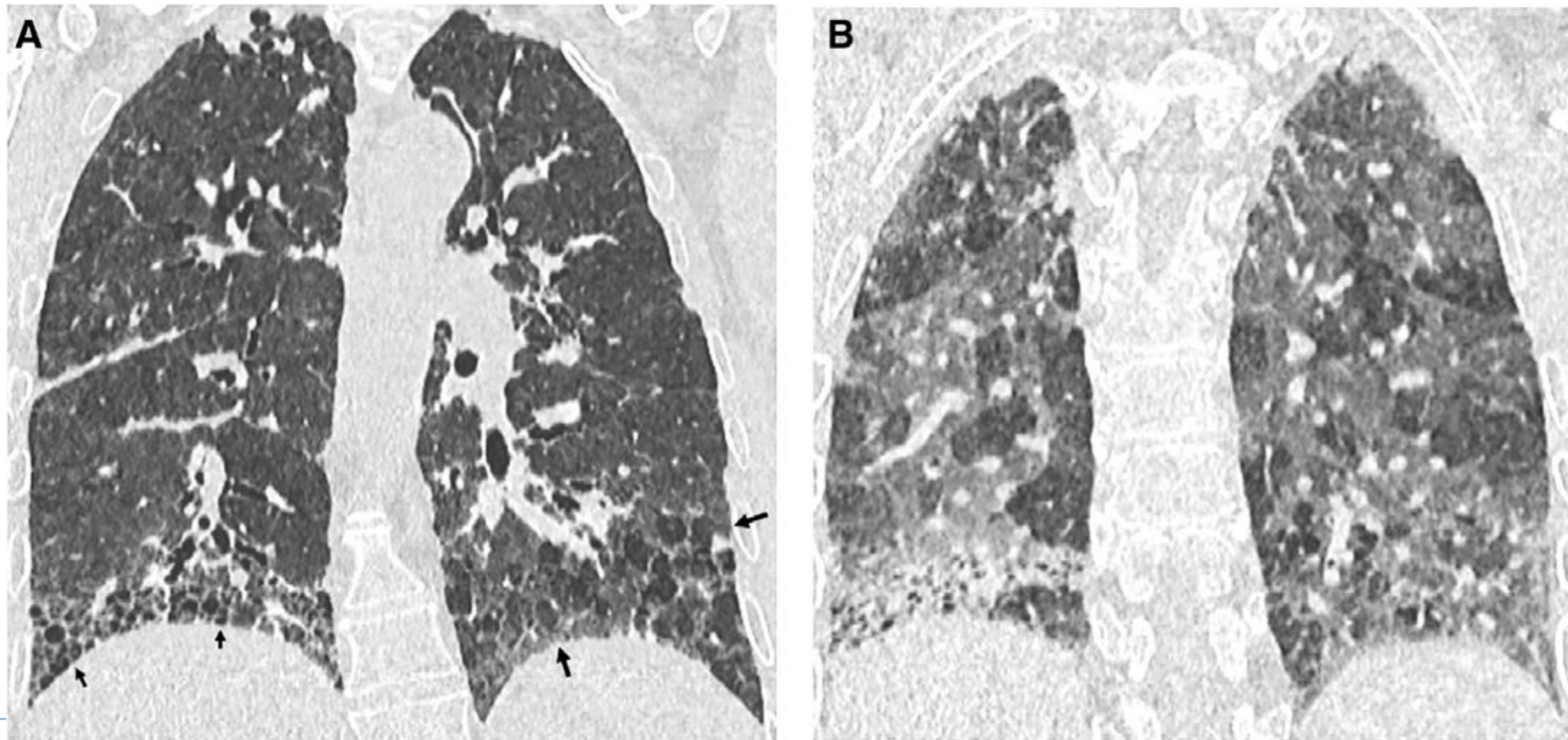


UIP VS. NSIP

	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	Most prominent feature
Traction bronchiectasis	Common (subpleural)	Common (bronchovascular bundle)
Honeycombing	Common; subpleural and basal	Uncommon at presentation

Fibrotic hypersensitivity pneumonitis

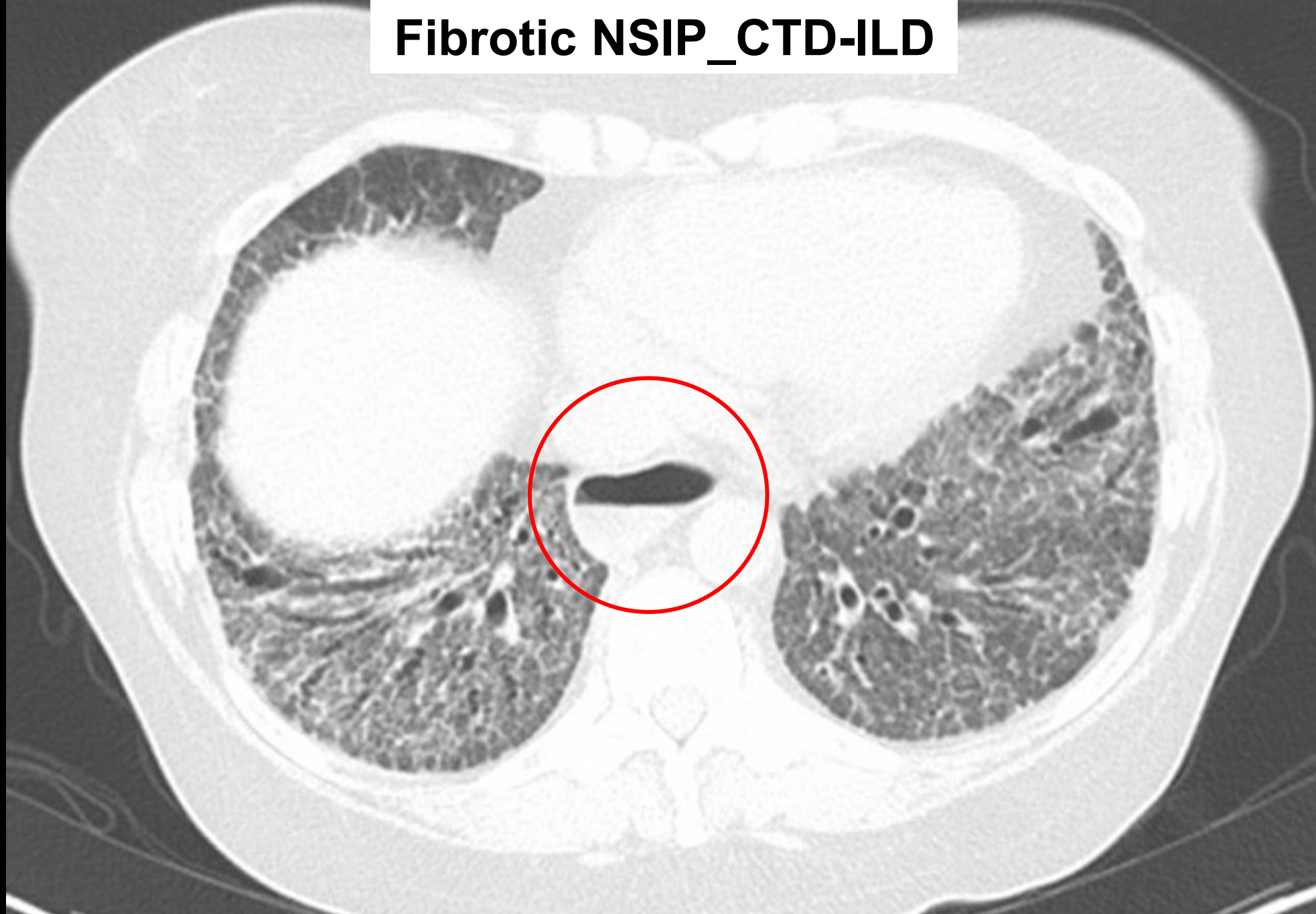
- **Bronchiolocentric interstitial pneumonia**
- **Air-trapping or Three-density sign, Diffuse or mid-to-upper lung predominancy**
- Centrilobular nodules, central peribronchovascular reticulation and distortion
- Traction bronchiectasis & Honeycombing: may be present but do not predominate



Connective tissue disease-ILD

- **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

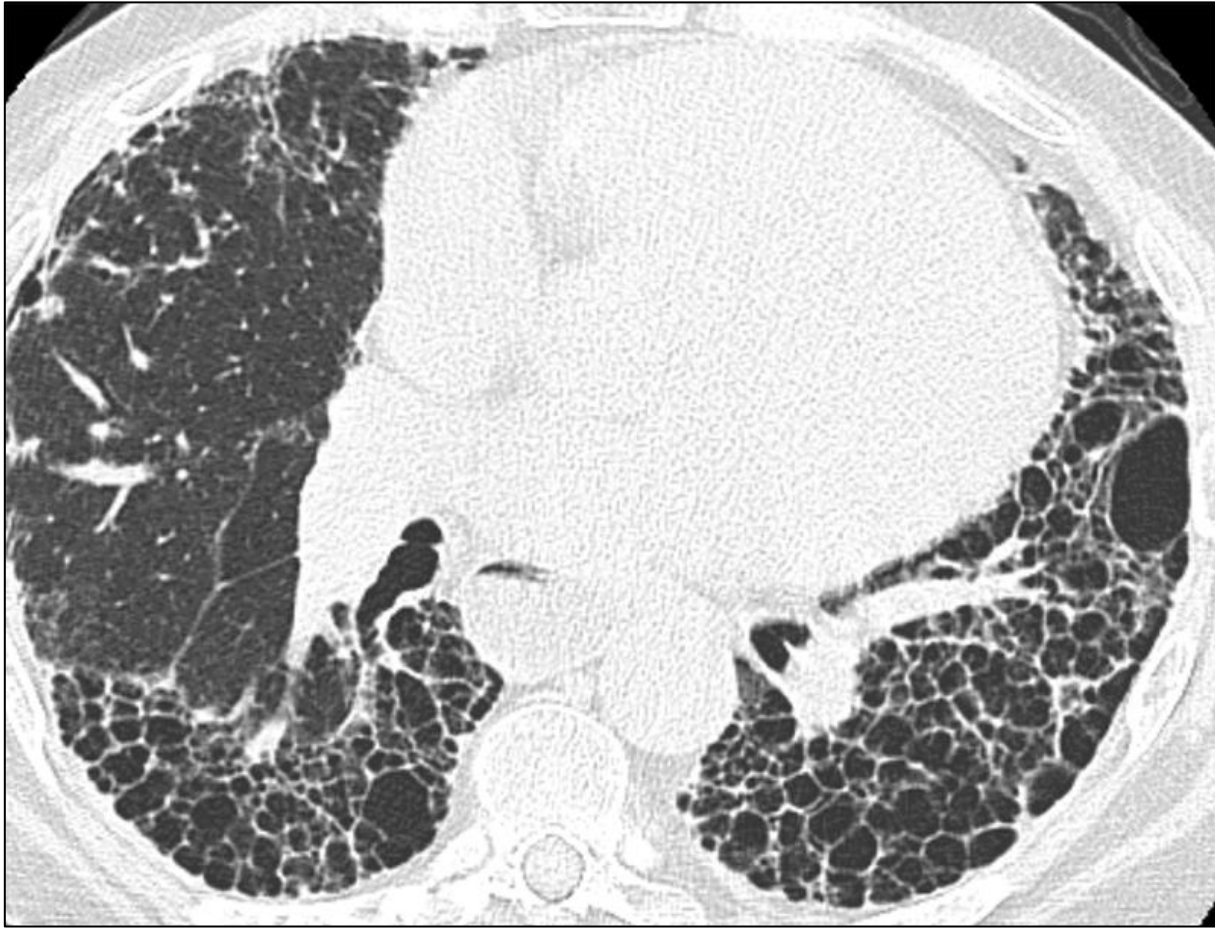


Anterior upper lobe sign

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

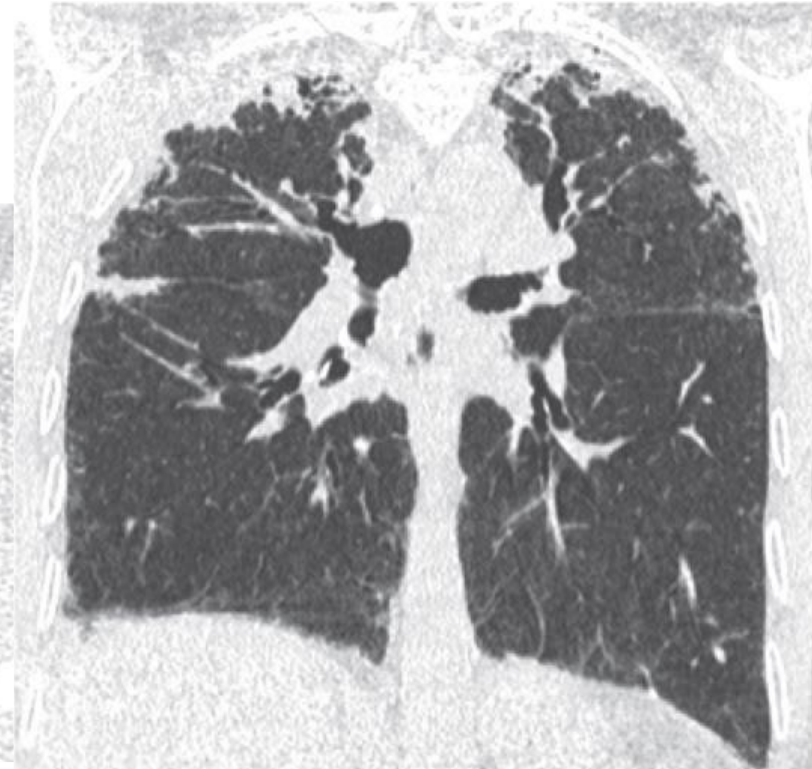
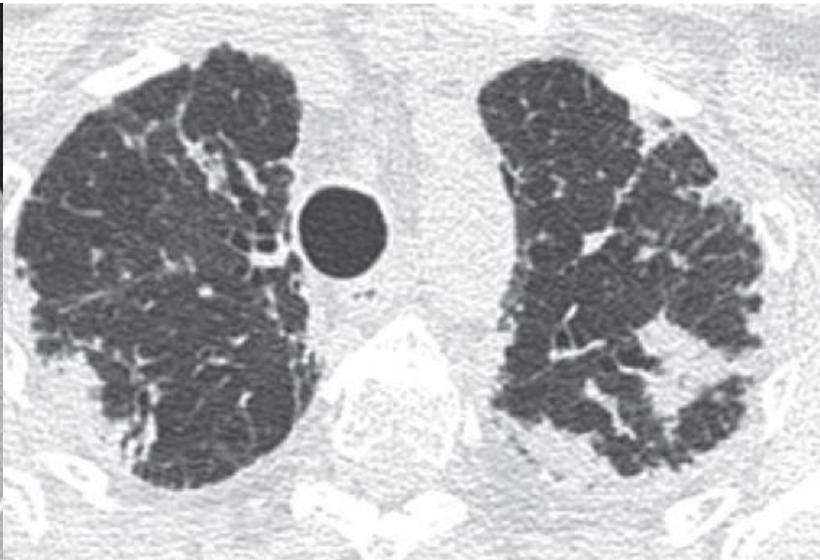
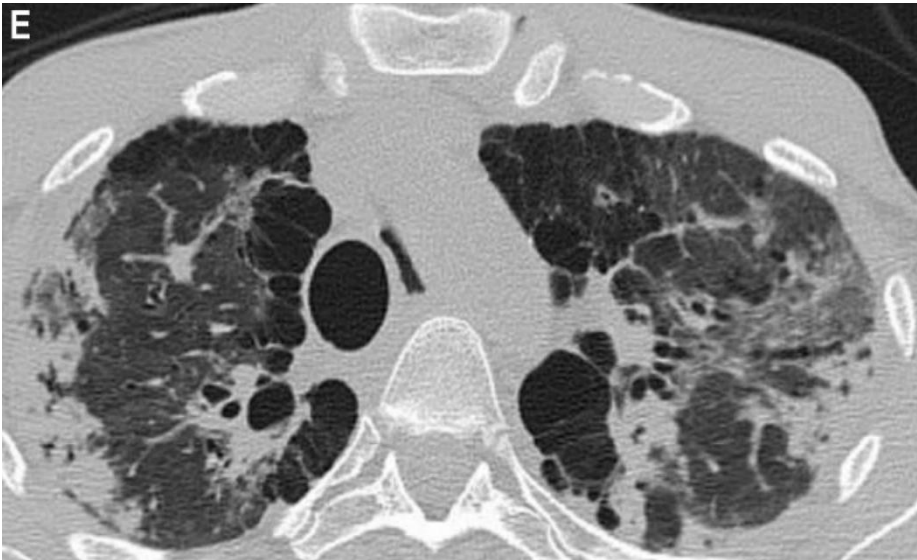


Straight-edge sign



Pleuroparenchymal fibroelastosis (PPFE)

- Irregular visceral pleural thickening and merge with fibrotic changes in the subjacent lung
- “Tags” in the upper lungs (triangle)
- Substantial upper lobes volume loss, architectural distortion
- Pneumothorax, pneumomediastinum



Asbestosis & End-stage sarcoidosis



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



- Middle to upper lungs
- Fibrotic distortion with peribronchovascular distribution & perilymphatic nodules
- Large cyst, honeycomb-like cysts

Take-home message

- **ILD classification & ILA**
- **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...

Thank you for your attentions
