

# Interstitial Lung Disease

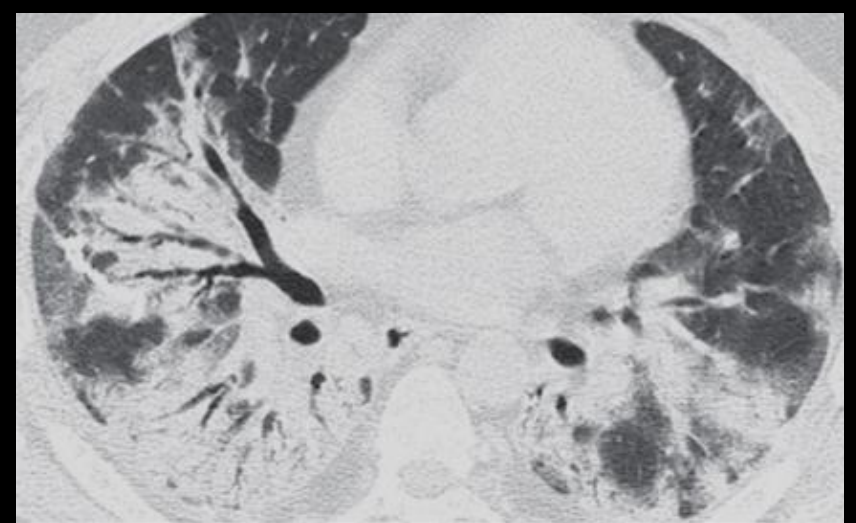
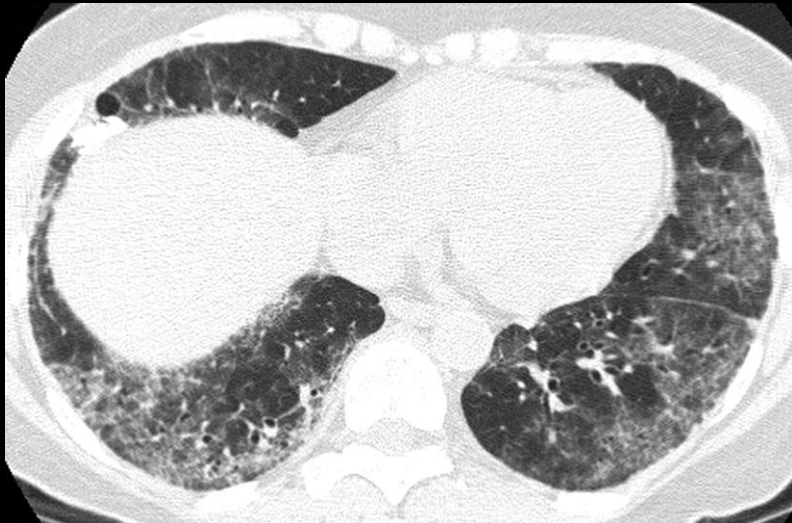
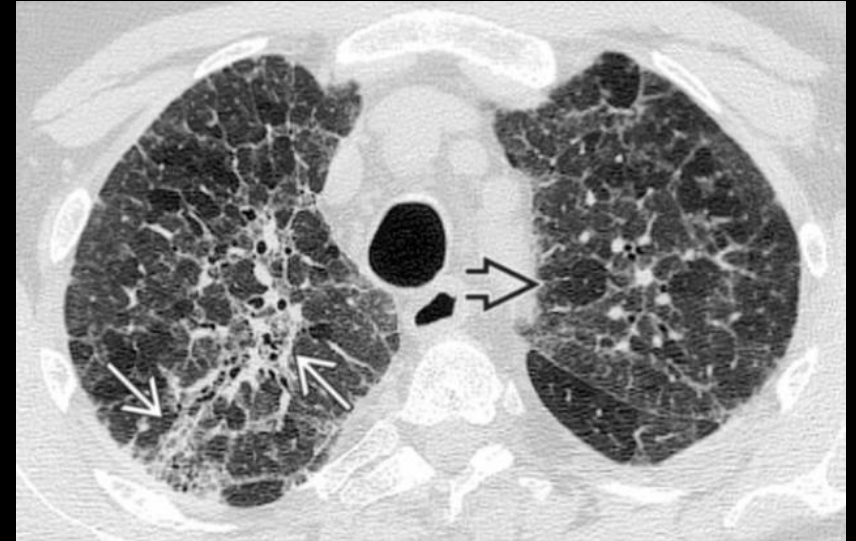
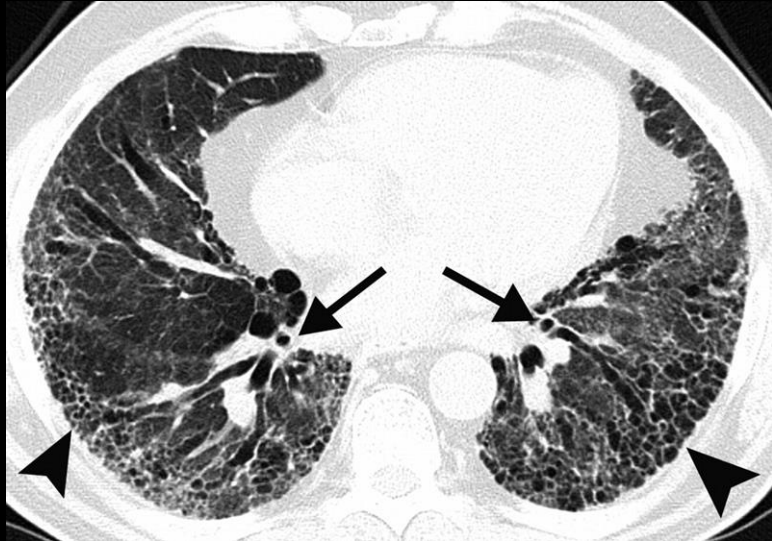
## The Big 4

David L. Smith, MD

Professor of Radiology  
Professor of Medicine  
Professor of Cell Biology & Anatomy

**LSU Health**  
NEW ORLEANS

University   
Medical Center  
New Orleans  
LCMC Health



# ILD: The Big 4

UIP

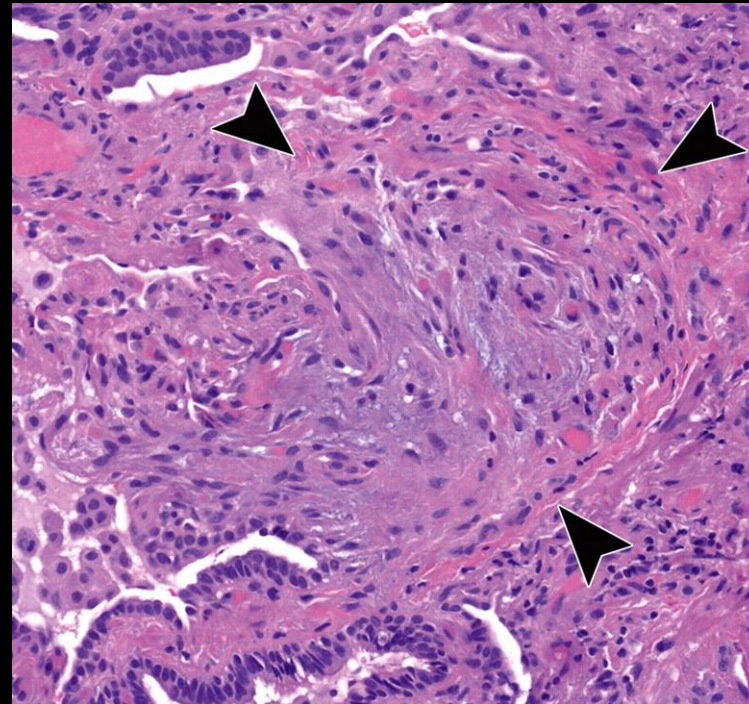
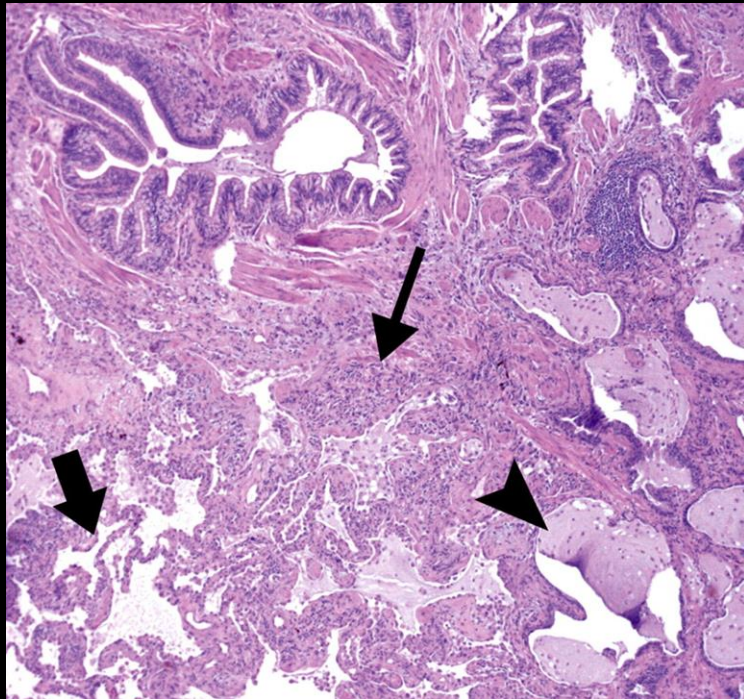
NSIP

HP

OP

# Usual Interstitial Pneumonia (UIP)

A histologic pattern of pulmonary fibrosis characterized by temporal and spatial heterogeneity, with established fibroblastic foci and architectural distortion interspersed among normal lung.



# Usual Interstitial Pneumonia (UIP)

## UIP (per ATS 2018)

- A. Honeycombing
- B. Reticulation
- C. Subpleural, basal predominance
- D. Absence of inconsistent features

## Probable UIP (per ATS 2018)

B-D, but no honeycombing

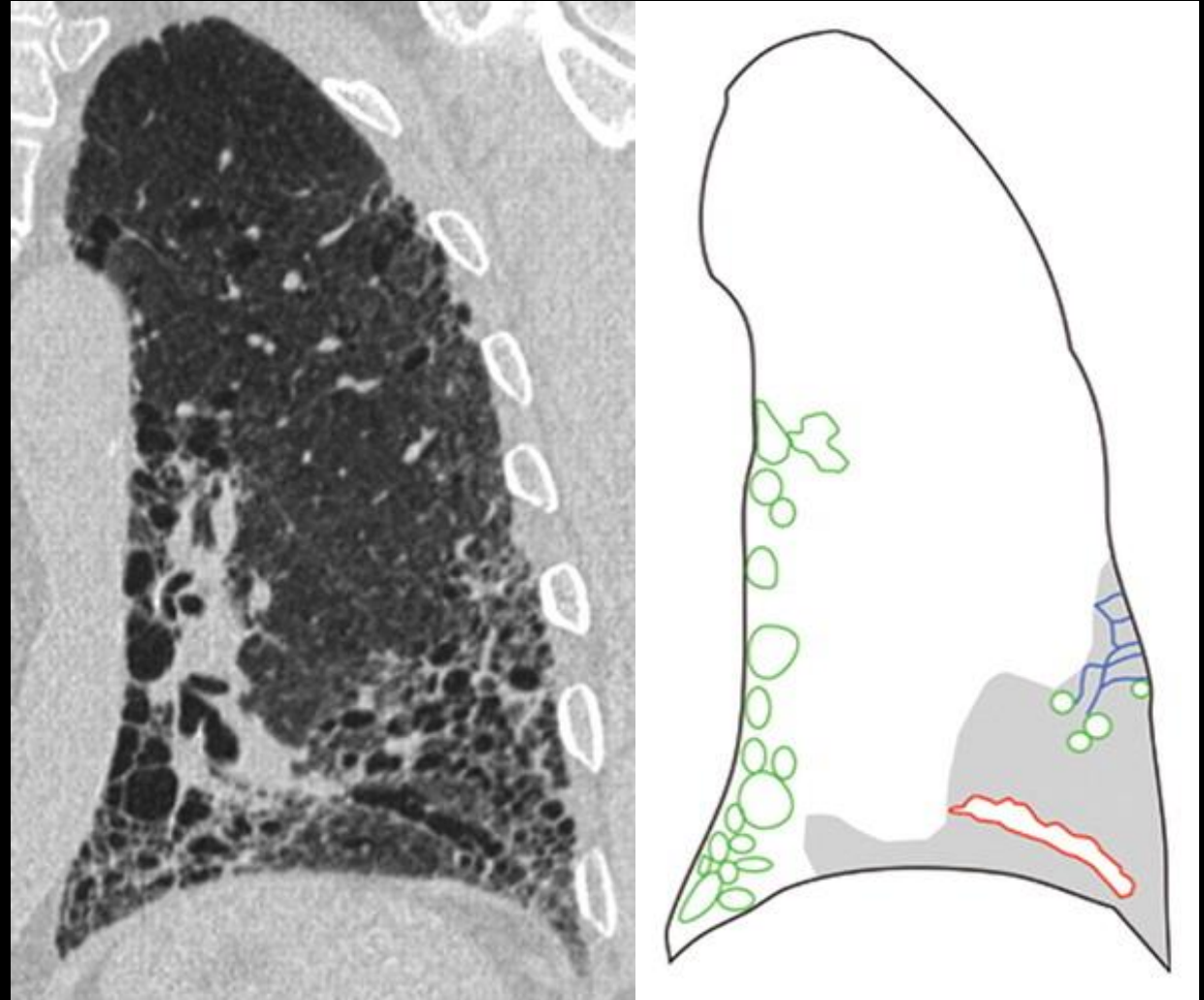
# Usual Interstitial Pneumonia (UIP)

## UIP (per ATS 2018)

- A. Honeycombing (green)
- B. Reticulation
- C. Subpleural, basal predominance
- D. Absence of inconsistent features

## Probable UIP

B-D, but no honeycombing



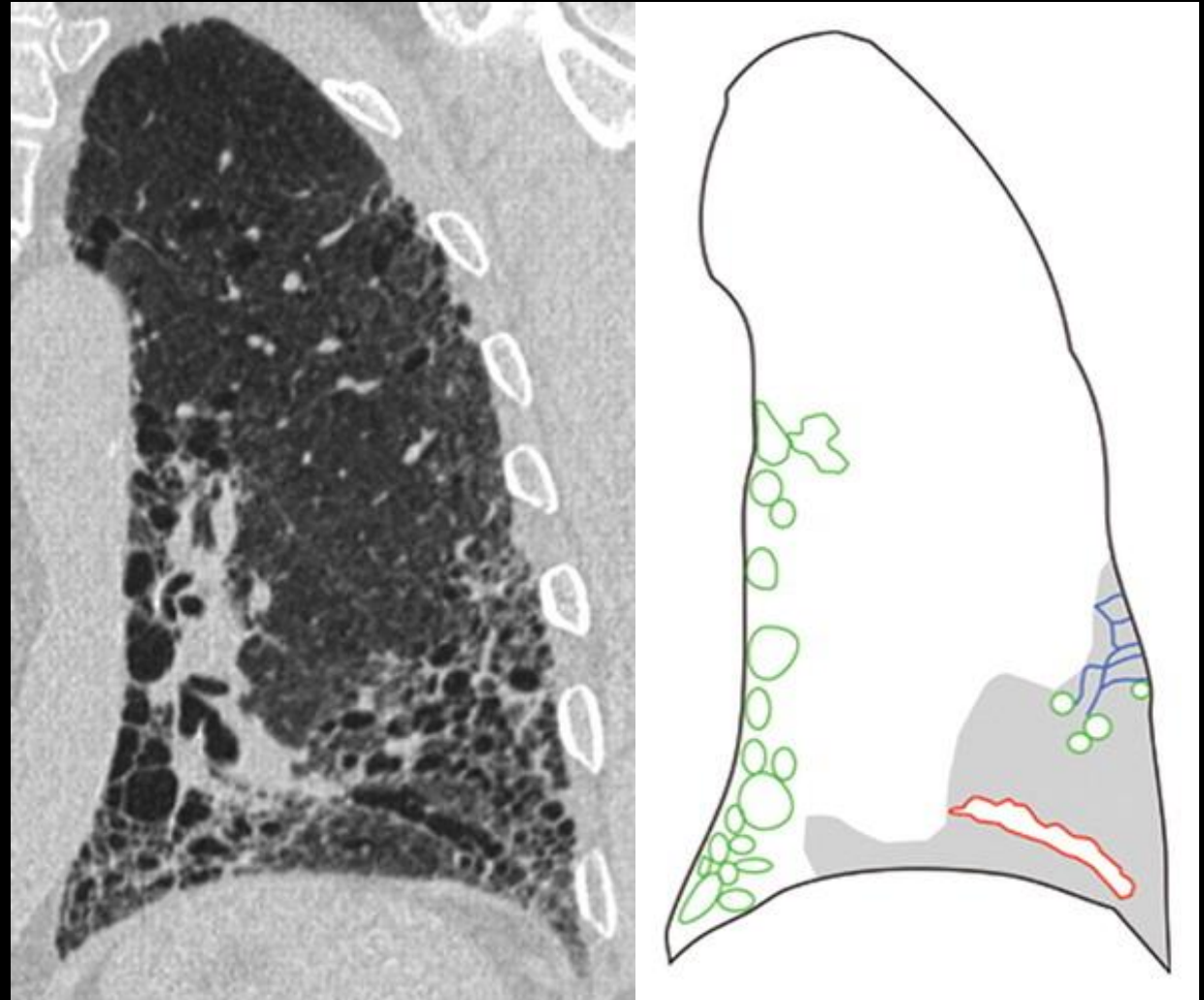
# Usual Interstitial Pneumonia (UIP)

## UIP (per ATS 2018)

- A. Honeycombing
- B. Reticulation (blue)
- C. Subpleural, basal predominance
- D. Absence of inconsistent features

## Probable UIP

B-D, but no honeycombing



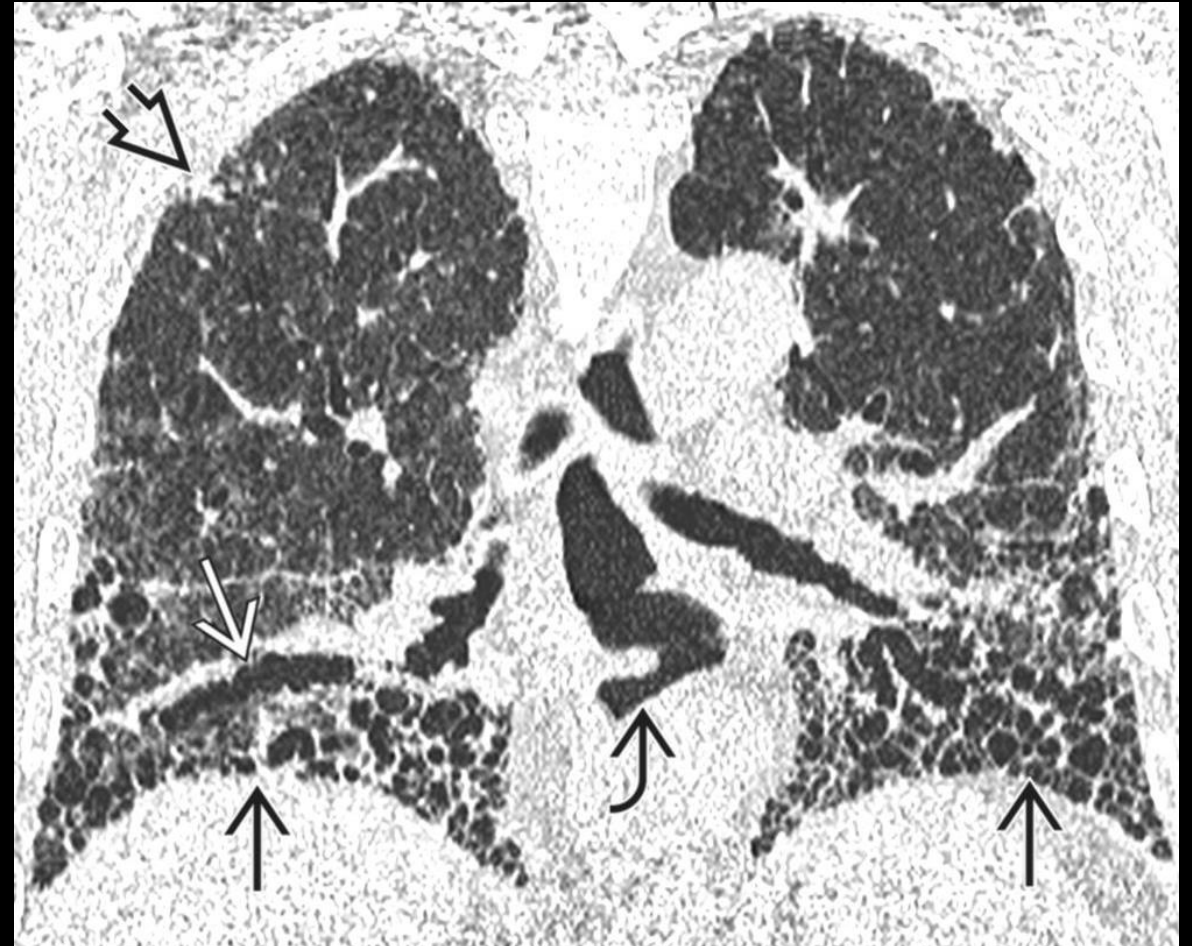
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## Probable UIP

B-D, but no honeycombing

- 1) Upper or mid-lung predominance
- 2) Peribronchovascular predominance
- 3) Extensive ground glass (extent > reticulation)
- 4) Profuse micronodules
- 5) Discrete (nonclustered) cysts
- 6) Diffuse mosaic attenuation/air trapping
- 7) Consolidation in bronchopulmonary segments/lobes



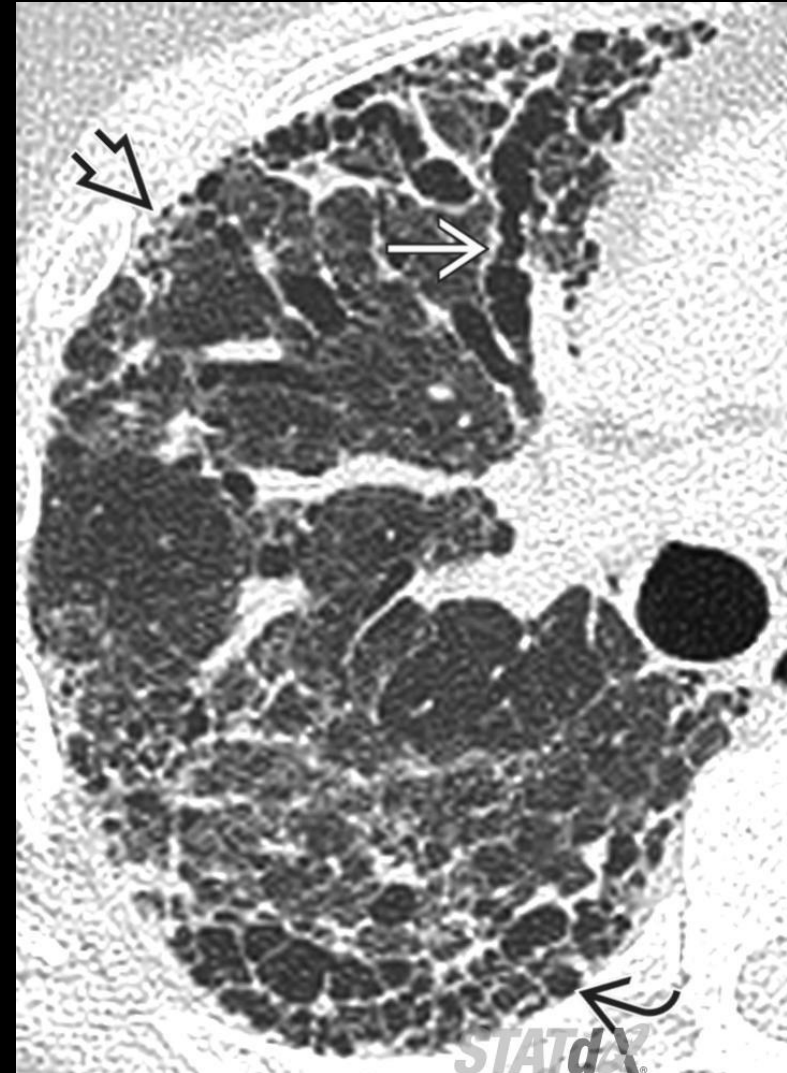
# Usual Interstitial Pneumonia (UIP)

## UIP (per ATS 2018)

- A. Honeycombing (curved arrow)
- B. Reticulation (open arrow)
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## Probable UIP

B-D, but no honeycombing



# Usual Interstitial Pneumonia (UIP)

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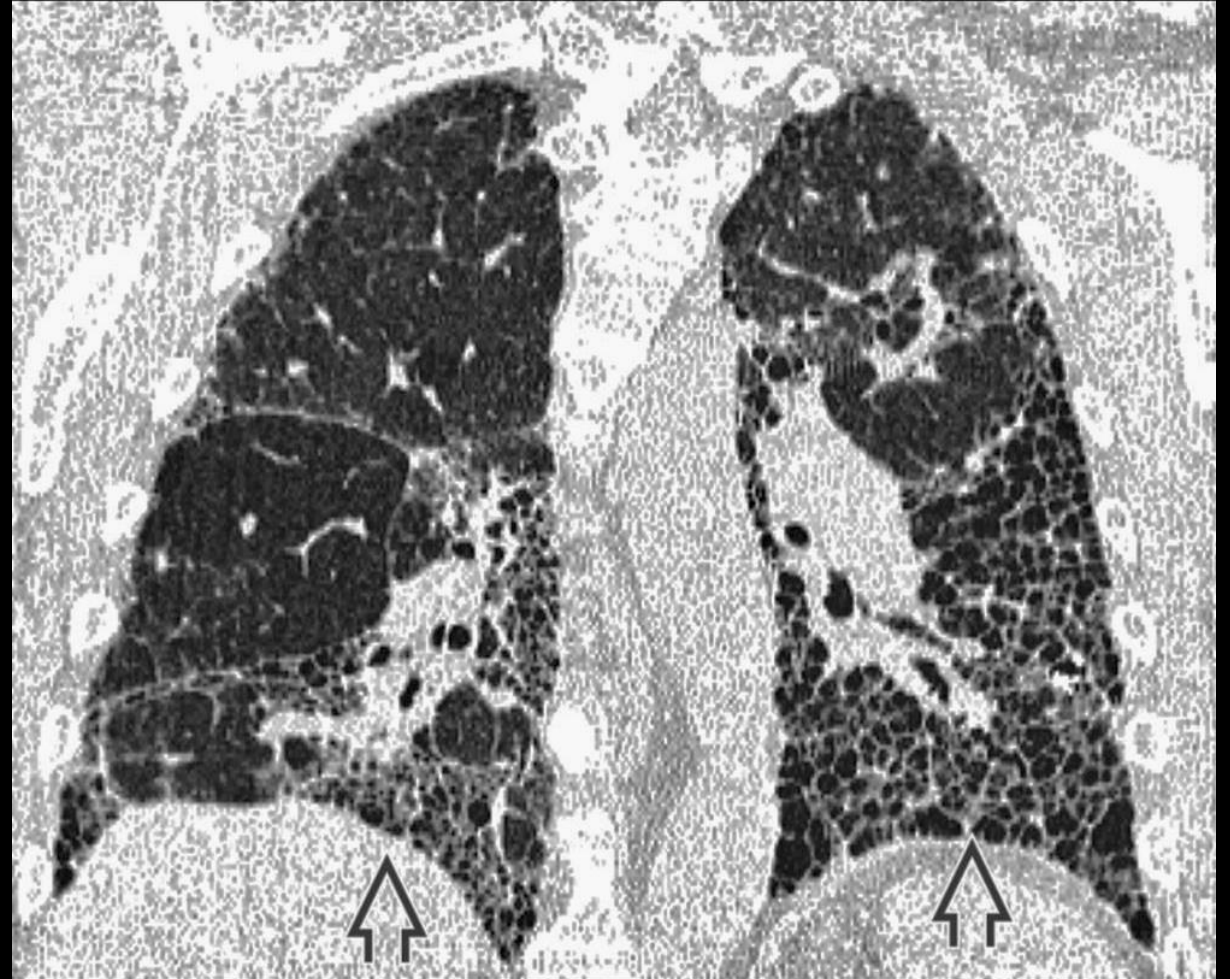
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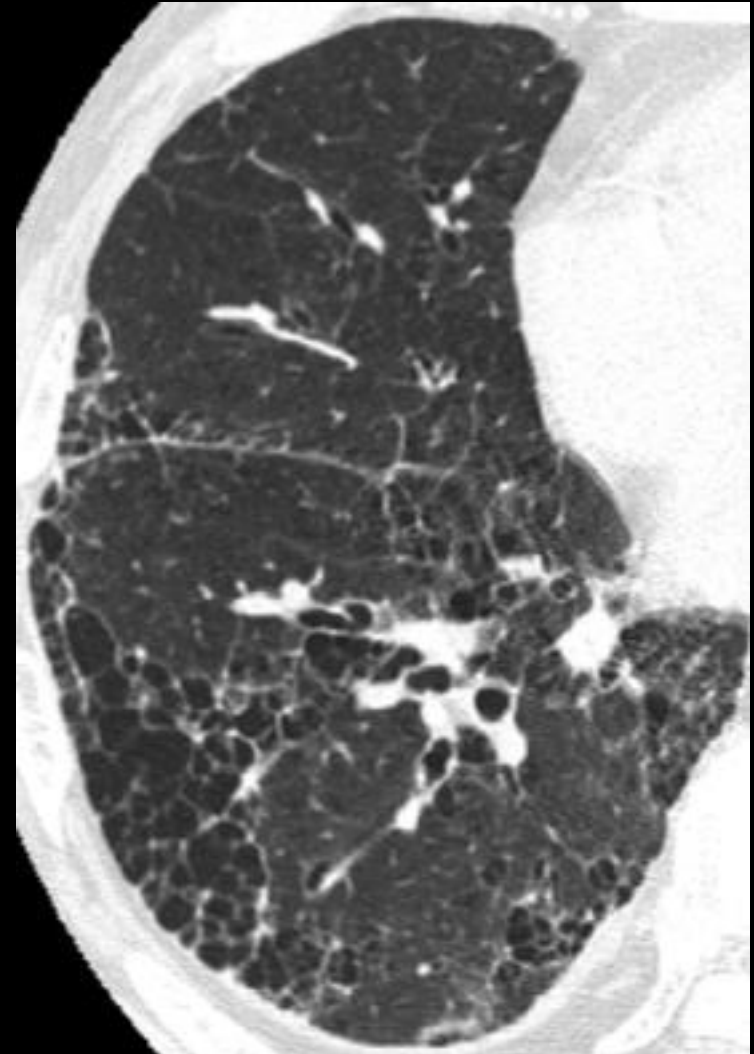
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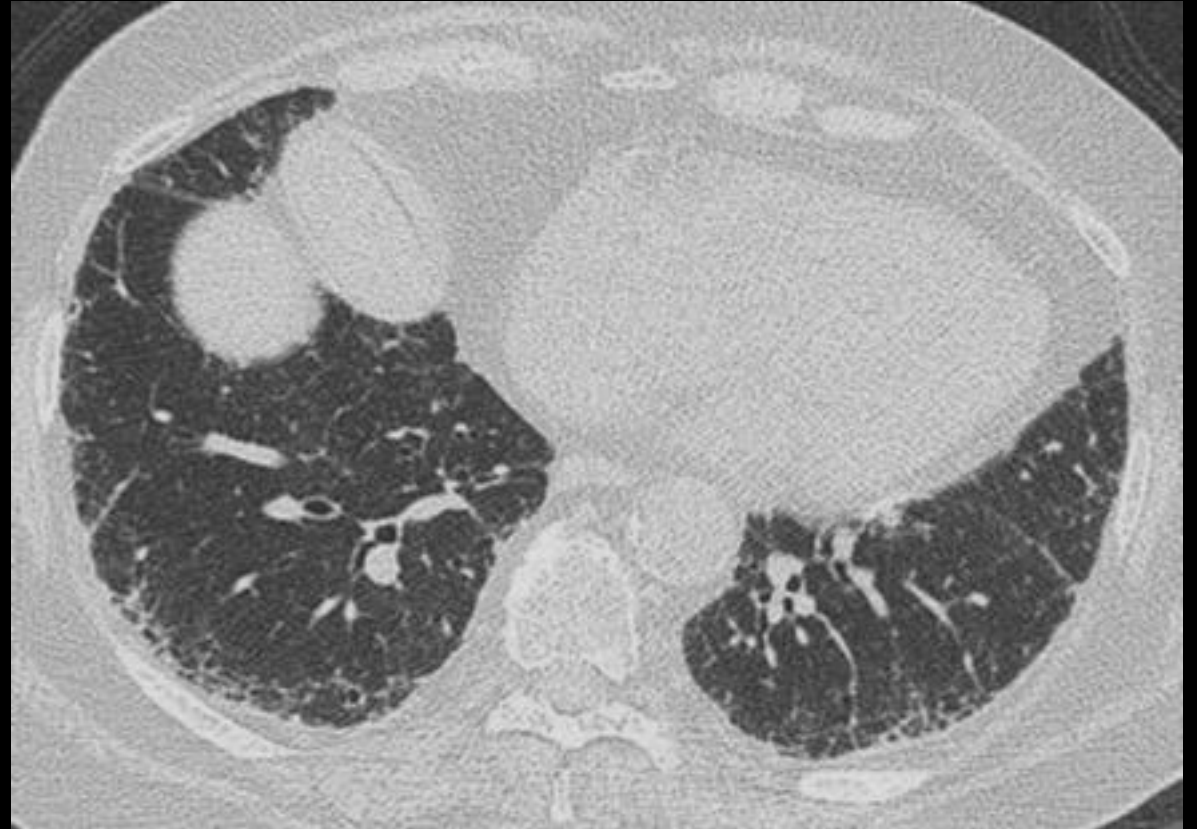
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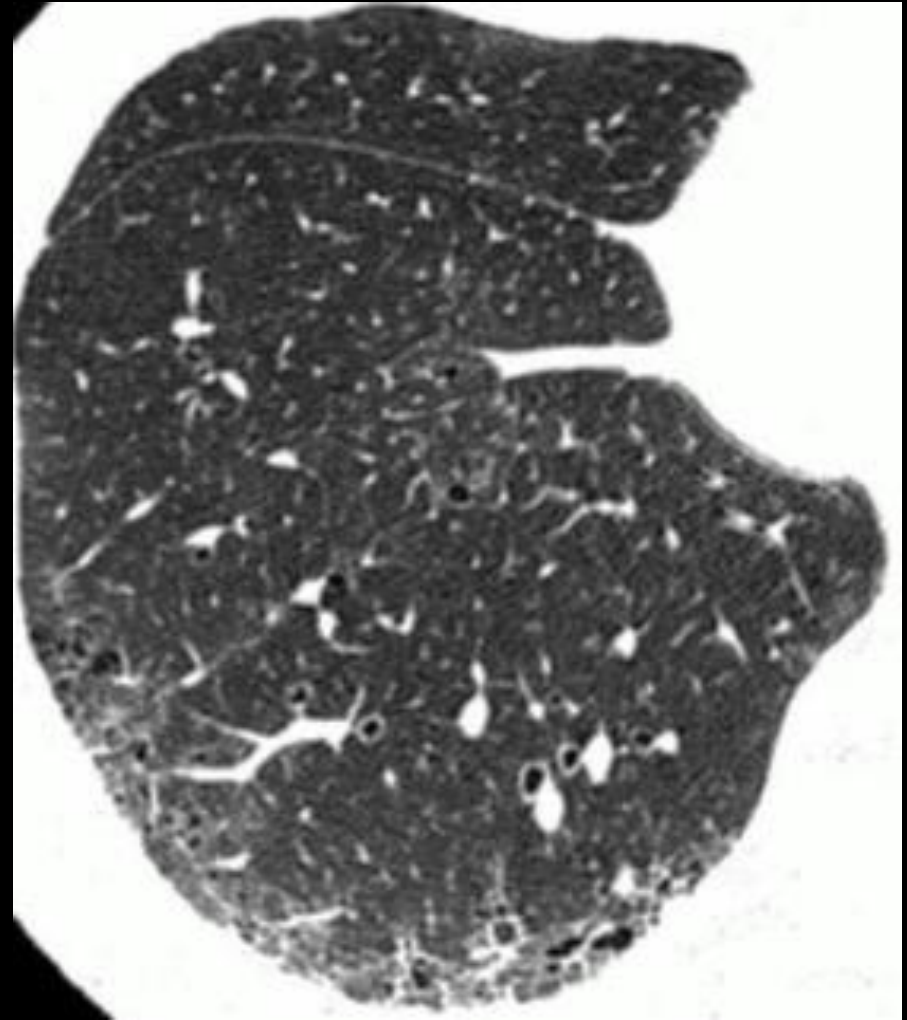
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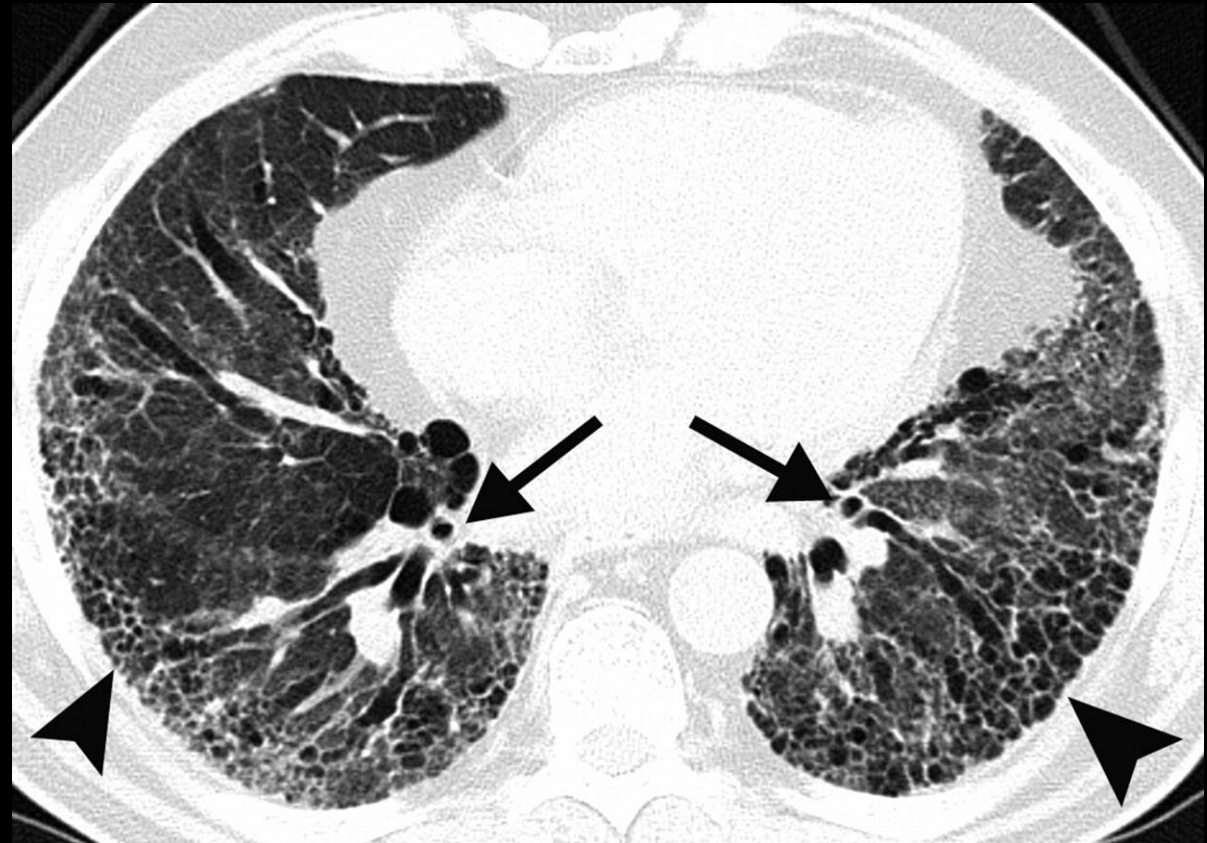
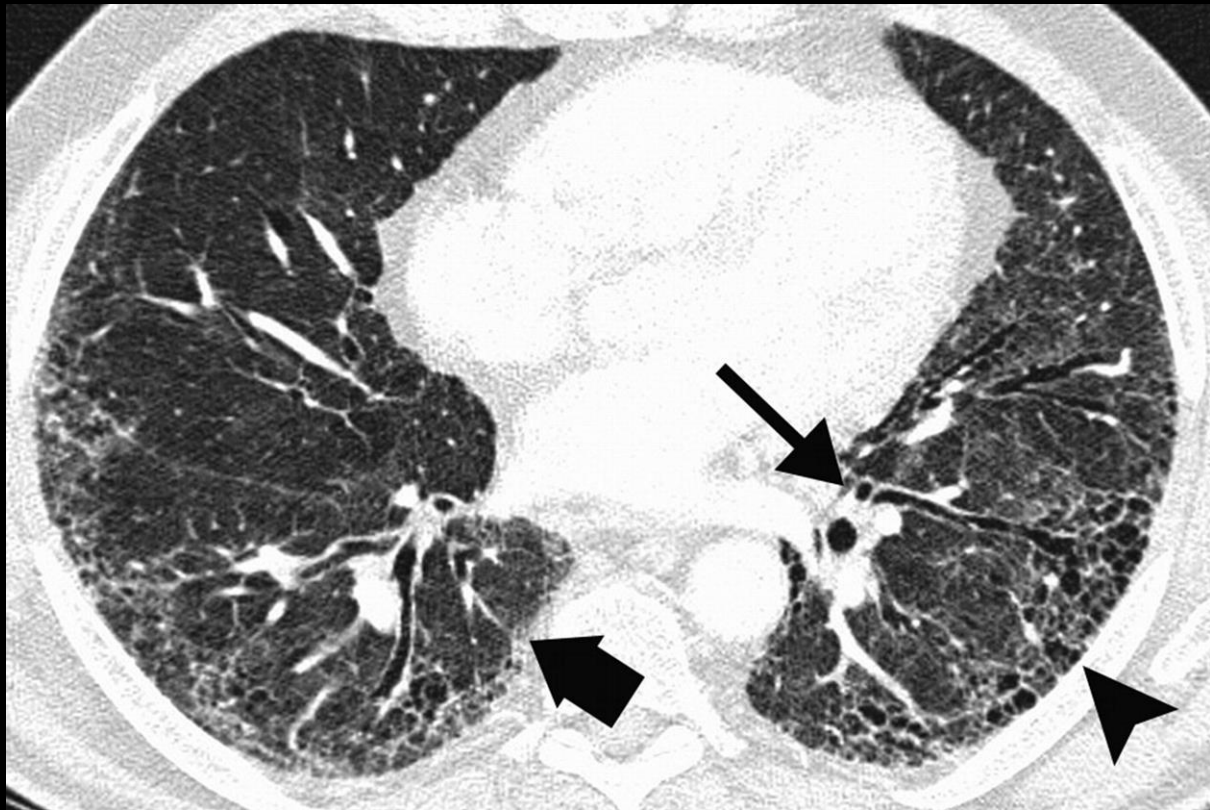
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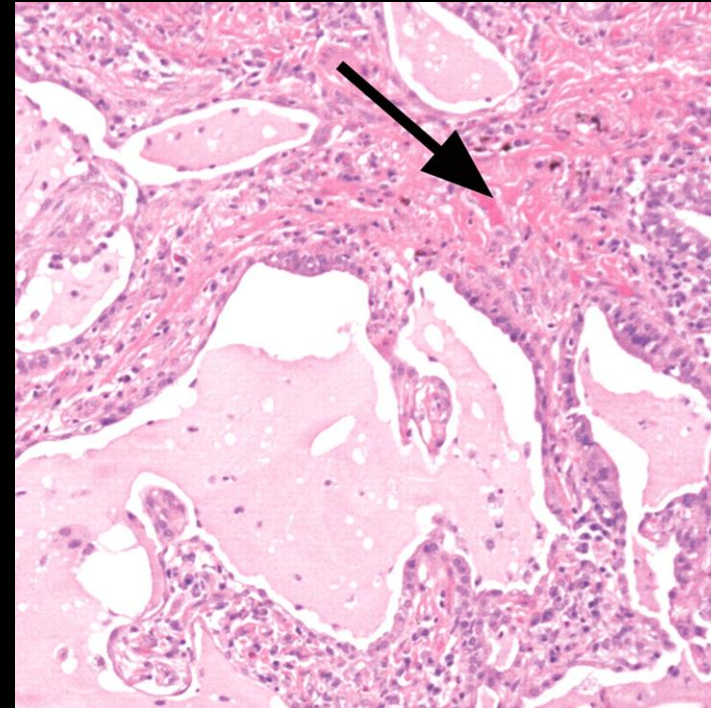
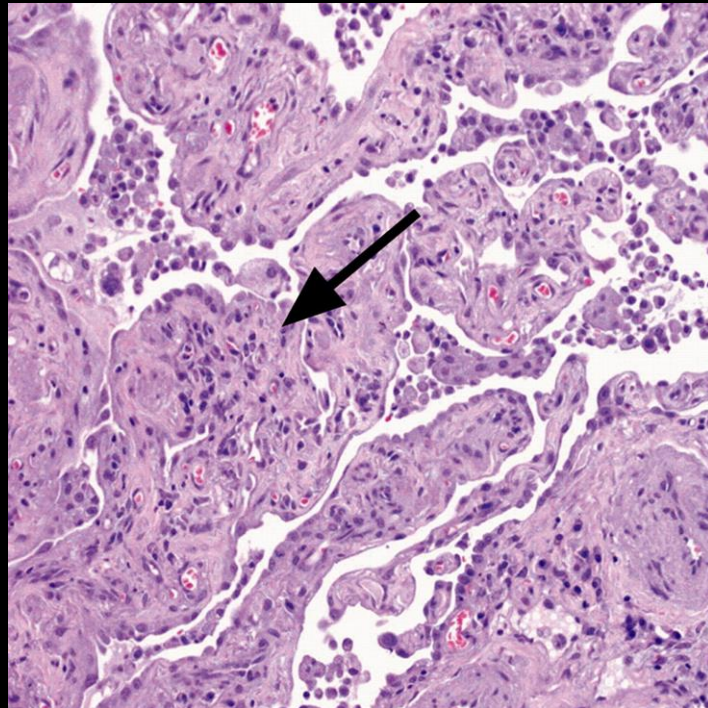


# Usual Interstitial Pneumonia (UIP)

CT Finding	Sensitivity (%)	Specificity (%)	Accuracy (%)
<b>Diagnosis of UIP</b>			
Upper lung irregular lines	83	57	69
Traction bronchiolectasis	67	94	82
Reticular opacity-to-ground-glass opacity ratio of 1 or greater	93	54	71
Honeycombing of 8% or greater	14	100	63

# Nonspecific Interstitial Pneumonia (NSIP)

A histologic pattern of spatial and temporal homogeneity of chronic interstitial inflammation (cellular NSIP) and/or interstitial fibrosis (fibrotic NSIP)



# Nonspecific Interstitial Pneumonia (NSIP)

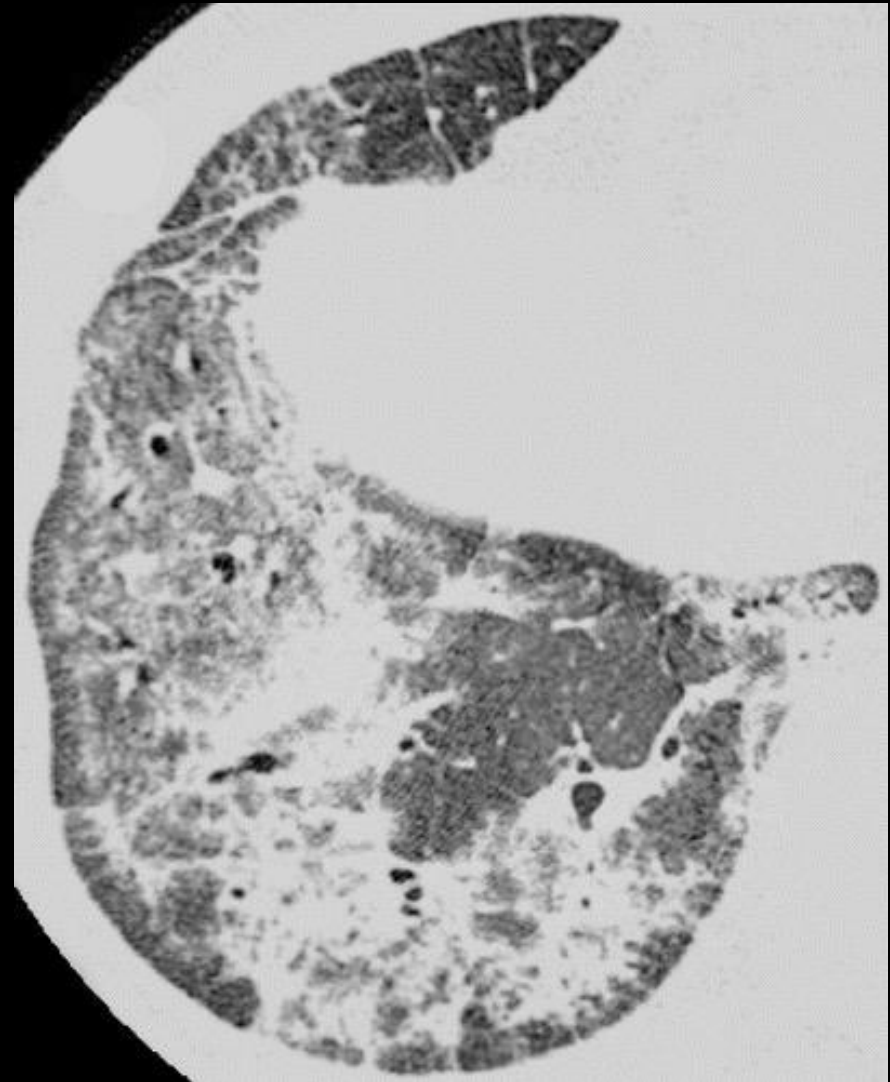
## Features

- A. Bibasilar predominance
- B. Ground glass opacity
- C. Reticulation
- D. Subpleural sparing (cellular?)
- E. Traction bronchiectasis (fibrotic)
- F. LITTLE, IF ANY, HONEYCOMBING

# Nonspecific Interstitial Pneumonia (NSIP)

## Features

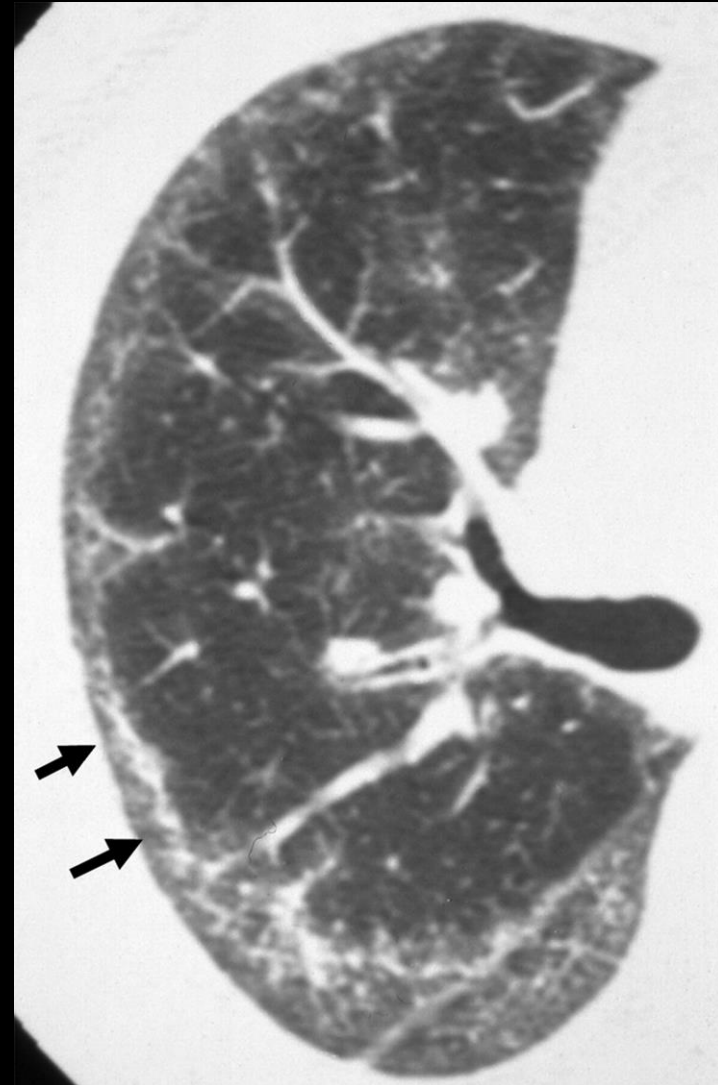
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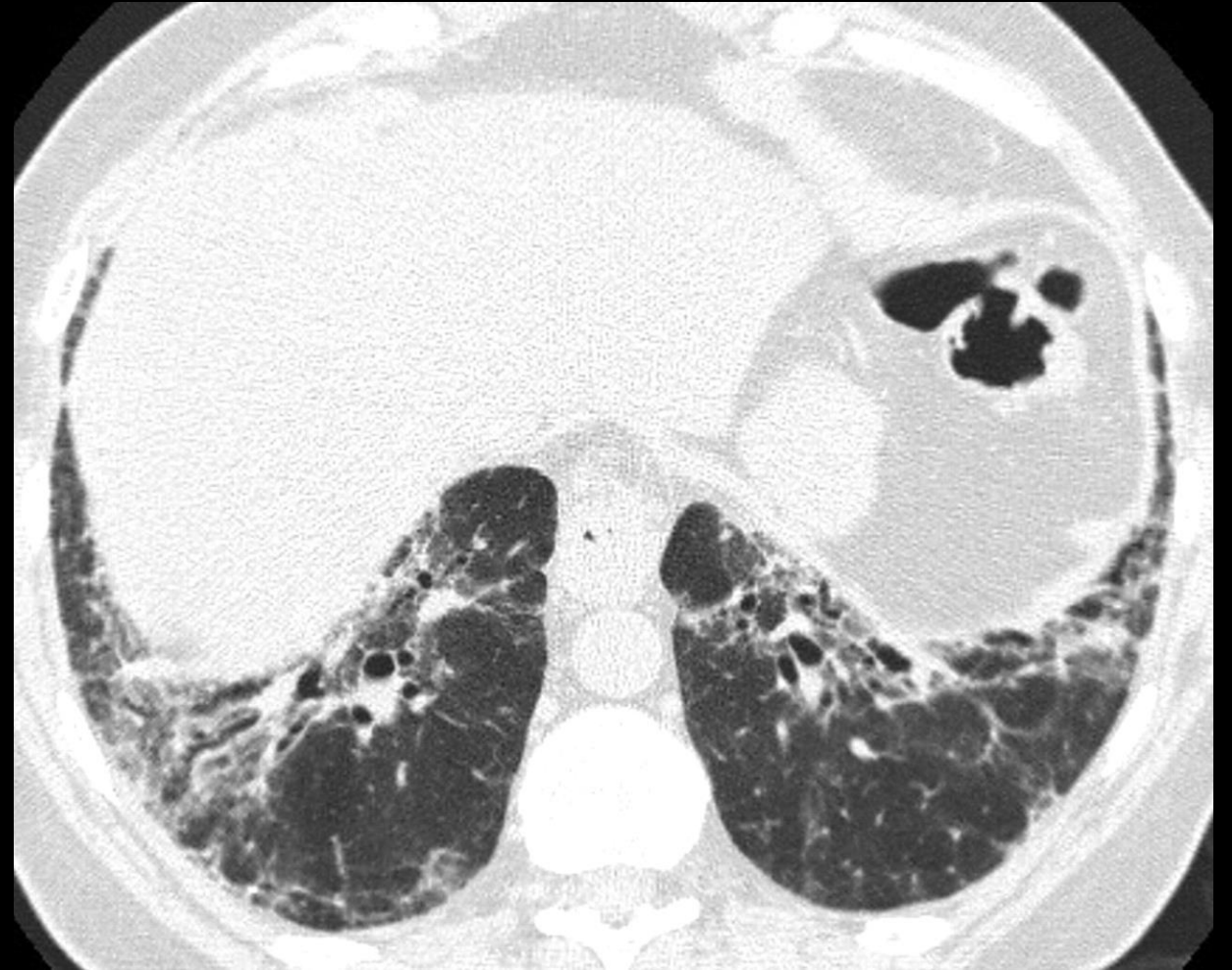
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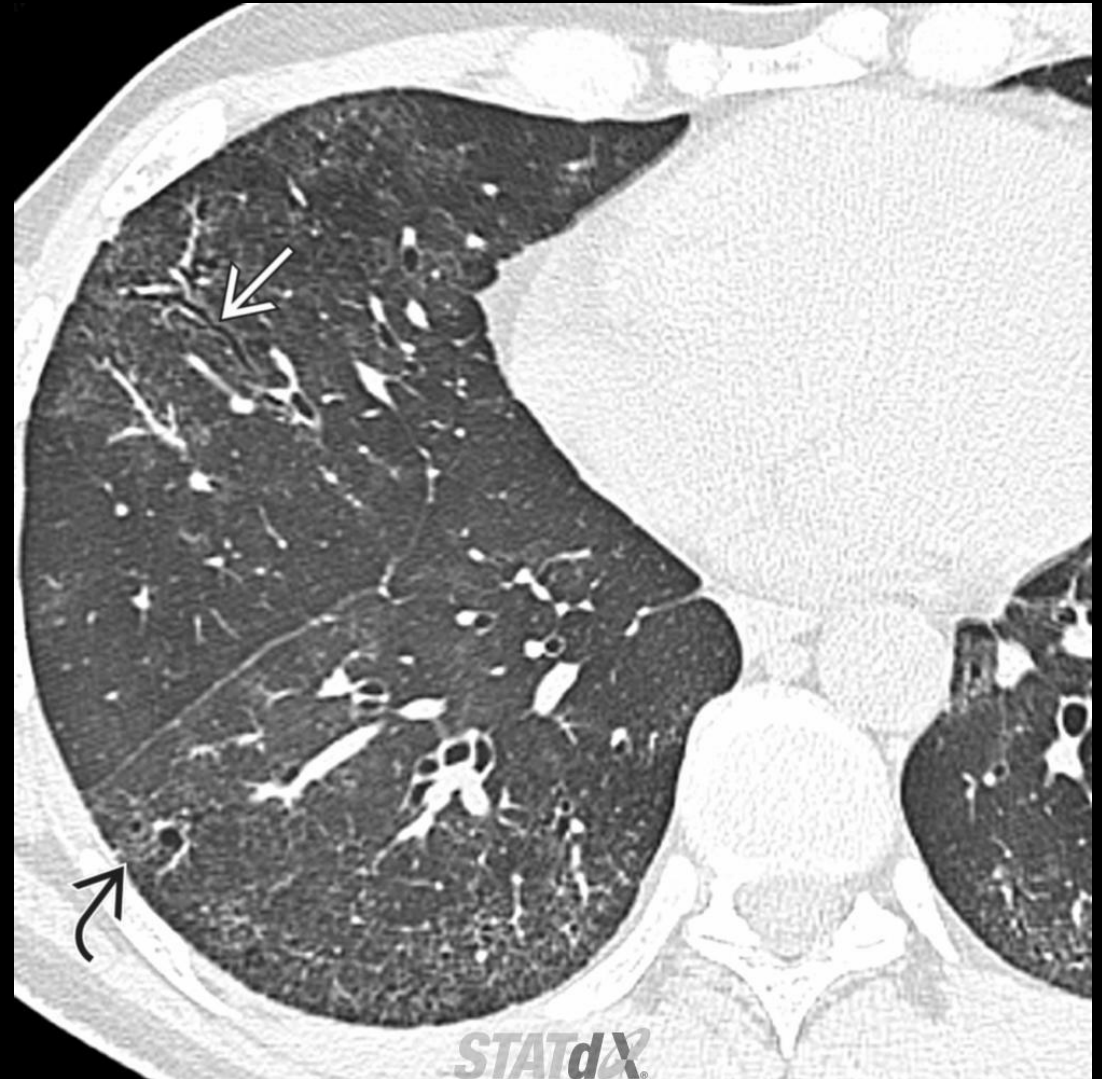
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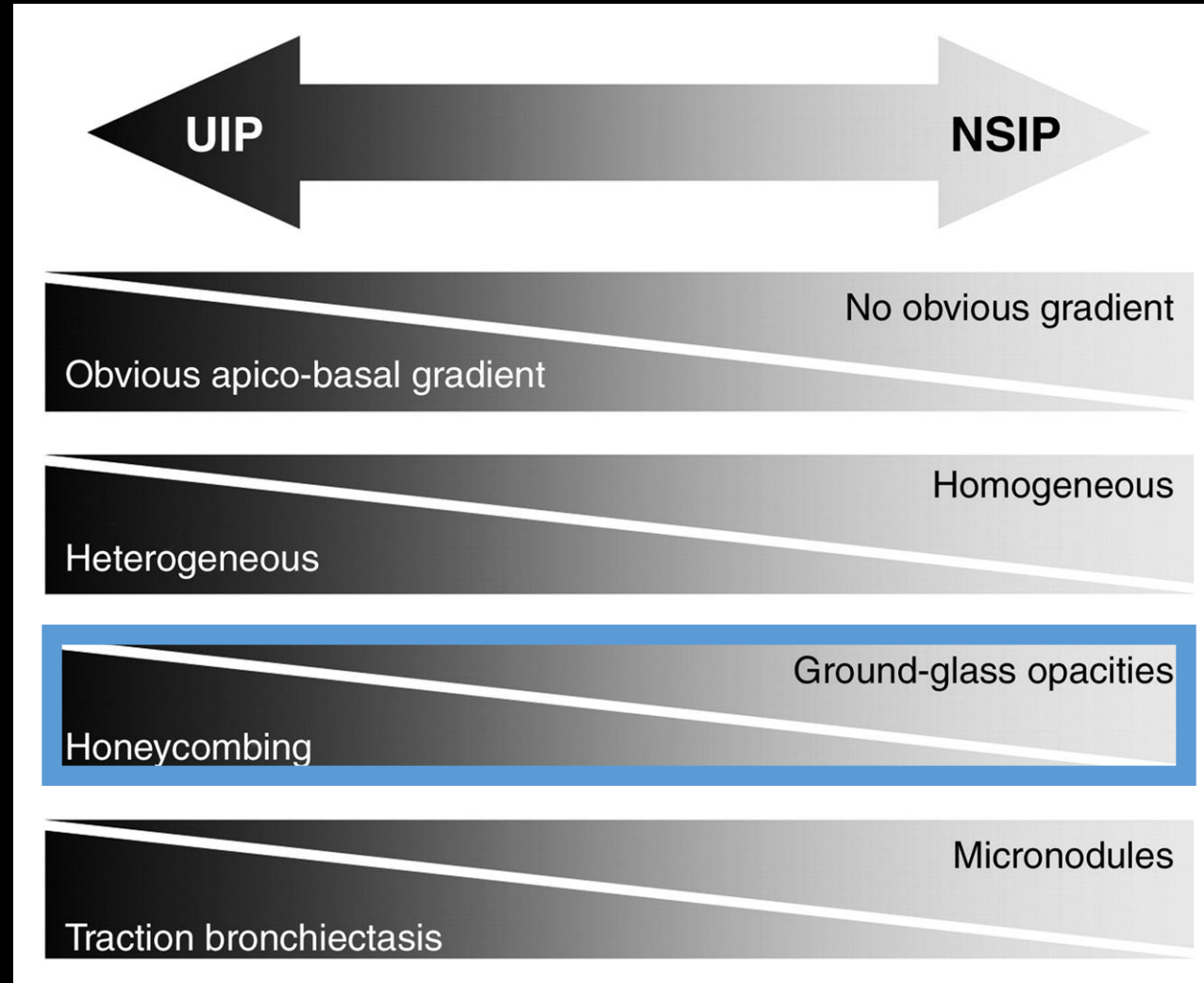
# Nonspecific Interstitial Pneumonia (NSIP)

CT Finding	Sensitivity (%)	Specificity (%)	Accuracy (%)
Diagnosis of NSIP			
Subpleural sparing	41	100	67
Nodules	22	100	56
Peribronchovascular distribution	37	95	63
Ground-glass appearance of 15% or greater	15	95	50

# Differentiating UIP and NSIP



# Differentiating UIP and NSIP



# Differentiating UIP and NSIP

CT Finding	Pathologic UIP Pattern	Pathologic NSIP Pattern	PValue	All	Interobserver Variability		Predictor of Outcome	
					rValue	PValue	Hazard Ratio	95% Confidence Interval
Spared area	61.0 ± 16.0*	64.1 ± 13.4*	.52	62.0 ± 15.1*	0.53	<.001	0.99	0.97, 1.01
Ground-glass attenuation without traction bronchiectasis	4.9 ± 6.2*	7.2 ± 6.2*	.003†	5.7 ± 6.3*	0.57	<.001	0.97	0.93, 1.02
Ground-glass attenuation with traction bronchiectasis	13.5 ± 8.8*	12.6 ± 8.1*	.69	13.2 ± 8.5*	0.67	<.001	1.01	0.98, 1.04
Air-space consolidation	4.4 ± 5.6*	5.7 ± 5.4*	.14	4.9 ± 5.6*	0.83	<.001	1.03	0.98, 1.08
Honeycombing	1.5 ± 3.3*	0.2 ± 1.2*	<.001†	1.0 ± 2.8*	0.44	<.001	1.19	1.11, 1.27
Intralobular reticular opacity	9.9 ± 5.5*	5.2 ± 4.3*	<.001†	8.3 ± 5.6*	0.56	<.001	1.07	1.03, 1.12
Emphysema	1.7 ± 4.4*	1.8 ± 4.0*	.51	1.8 ± 4.3*	0.82	<.001	0.94	0.86, 1.02
Segments of traction bronchiectasis	13.8 ± 4.0‡	11.9 ± 4.0‡	.01	13.1 ± 4.1‡	0.68	<.001	1.19	1.08, 1.31
Subpleural spared area	7§	12§	.007†	19§	0.21	.19	0.09	0.01, 0.67
Upper subpleural linear shadow	63§	12§	<.001†	75§	0.43	.005	2.99	1.44, 6.19
Lower predominance	57§	33§	.34	90§	0.40	.002	0.60	0.32, 1.12
Peripheral predominance	59§	17§	<.001†	76§	0.57	<.001	1.90	0.97, 3.73
Peribronchovascular predominance	11§	16§	.003†	27§	0.81	<.001	0.49	0.21, 1.16
Asymmetry	10§	2§	.22	12§	0.19	.019	2.25	1.01, 5.05

# Differentiating UIP and NSIP

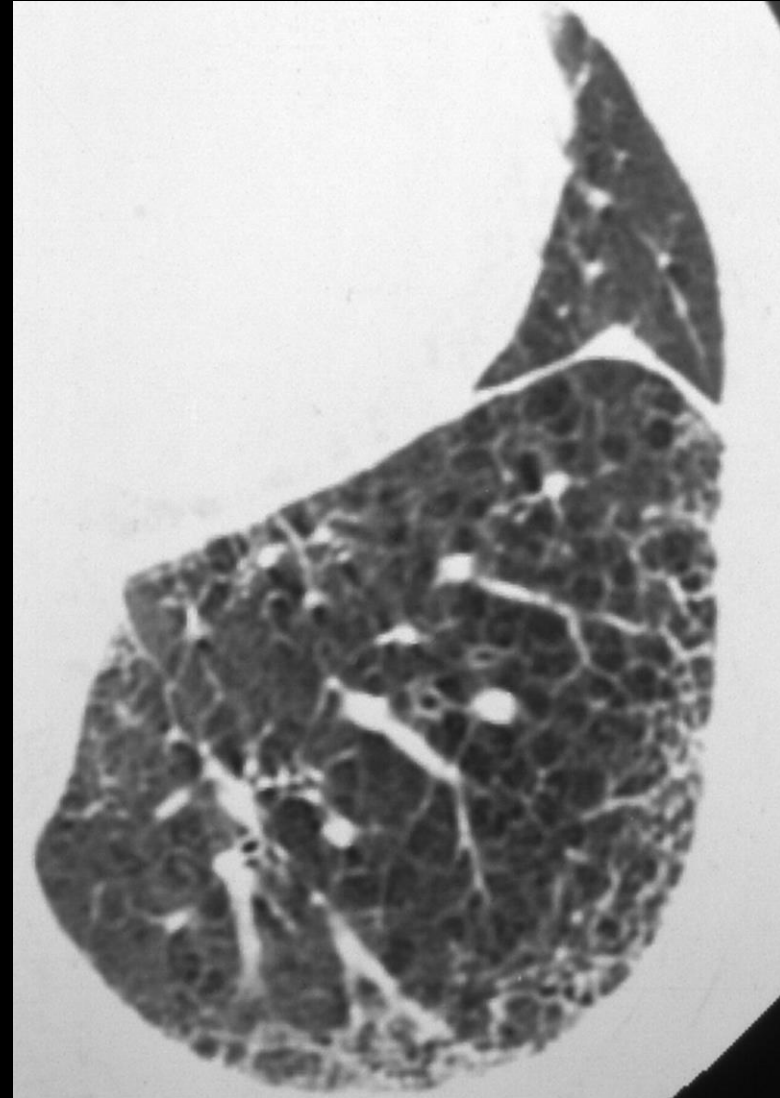
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<b>UIP</b>			



# Differentiating UIP and NSIP

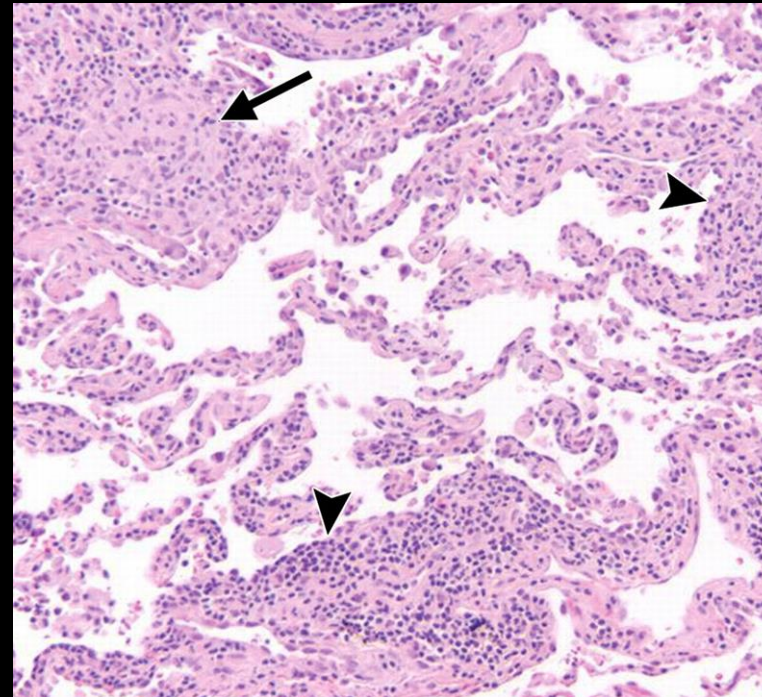
BE CAREFUL IN THE SETTING OF  
EMPHYSEMA!

This is NSIP with emphysema, not  
honeycombing of UIP ☹️



# Hypersensitivity Pneumonitis (HP)

A histologic pattern of cellular bronchiolitis, diffuse interstitial inflammatory (lymphocyte-predominant) infiltrates, poorly circumscribed interstitial nonnecrotizing granulomas, and alveolar or interstitial giant cells



# Hypersensitivity Pneumonitis (HP)

## Acute HP

- A. Centrilobular ground glass nodules
- B. Bilateral ground glass opacity
- C. Relative basilar sparing

## Subacute HP

- A. Centrilobular ground glass nodules
- B. Patchy ground glass
- C. Air trapping (mosaicism)

## Chronic HP

- A. Fibrosis (and its sequelae, including honeycombing) with relative basilar sparing
- B. Residua of Subacute HP

# Hypersensitivity Pneumonitis (HP)

OLD WAY

Acute HP

Subacute HP

Chronic HP



NEW WAY

Nonfibrotic HP

Fibrotic HP

# Hypersensitivity Pneumonitis (HP)

**Table 5. Chest HRCT Scan Features of the Nonfibrotic HP Pattern**

HRCT Pattern	Typical HP	Compatible with HP	Indeterminate for HP
Description	The “typical HP” pattern is suggestive of a diagnosis of HP. It requires <i>a</i> ) at least one HRCT abnormality indicative of parenchymal infiltration and <i>b</i> ) at least one HRCT abnormality indicative of small airway disease, both in a diffuse distribution	“Compatible-with-HP” patterns are nonspecific patterns that have been described in HP	N/A
Relevant radiological findings	<p>HRCT abnormalities indicative of parenchymal infiltration:</p> <ul style="list-style-type: none"> <li>• GGOs</li> <li>• Mosaic attenuation*</li> </ul> <p>HRCT abnormalities indicative of small airway disease:</p> <ul style="list-style-type: none"> <li>• Ill-defined, centrilobular nodules</li> <li>• Air trapping</li> </ul> <p>Distribution of parenchymal abnormalities:</p> <ul style="list-style-type: none"> <li>• Craniocaudal: diffuse (with or without some basal sparing)</li> <li>• Axial: diffuse</li> </ul>	<p>Parenchymal abnormalities:</p> <ul style="list-style-type: none"> <li>• Uniform and subtle GGOs</li> <li>• Airspace consolidation</li> <li>• Lung cysts</li> </ul> <p>Distribution of parenchymal abnormalities:</p> <ul style="list-style-type: none"> <li>• Craniocaudal: diffuse (variant: lower lobe predominance)</li> <li>• Axial: diffuse (variant: peribronchovascular)</li> </ul>	N/A

# Hypersensitivity Pneumonitis (HP)

**Table 6. Chest HRCT Scan Features of the Fibrotic HP Pattern**

HRCT Pattern	Typical HP	Compatible with HP	Indeterminate for HP
Description	The “typical HP” pattern is suggestive of a diagnosis of HP. It requires <i>a</i> ) an HRCT pattern of lung fibrosis (as listed below) in one of the distributions and <i>b</i> ) at least one abnormality that is indicative of small airway disease	“Compatible-with-HP” patterns exist when the HRCT pattern and/or distribution of lung fibrosis varies from that of the typical HP pattern; the variant fibrosis should be accompanied by signs of small airway disease	The “indeterminate-for-HP” pattern exists when the HRCT is neither suggestive nor compatible with a typical and probable HP pattern
Relevant radiological findings	<p>HRCT abnormalities indicative of lung fibrosis are most commonly composed of</p> <p>irregular linear opacities/coarse reticulation with lung distortion; traction bronchiectasis and honeycombing may be present but do not predominate</p> <p>The distribution of fibrosis may be:</p> <ul style="list-style-type: none"> <li>• Random both axially and craniocaudally or</li> <li>• Mid lung zone–predominant or</li> <li>• Relatively spared in the lower lung zones</li> </ul> <p>HRCT abnormalities indicative of small airway disease:</p> <ul style="list-style-type: none"> <li>• Ill-defined, centrilobular nodules and/or GGOs</li> <li>• Mosaic attenuation, three-density pattern,* and/or air trapping (often in a lobular distribution)</li> </ul>	<p>Variant patterns of lung fibrosis:</p> <ul style="list-style-type: none"> <li>• UIP pattern: basal and subpleural distribution of honeycombing with/without traction bronchiectasis (per 2018 diagnosis of IPF guidelines [20])</li> <li>• Extensive GGOs with superimposed subtle features of lung fibrosis</li> </ul> <p>Variant (predominant) distributions of lung fibrosis:</p> <ul style="list-style-type: none"> <li>• Axial: peribronchovascular, subpleural areas</li> <li>• Craniocaudal: upper lung zones</li> </ul>	<p>Lone patterns (i.e., not accompanied by other findings suggestive of HP) of:</p> <ul style="list-style-type: none"> <li>• UIP pattern (as per 2018 IPF diagnosis guidelines [20])</li> <li>• Probable UIP pattern (as per 2018 IPF diagnosis guidelines [20])</li> <li>• Indeterminate pattern for UIP (as per 2018 IPF diagnosis guidelines [20])</li> <li>• Fibrotic NSIP pattern</li> <li>• Organizing pneumonia–like pattern</li> <li>• Truly indeterminate HRCT pattern</li> </ul>
		<p>HRCT abnormalities indicative of small airway disease:</p> <ul style="list-style-type: none"> <li>• Ill-defined centrilobular nodules, or</li> <li>• Three-density pattern* and/or air trapping</li> </ul>	

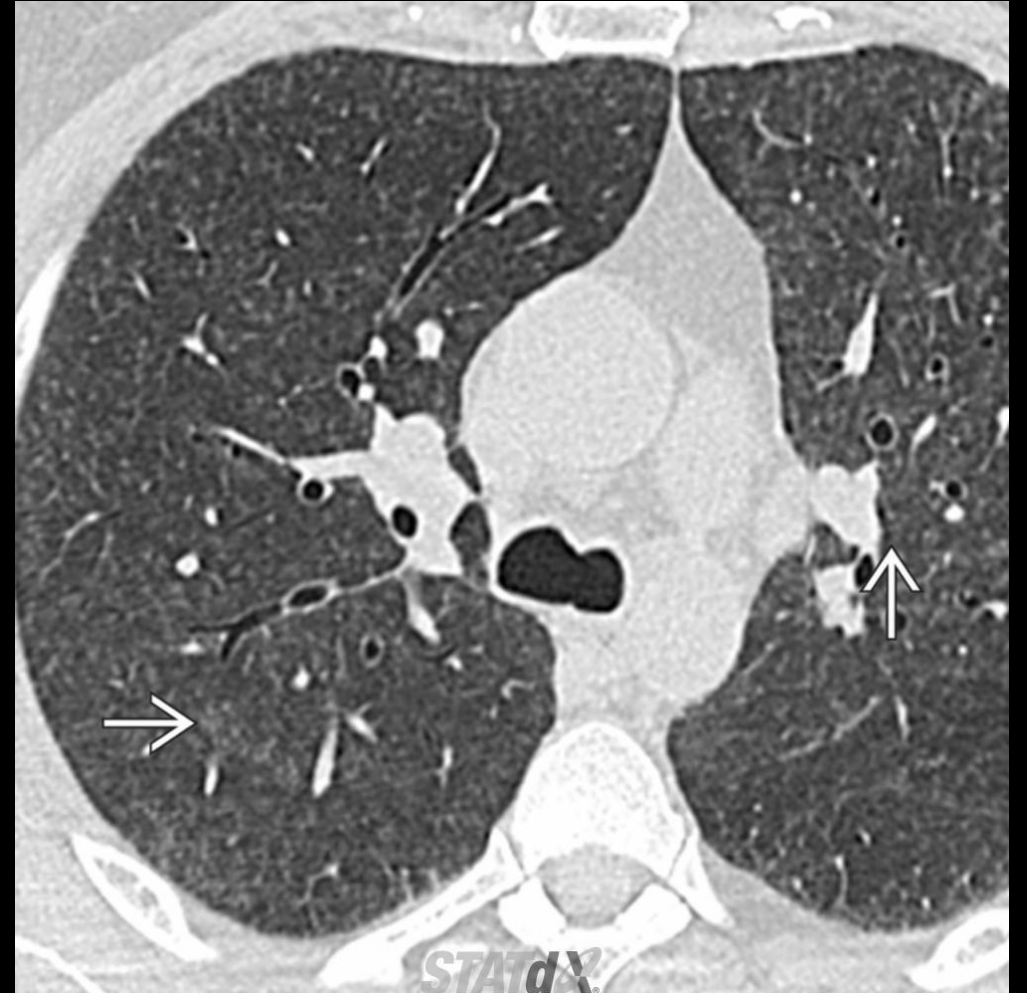
# Hypersensitivity Pneumonitis (HP)

## Acute HP

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- C. Relative basilar sparing

## Subacute HP

- A. Centrilobular ground glass nodules
- B. Patchy ground glass
- C. Air trapping (mosaic attenuation)



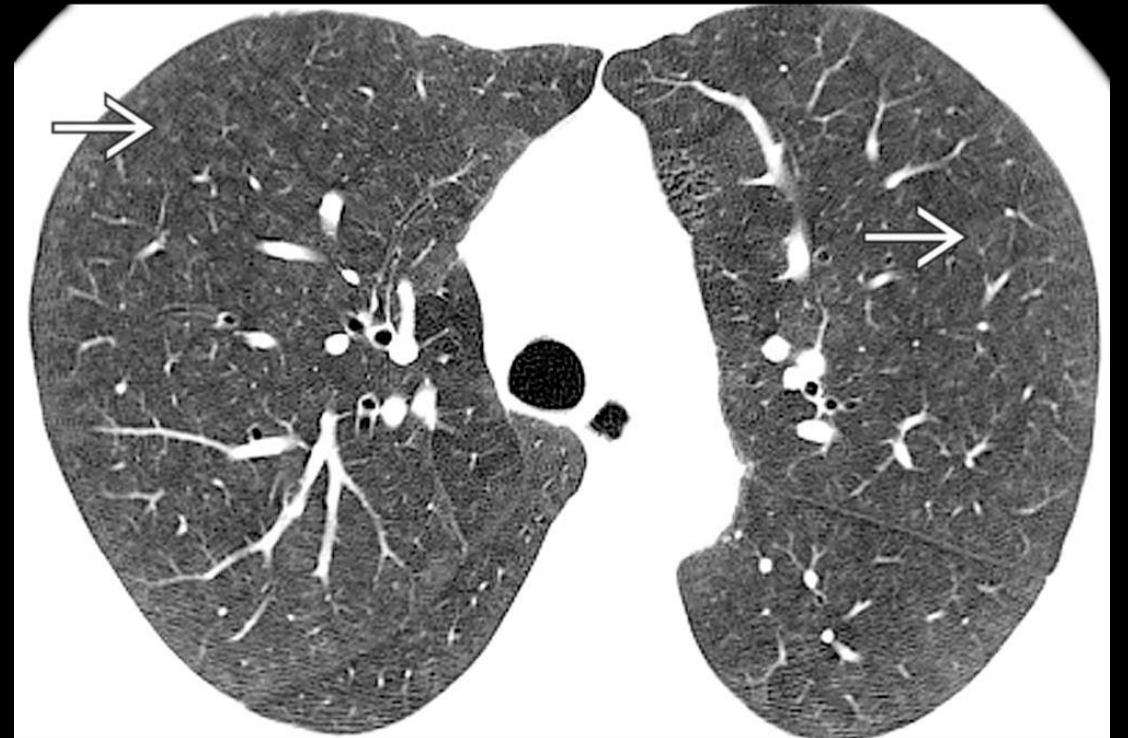
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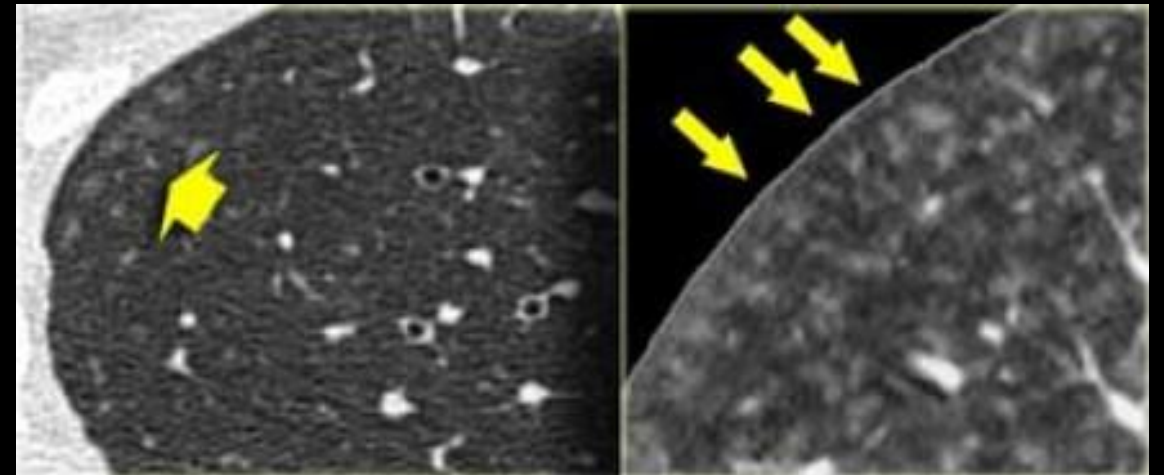
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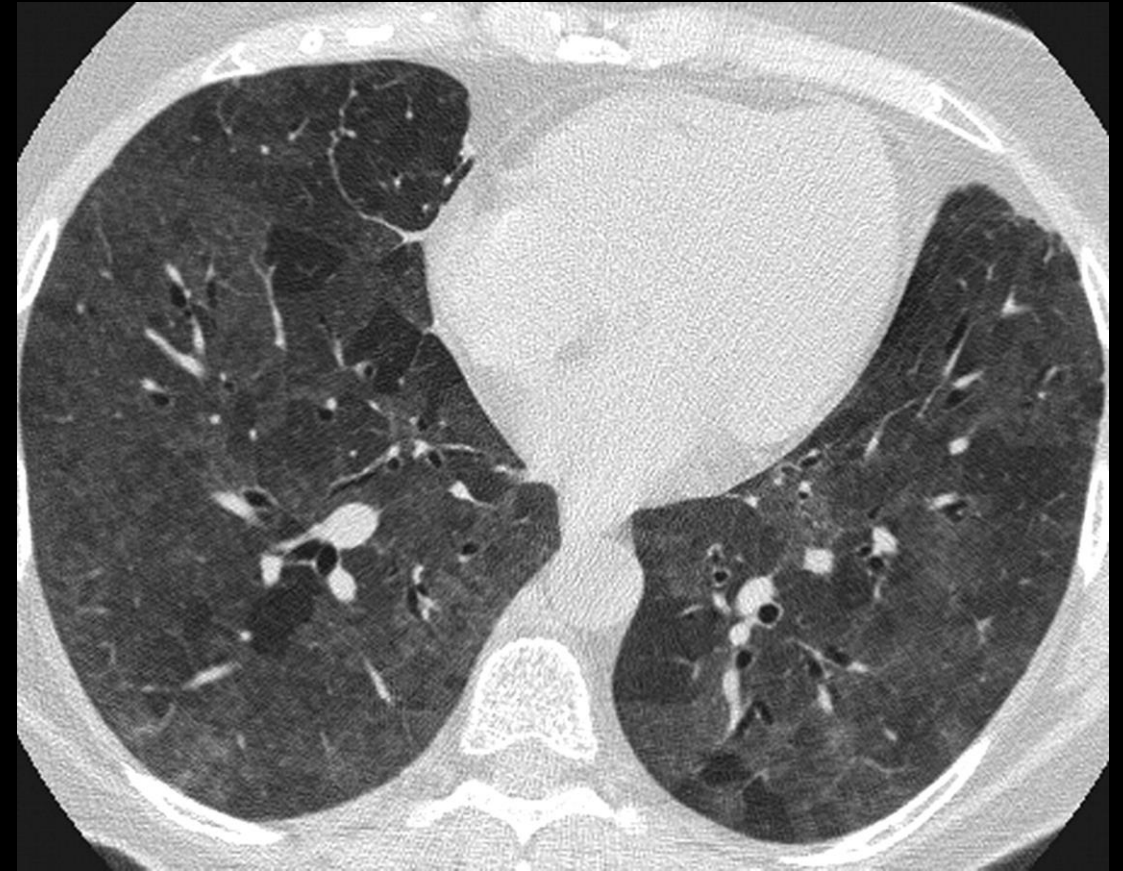
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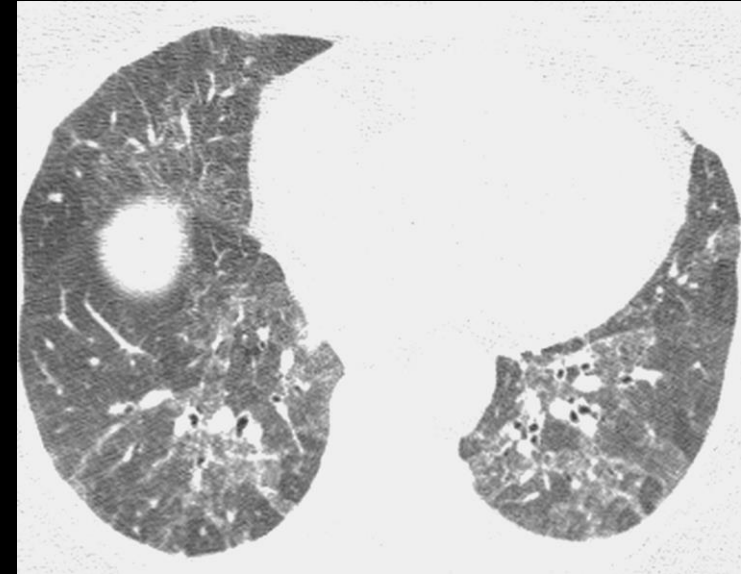
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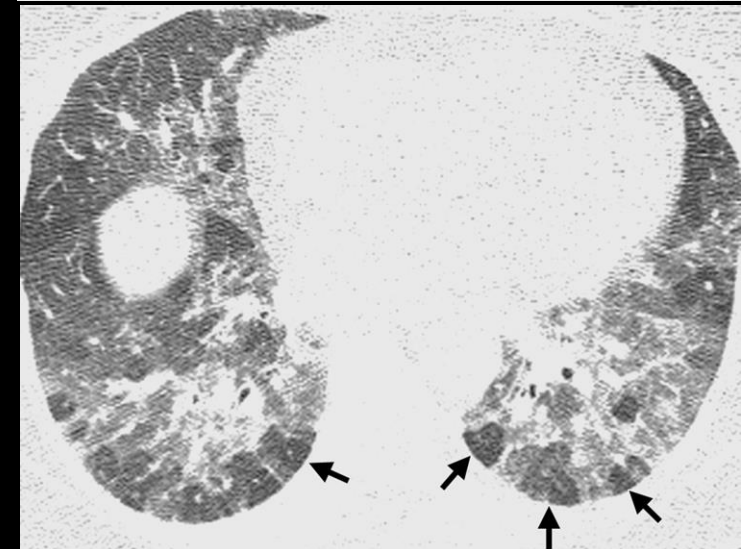
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Three density sign: Co-existence of ground glass, air trapping, and normal lung



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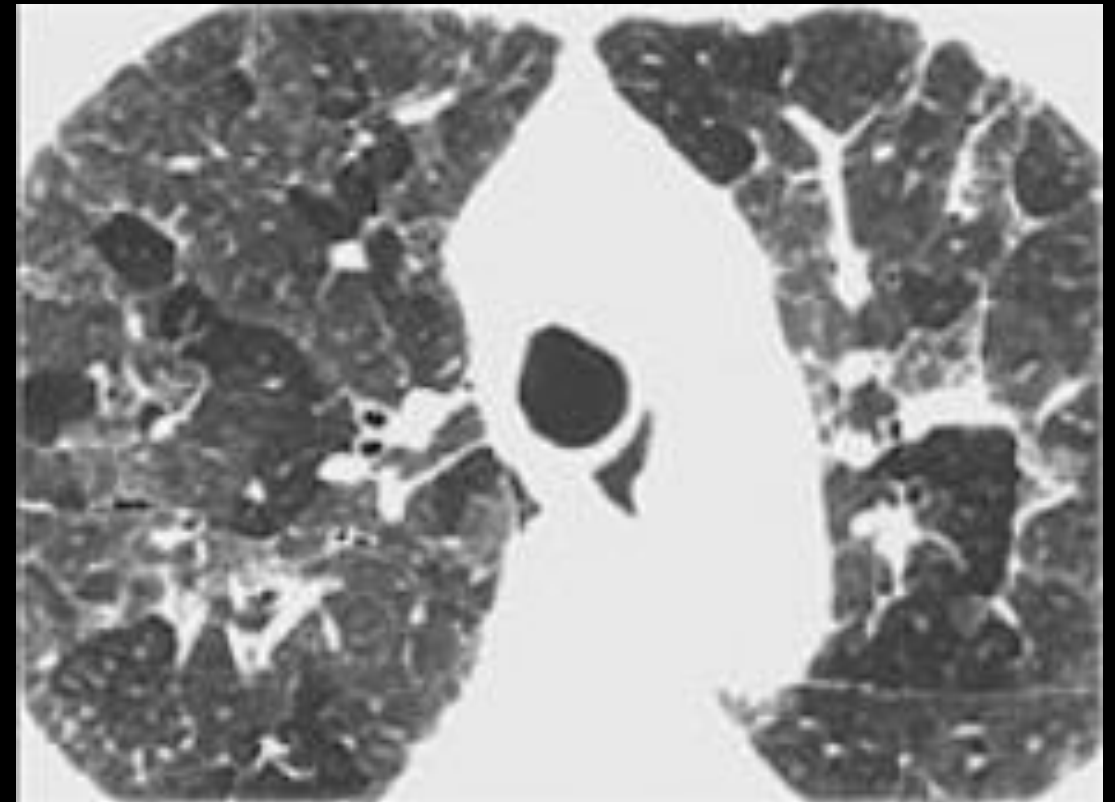
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- B. Bilateral ground glass opacity
- C. Relative basilar sparing

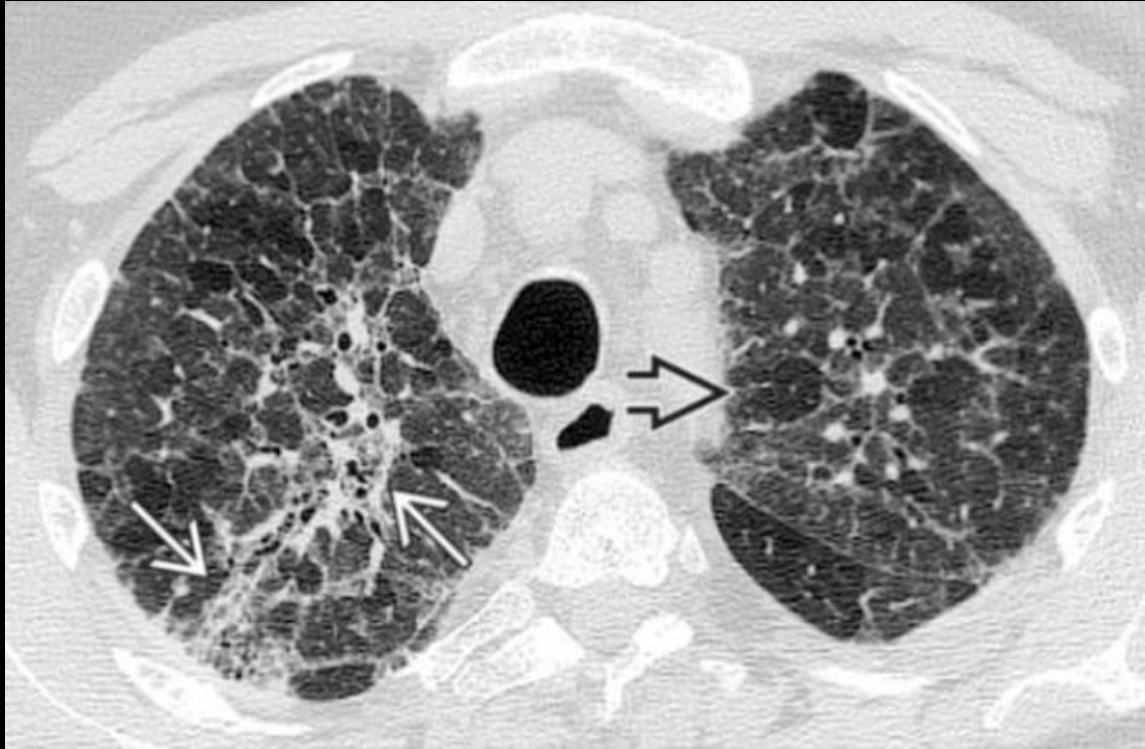
## Subacute HP

- A. Centrilobular ground glass nodules
- B. Patchy ground glass
- C. Air trapping (mosaicism)

Three density sign: Co-existence of ground glass, air trapping, and normal lung



# Hypersensitivity Pneumonitis (HP)



## Chronic HP

- A. Fibrosis (and its sequelae, including honeycombing)
- B. Residua of Subacute HP, including the Three density sign



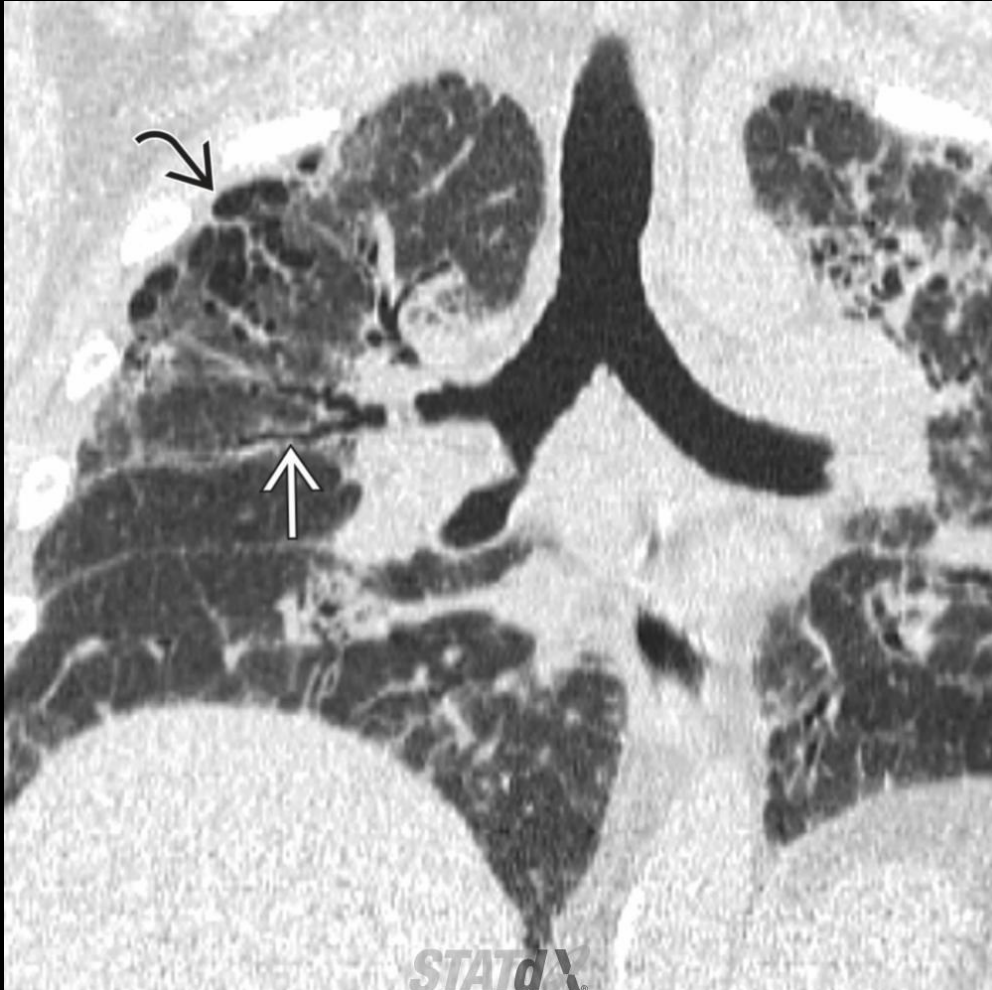
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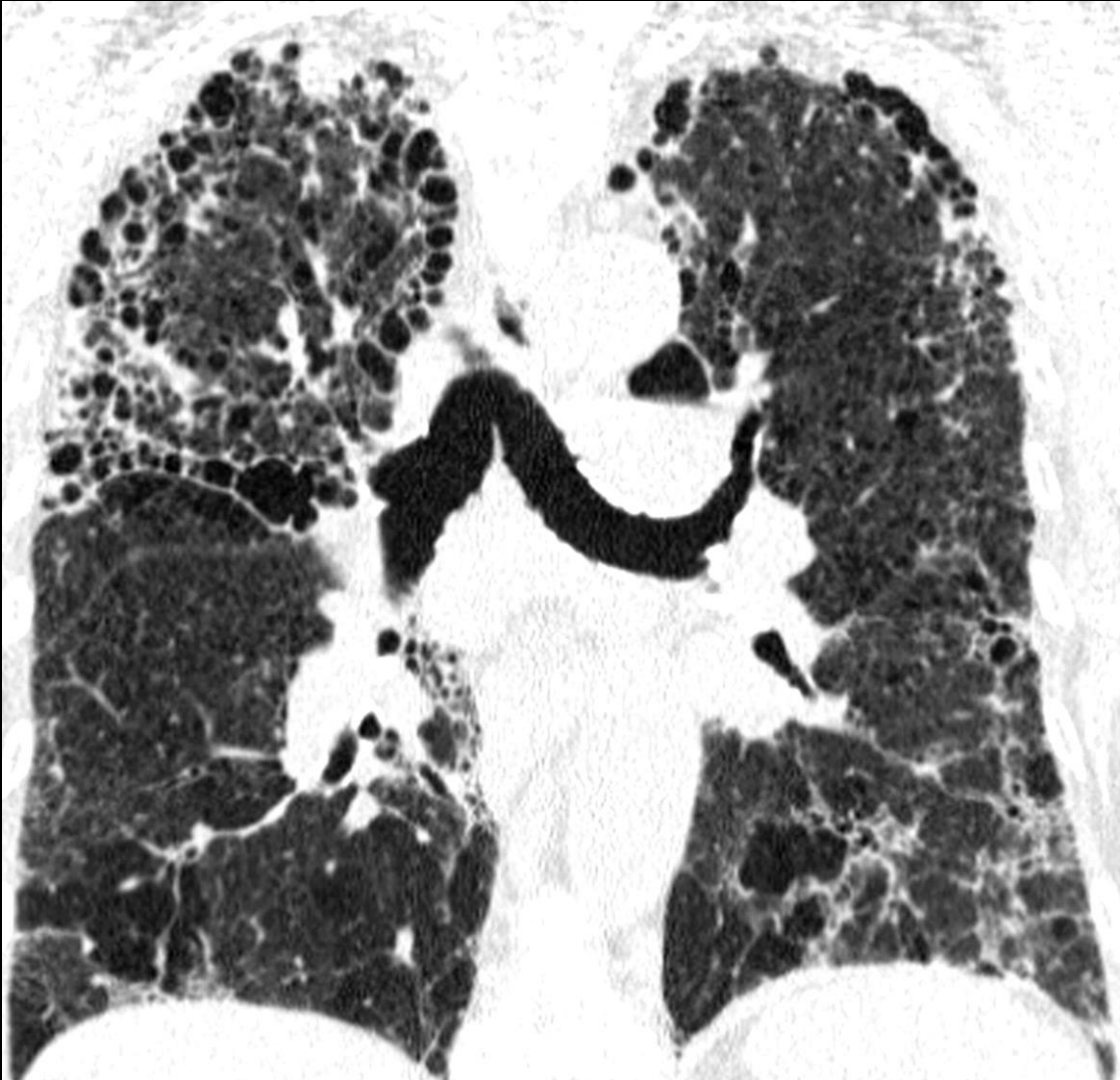
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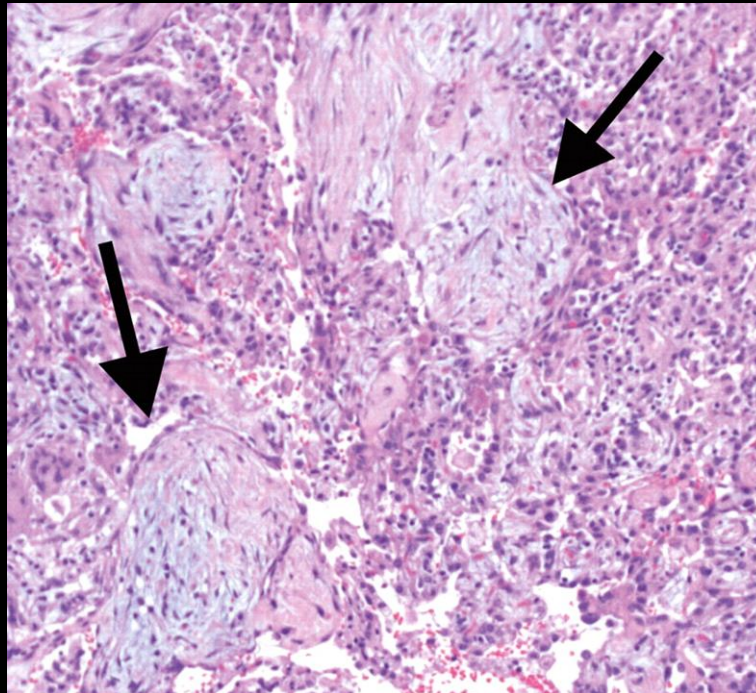


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# Organizing Pneumonia (OP)

A histologic pattern of patchy but temporally uniform intraluminal organizing fibrosis in distal airspaces. Endoluminal buds of granulation tissue (Masson bodies) are characteristic.



# Organizing Pneumonia (OP)

## Forms and features:

- A. Nodular
- B. Peripheral
- C. Peribronchovascular
- D. Bandlike
- E. Atoll or Reverse Halo sign
- F. Perilobular sign
- G. Progressive fibrosis

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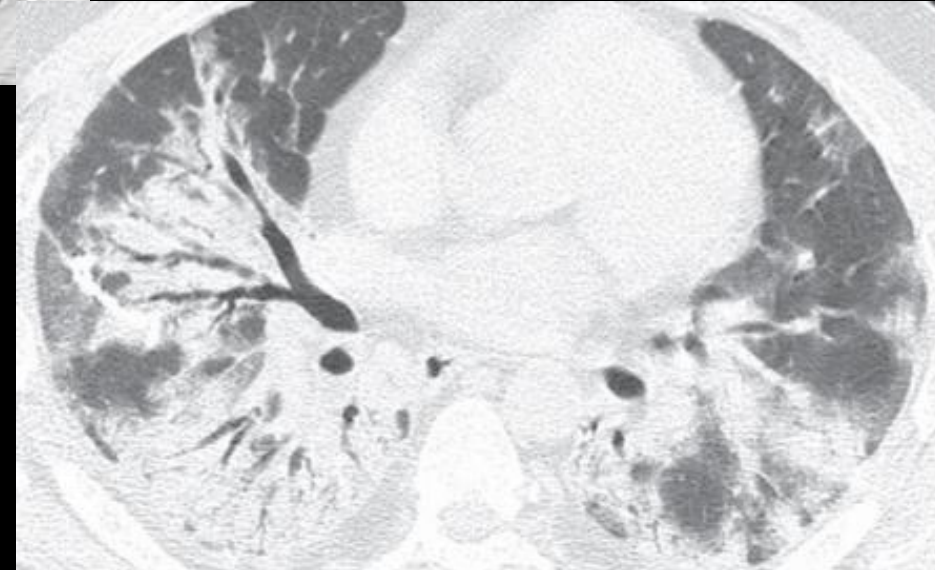
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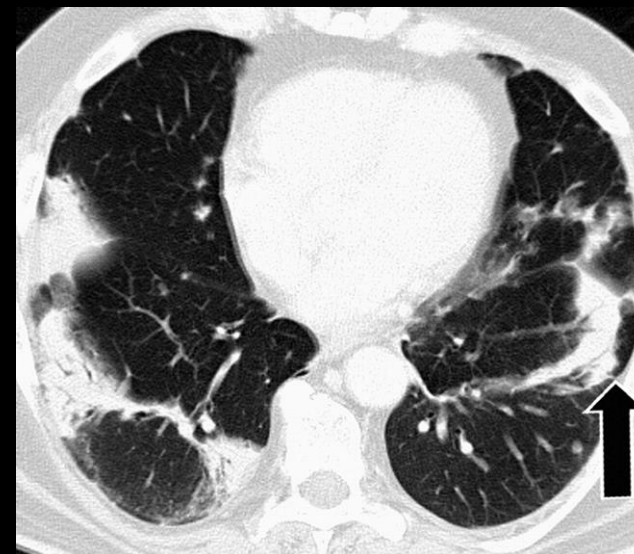




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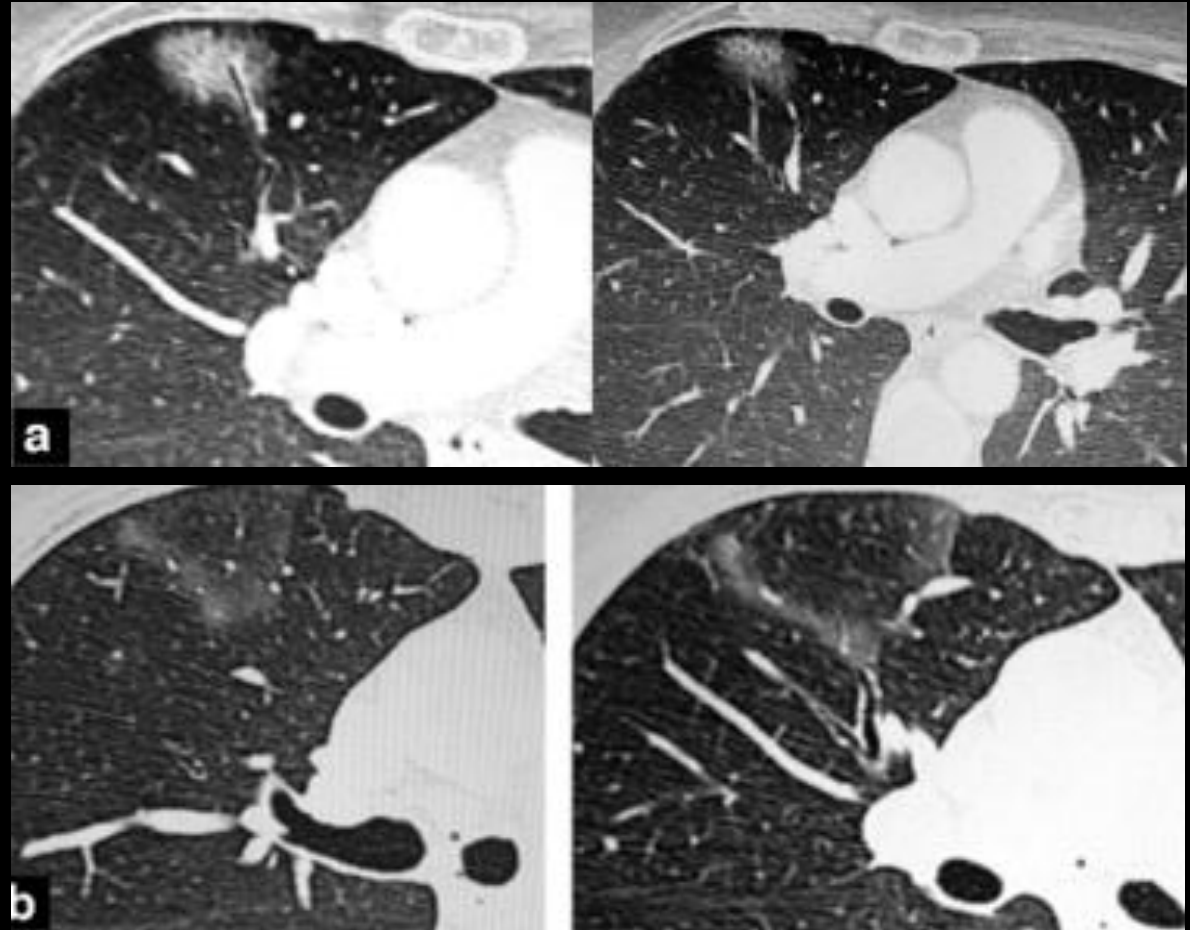
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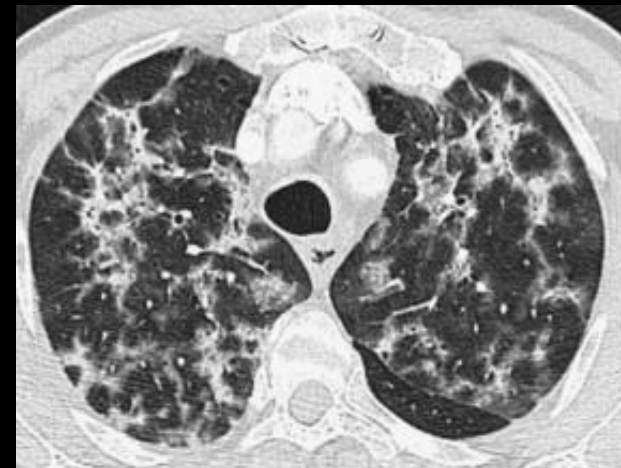
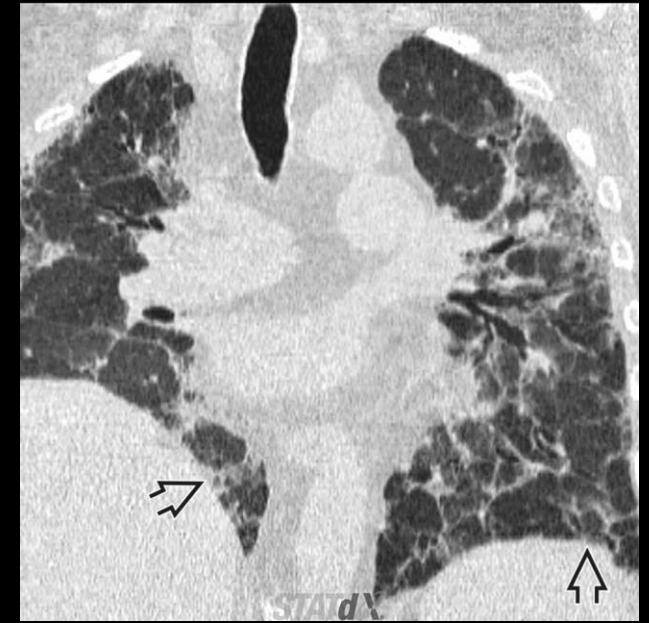
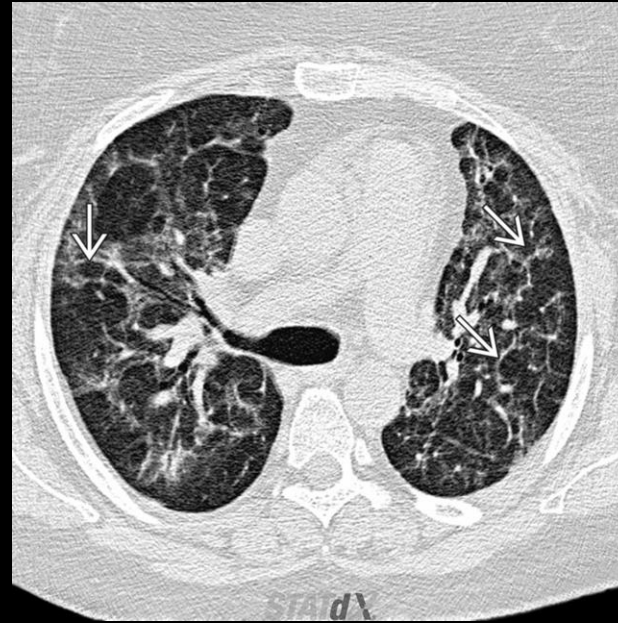
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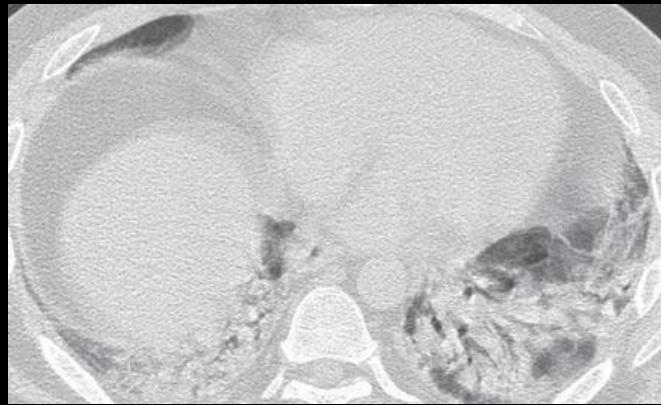
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# Organizing Pneumonia (OP)



# Organizing Pneumonia (OP)

Parameter	Initial CT	
	No. (%) of Patients	Average Extent (%)
<b>Pattern</b>		
Consolidation	17 (77)	27
Ground-glass opacification	19 (86)	33
Nodule	7 (32)	10
Reticulation	4 (18)	10
Honeycombing	0	0
<b>Distribution</b>		
Lower lung predominance	12 (55)	NA
Subpleural	9 (41)	NA
Peribronchovascular	5 (23)	NA

Note— NA = not applicable.

# ILD : The Big 4

UIP

NSIP

HP

OP





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# Questions?