

# FIBROSING INTERSTITIAL LUNG DISEASES OF IDIOPATHIC AND EXOGENOUS ORIGIN. PHENOTYPE APPROACH.

Conference and Postgradual Course

## How to diagnose UIP

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# Usual Interstitial Pneumonia (UIP) – Idiopathic Pulmonary Fibrosis (IPF)

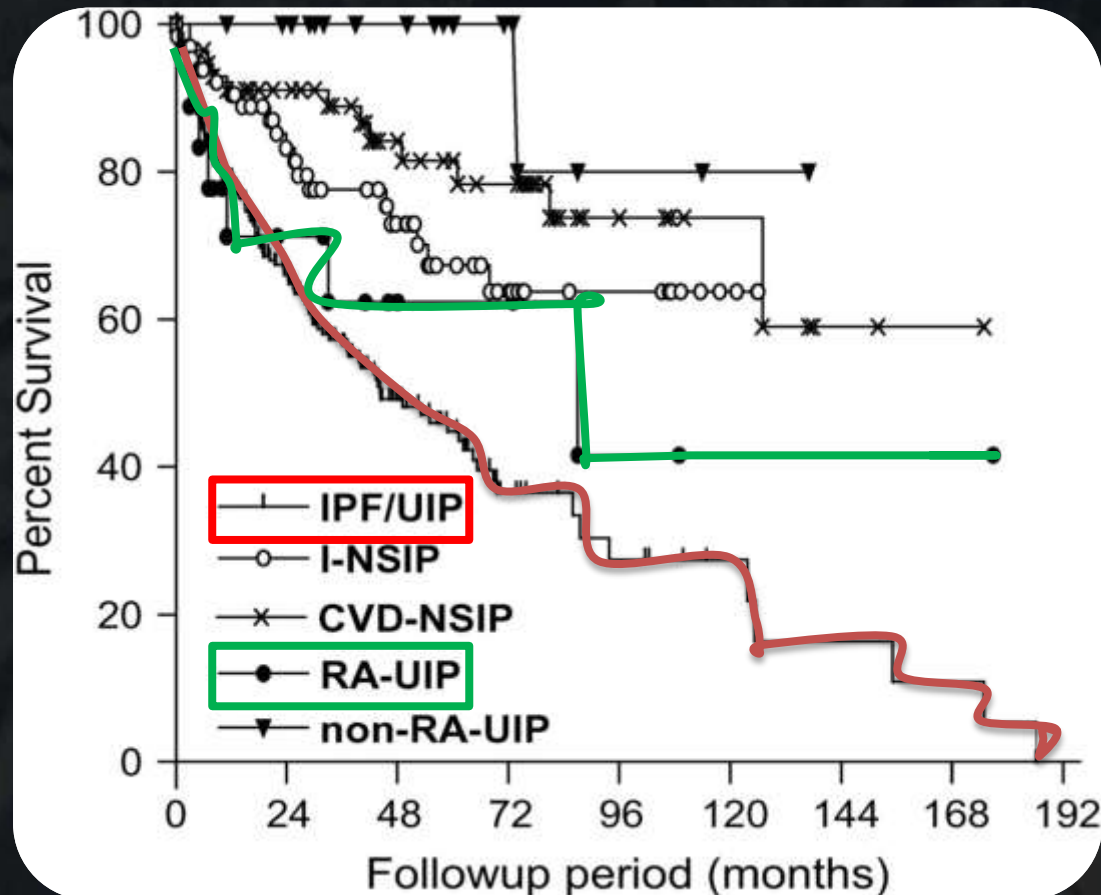
- The most common and progressive fibrotic lung disease

	CLINICAL-RADIOLOGIC-PATHOLOGIC DIAGNOSES	ASSOCIATED MORPHOLOGIC PATTERNS
Chronic Fibrosing IIP	Idiopathic Pulmonary Fibrosis	Usual Interstitial Pneumonia
	Idiopathic Nonspecific Interstitial Pneumonia	Nonspecific Interstitial Pneumonia
Smoking related IIP	Respiratory Bronchiolitis Interstitial Lung Disease	Respiratory Bronchiolitis
	Desquamative Interstitial Pneumonia	Desquamative Interstitial Pneumonia
Acute/subacute IP	Cryptogenic Organizing Pneumonia	Organizing Pneumonia
	Acute Interstitial Pneumonia	Diffuse Alveolar Damage

# Usual Interstitial Pneumonia (UIP) – Idiopathic Pulmonary Fibrosis (IPF)

- The most common and progressive fibrotic lung disease
- Idiopathic (IPF) or secondary to other conditions
  - ✓ Connective tissue disease
  - ✓ Chronic hypersensitivity pneumonitis
  - ✓ Asbestosis
  - ✓ Familiar interstitial lung disease
  - ✓ Vasculitis
  - ✓ Hermansky Pudlak syndrome

# IPF prognosis worse than any other fibrotic lung diseases



# Usual Interstitial Pneumonia (UIP) – Idiopathic Pulmonary Fibrosis (IPF)

- The most common and progressive fibrotic lung disease
- Idiopathic (IPF) or secondary to other conditions
- Morphologic features:
  - Sometimes recognizable on CT: «definite» UIP pattern
  - Sometimes the CT pattern is «possible» or «inconsistent» with UIP

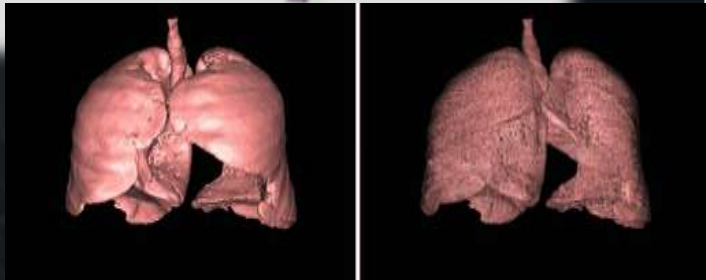
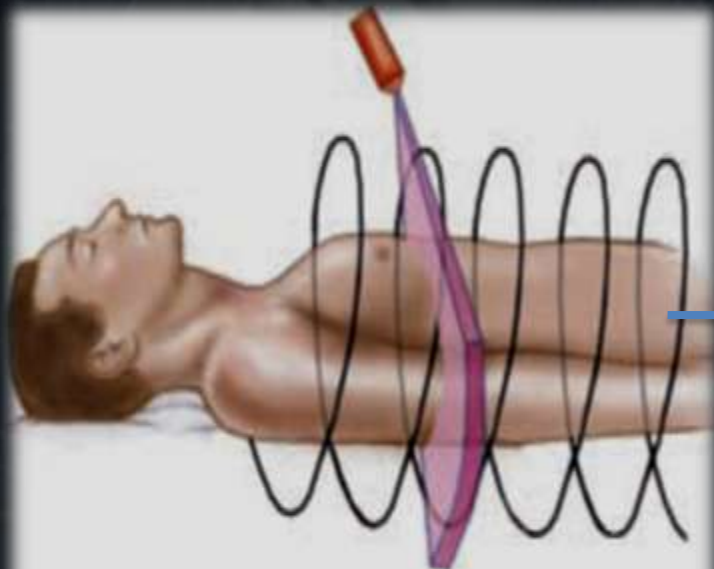


# Learning objectives:

- Key principles for radiologic assessment of UIP
- Identify the CT features of UIP
- Differential diagnosis
- Longitudinal evaluation
  - Acute complications, comorbidities

# Optimal quality CT

Thin-section reconstruction (< 2mm), high spatial reconstruction algorithm, suspended deep inspiration



- Comparison between CT and radiography for qualitative evaluation
- Identification of subtle findings
- Quantification of patchy disease
- Multiplanar reformation

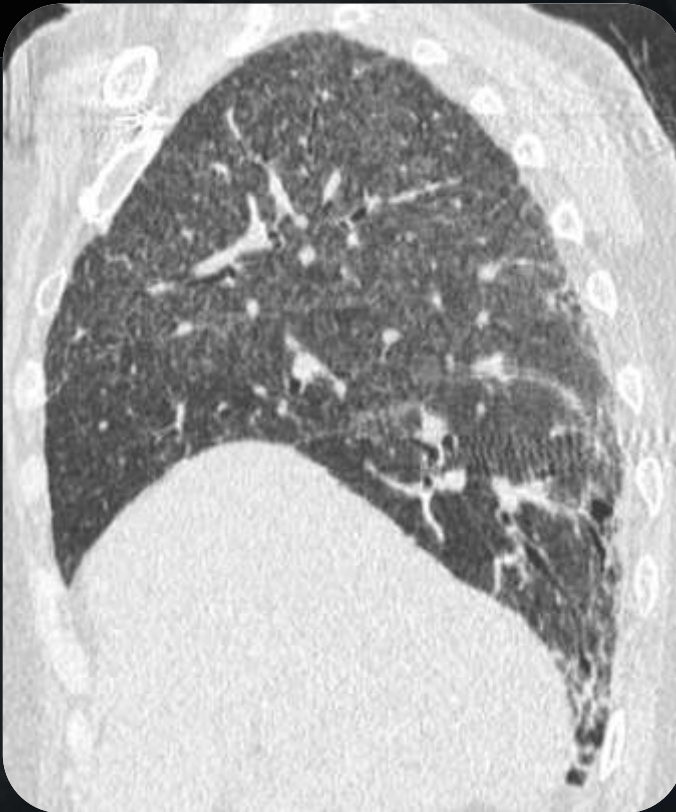




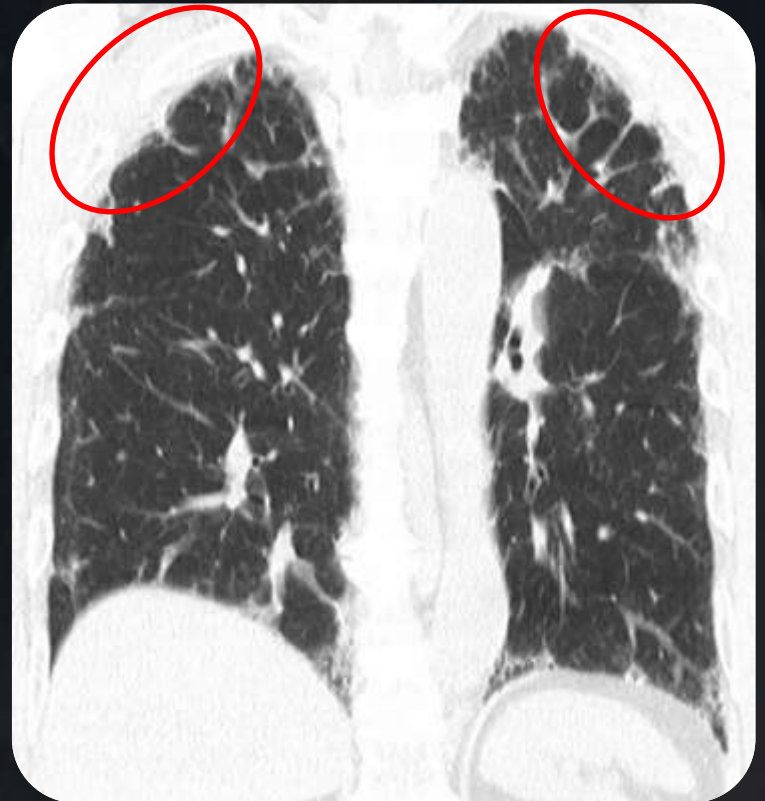


Better delineation of disease extent

Simplify analysis of disease distribution

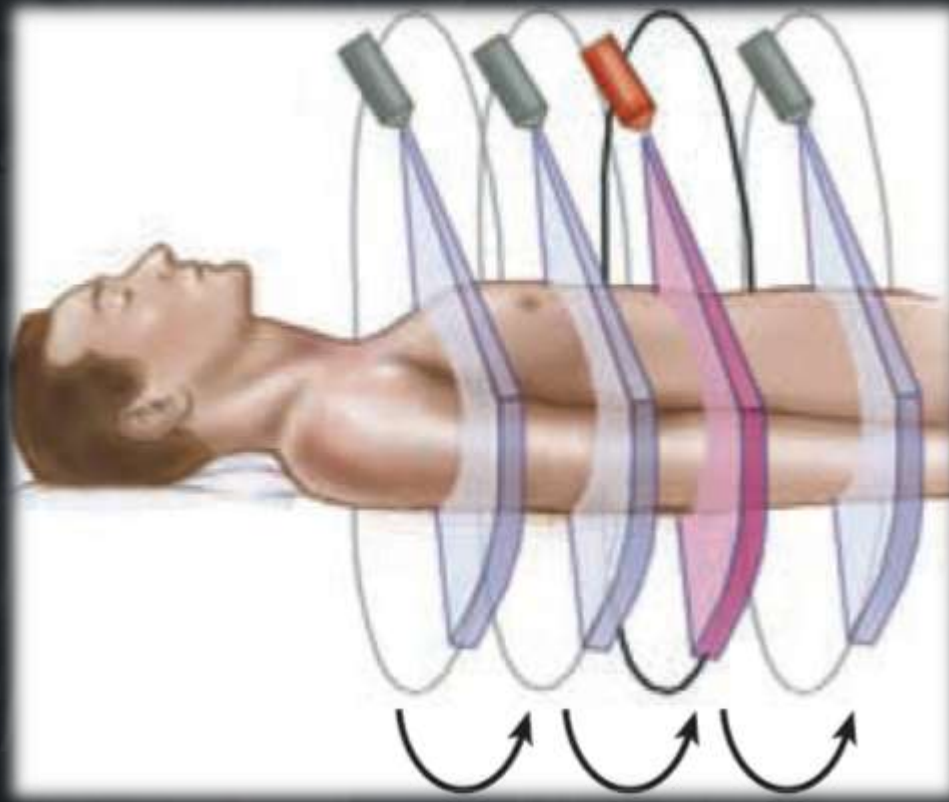


Chronic HP



PPFE

Interspaced HRCT (standard) protocol for younger patients  
(eg <40 yrs old)

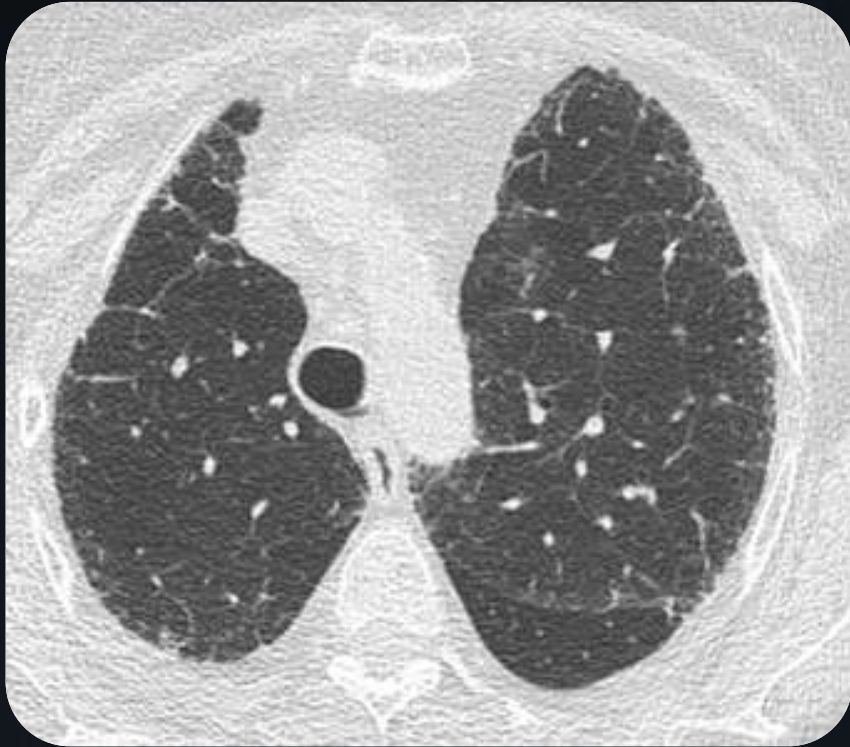


# Supine or Prone?





# Expiratory CT scanning



insp



exp



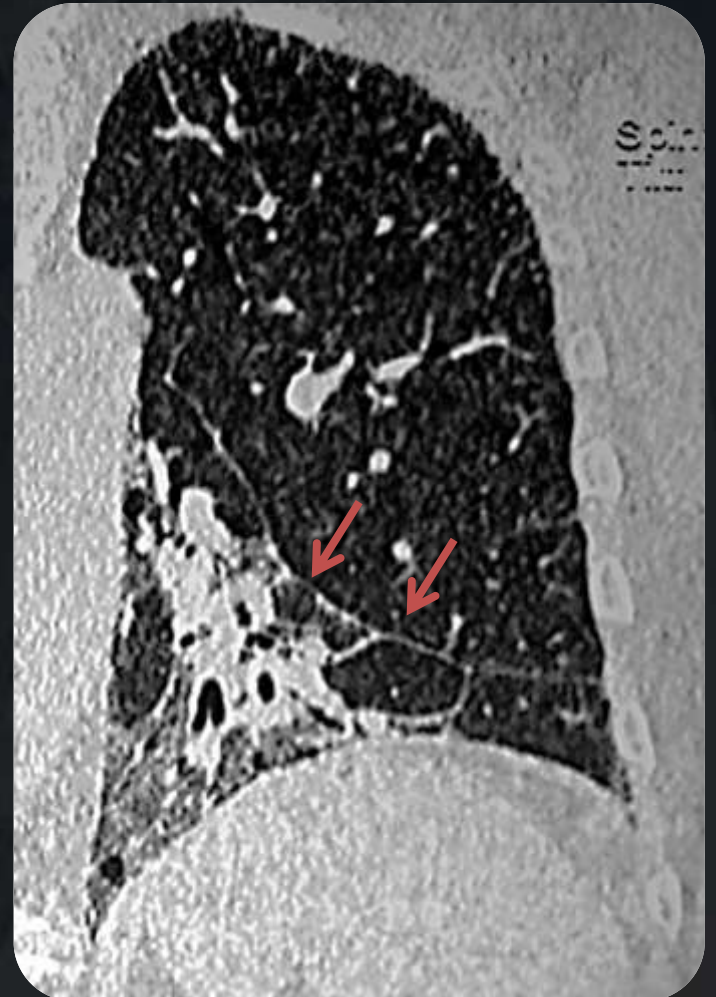
# Systematic approach to CT

- Evaluation of image quality
- Precise description of specific disease features using standard terminology
- Disease distribution
- Is it a fibrotic ILD or non-fibrotic ILD?

# Reminder: CT features of fibrosis

(++++ = complete certainty)

- Honeycombing  
+++(+)
- Traction bronchiectasis  
++(+)
- Volume loss  
+



# Systematic approach to CT

- Evaluation of image quality
- Precise description of specific disease features using standard terminology
- Disease distribution
- Is it a fibrotic ILD or non-fibrotic ILD?
  - If so, is it definite UIP?
  - If no, is possible or inconsistent?
  - what are the alternatives (e.g. fibrotic sarcoid, PPFE etc.)?

# An Official ATS/ERS/JRS/ALAT Statement: Idiopathic pulmonary fibrosis: Evidence-based Guidelines for Diagnosis and Management

## UIP Pattern (All Four Features)

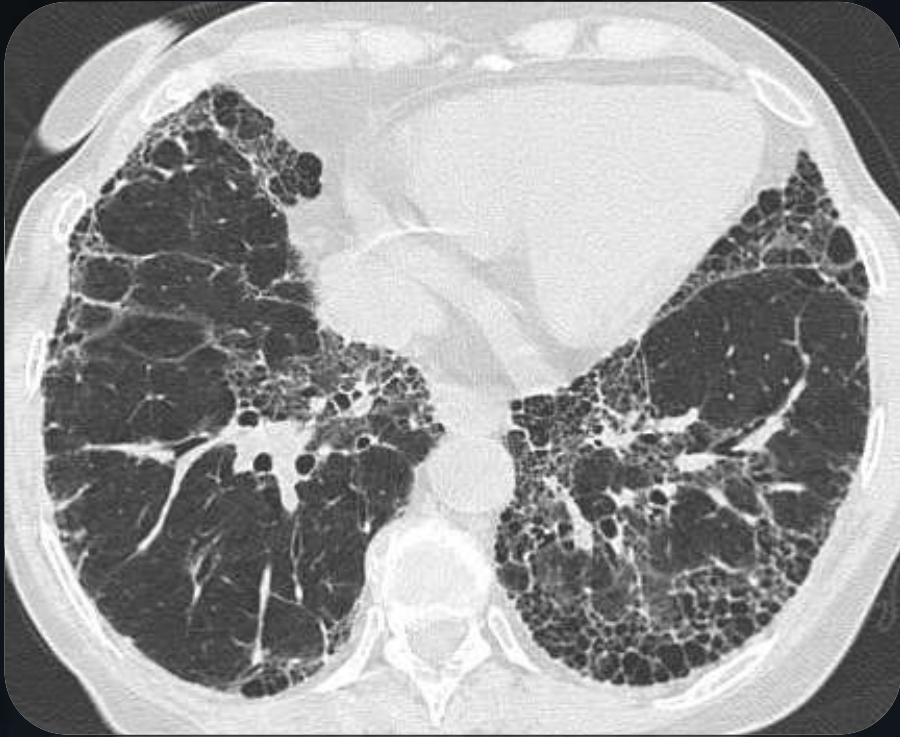
- Subpleural, basal predominance
- Reticular abnormality
- Honeycombing with or without traction bronchiectasis
- Absence of features listed as inconsistent with UIP pattern (see third column)



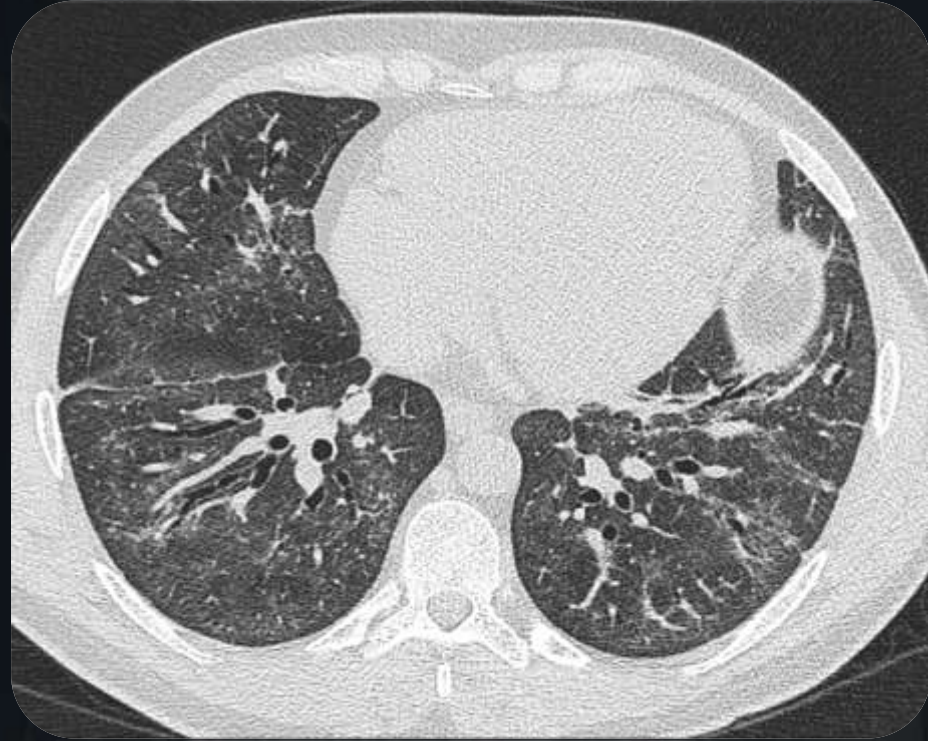
*Raghu G et al, AJRCCM 2011*



# CT patterns



UIP pattern

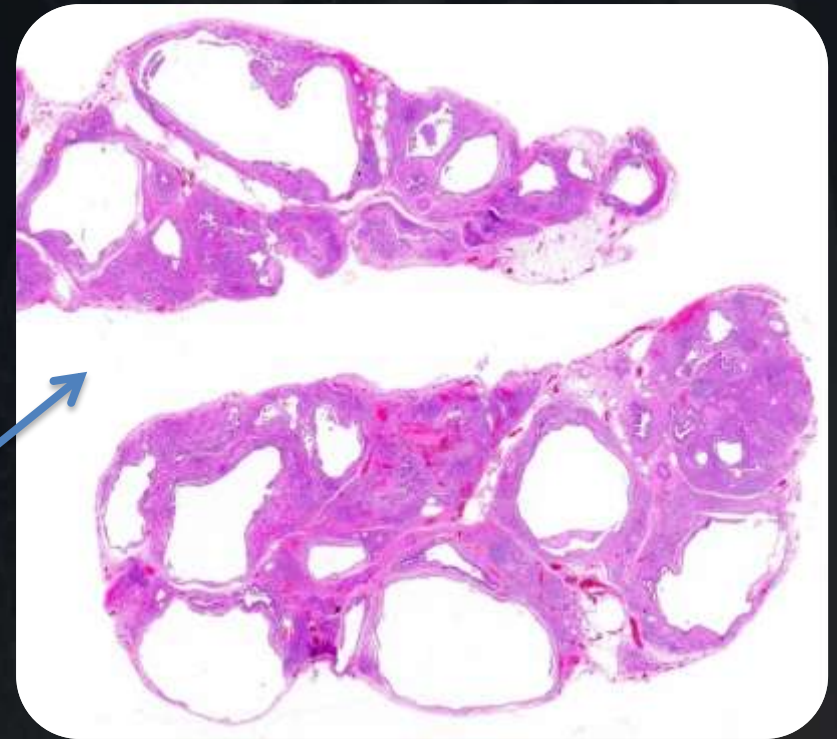
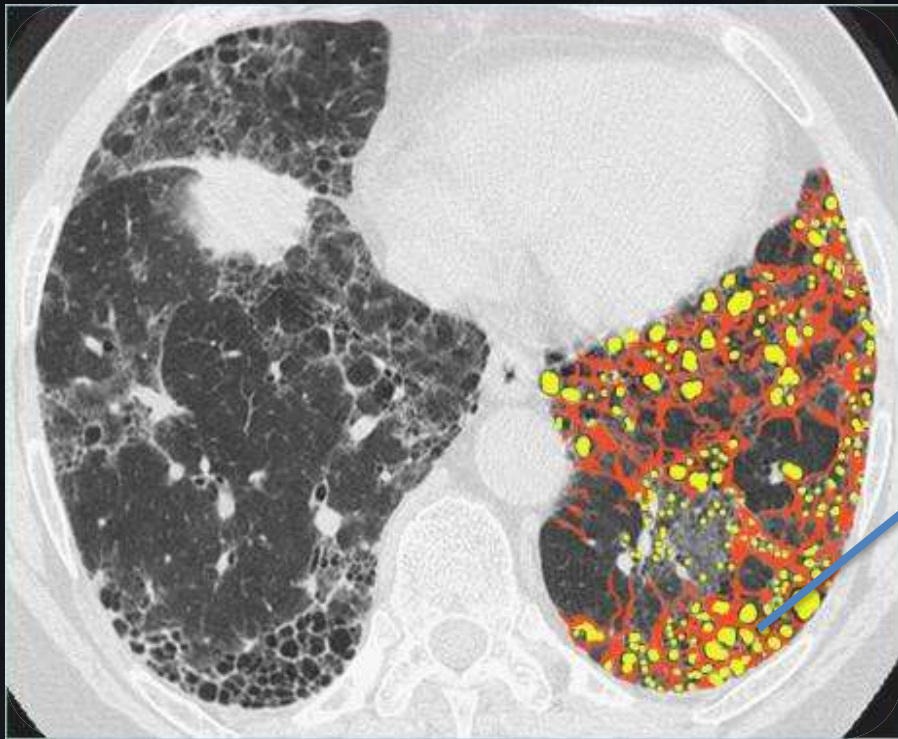


NSIP pattern

# Subpleural basal honeycombing $\equiv$ UIP

Clustered cystic airspaces, typically of comparable diameters of the order of 3–10 mm but occasionally as large as 2.5 cm

*Hansell DM et al, Radiology 2008*

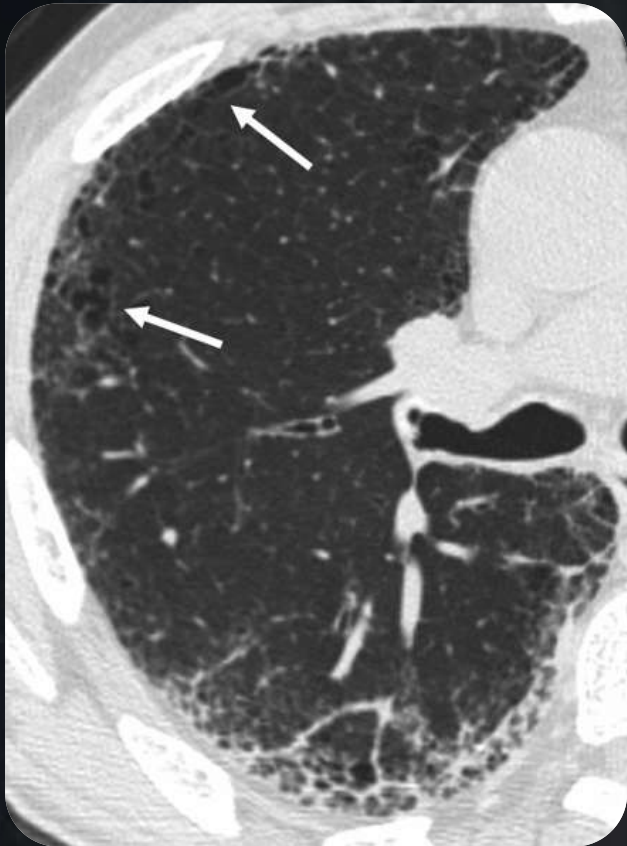




# Honeycombing:

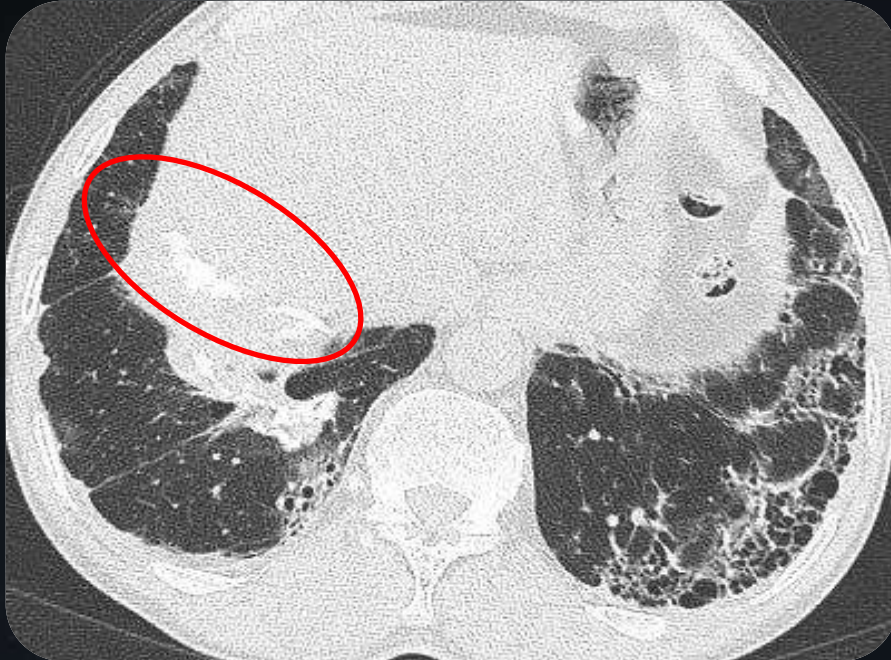
in 29% disagreement on presence /absence

*Watadani T et al Radiology 2013*

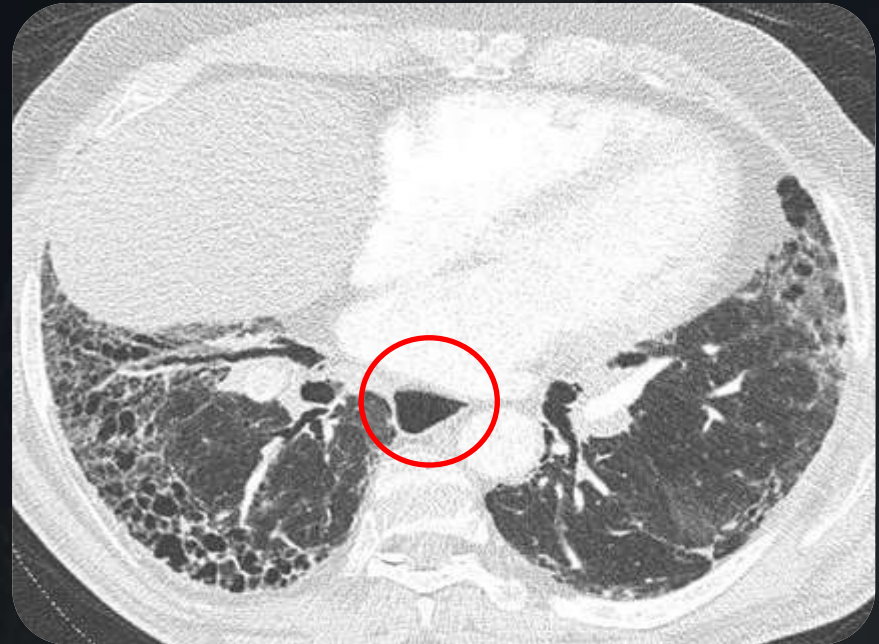


UIP pattern also in.....

do not overlook ancillary findings in the other thoracic structures!



Definite UIP pattern in asbestosis



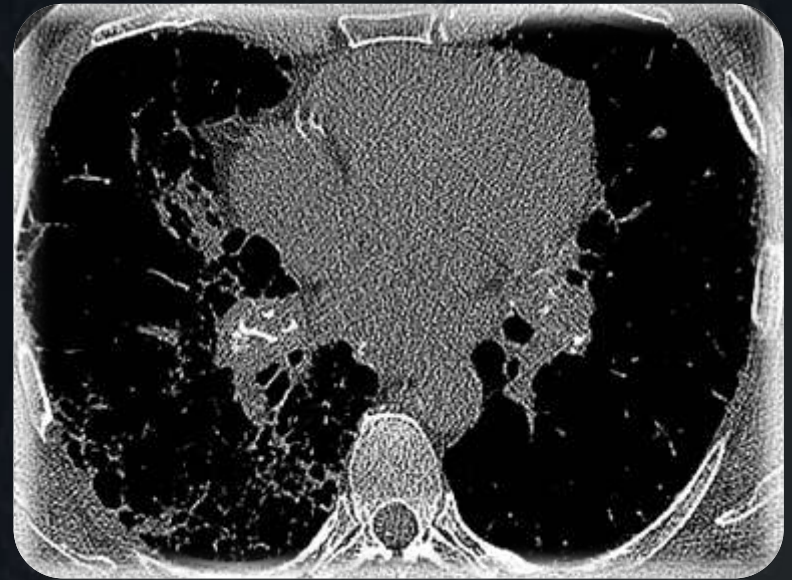
Definite UIP pattern in SSc



# An Official ATS/ERS/JRS/ALAT Statement: Idiopathic pulmonary fibrosis: Evidence-based Guidelines for Diagnosis and Management

## UIP Pattern (All Four Features)

- Subpleural, basal predominance
- Reticular abnormality
- Honeycombing with or without traction bronchiectasis
- Absence of features listed as inconsistent with UIP pattern (see third column)

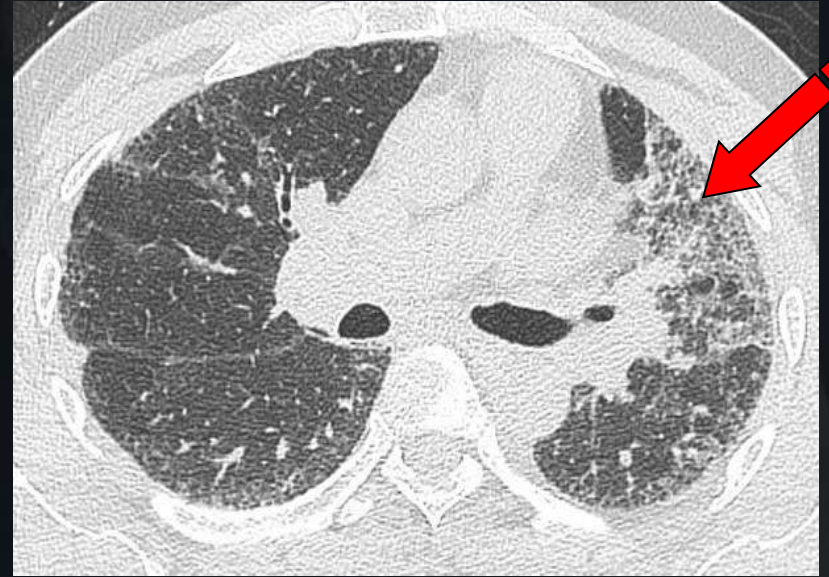
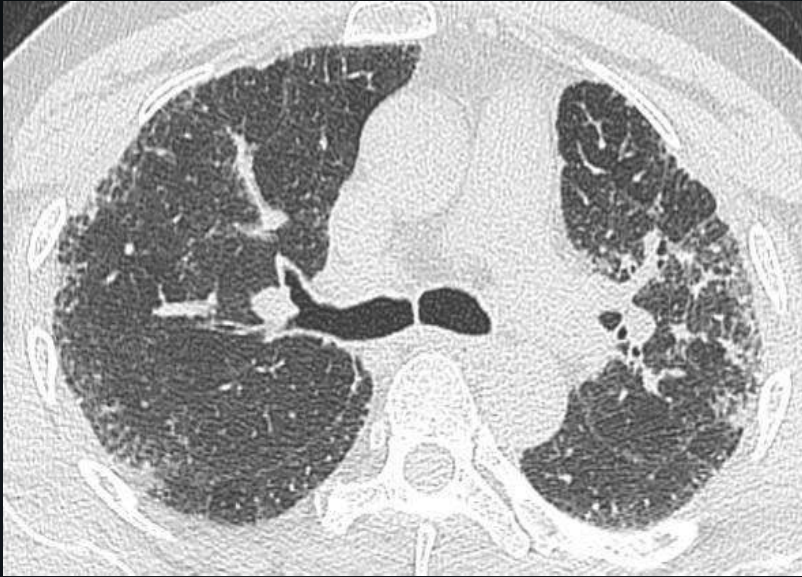


*<5% of cases, a definite UIP pattern on CT may end up with another diagnosis at MDT.....*

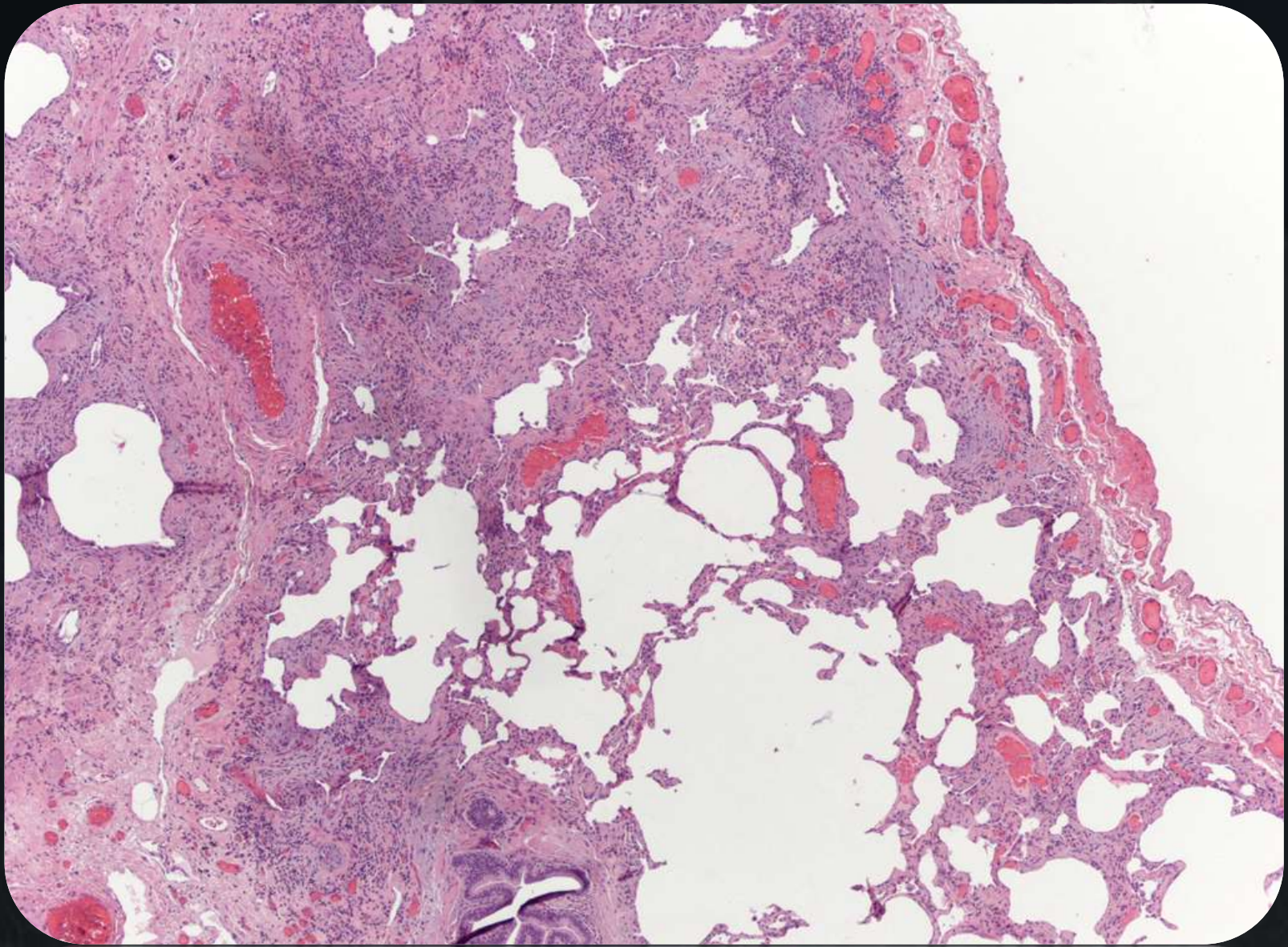
# Accuracy of a CT diagnosis of UIP/IPF

Study	Correctness of confident first choice CT diagnosis - 'definite UIP'	% of cases of UIP without a confident CT diagnosis - 'atypical UIP'
<i>JR Mathieson et al, Radiology, 1989;171:111</i>	95%	<b>38%</b>
<i>KS Lee et al, Radiology 1994;191:669</i>	100%	<b>39%</b>
<i>S Swenson et al, Radiology, 1997;205:229</i>	100%	<b>33%</b>
<i>GW Hunninghake, AJRCCM, 2001;164:193</i>	96%	<b>52%</b>

46 years old man with known fibrotic ILD and decreasing DLco

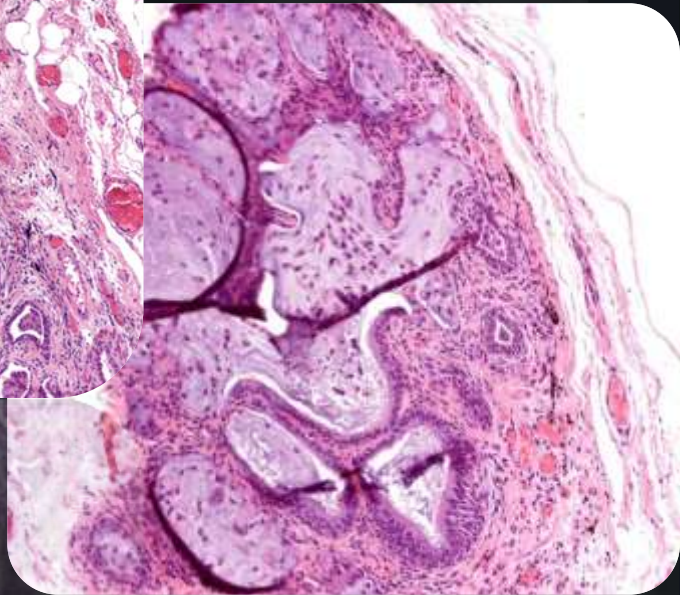
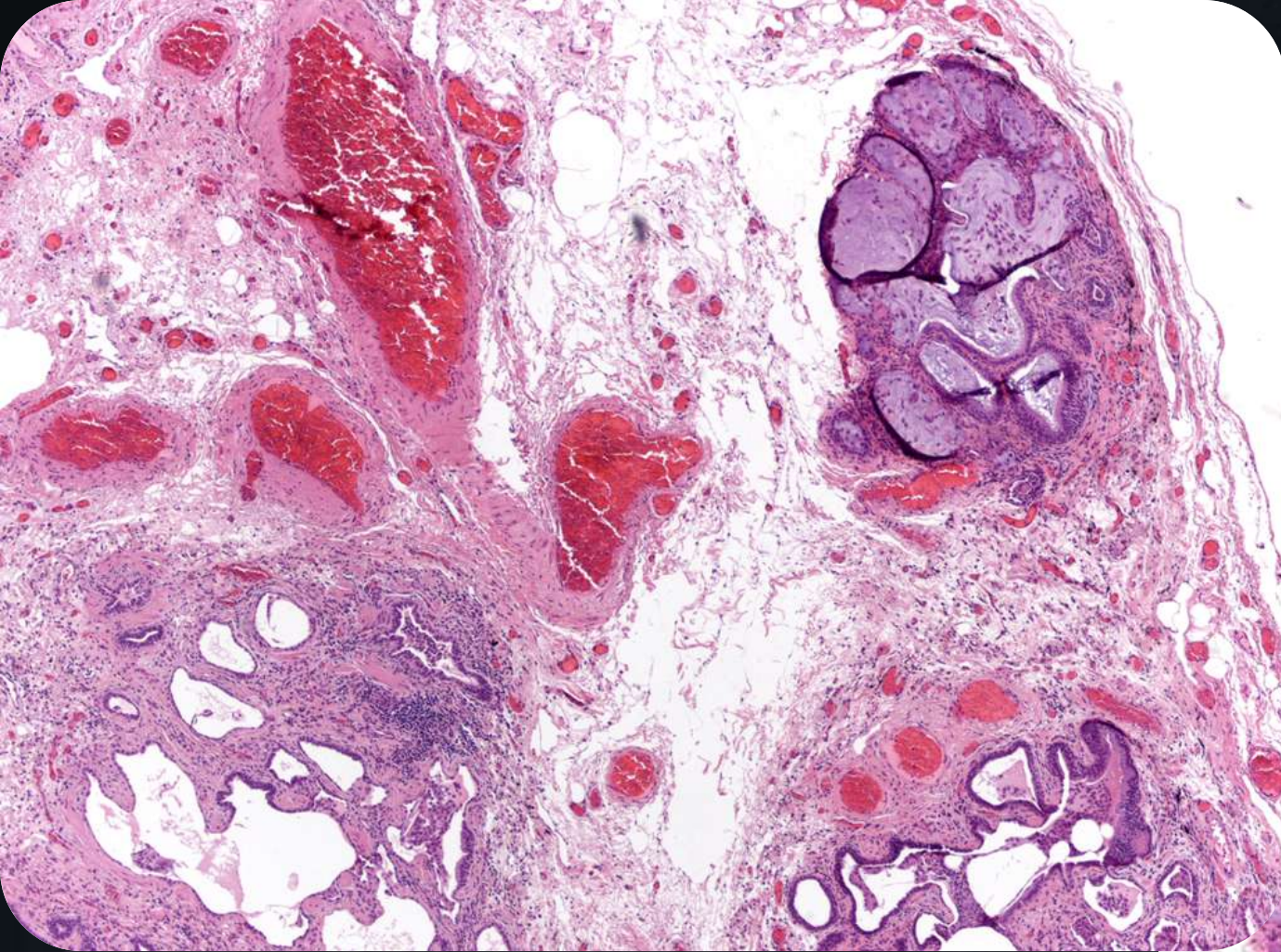






Patchwork pattern: scar-normal





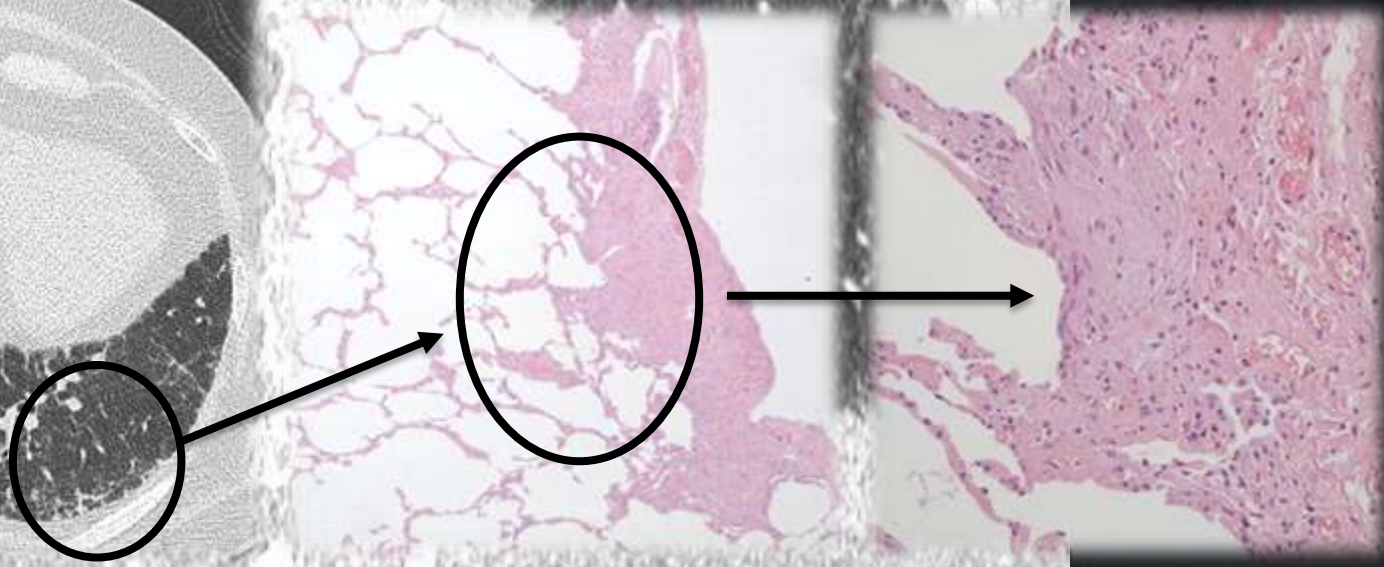
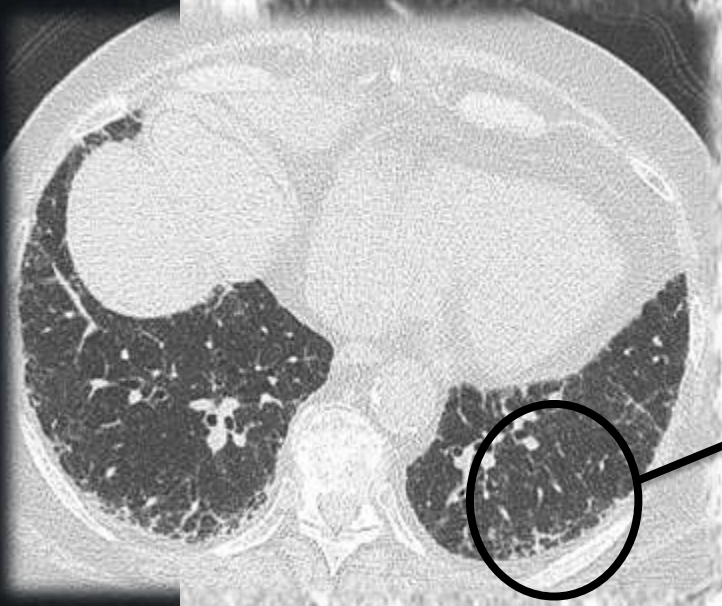
Honeycombing (probably too small to be seen on HRCT)  
+ some fibroblastic foci



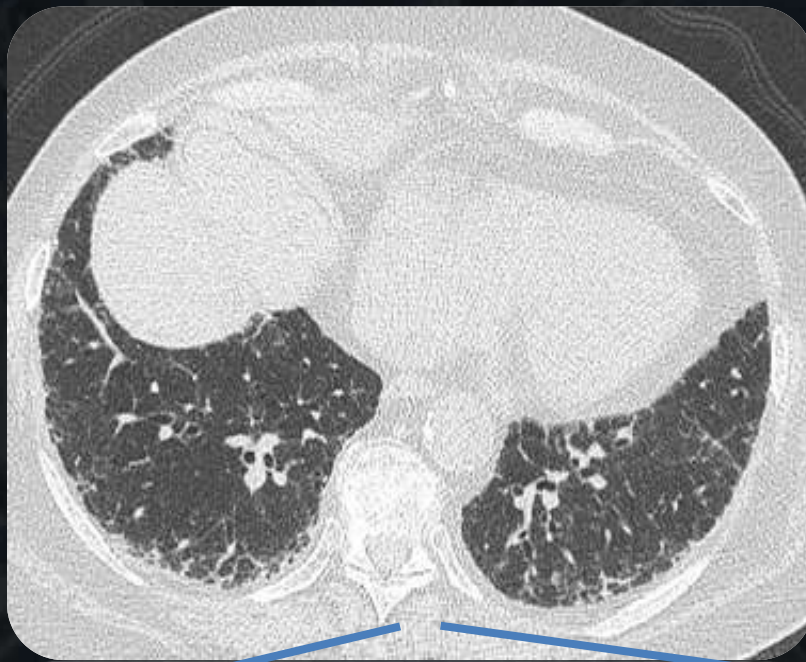
# An Official ATS/ERS/JRS/ALAT Statement: Idiopathic pulmonary fibrosis: Evidence-based Guidelines for Diagnosis and Management

**TABLE 4. HIGH-RESOLUTION COMPUTED TOMOGRAPHY CRITERIA FOR UIP PATTERN**

UIP Pattern (All Four Features)	Possible UIP Pattern (All Three Features)	Inconsistent with UIP Pattern (Any of the Seven Features)
<ul style="list-style-type: none"> <li>• Subpleural, basal predominance</li> <li>• Reticular abnormality</li> <li>• Honeycombing with or without traction bronchiectasis</li> <li>• Absence of features listed as inconsistent with UIP pattern (see third column)</li> </ul>	<ul style="list-style-type: none"> <li>• Subpleural, basal predominance</li> <li>• Reticular abnormality</li> <li>• Absence of features listed as inconsistent with UIP pattern (see third column)</li> </ul>	<ul style="list-style-type: none"> <li>• Upper or mid-lung predominance</li> <li>• Peribronchovascular predominance</li> <li>• Extensive ground glass abnormality (extent &gt; reticular abnormality)</li> <li>• Profuse micronodules (bilateral, predominantly upper lobes)</li> <li>• Discrete cysts (multiple, bilateral, away from areas of honeycombing)</li> <li>• Diffuse mosaic attenuation/air-trapping (bilateral, in three or more lobes)</li> <li>• Consolidation in bronchopulmonary segment(s)/lobe(s)</li> </ul>







Male gender  
Current or former smoker  
Older age (>70 yrs)  
Low-inspiratory squeaks  
Neutrophils on BAL



Very high likelihood of IPF  
(PPV 95%)

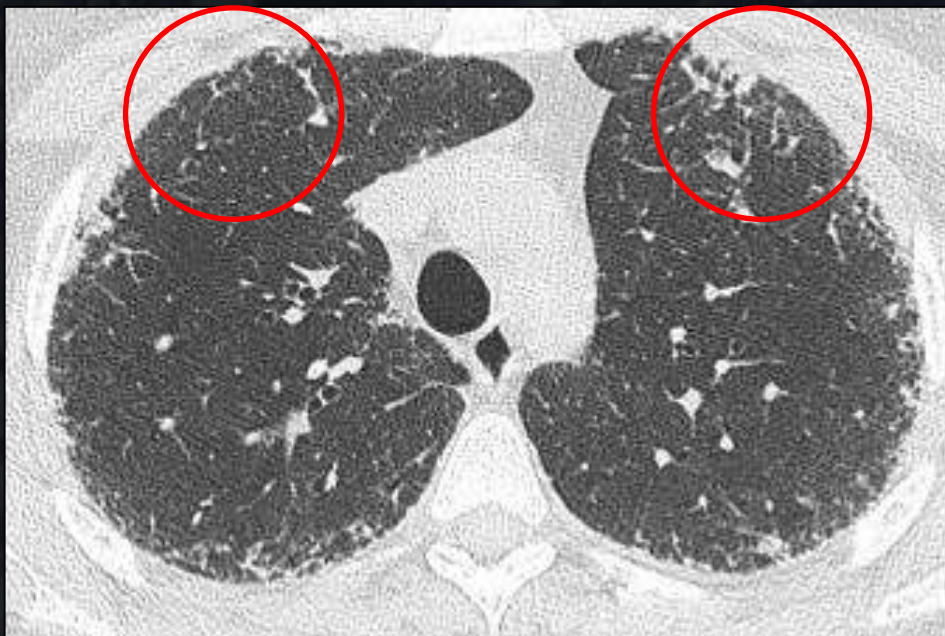
*Fell CD et al, AJRCCM 2010*

Female gender  
Younger age  
Non smoker

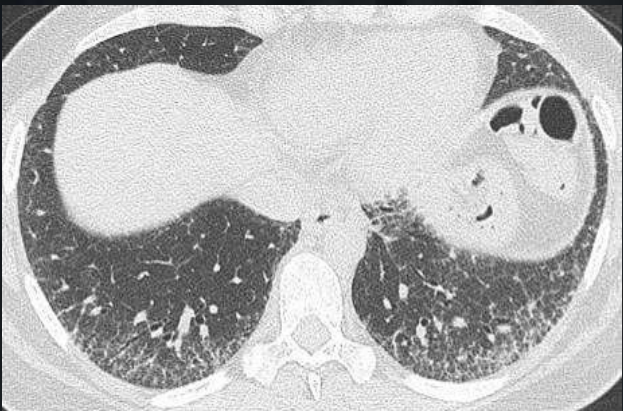
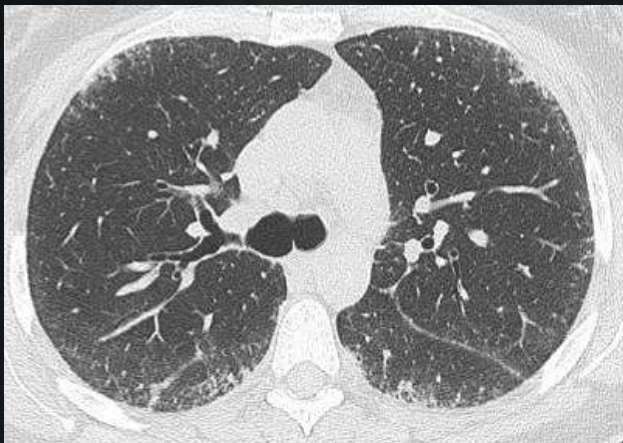
Mid-inspiratory squeaks  
Positive serologies  
Lymphocytosis on BAL  
Skin findings



More likely idiopathic or secondary NSIP



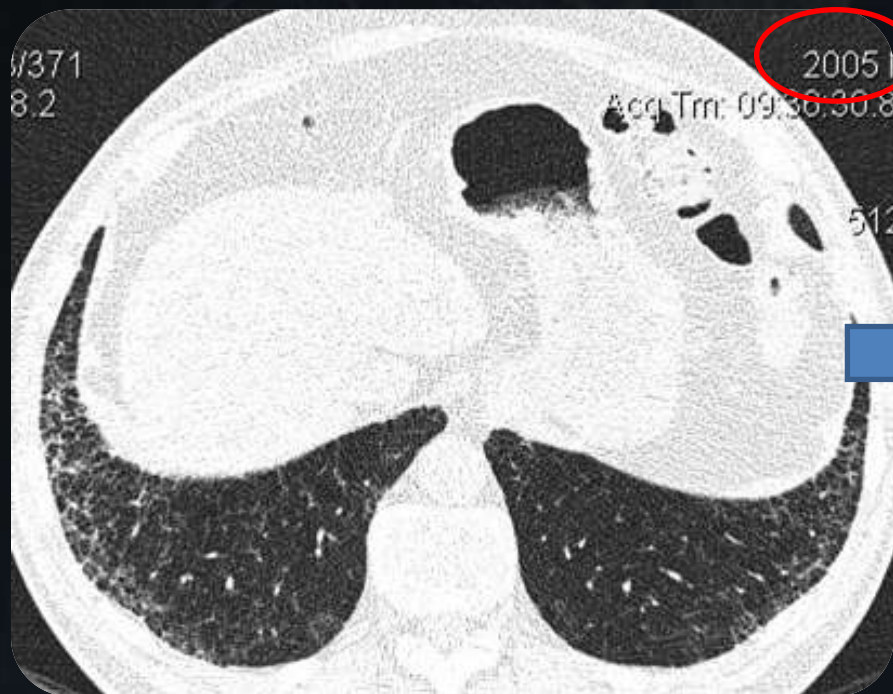


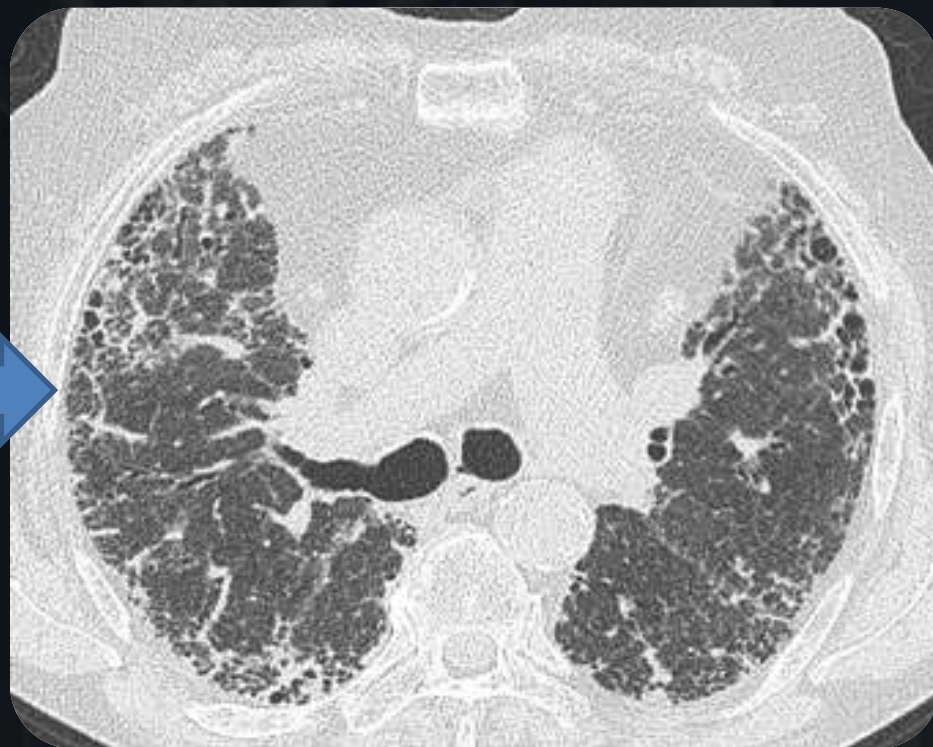
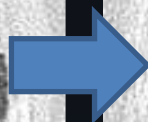
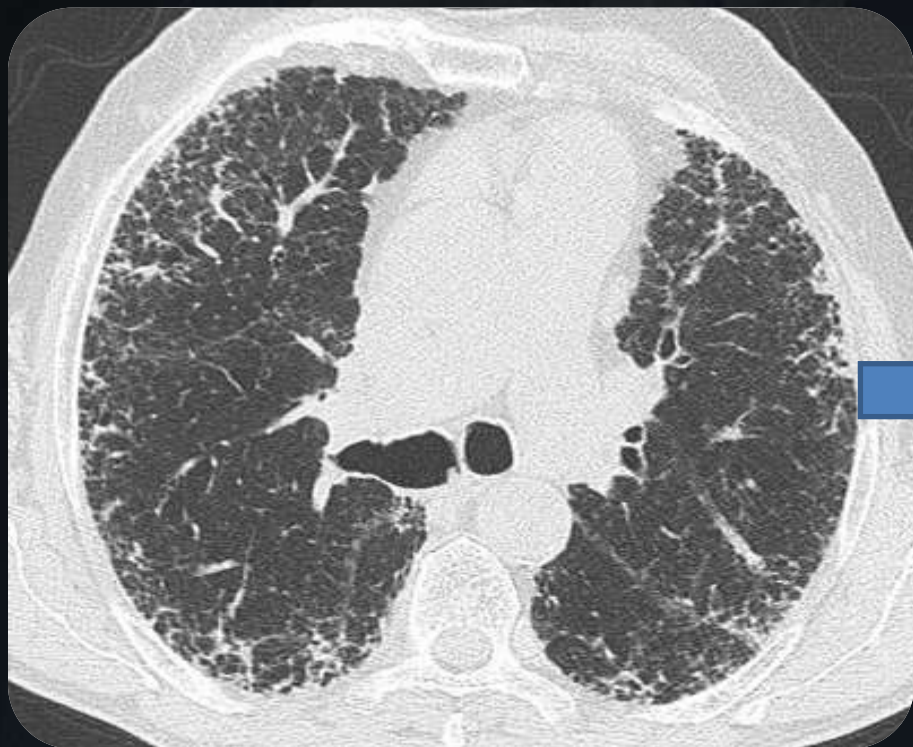


6 months  
later









*1 year later...*

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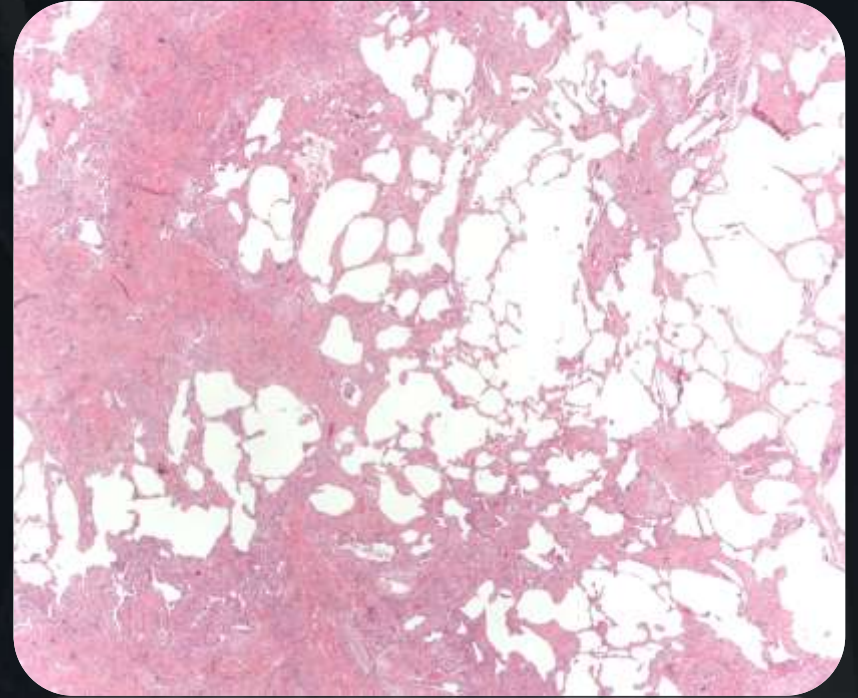
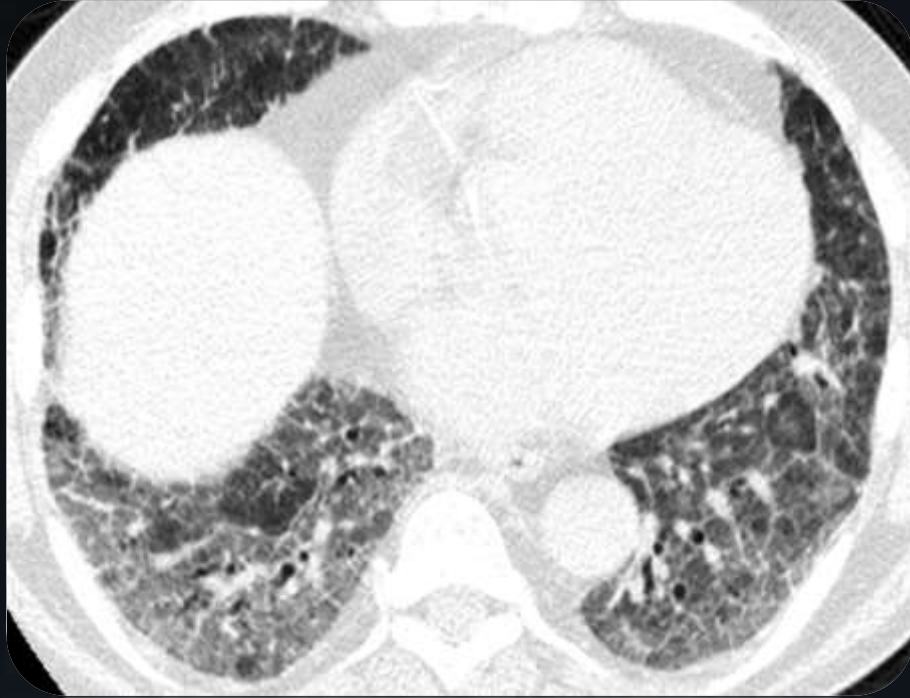
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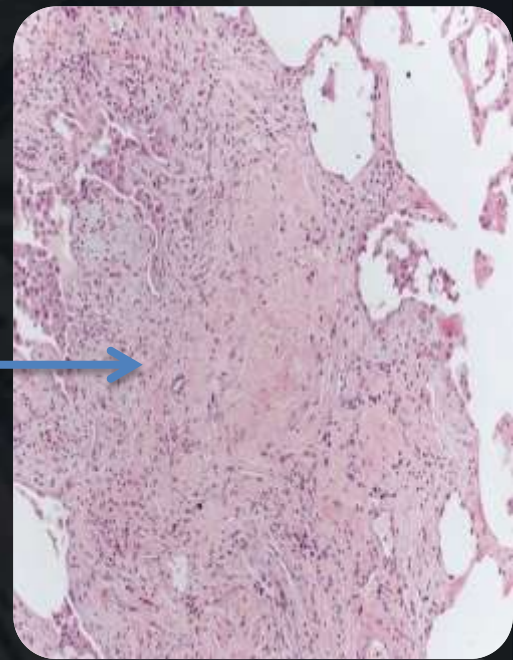
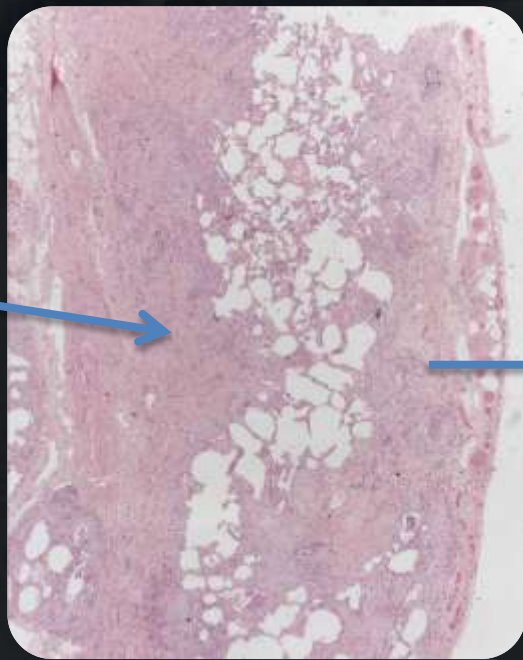
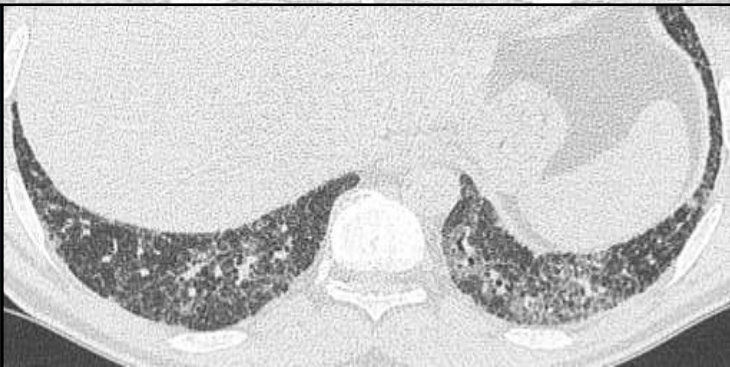
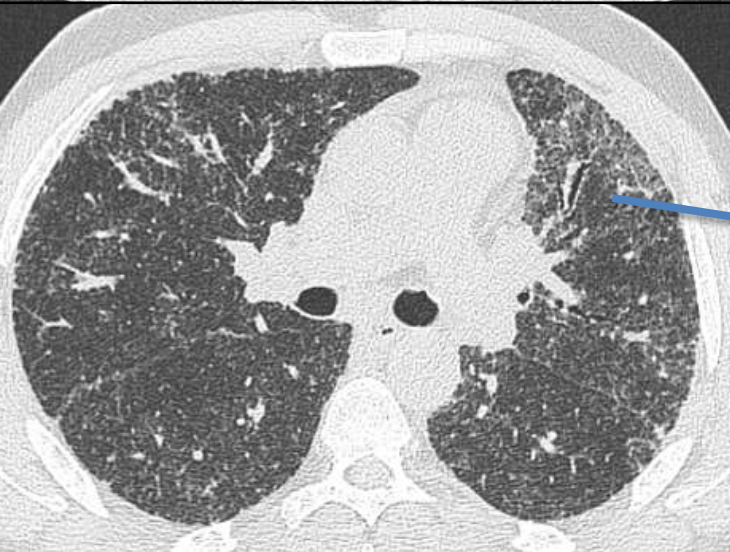
# Spectrum of atypical radiologic appearances of biopsy proven UIP

Most common radiologic diagnoses in 34 patients with biopsy proven UIP whose CT does not meet radiologic criteria for definite UIP (i.e. basal, subpleural honeycombing).....

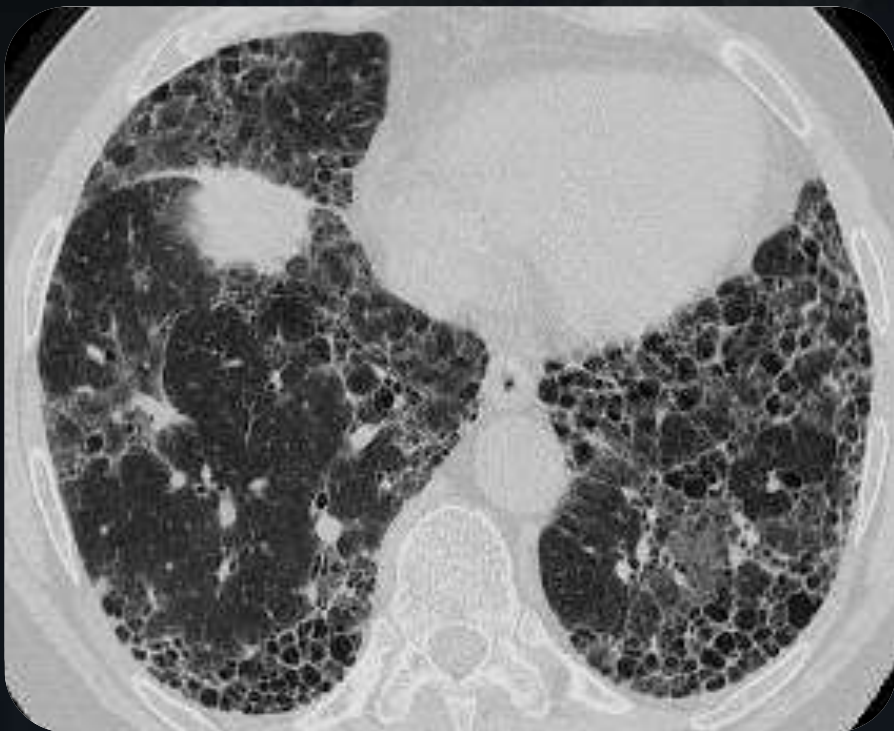
- **NSIP** 18
- **CHP** 4
- **Sarcoidosis** 3
- **OP** 1
- **Other** 8





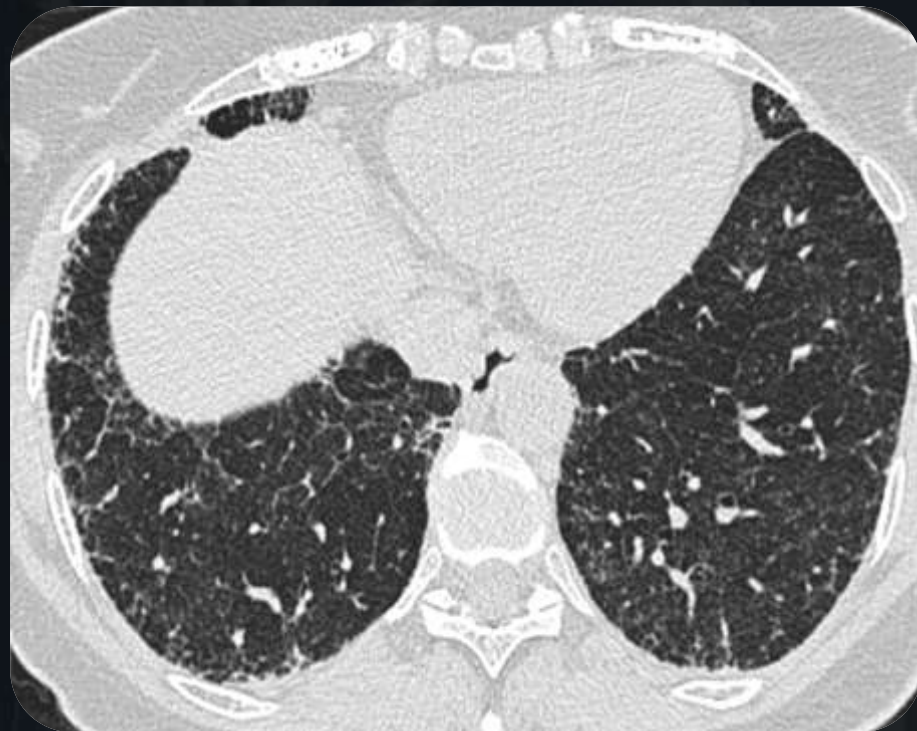






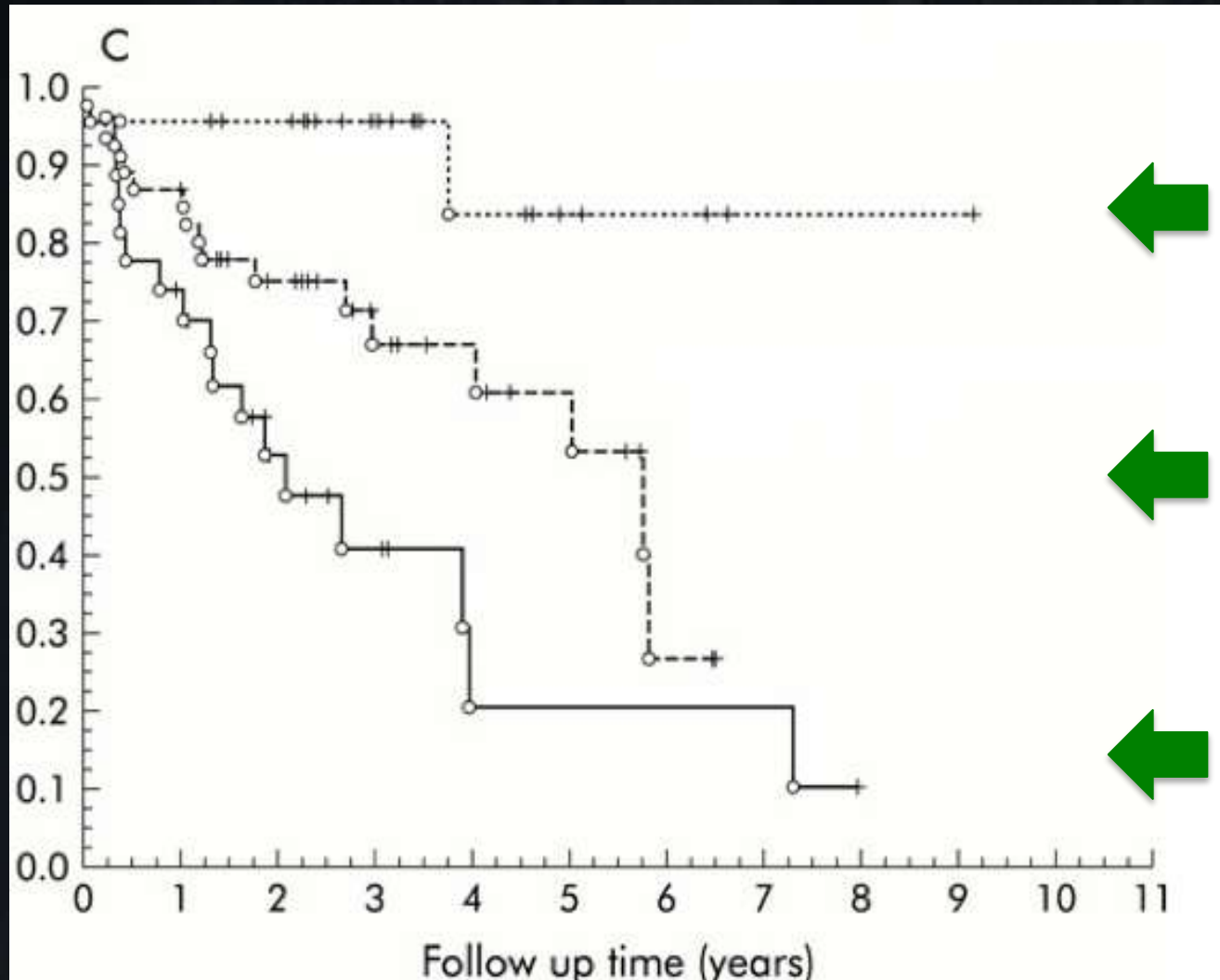
Definite UIP

VS



Non definite UIP

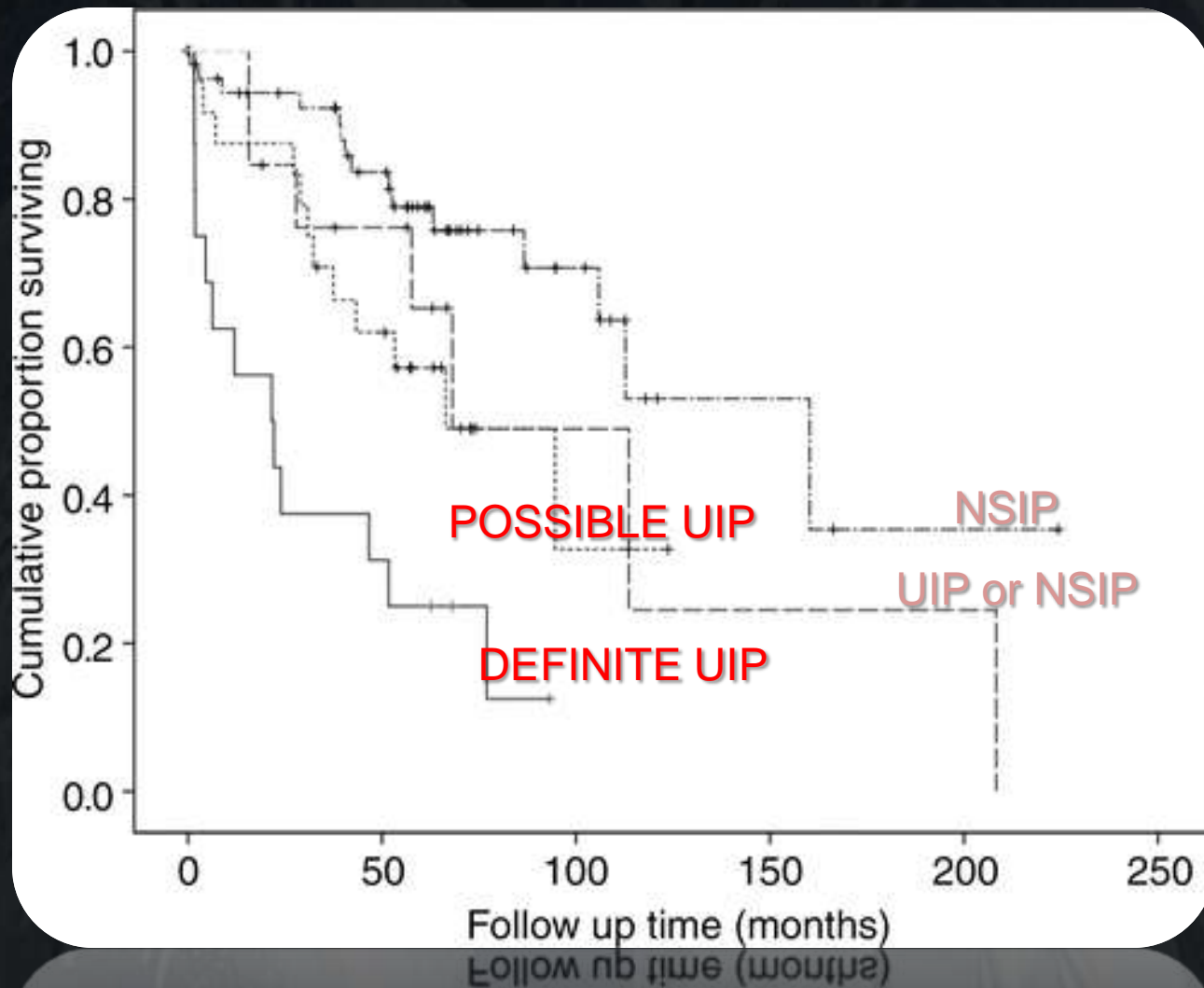
# Radiologic-pathologic discordance in IPF may confer a more favorable prognosis.....



← Radiology: NSIP  
Histology: NSIP

← Radiology: NSIP  
Histology: UIP  
(discordant)

← Radiology: UIP  
Histology: UIP  
(concordant)





# Key points

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UIP



UIP or fibrotic NSIP



NSIP or chronic hypersensitivity pneumonitis

- ✓ Do not downstage the «possible UIP» pattern
- ✓ Follow-up changes may be important, particularly when baseline CT is not diagnostic and surgical lung biopsy is not feasible