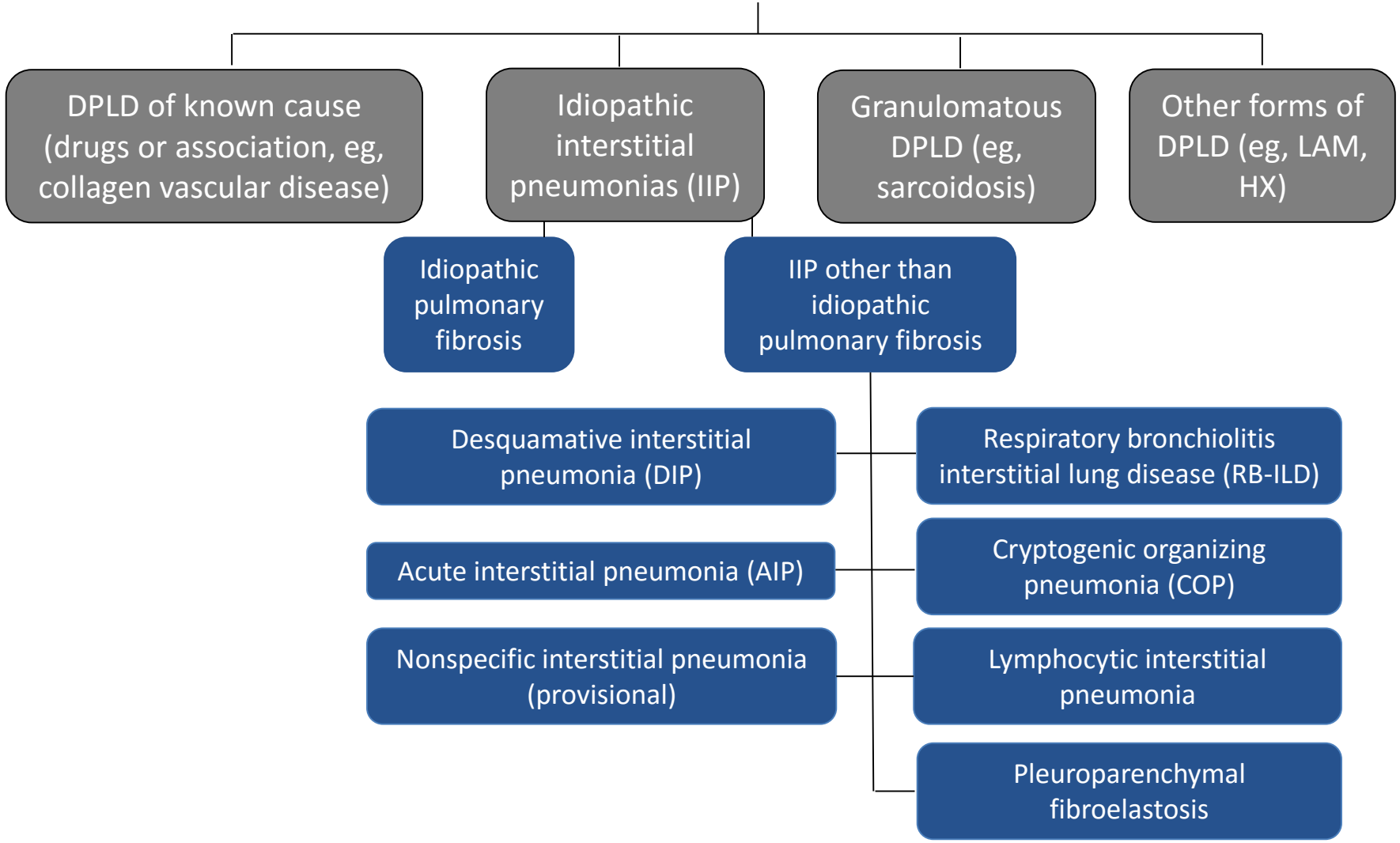
PILOT™ Education Transforming PULMONARY CARE

Thoracic Radiology

www.PILOTforPulmonary.org

Diffuse Parenchymal Lung Disease (DPLD)



Travis WD, et al. ATS/ERS Committee on Idiopathic Interstitial Pneumonias. *Am J Respir Crit Care Med*. 2013;188(6):733-748.

Major Idiopathic Interstitial Pneumonias

Category	Clinical-Radiologic-Pathologic Diagnosis	Associated Radiographic and/or Pathologic Pattern
Chronic fibrosing	IPF	UIP
	Idiopathic nonspecific interstitial pneumonia (iNSIP)	NSIP
Smoking-related	Respiratory bronchiolitis-ILD (RB-ILD)	Respiratory bronchiolitis
	Desquamative interstitial pneumonia (DIP)	Desquamative interstitial pneumonia
Acute/ subacute	Cryptogenic organizing pneumonia (COP)	Organizing pneumonia
	Acute interstitial pneumonia (AIP)	Diffuse alveolar damage

Etiologies of Pulmonary Fibrosis

- Idiopathic pulmonary fibrosis (IPF)
- Connective tissue disease (may have NSIP)
- Occupational lung disease
- Chronic hypersensitivity pneumonitis (CHP)
- Sarcoidosis
- Drug-related fibrosis (esp bleomycin, MTX)
- Familial pulmonary fibrosis

Any of these may show UIP pattern on HRCT;
pulmonologist correlates clinical, imaging and pathology

Usual Interstitial Pneumonia (UIP)

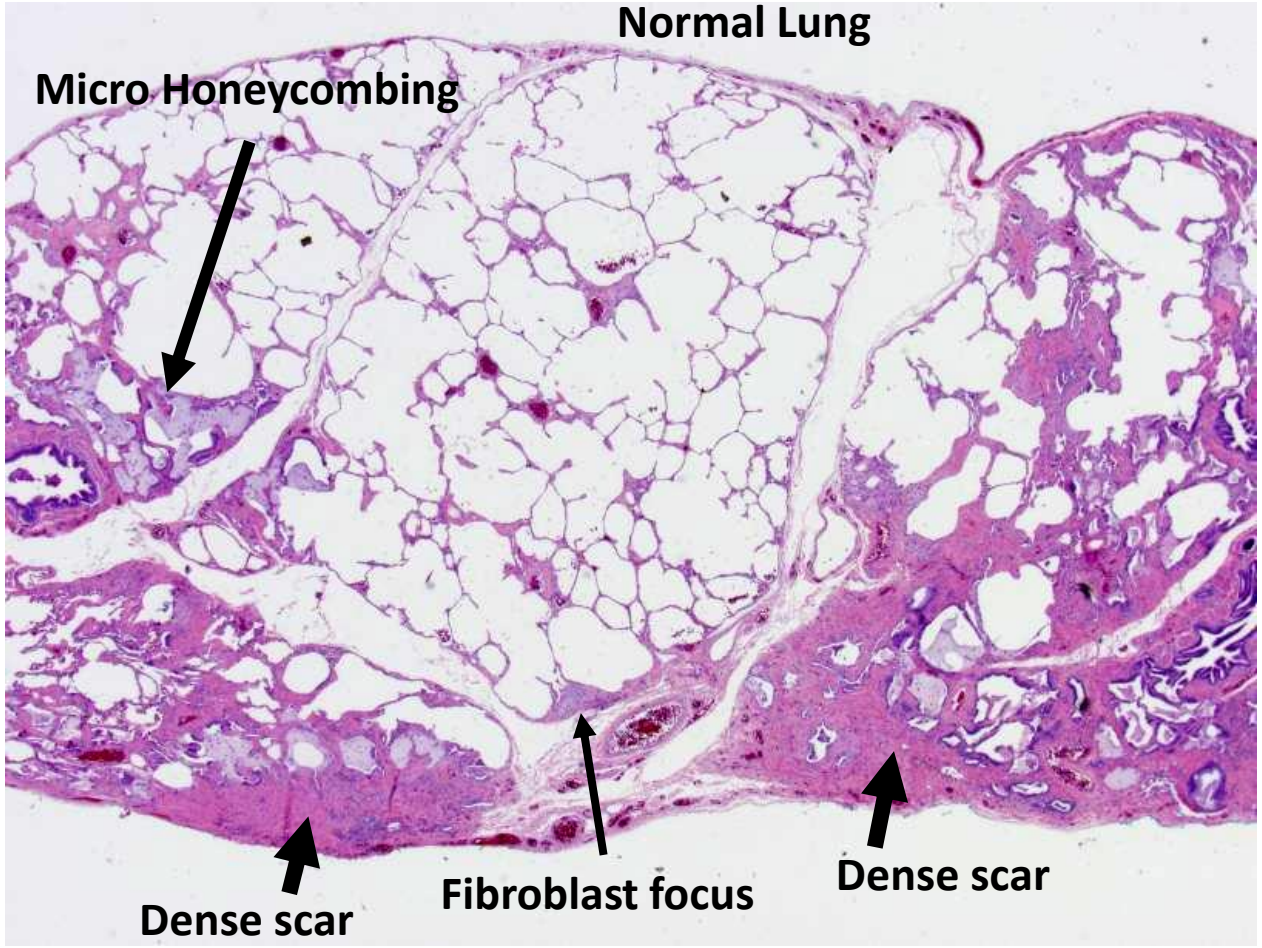
- Pattern of disease identified on HRCT and pathology
- Pathology – fibrotic lesions
  - Fibroblastic foci
  - Mature fibrosis
  - Honeycombing
- Heterogeneous *temporal* and *spatial* distribution

Radiologist identifies UIP, not IPF

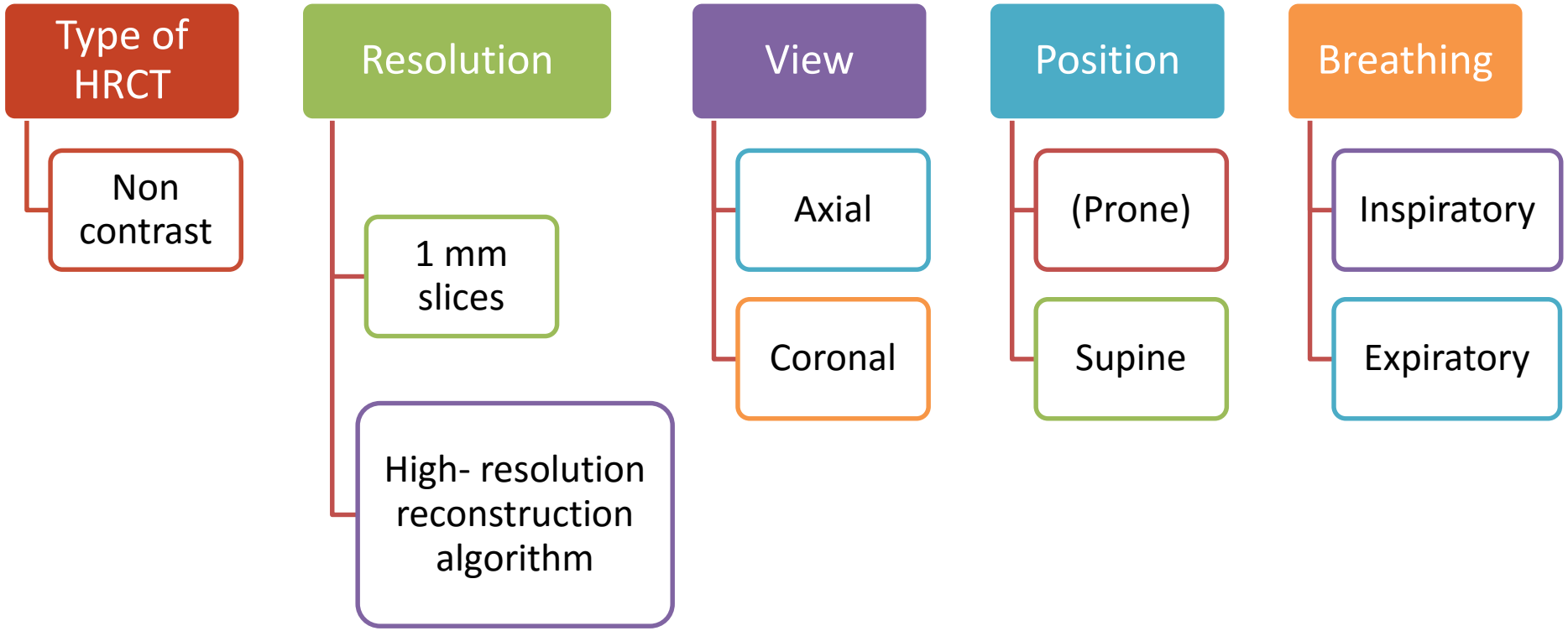
Histopathology

THIS is UIP

1. Temporal heterogeneity
2. Microscopic honeycombing
3. Dense subpleural pink scar
4. Fibroblast foci (at the edge of dense scar)



What are the features of an HRCT?



HRCT Scanning Parameters ATS Guidelines

1. Noncontrast examination
2. Volumetric acquisition with selection of:
   - Sub-millimetric collimation
   - Shortest rotation time
   - Highest pitch
   - Tube potential and tube current appropriate to patient size:
     - Typically 120 kVp and ≤ 240 mAs
     - Lower tube potentials (e.g., 100 kVp) with adjustment of tube current encouraged for thin patients
   - Use of techniques available to avoid unnecessary radiation exposure (e.g., tube current modulation)

HRCT Scanning Parameters ATS Guidelines, cont.

3. Reconstruction of thin-section CT images (≤ 1.5 mm):
   - Contiguous or overlapping
   - Using a high-special-frequency algorithm
   - Iterative reconstruction algorithm if validated on the CT unit (if not, filtered back projection)
4. **Number of acquisitions:**
   - **Supine: inspiratory (volumetric)**
   - **Supine: expiratory (can be volumetric or sequential)**
   - **Prone: only inspiratory scans (can be sequential or volumetric); optional**
   - **Inspiratory scans obtained at full inspiration**
5. Recommended radiation dose for the inspiratory volumetric acquisition:
   - 1-3 mSv (i.e., “reduced” dose)
   - Strong recommendation to avoid “ultra-low-dose CT” (<1 mSv)

Diagnostic criteria for idiopathic pulmonary fibrosis: a Fleischner Society White Paper

David A Lynch, Nicola Sverzellati, William D Travis, Kevin K Brown, Thomas V Colby, Jeffrey R Galvin, Jonathan G Goldin, David M Hansell, Yoshikazu Inoue, Takeshi Johkoh, Andrew G Nicholson, Shandra L Knight, Suhail Raoof, Luca Richeldi, Christopher J Ryerson, Jay H Ryu, Athol U Wells

This Review provides an updated approach to the diagnosis of idiopathic pulmonary fibrosis (IPF), based on a systematic search of the medical literature and the expert opinion of members of the Fleischner Society. A checklist is provided for the clinical evaluation of patients with suspected usual interstitial pneumonia (UIP). The role of CT is expanded to permit diagnosis of IPF without surgical lung biopsy in select cases when CT shows a probable UIP pattern. Additional investigations, including surgical lung biopsy, should be considered in patients with either clinical or CT findings that are indeterminate for IPF. A multidisciplinary approach is particularly important when deciding to perform additional diagnostic assessments, integrating biopsy results with clinical and CT features, and establishing a working diagnosis of IPF if lung tissue is not available. A working diagnosis of IPF should be reviewed at regular intervals since the diagnosis might change. Criteria are presented to establish confident and working diagnoses of IPF.

Lancet Respir Med 2017

Published Online
November 15, 2017
http://dx.doi.org/10.1016/S2213-2600(17)30433-2

See Online/Comment
http://dx.doi.org/10.1016/S2213-2600(17)30443-5

Department of Radiology (Prof D A Lynch MB), **Department of Medicine** (Prof K K Brown MD), **and Library and Knowledge**

Diagnostic Categories of UIP Based on CT Patterns

UIP	Probable UIP	Indeterminate for UIP	Alternative Diagnosis
Subpleural and basal predominant; distribution is often heterogeneous*	Subpleural and basal predominant; distribution is often heterogeneous	Subpleural and basal predominant	Findings suggestive of another diagnosis, including: • CT features: ° Cysts ° Marked mosaic attenuation ° Predominant GGO ° Profuse micronodules ° Centrilobular nodules ° Nodules ° Consolidation • Predominant distribution: ° Peribronchovascular ° Perilymphatic ° Upper or mid-lung • Other: ° Pleural plaques (consider asbestosis) ° Dilated esophagus (consider CTD) ° Distal clavicular erosions (consider RA) ° Extensive lymph node enlargement (consider other etiologies) ° Pleural effusions, pleural thickening (consider CTD/drugs)
Honeycombing with or without peripheral traction bronchiectasis or bronchiolectasis†	Reticular pattern with peripheral traction bronchiectasis or bronchiolectasis May have mild GGO	Subtle reticulation; may have mild GGO or distortion ("early UIP pattern") CT features and/or distribution of lung fibrosis that do not suggest any specific etiology ("truly indeterminate for UIP")	

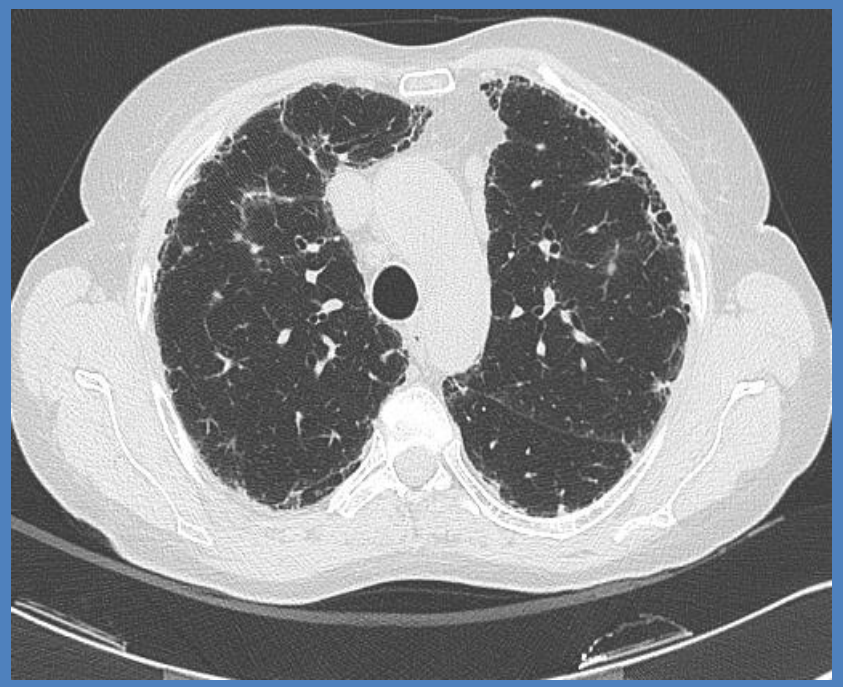
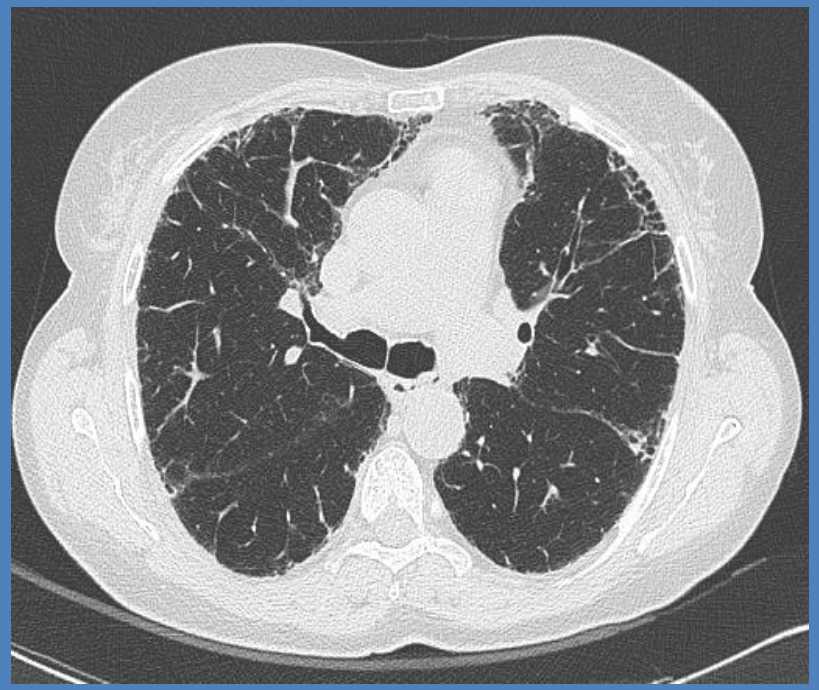
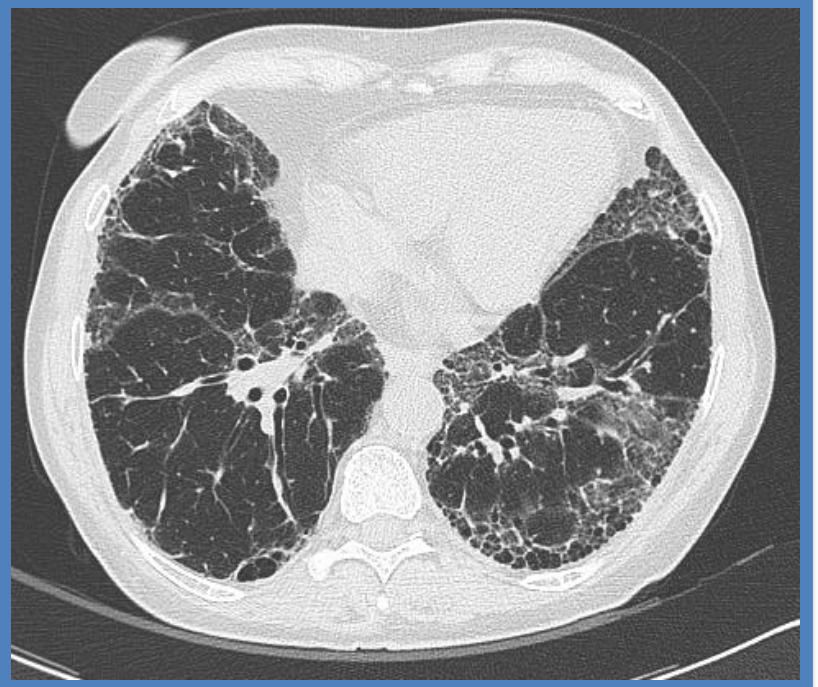
Raghu G, et al. *Am J Respir Crit Care Med*. 2018;198(5):e44-e68.

Histopathological Criteria for UIP

	Definite UIP-IPF	Probable UIP-IPF	Indeterminate for UIP-IPF	Features most consistent with an alternative diagnosis
General comments	Patients show features with all four criteria, and do not show features that might suggest an alternative diagnosis (eg, non-UIP)	Patients show either honeycomb fibrosis only, or a severe fibrosing process that falls short of showing all four criteria for definite UIP-IPF and do not show features that might suggest an alternative diagnosis	Patients show evidence of a fibrosing process but with features that are more in favour of either a non-UIP pattern, or UIP in a setting other than IPF	Patients show either a UIP pattern with ancillary features strongly suggesting an alternative diagnosis, or a non-UIP pattern (see cell below)
Specific criteria	Dense fibrosis causing architecture remodelling with frequent honeycombing; patchy lung involvement by fibrosis; subpleural or paraseptal distribution, or both; fibroblast foci at the edge of dense scars	Honeycomb fibrosis only or; dense fibrosis causing architecture remodelling with frequent honeycombing; patchy lung involvement by fibrosis; fibroblast foci at the edge of dense scars may or may not be present	Patients have less compelling histological changes than those classified by the final column (eg, occasional foci of centrilobular injury or scarring, rare granulomas or giant cells, only a minor degree of lymphoid hyperplasia or diffuse inflammation, or diffuse homogenous fibrosis favouring fibrotic non-specific interstitial pneumonia); these features, and the differential diagnoses they call to mind, become part of the multidisciplinary discussion and decision with regard to a multidisciplinary diagnosis of IPF, or not	Non-UIP pattern: patients with features of other fibrotic disorders—eg, fibrotic hypersensitivity pneumonitis, fibrotic non-specific interstitial pneumonia, fibrosing organising pneumonia, pleuroparenchymal fibroelastosis, pulmonary Langerhans cell histiocytosis, or smoking-related interstitial fibrosis; UIP pattern with ancillary features strongly suggesting an alternative diagnosis: eg, prominent diffuse alveolar damage or organising pneumonia (consider acute exacerbation of UIP), granulomas, (consider hypersensitivity pneumonitis, sarcoid, infection), marked interstitial inflammatory cell infiltrate away from areas of UIP (consider hypersensitivity pneumonitis)

Lynch DA, et al. *Lancet Respir Med*: 2018;6(2):138-153.

Typical UIP CT Pattern

DISTRIBUTION

Basal (occasionally diffuse) and subpleural predominant

Distribution is often heterogeneous

CT FEATURES

Honeycombing

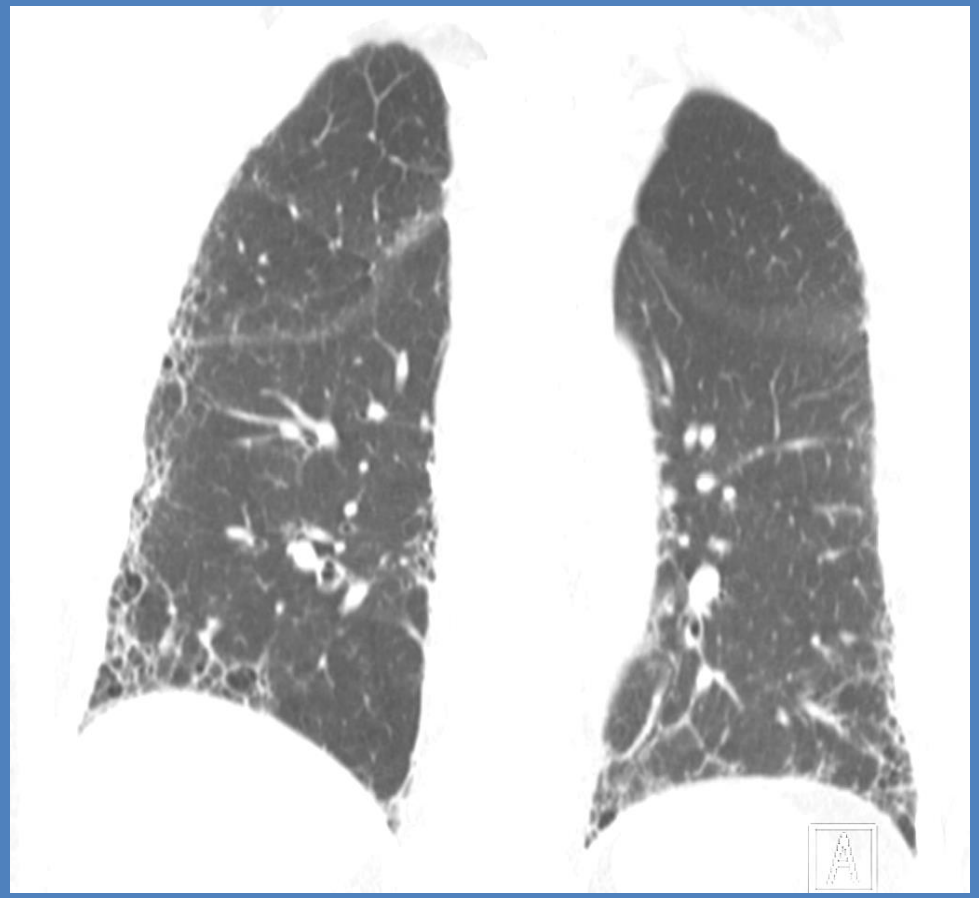
Reticular pattern

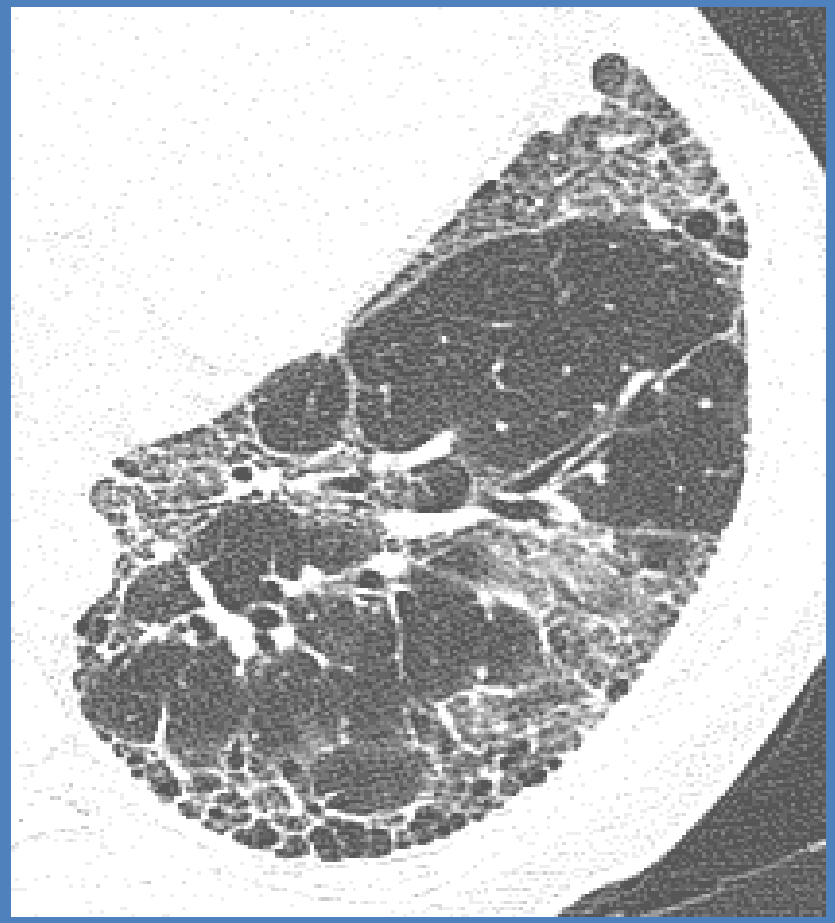
Traction bronchiectasis/bronchiolectasis

Absence of non-UIP features

Images courtesy of D. Lynch

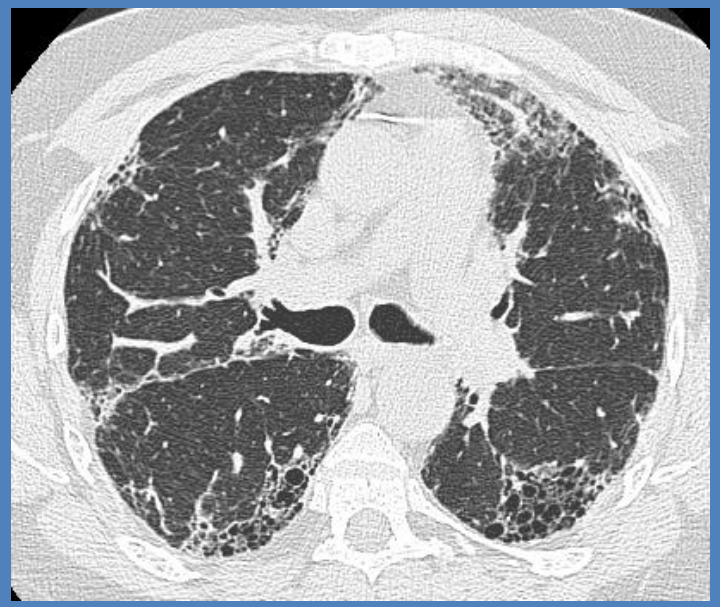
Typical UIP CT Pattern

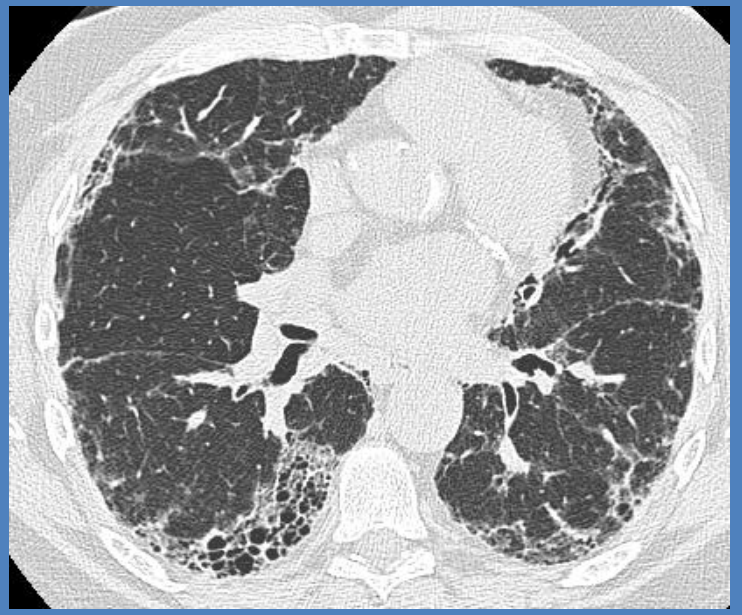


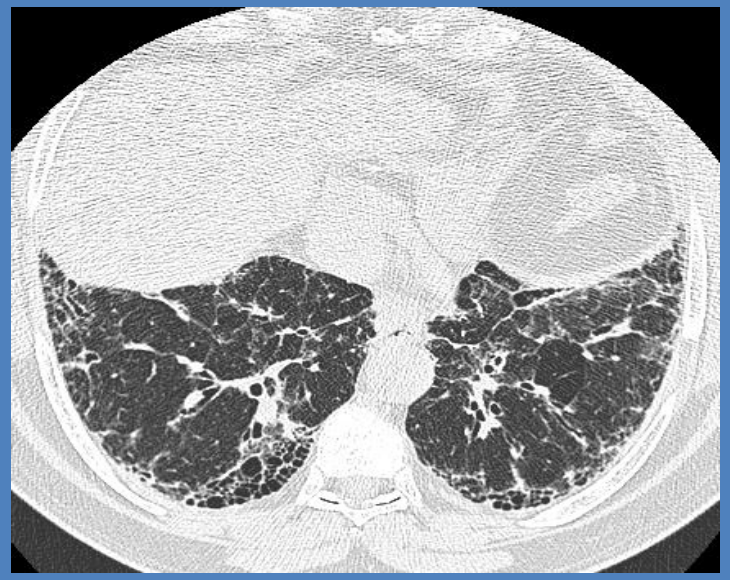


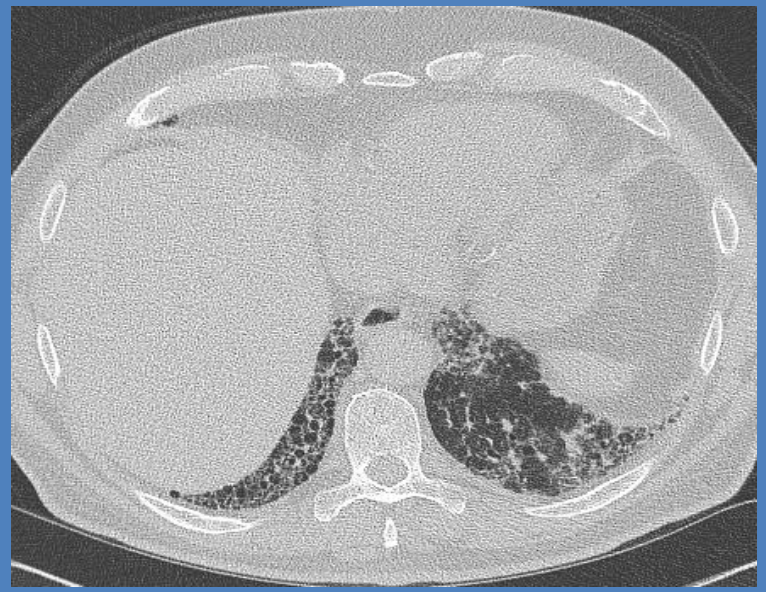
Images courtesy of D. Lynch

UIP



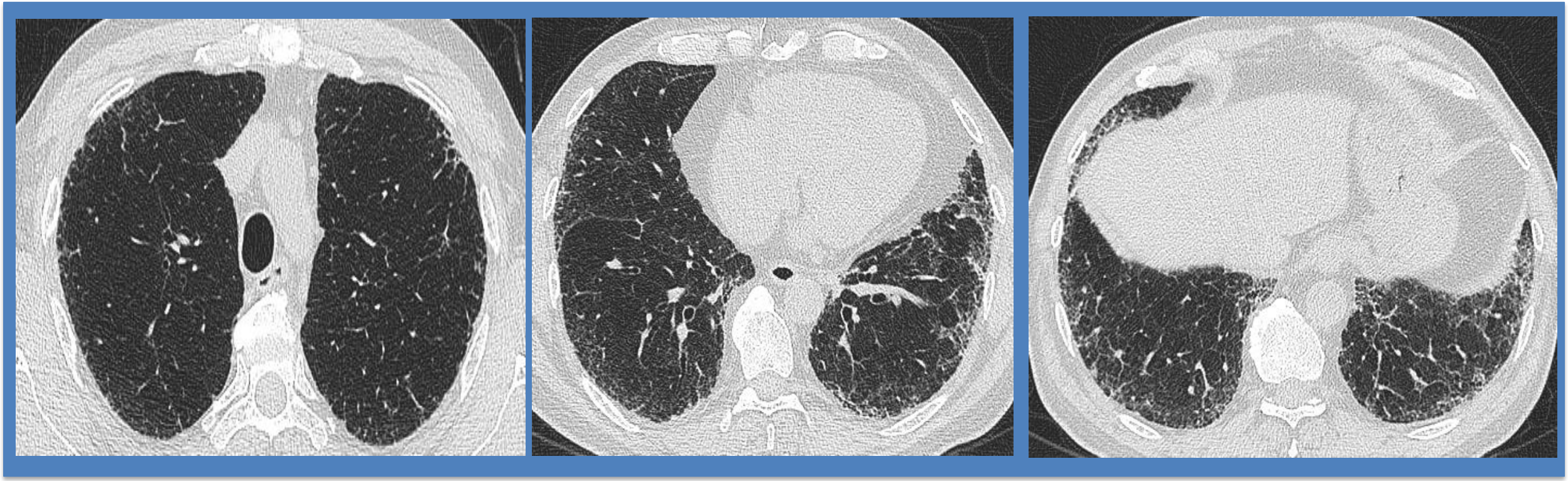






Images courtesy of D. Lynch

Probable UIP CT Pattern



DISTRIBUTION

Basal and subpleural predominant

Distribution is often heterogeneous

CT FEATURES

Reticular pattern

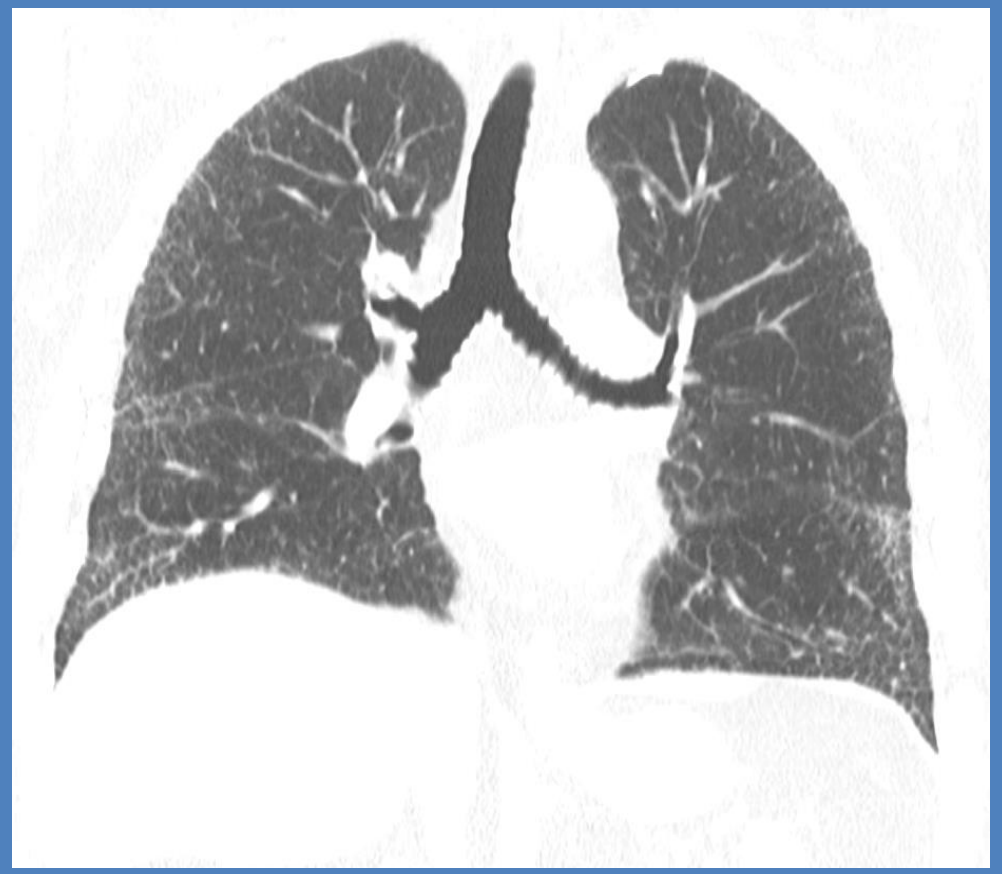
Traction bronchiectasis/bronchiolectasis

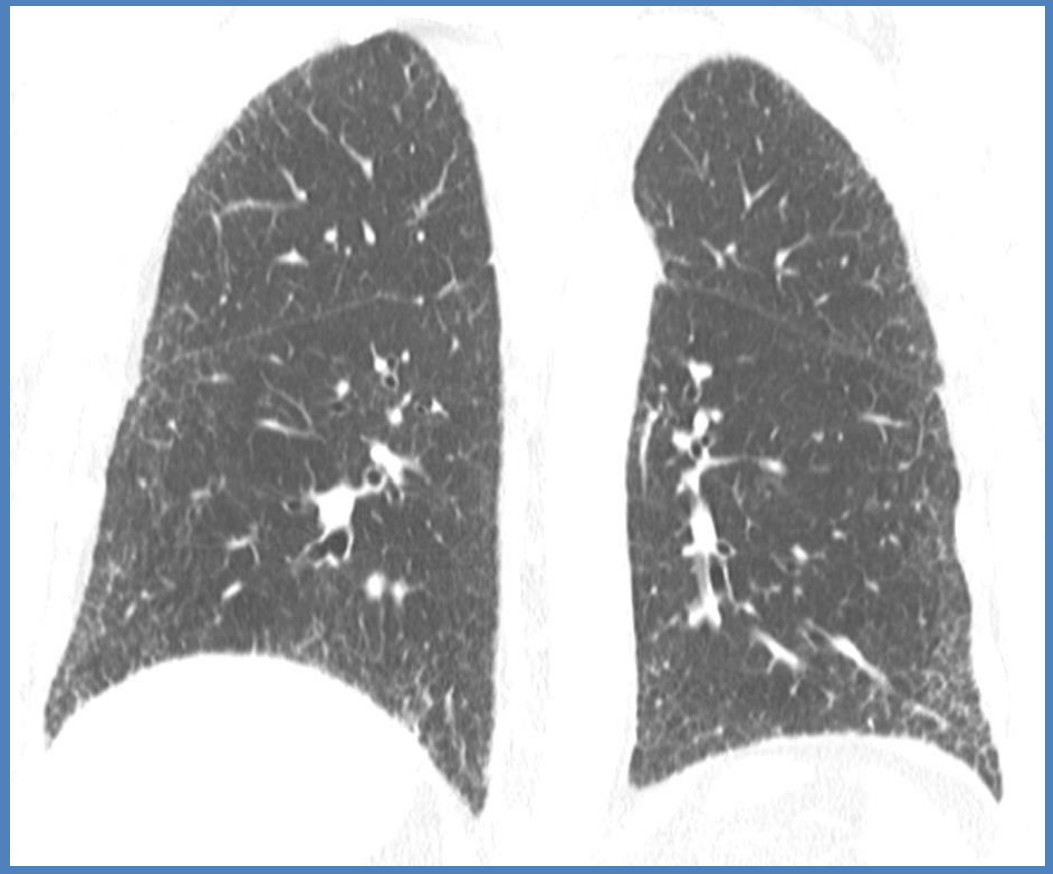
No honeycombing

Absence of non-UIP features

Images courtesy of D. Lynch

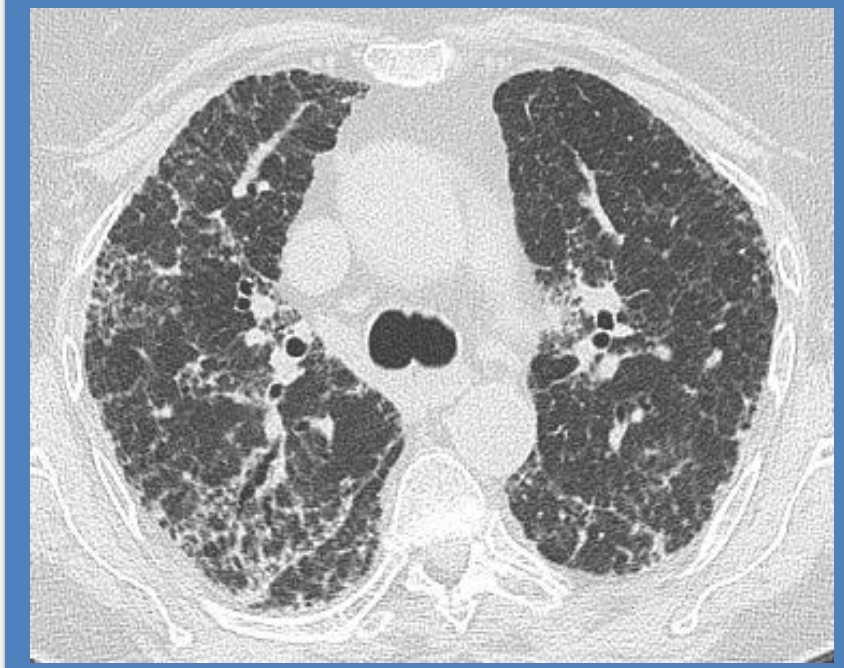
Probable UIP CT Pattern

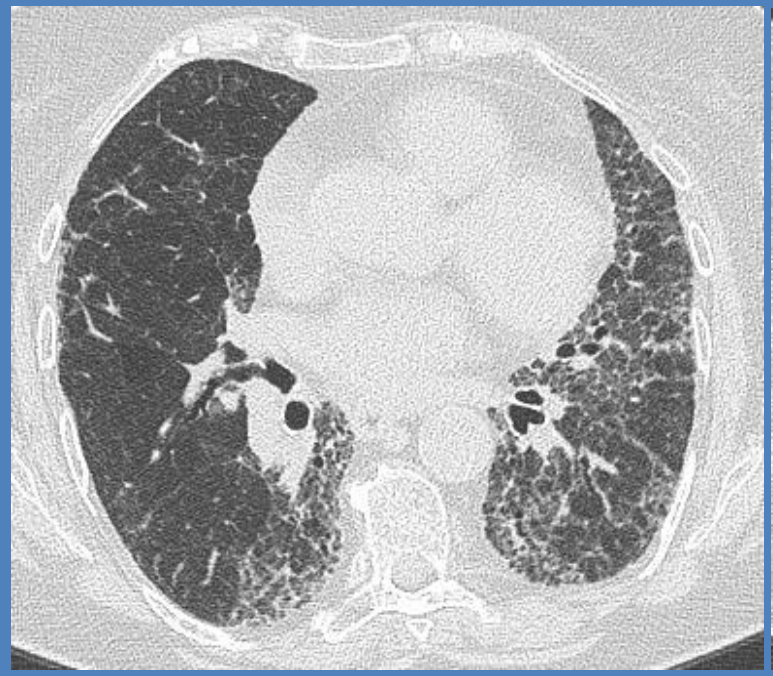


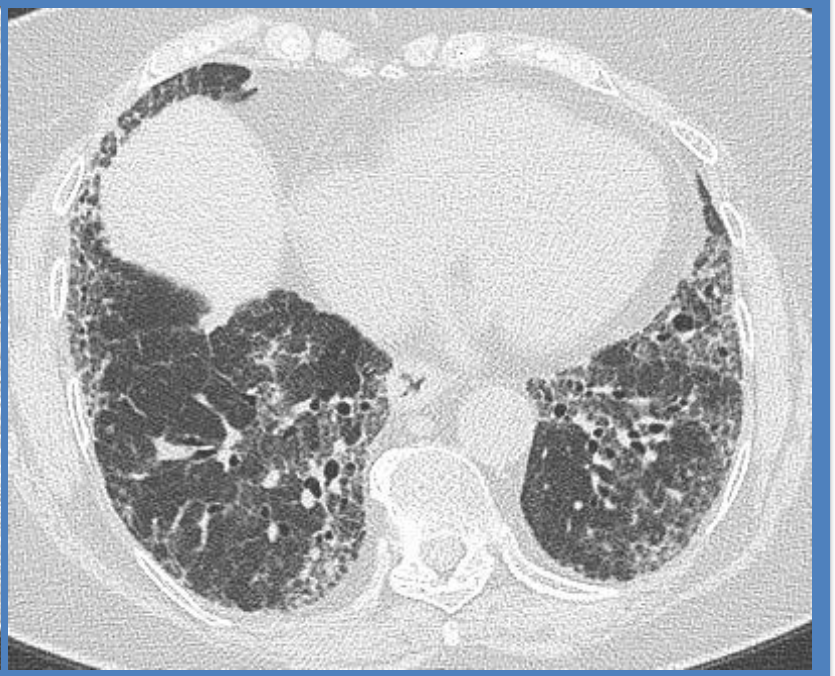


Images courtesy of D. Lynch

CT Pattern Indeterminate for UIP







DISTRIBUTION

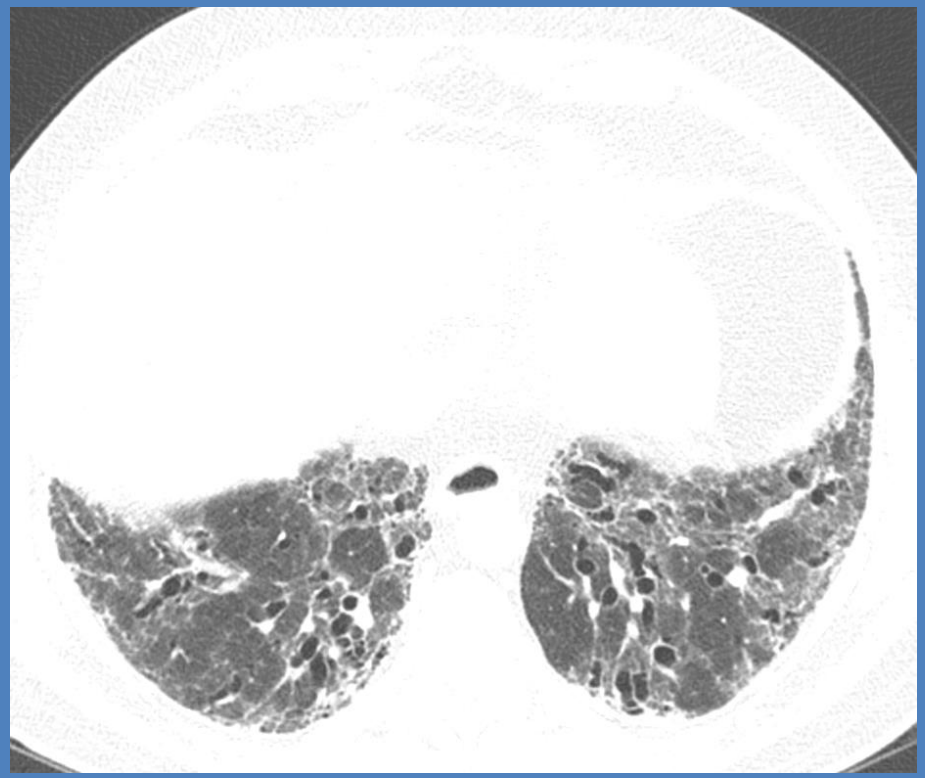
Variable or diffuse

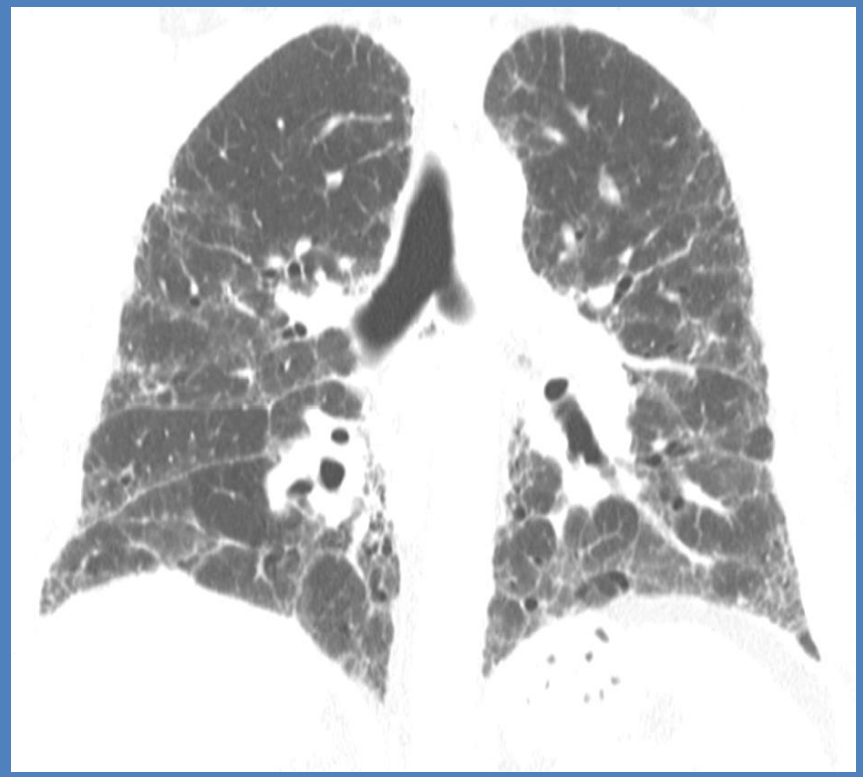
CT FEATURES

Evidence of fibrosis with some inconspicuous features suggestive of non-UIP pattern

Images courtesy of D. Lynch

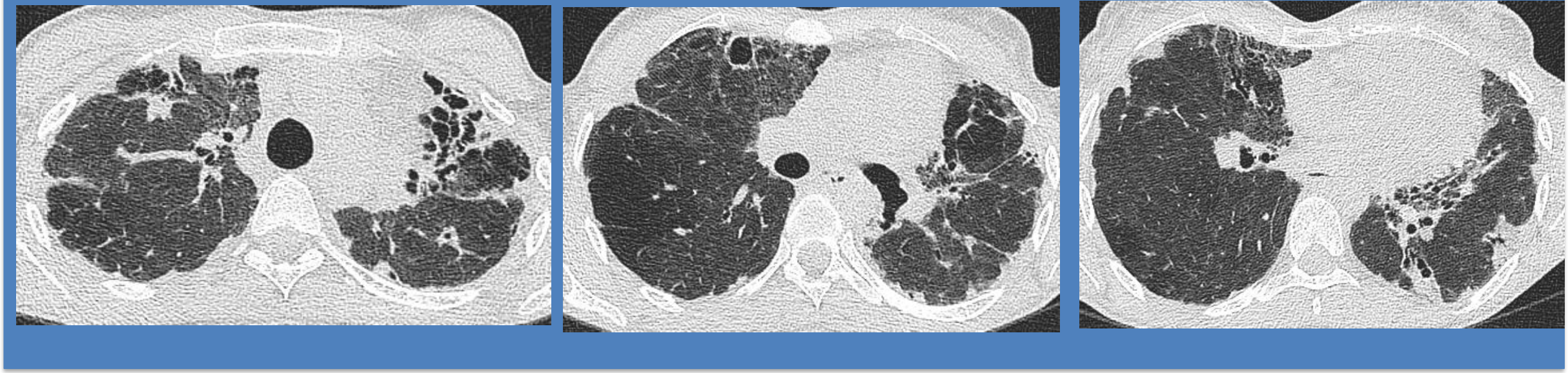
CT Pattern Indeterminate for UIP





Images courtesy of D. Lynch

CT Pattern Most Consistent with Alternative Diagnosis



DISTRIBUTION

Upper- or mid-lung predominant fibrosis

Peribronchovascular predominance with subpleural sparing

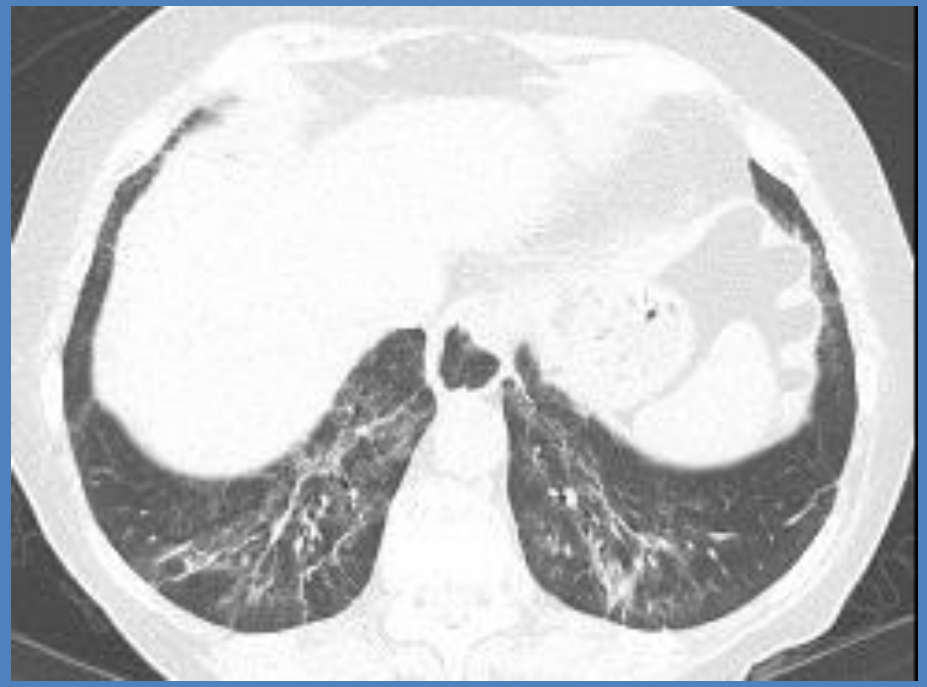
CT FEATURES

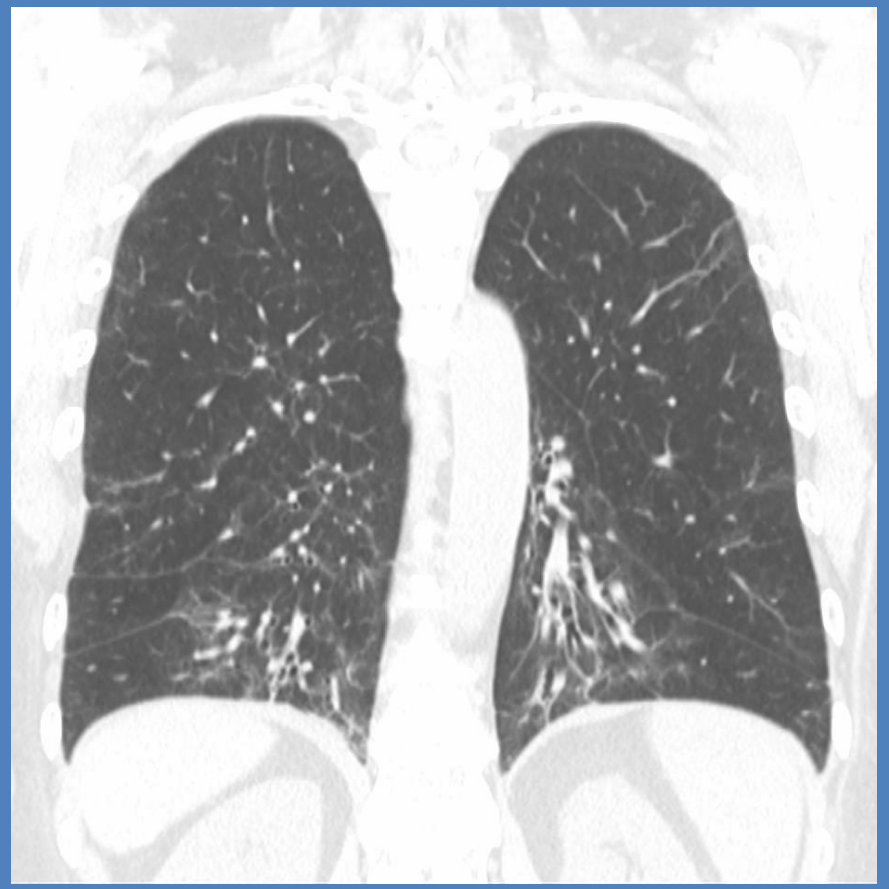
Any of the following:

- Predominant consolidation
- Extensive pure ground glass opacity (without acute exacerbation)
- Extensive mosaic attenuation with extensive sharply defined lobular air trapping on expiration
- Diffuse nodules or cysts

Images courtesy of D. Lynch

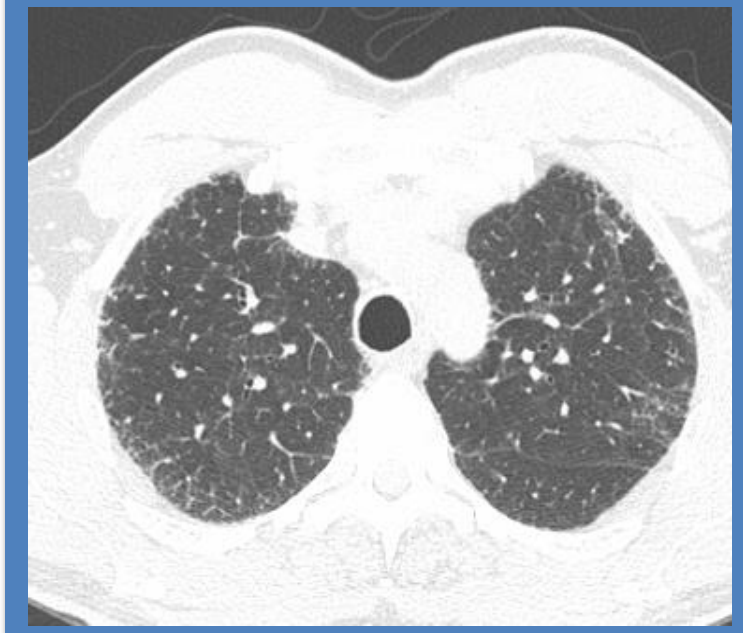
NSIP

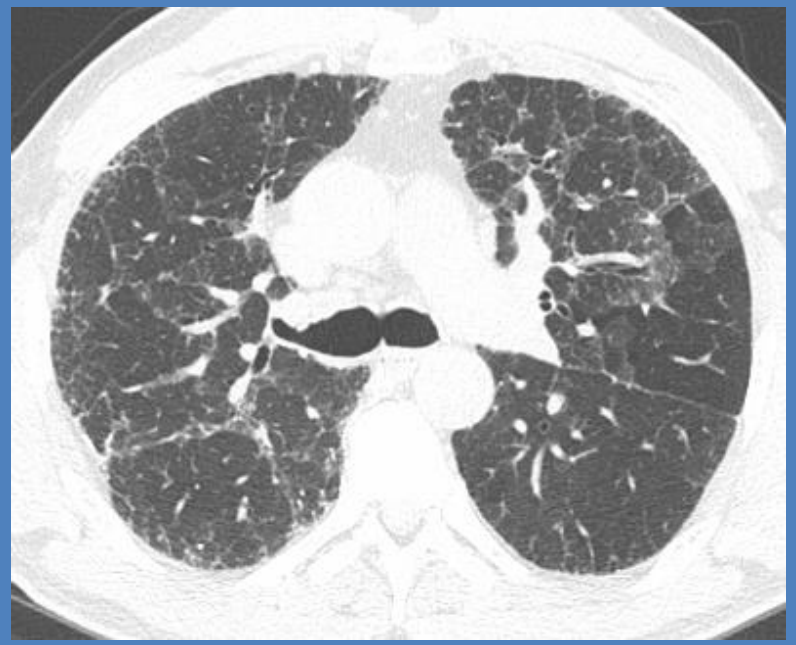


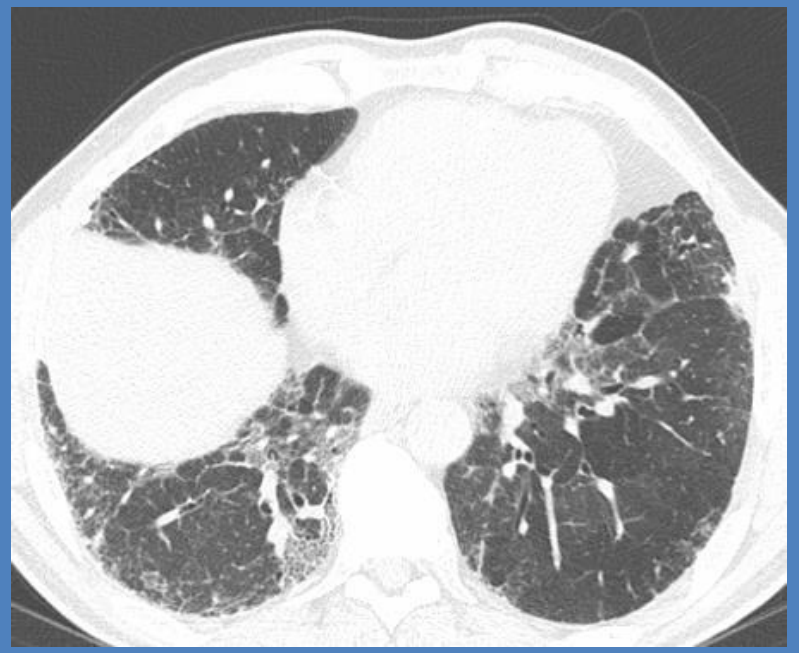


Images courtesy of D. Lynch

Fibrotic HP







DISTRIBUTION

Upper-, mid- or lower-lung predominant

Peribronchovascular, subpleural or diffuse

CT FEATURES

Reticular abnormality

Traction bronchiectasis

Lobar volume loss

± Ground glass

± Mosaic attenuation

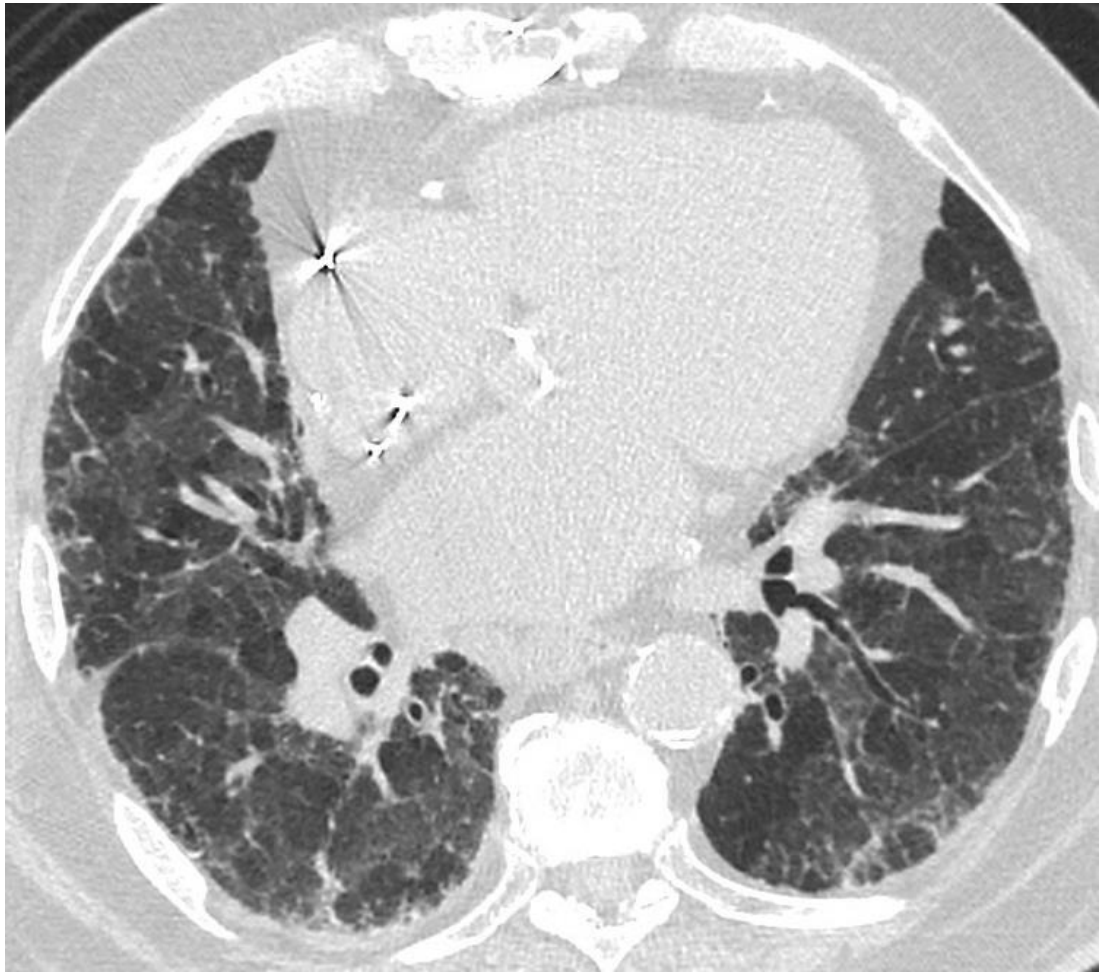
± Expiratory air trapping

± Honeycombing

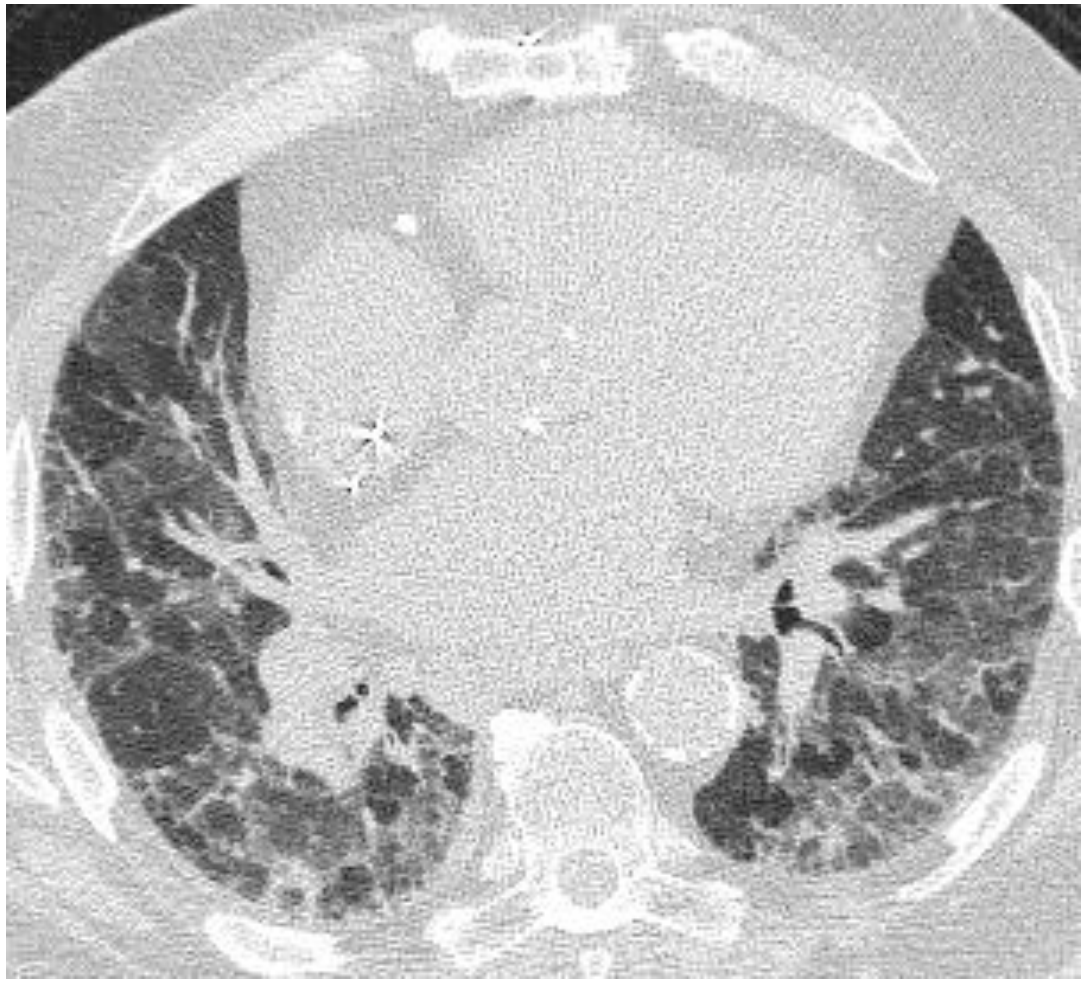
Images courtesy of D. Lynch

Fibrotic HP

Lobular Air Trapping on Expiratory Images



Inspiratory



Expiratory

Images courtesy of L. Heyneman, MD

Pathways to Confident Diagnosis of IPF

- When can one make a confident diagnosis of IPF without biopsy?
  - Clinical context of IPF, with CT pattern of definite or probable UIP
- When is a diagnostic biopsy necessary to make a confident diagnosis of IPF?
  - Clinical context of IPF with CT pattern either indeterminate or suggestive of an alternative diagnosis
  - Clinical context indeterminate for IPF (eg, potential relevant exposure) with any CT pattern

AMERICAN THORACIC SOCIETY DOCUMENTS

Diagnosis of Idiopathic Pulmonary Fibrosis

An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline

Ganesh Raghu, Martine Remy-Jardin, Jeffrey L. Myers, Luca Richeldi, Christopher J. Ryerson, David J. Lederer, Juergen Behr, Vincent Cottin, Sonye K. Danoff, Ferran Morell, Kevin R. Flaherty, Athol Wells, Fernando J. Martinez, Arata Azuma, Thomas J. Bice, Demosthenes Bouros, Kevin K. Brown, Harold R. Collard, Abhijit Duggal, Liam Galvin, Yoshikazu Inoue, R. Gisli Jenkins, Takeshi Johkoh, Ella A. Kazerooni, Masanori Kitaichi, Shandra L. Knight, George Mansour, Andrew G. Nicholson, Sudhakar N. J. Pipavath, Ivette Buendía-Roldán, Moisés Selman, William D. Travis, Simon Walsh, and Kevin C. Wilson; on behalf of the American Thoracic Society, European Respiratory Society, Japanese Respiratory Society, and Latin American Thoracic Society

This official clinical practice guideline of the American Thoracic Society (ATS), European Respiratory Society (ERS), Japanese Respiratory Society (JRS), and Latin American Thoracic Society (ALAT) was approved by the ATS, JRS, and ALAT May 2018, and the ERS June 2018

Background: This document provides clinical recommendations for the diagnosis of idiopathic pulmonary fibrosis (IPF). It represents a collaborative effort between the American Thoracic Society, European Respiratory Society, Japanese Respiratory Society, and Latin American Thoracic Society.

Methods: The evidence syntheses were discussed and recommendations formulated by a multidisciplinary committee of IPF experts. The evidence was appraised and recommendations were formulated, written, and graded using the Grading of Recommendations, Assessment, Development, and Evaluation approach.

Results: The guideline panel updated the diagnostic criteria for IPF. Previously defined patterns of usual interstitial pneumonia (UIP) were refined to patterns of UIP, probable UIP, indeterminate, and alternate diagnosis. For patients with newly detected interstitial lung disease (ILD) who have a high-resolution computed tomography scan pattern of probable UIP, indeterminate, or an alternative diagnosis, conditional recommendations were made for performing BAL and surgical lung biopsy; because of lack of evidence, no recommendation was made for or against performing transbronchial lung biopsy or lung cryobiopsy. In contrast, for patients with newly detected ILD who have a high-resolution computed tomography scan pattern of UIP, strong recommendations were made against performing surgical lung biopsy, transbronchial lung biopsy, and lung cryobiopsy, and a conditional recommendation was made against performing BAL. Additional recommendations included a conditional recommendation for multidisciplinary discussion and a strong recommendation against measurement of serum biomarkers for the sole purpose of distinguishing IPF from other ILDs.

Conclusions: The guideline panel provided recommendations related to the diagnosis of IPF.

Keywords: idiopathic pulmonary fibrosis; interstitial lung disease; pulmonary fibrosis

How do the Updated ATS/ERS/JRS/ALAT Diagnostic Guidelines Differ from Fleischner?

- Both are evidence based
  - ATS guidelines are clinical practice guidelines using GRADE methodology,
  - Fleischner is expert consensus but with systematic literature search based on key questions
- Radiologic categories are essentially the same
- ATS **suggests** surgical biopsy in subjects with ILD of unknown cause who have probable, indeterminate or alternative diagnosis (conditional recommendation)
- ATS **suggests** BAL in the same population
- ATS does not clearly include the concept of “working” or “provisional” diagnosis of IPF

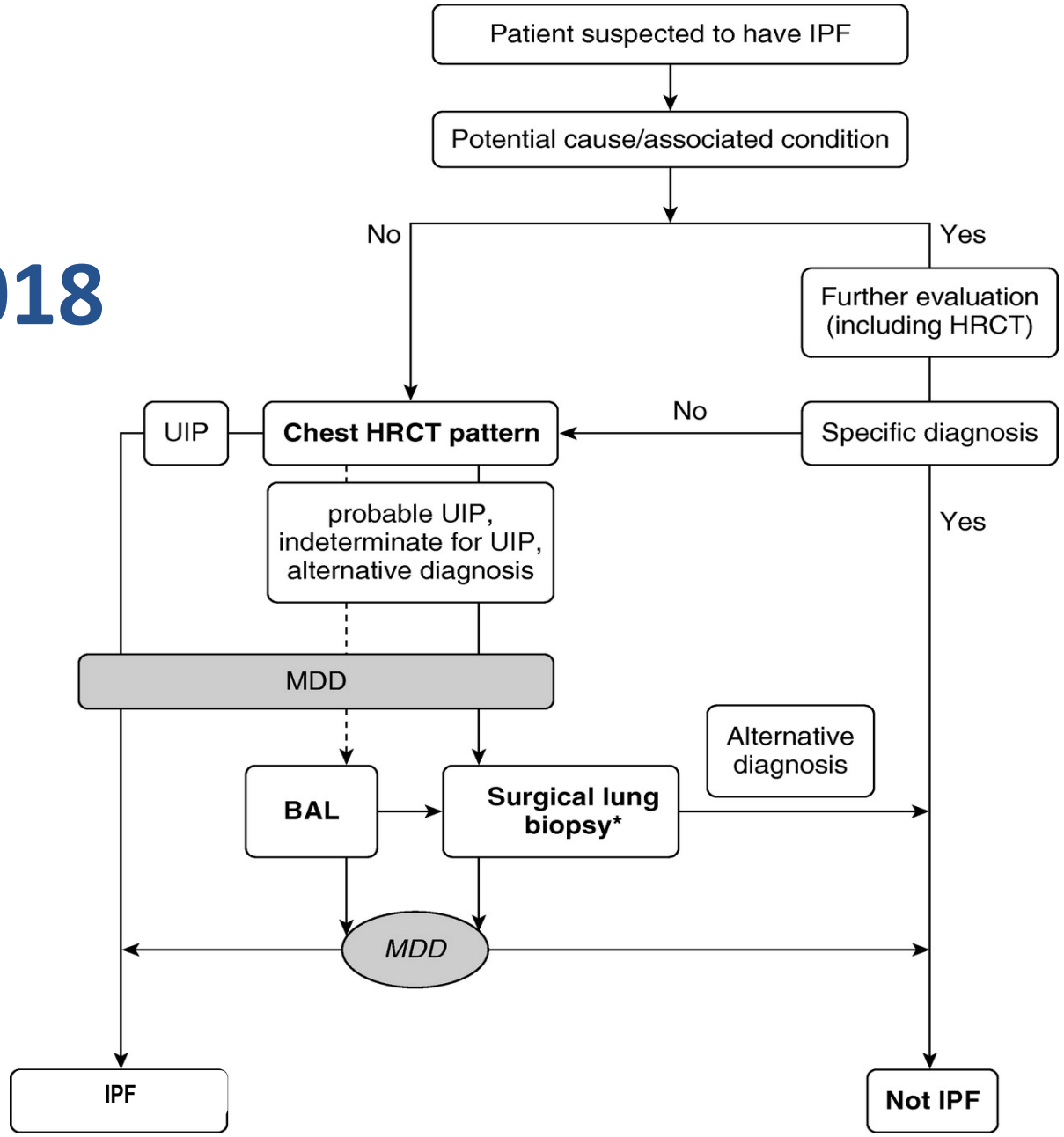
The Reality

- CT patterns provide valuable information on the **probability** of histologic UIP and IPF

Typical UIP	~ 90%
Probable UIP	~ 80%
Indeterminate	~ 50%
Alternative diagnosis	~ 50%

- These probabilities should be integrated with clinical probability in deciding on further diagnostic management

IPF Diagnosis: Flow Diagram- ATS Guidelines, 2018



Raghu G et al. *Am J Respir Crit Care Med*. 2018;198(5):e44-e68.

IPF Diagnosis-ATS Guidelines, 2018

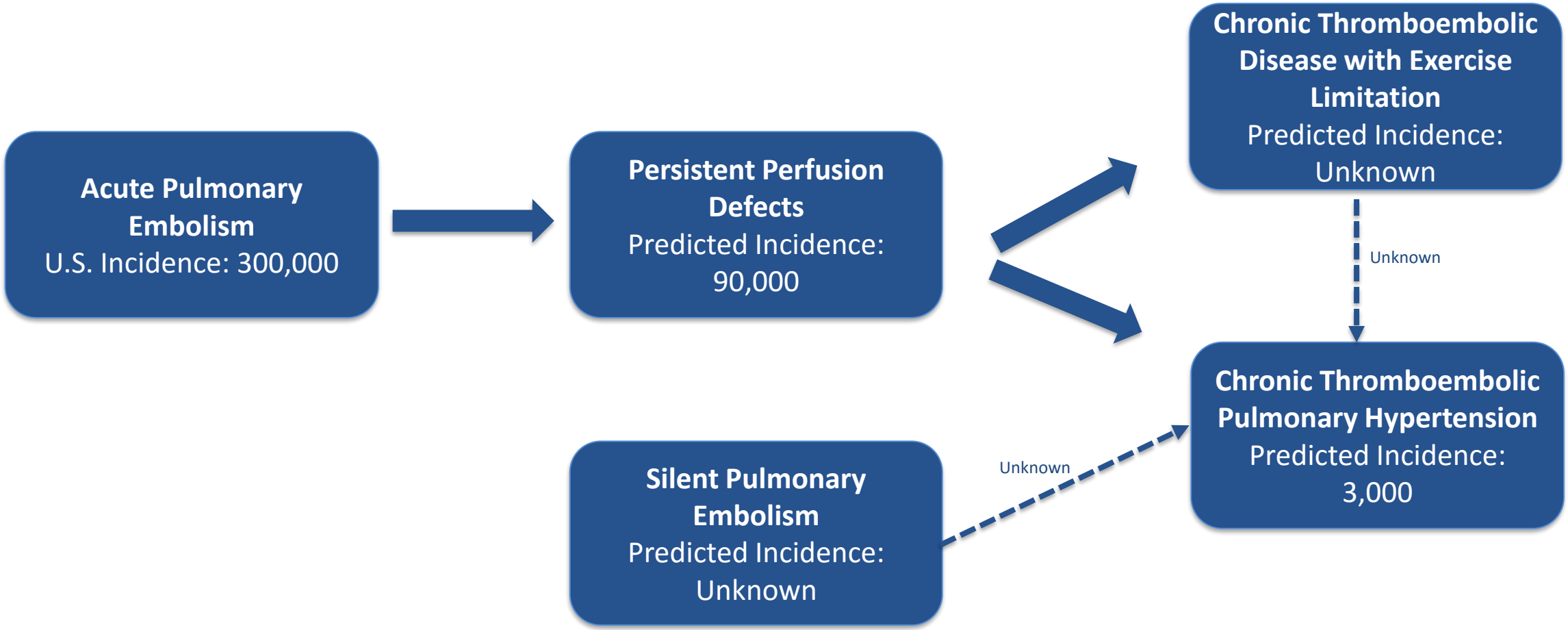
IPF suspected*		Histopathology pattern			
		UIP	Probable UIP	Indeterminate for UIP	Alternative diagnosis
HRCT pattern	UIP	IPF	IPF	IPF	Non-IPF dx
	Probable UIP	IPF	IPF	IPF (Likely)**	Non-IPF dx
	Indeterminate for UIP	IPF	IPF (Likely)**	Indeterminate for IPF***	Non-IPF dx
	Alternative diagnosis	IPF (Likely)** /non-IPF dx	Non-IPF dx	Non-IPF dx	Non-IPF dx

Important Points

- IPF is a clinical diagnosis
  - Pulmonology → ILD
  - Radiology → UIP
  - (Pathology) → UIP
- Using the guideline-based vocabulary will facilitate a guideline-based diagnosis
  - New guidelines
- Biopsy is not necessary for IPF diagnosis with definite **or probable UIP, if the clinical context is appropriate**

CTEPH

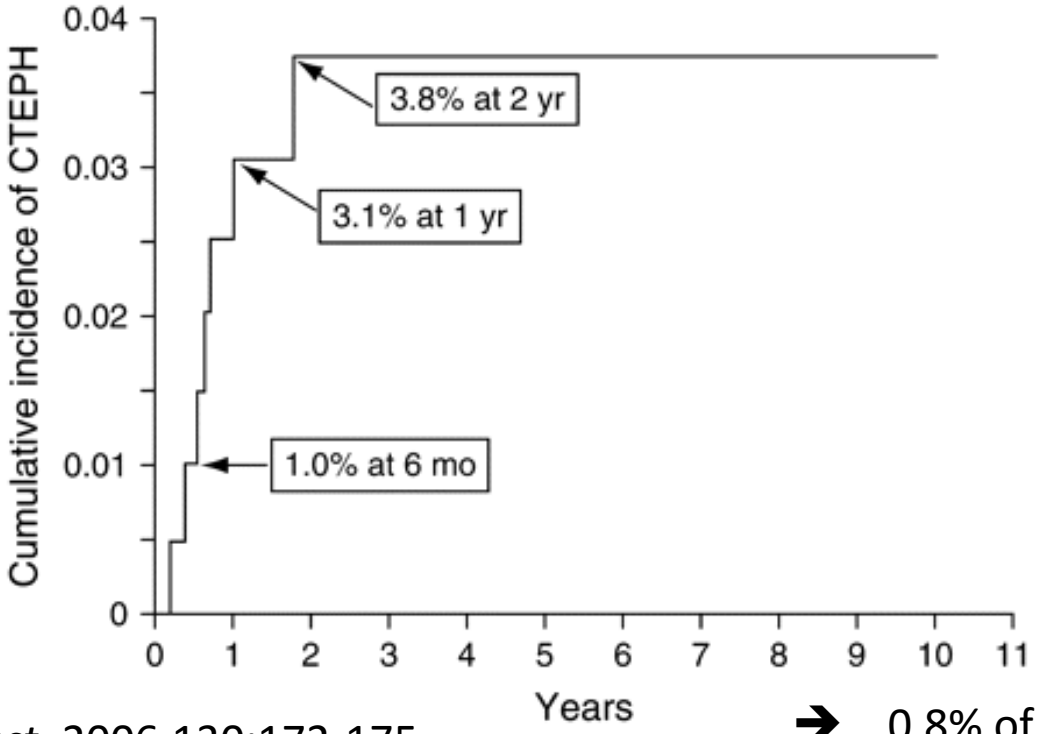
Acute PE May Fail to Resolve Leading to CTEPH



Estimates of the annual U.S. incidence of chronic thromboembolic pulmonary hypertension based on the U.S. annual incidence of pulmonary embolism

Fernandes T, et al. *Thromb Res*. 2018;164:145-9.

Cumulative Incidence of CTEPH After a First Episode of Pulmonary Embolism Without Prior Deep-Vein Thrombosis



- Becattini P, et al. *Chest.* 2006;130:172-175. ➔ 0.8% of 259 patients
- Miniati M, et al. *Medicine*. 2006;85:253-262. ➔ 0.8% of 259 patients
- Klok F, et al. *Haematologica*. 2010; 95:970-975. ➔ 0.57-1.5% of 866 patients
- Korkmaz A, et al. *Clin Appl Thromb Hemost*. 2012;18:281-288. ➔ 4.6% of 291 patients

Pengo V, et al. *N Engl J Med.* 2004;350:2257-2264.

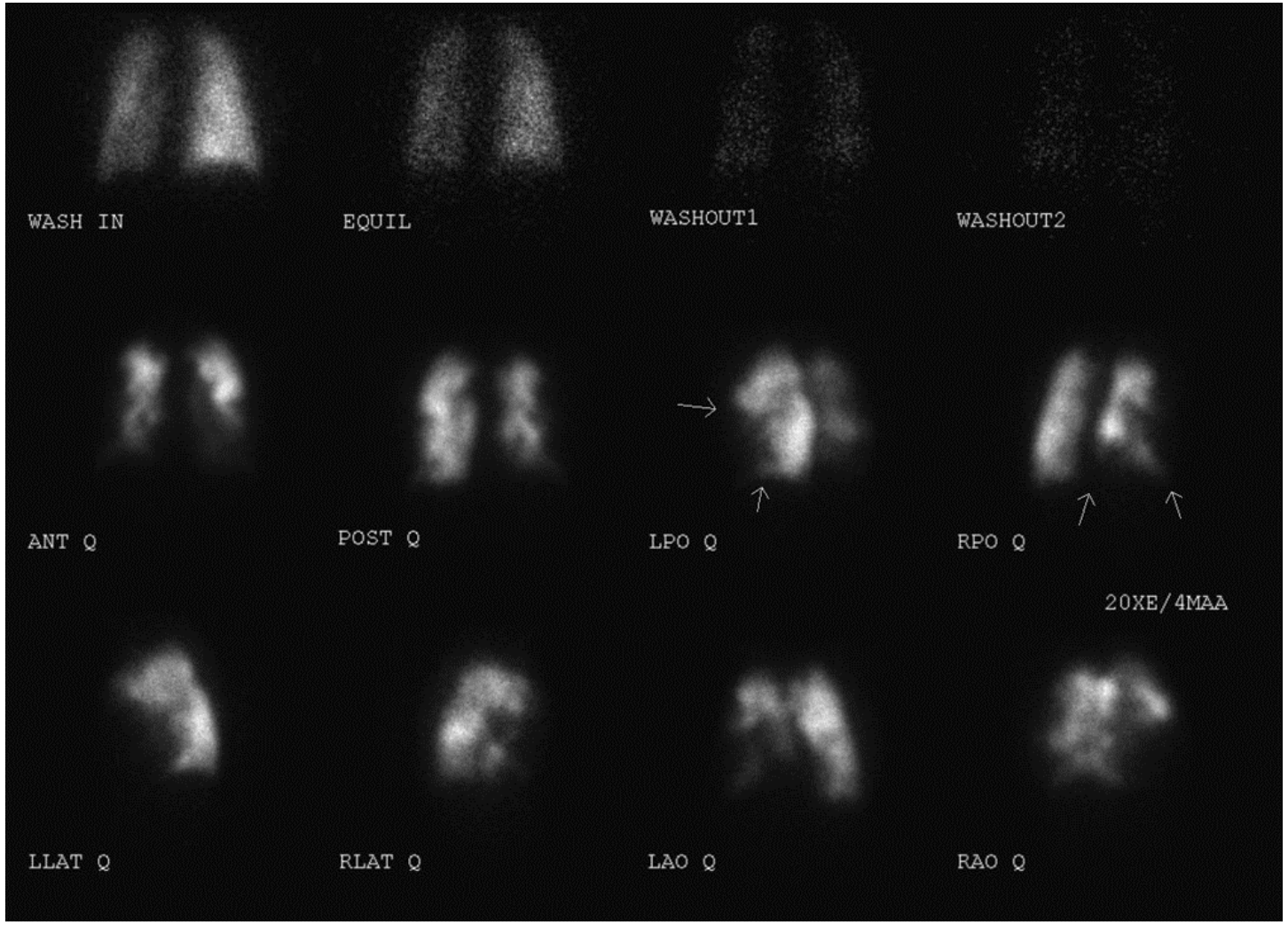
Identified Risk Factors for CTEPH

Table 1
Selected risk factors and univariate odds ratios for CTEPH.

Risk factor	Comparator	OR (95% CI)	Ref.
VA shunt	IPAH	19.49 (2.47–2520)	[33]
Splenectomy	IPAH	22.09 (2.97–2824)	[33]
Massive/submassive PE	IPAH	13.03 ($p = .004$)[a]	[18]
VTE history	IPAH	49.01 ($p < .001$)[a]	[18]
Recurrent VTE	IPAH	45.02 (21.00–114.73)	[33]
Thyroid replacement	IPAH	5.41 (2.70–12.23)	[33]
Hypothyroidism	Resolved PE	4.3 (1.4–13.0)	[15]
Prior VTE	IPAH	19.36 (11.66–33.79)	[33]
APS/lupus AC	IPAH	3.28 (1.58–7.50)	[33]
Non-blood group O	IPAH	3.12 ($p < .001$)[a]	[18]
Malignancy	IPAH	1.99 (1.01–4.26)	[33]
Unprovoked PE	Resolved PE	20.0 (2.7– > 100)	[15]
RV dysfunction at diagnosis	Resolved PE	4.1 (1.4–12.0)	[15]
Symptoms > 2 weeks prior to PE diagnosis	Resolved PE	7.9 (3.3–19.0)	[15]
Age > 60 years	Resolved PE	2.9 (1.2–7.2)	[15]

[a] Adjusted for age and sex.

VQ Scan Remains Screening Test of Choice



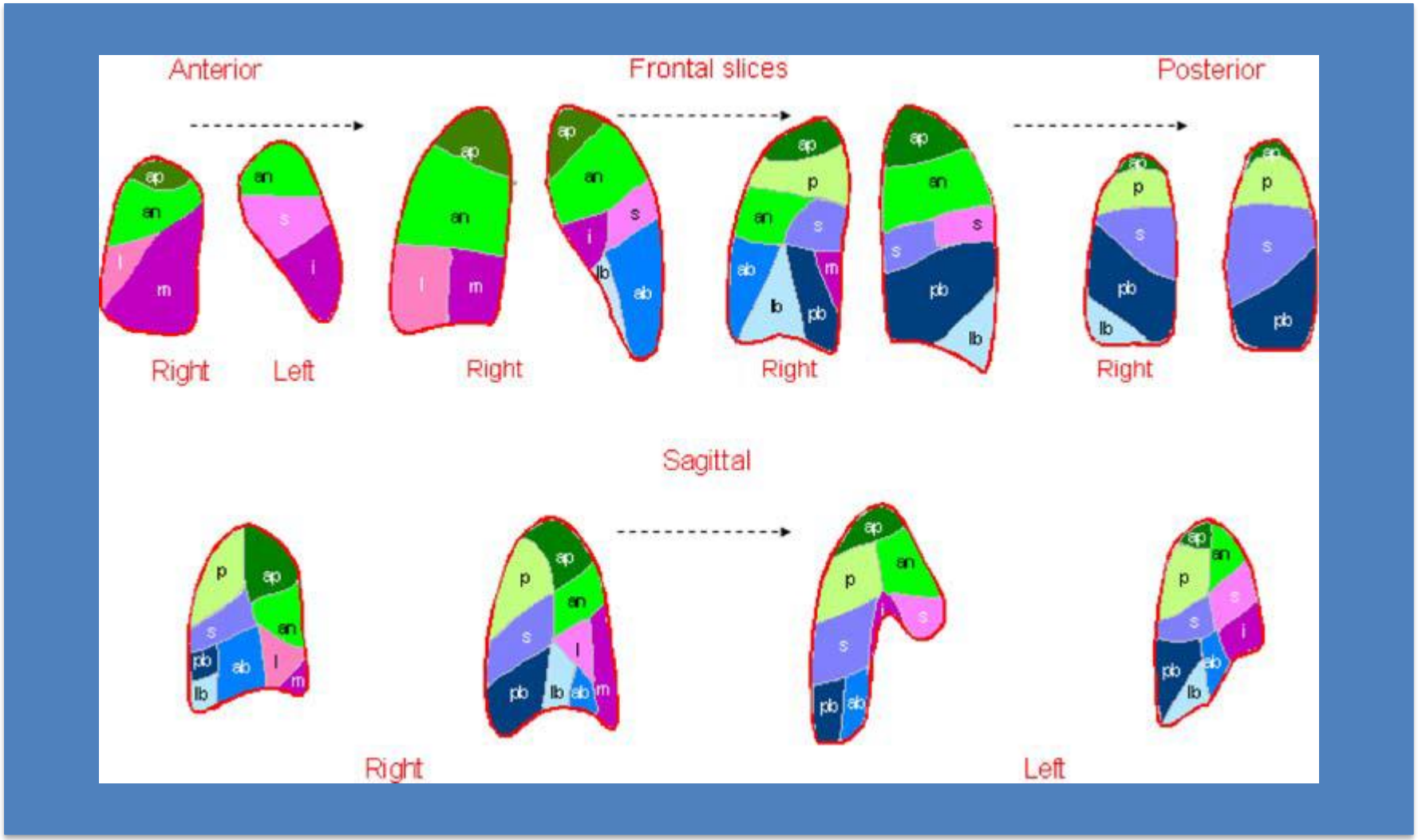
V/Q Scanning Basic Principles

- V/Q scanning exploits the unique pulmonary arterial segmental anatomy. Each bronchopulmonary segment is supplied by a single end artery.

- In principle, conical bronchopulmonary segments have their apex towards the hilum and base projecting onto the pleural surface.

- Occlusive thrombi affecting individual pulmonary arteries therefore produce characteristic lobar, segmental or subsegmental peripheral wedge-shaped defects with the base projecting to the lung periphery.

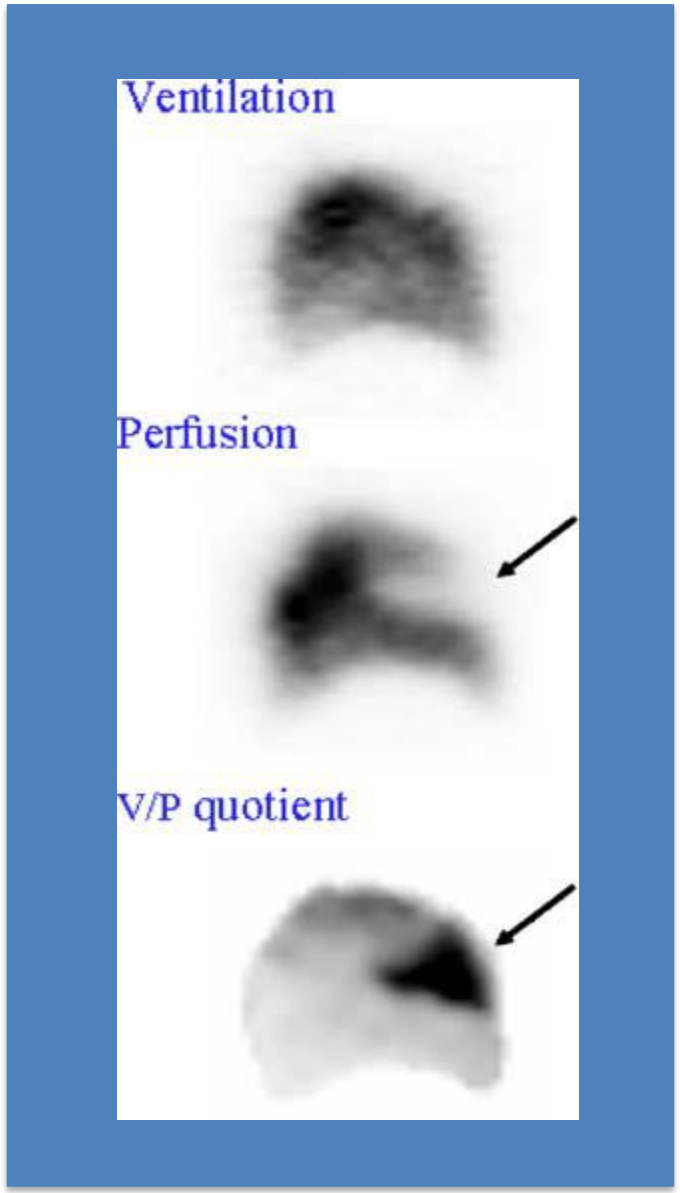
Anatomy


Anterior
Frontal slices
Posterior
ap
an
s
i
l
m
Right
Left
Right
Right
Right
p
lb
pb
ab
Sagittal
Right
Left

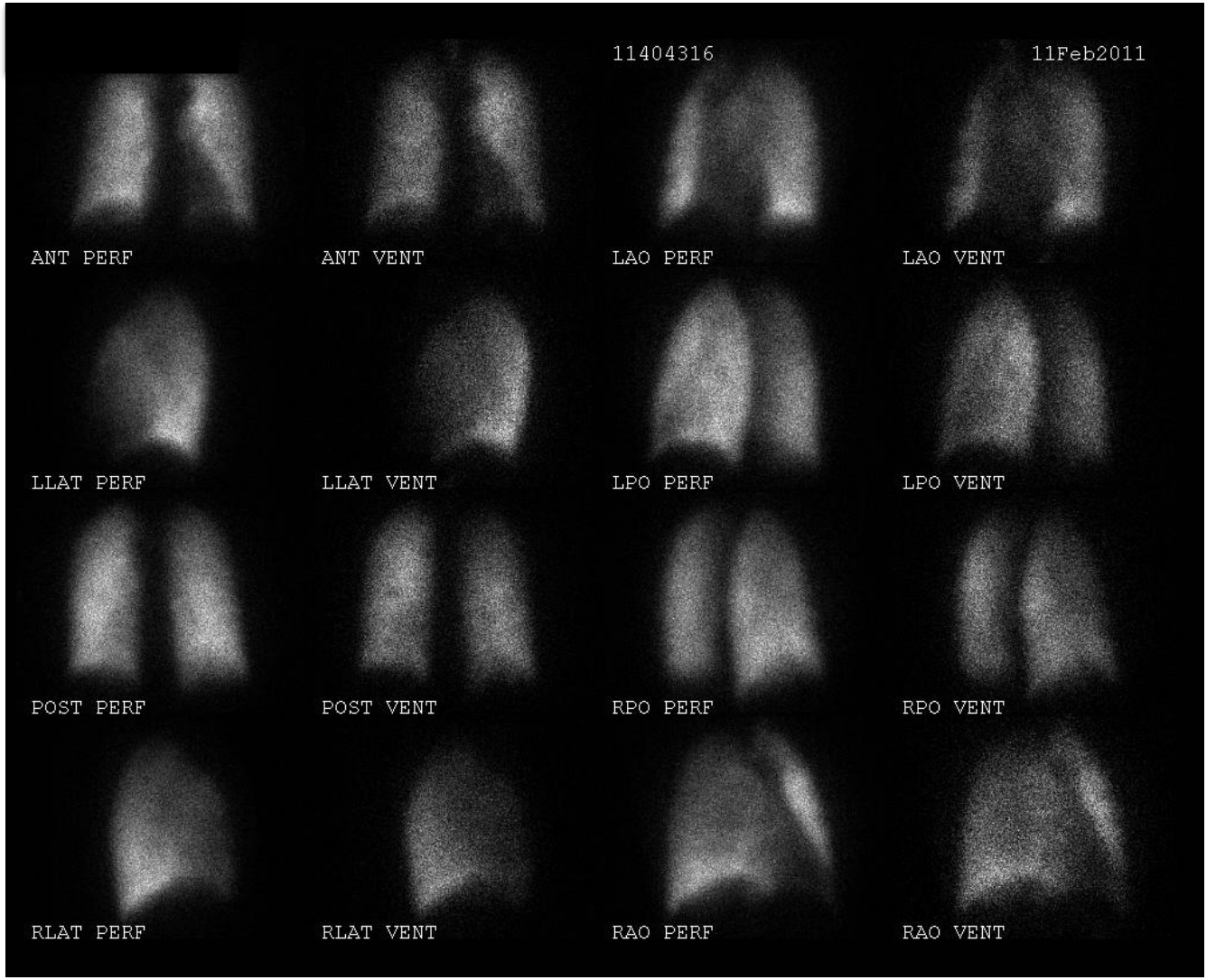
V/Q Mismatch

- Within bronchopulmonary segment(s) affected by PE, ventilation is usually preserved.
- This pattern of preserved ventilation and absent perfusion within a lung segment gives rise to the fundamental rubric for PE diagnosis using V/Q scanning known as V/Q mismatch.
- It is generally accepted that a normal pulmonary perfusion pattern excludes acute and chronic PE.

Typical Defect

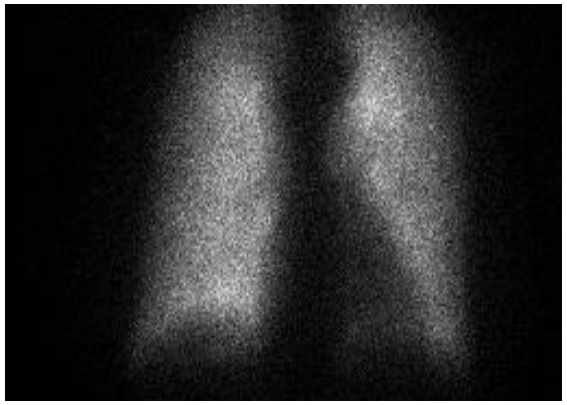


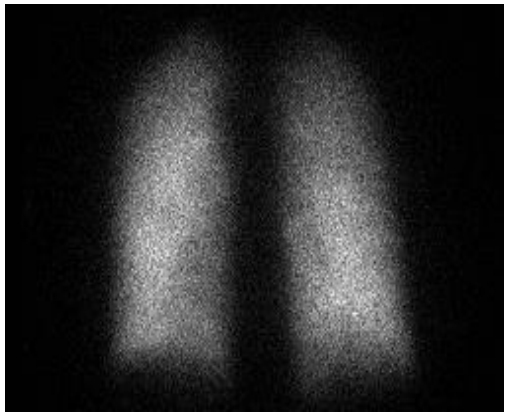
Normal VQ Scan: No Areas of VQ Mismatch



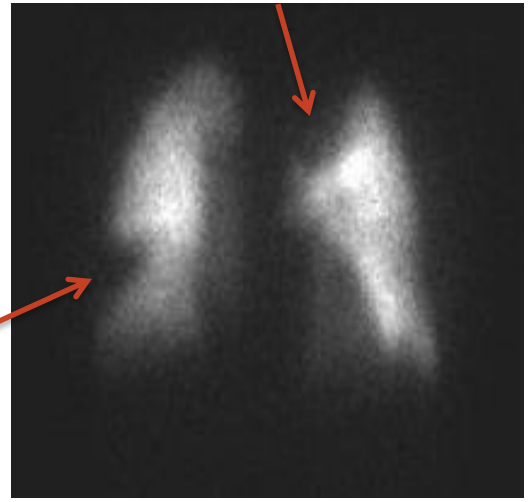
Unmatched Perfusion Defects

Ventilation

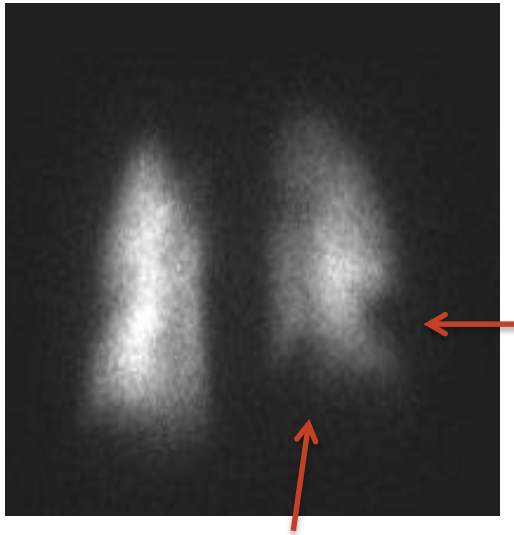




Perfusion



Anterior



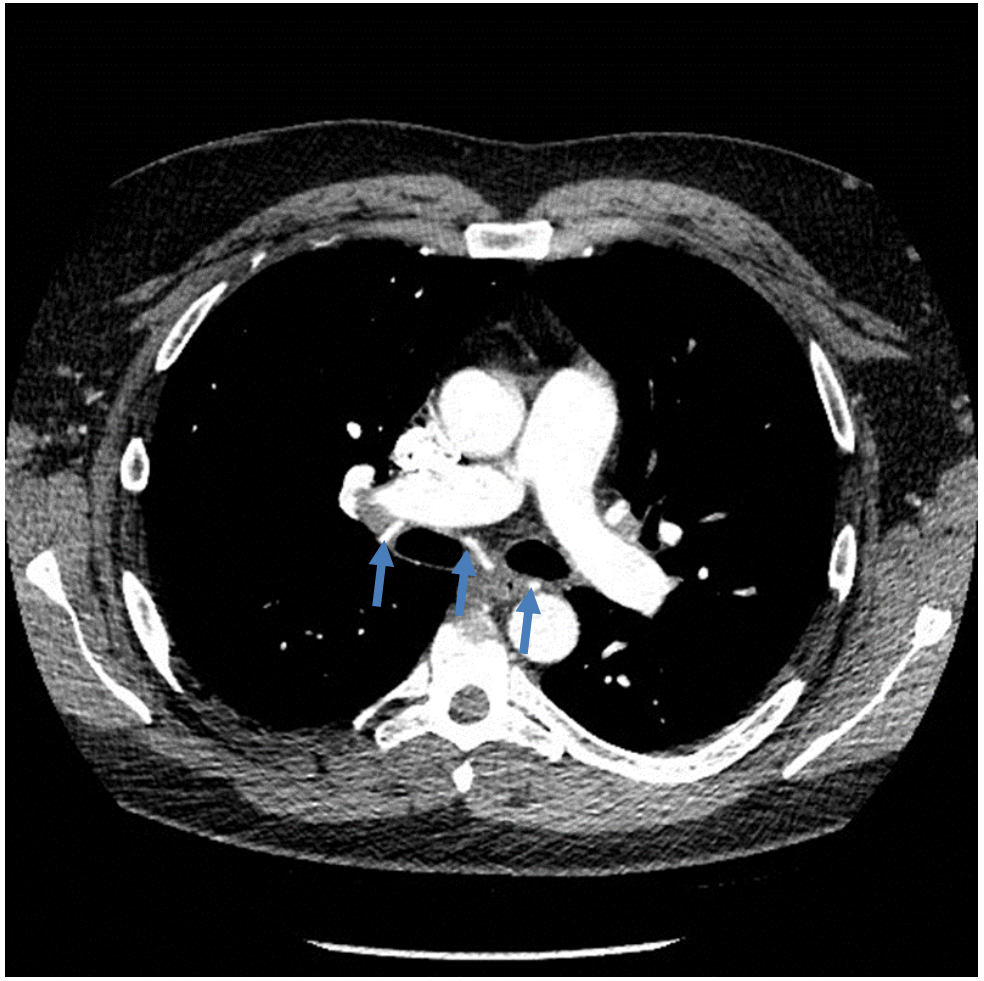
Posterior

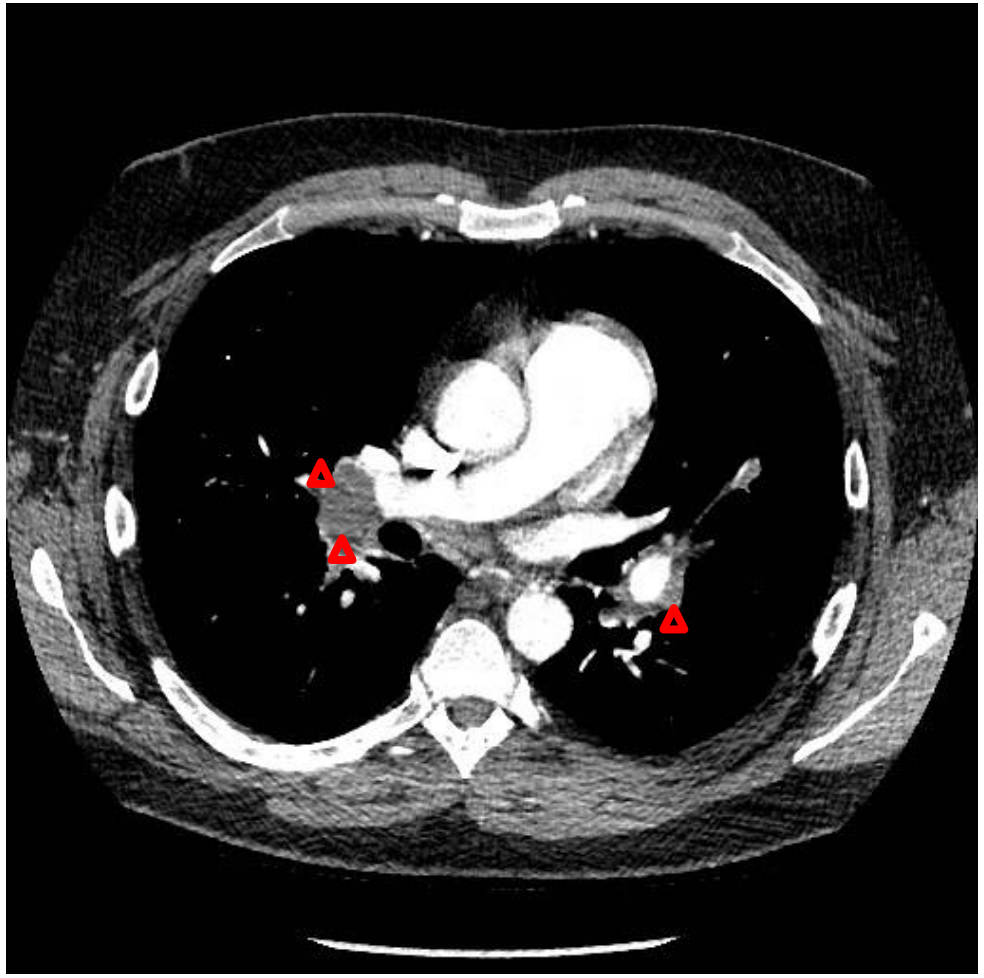
Further Imaging

- Unmatched perfusion defects on V/Q is very suggestive of CTEPH but does not confirm the diagnosis.

- Other imaging (CTA, DSA or MRA) are required to confirm the diagnosis of CTEPH.

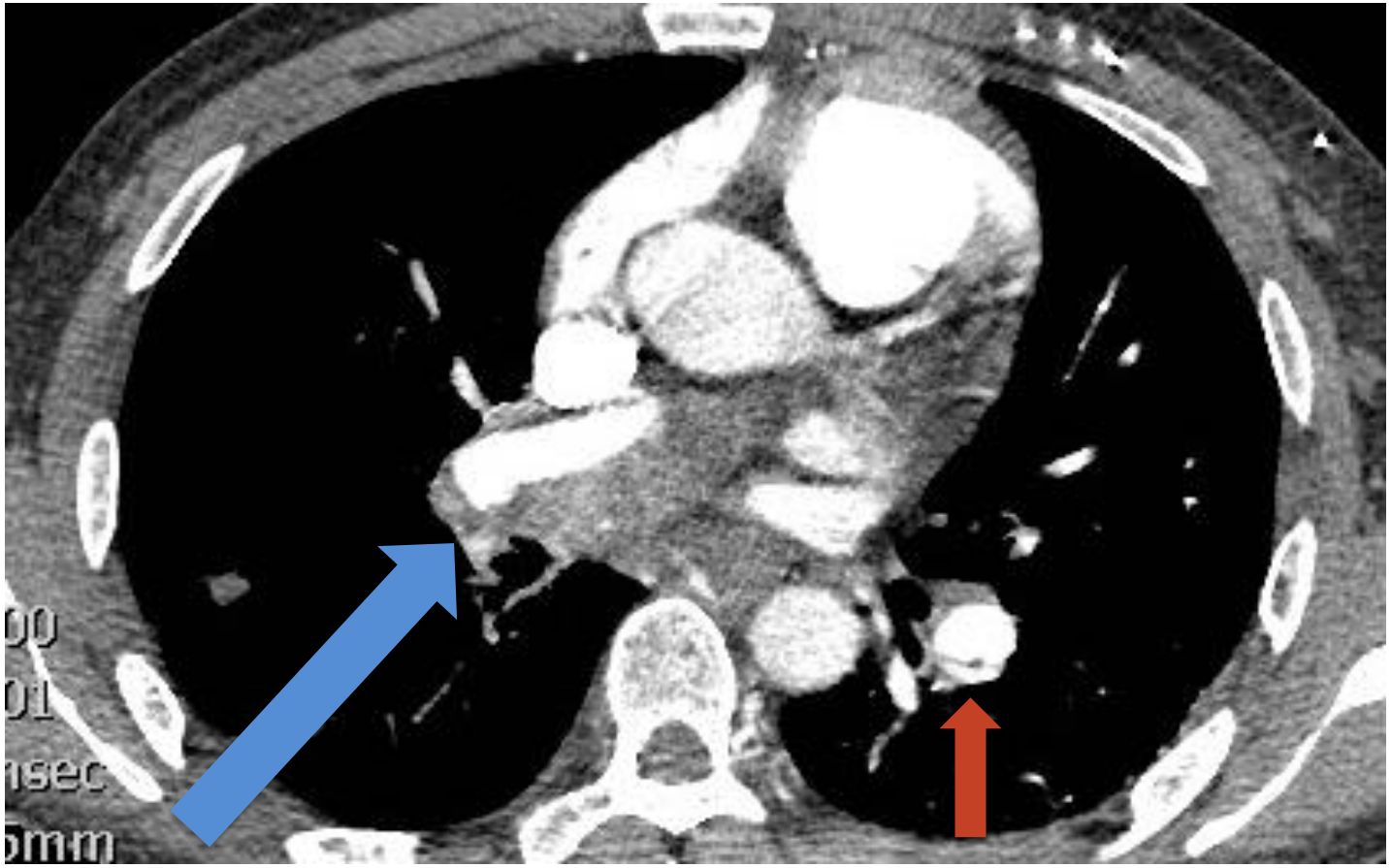
Clues to CTEPH Present on CT





Fernandes TM, et al. *Am J Respir Crit Care Med*. 2017;195(8):1066-1067.

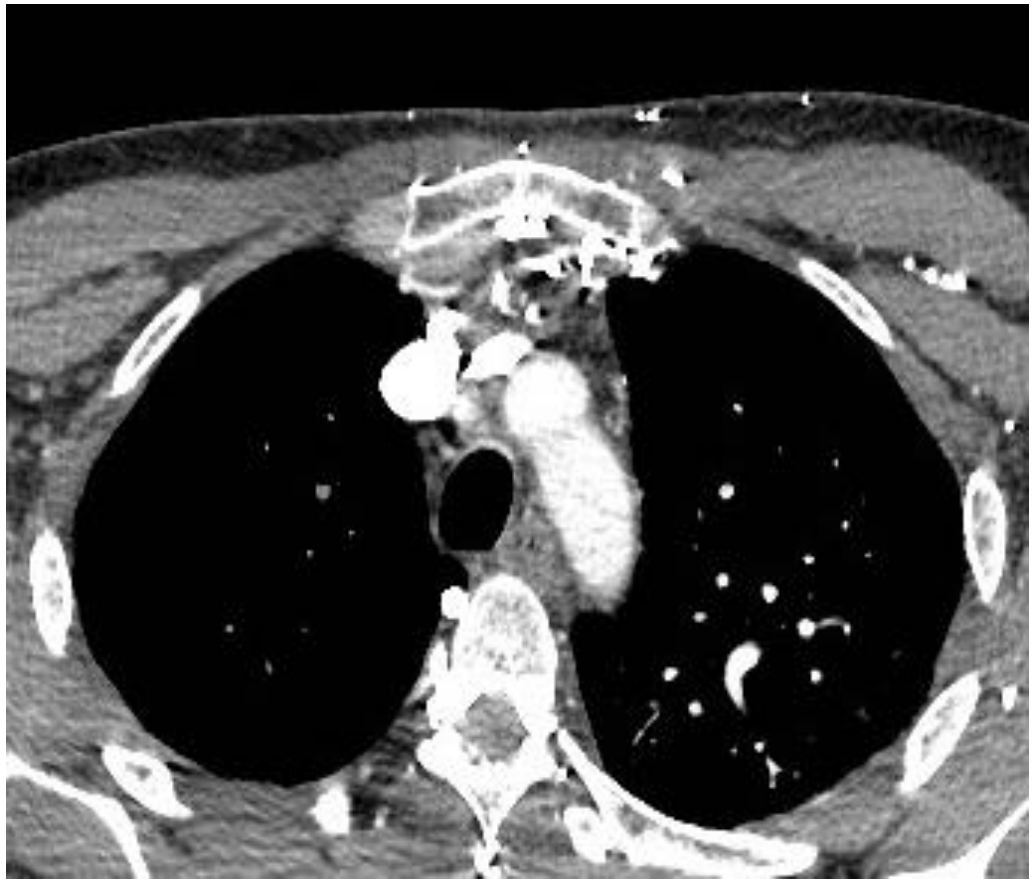
Web and Lining Thrombus



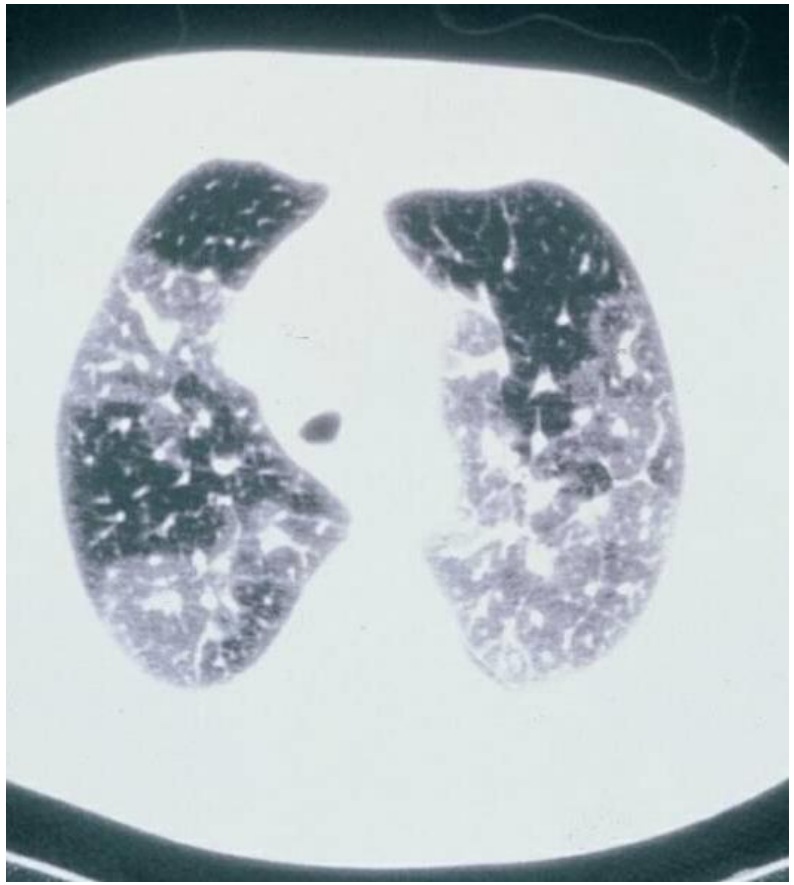
Lining thrombus

Web in left descending PA

Vessel Asymmetry

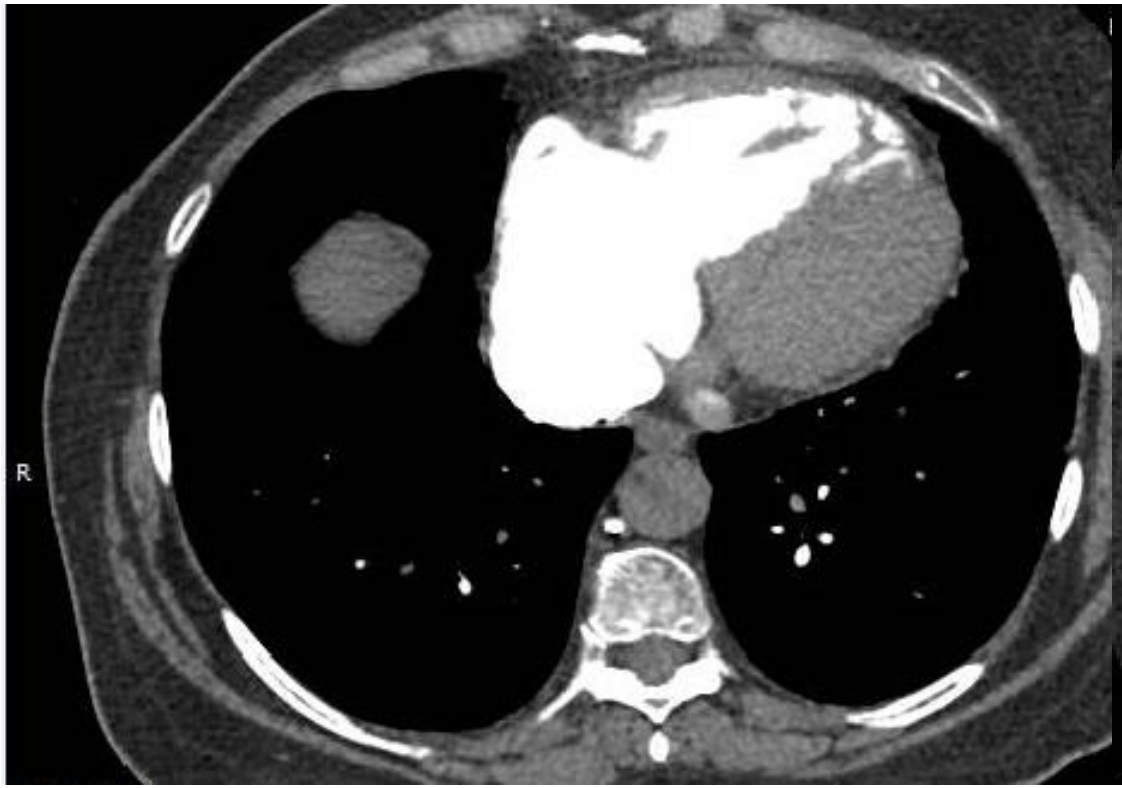


Mosaic Perfusion

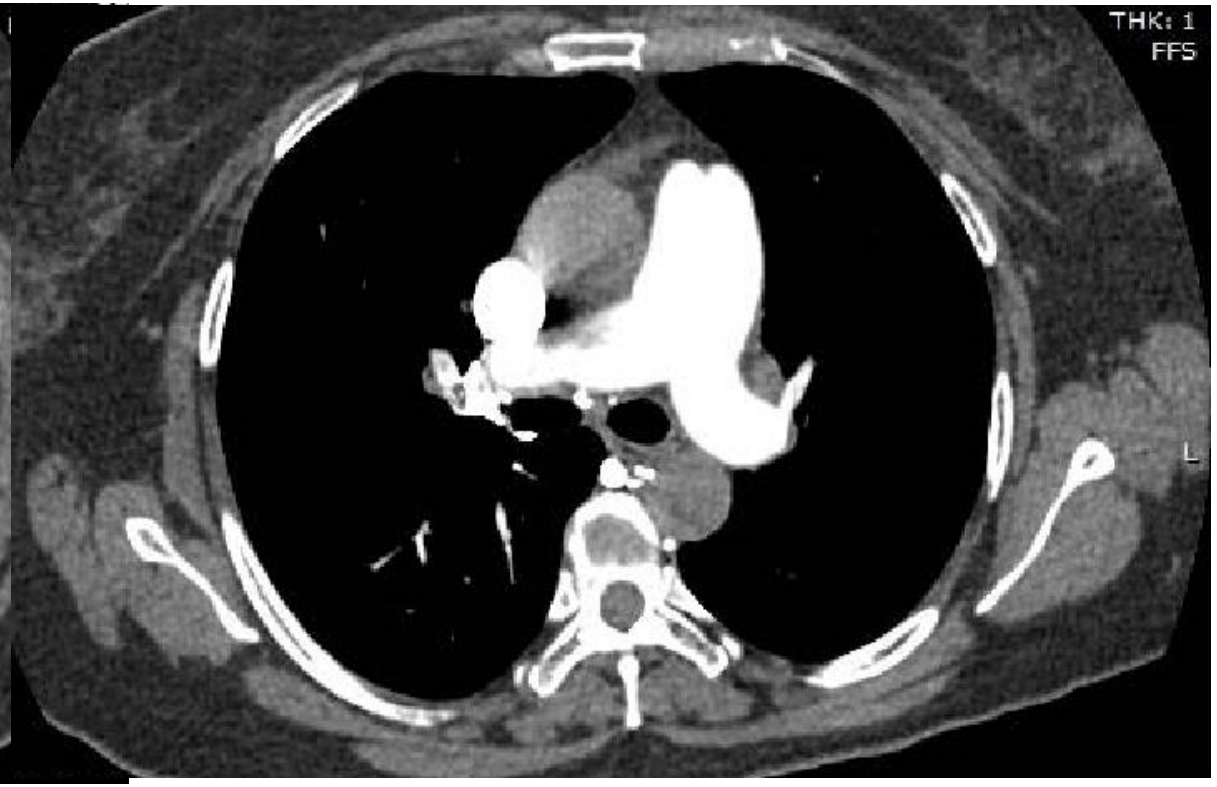


- May result from regional pulmonary vascular disease but not diagnostic
- White areas are the relatively hyperperfused regions of lung. May be confused with GGO

CT Findings
Signs of PA Hypertension

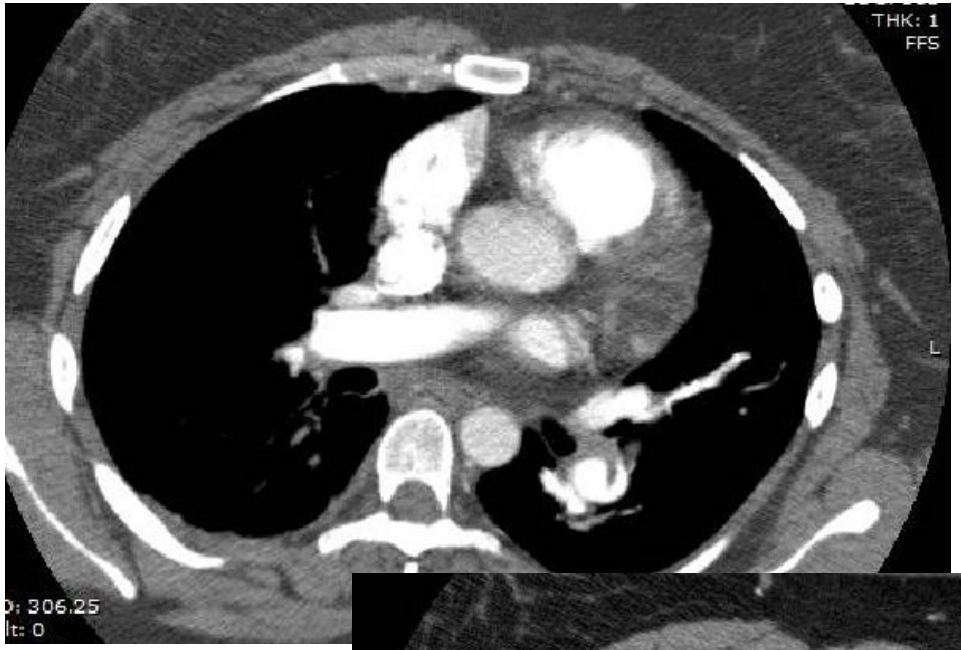


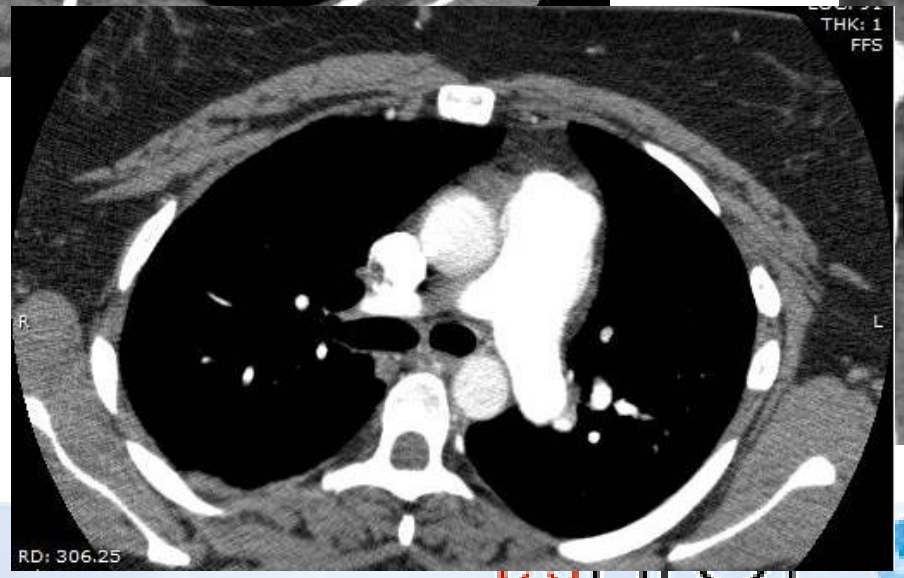
RV Hypertrophy

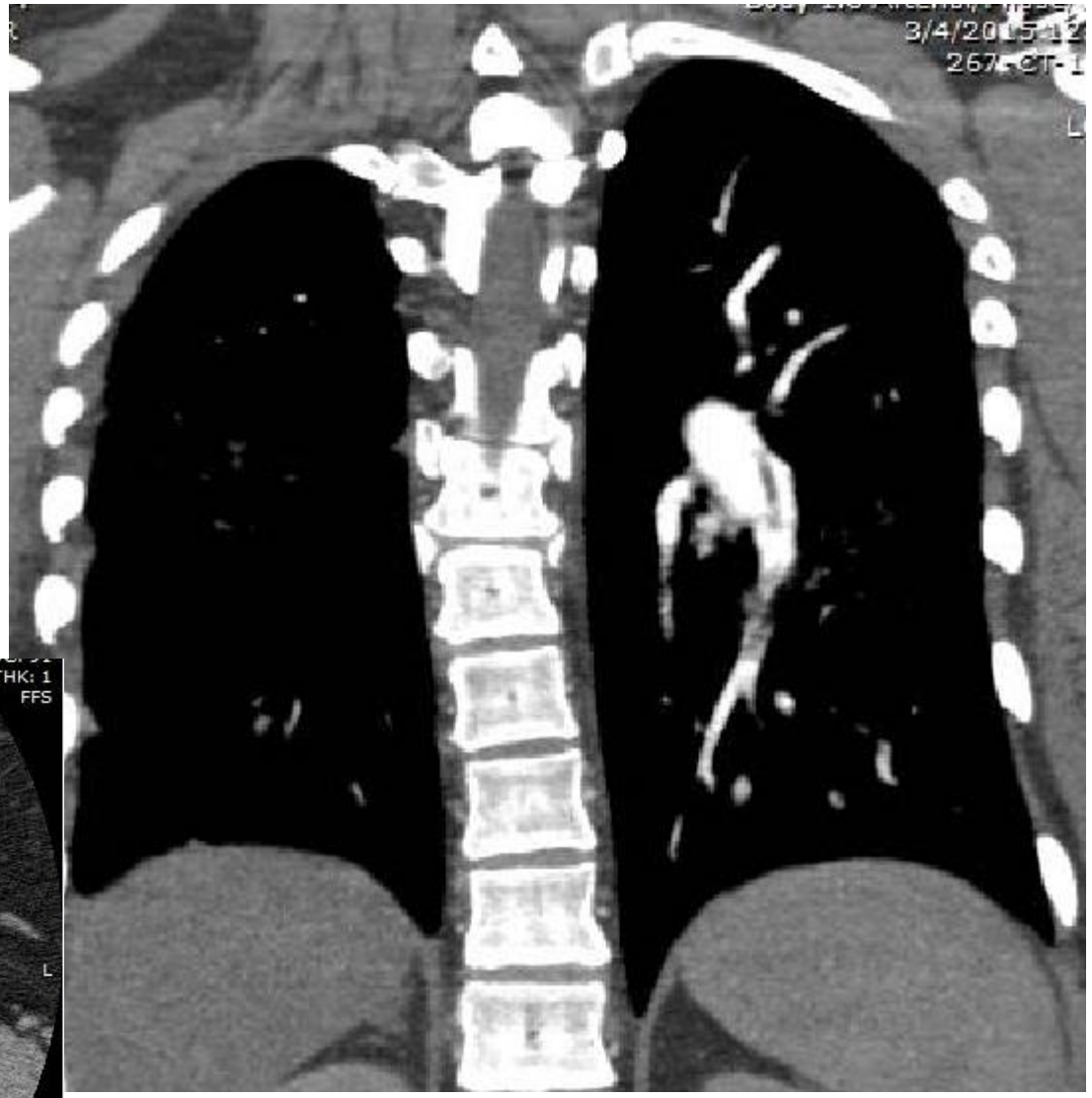


Enlarged PA with Collaterals

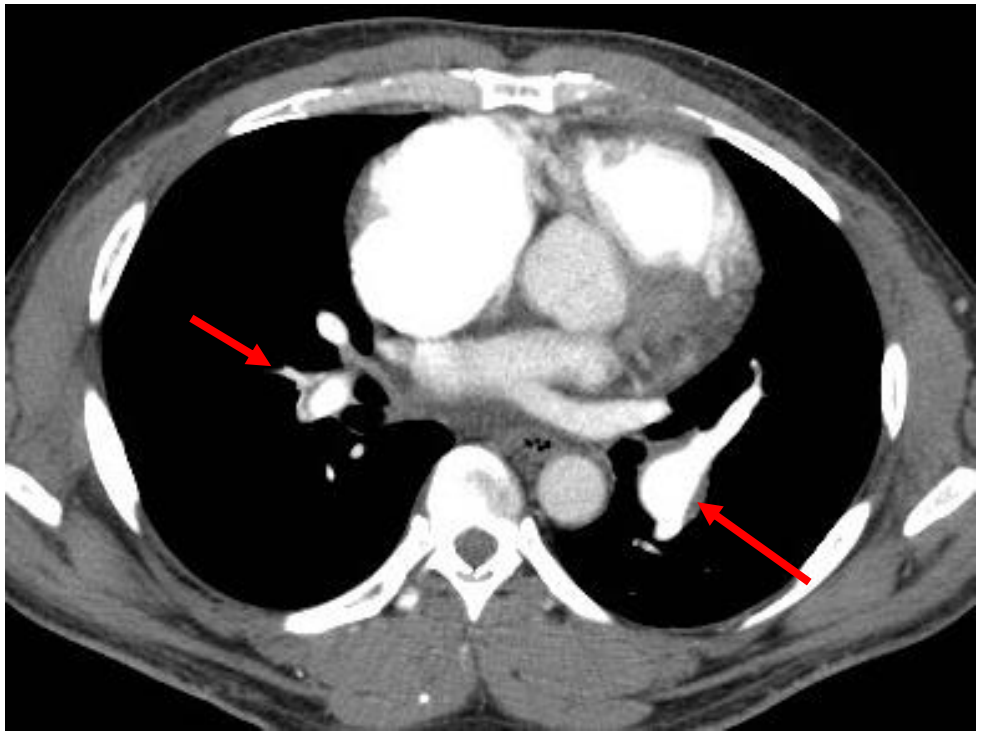
30-Year-Old Female

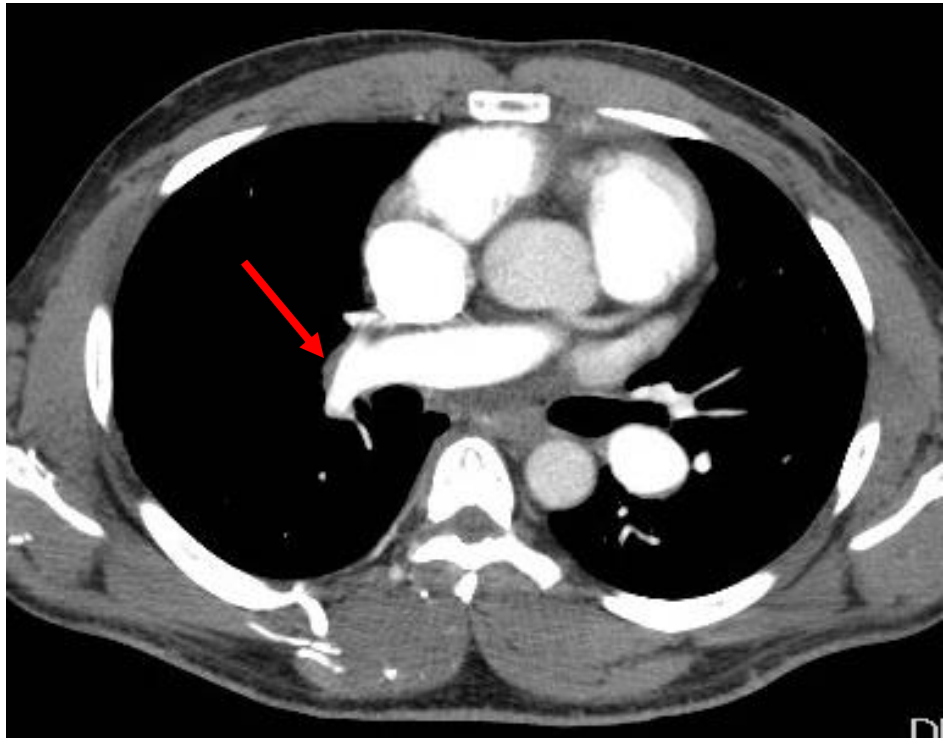






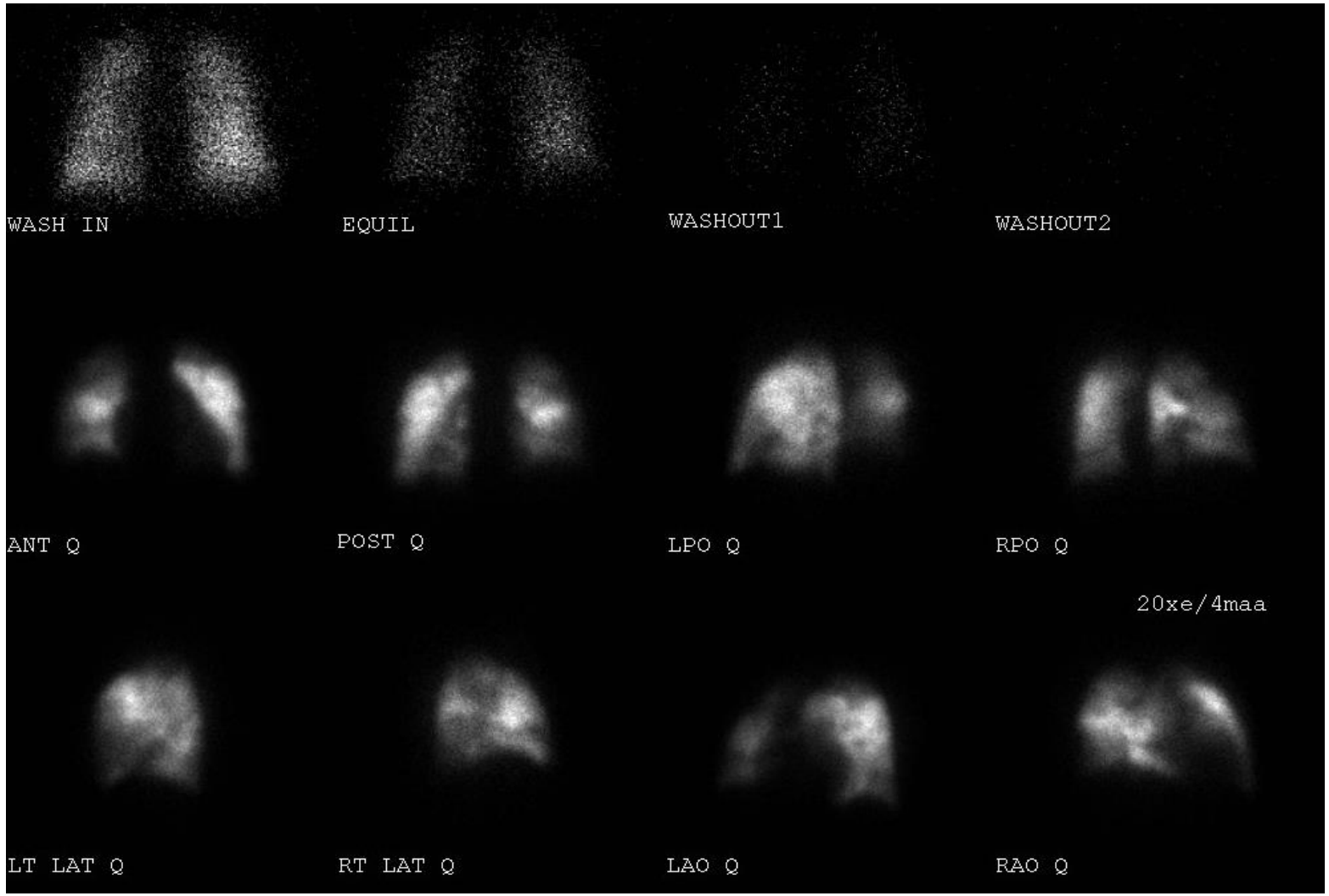
50-year-old Female with PH
CT Findings Reveal Eccentric Thrombus



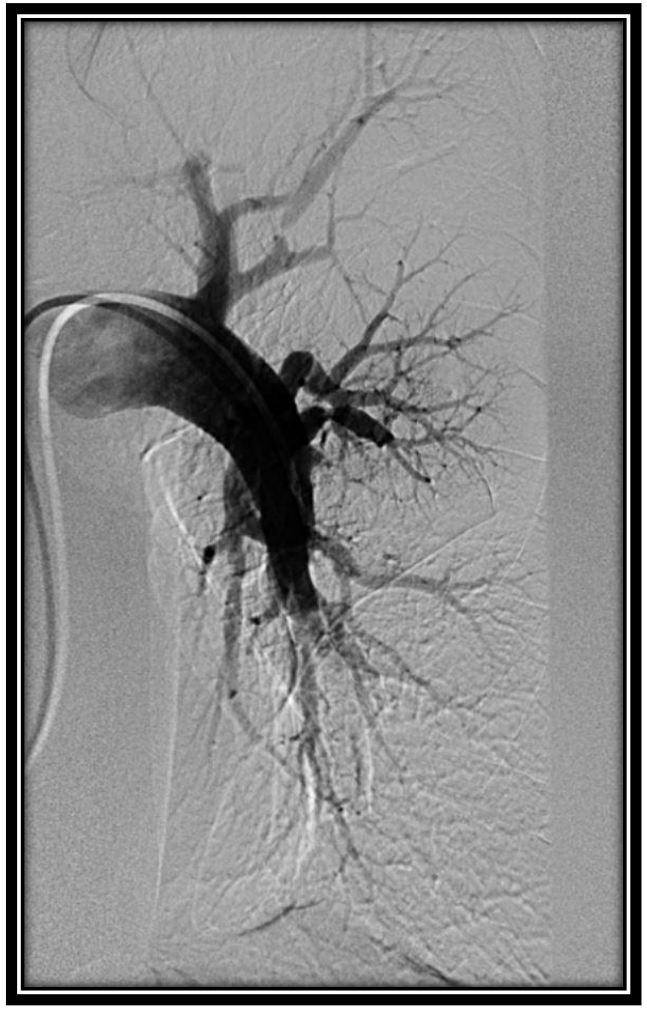


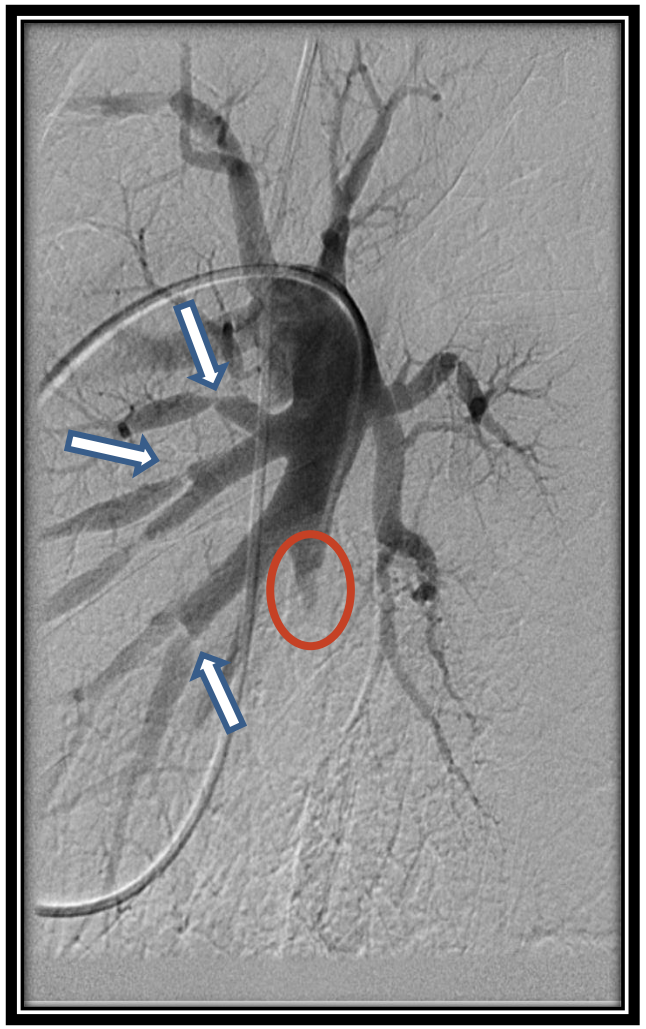
Red arrows indicating lining clot

Same Patient → Multifocal Clot More Obvious on V/Q



CTEPH Pulmonary Angiogram

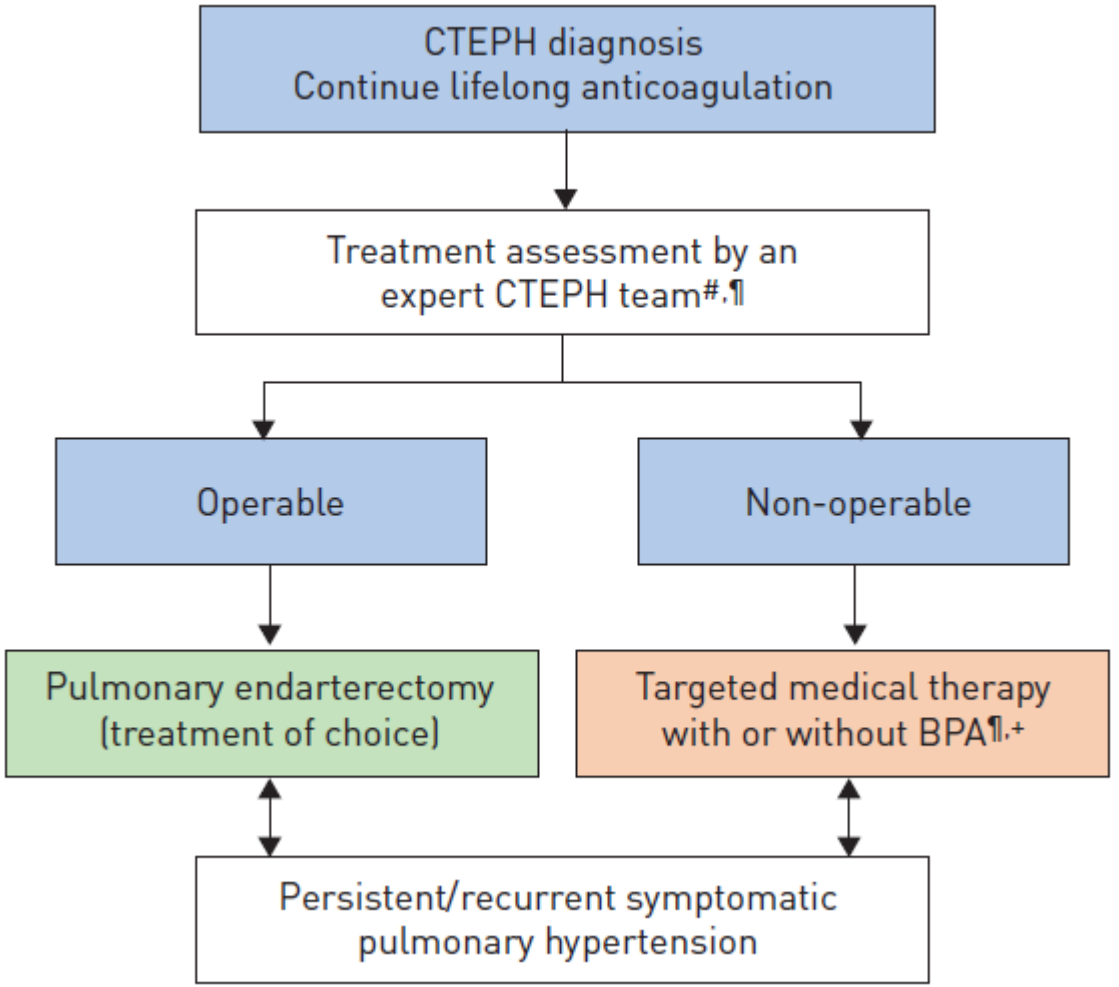




Arrows indicate “webs” or “bands”

Red circles indicate pouches

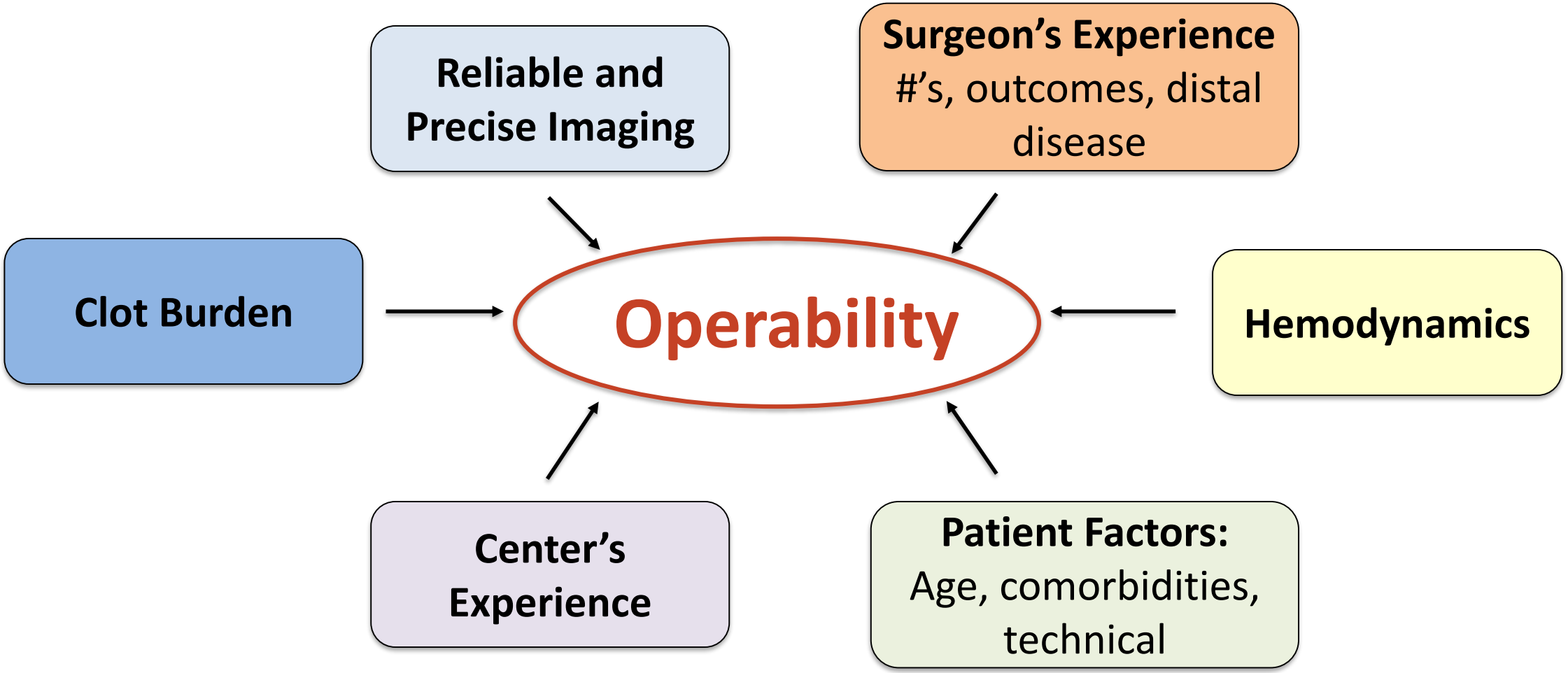
CTEPH Treatment Algorithm



- BPA: balloon pulmonary angioplasty
- #: multidisciplinary: pulmonary endarterectomy surgeon, PH expert, BPA interventionist and radiologist
- ¶: treatment assessment may differ depending on the level of expertise
- +: BPA without medical therapy can be considered in selected cases

Kim NH, et al. *Eur Resp J*. 2019;53(1):1801915.

PTE Operability Assessment



Favorable Risk-Benefit Assessment for Pulmonary Endarterectomy

Characteristics	Lower risk with predictable good long-term outcome	Higher risk with less predictable long-term outcome (not contraindications)
History	History of DVT/PE	No history of DVT/PE
Examination	No signs of right heart failure	Signs of right heart failure
Comorbidity	None	Significant concomitant lung or left heart disease
Functional limitation	Functional class II or III	Functional class IV
Imaging	Clear disease concordant on all images	Inconsistency on imaging modalities
Type of disease	Bilateral lower lobe disease	No disease appreciable in lower lobes
Haemodynamics	PVR <1000 dyn·s·cm^{-5}, in proportion to site and number of obstructions on imaging; higher PA pulse pressure	PVR >1200 dyn·s·cm^{-5}, out of proportion to site and number of obstructions on imaging; higher PA diastolic pressure

DVT: deep vein thrombosis; PE: pulmonary embolism; PVR: pulmonary vascular resistance; PA: pulmonary artery.

Kim NH, et al. *Eur Resp J*. 2019;53(1):1801915.

In Summary

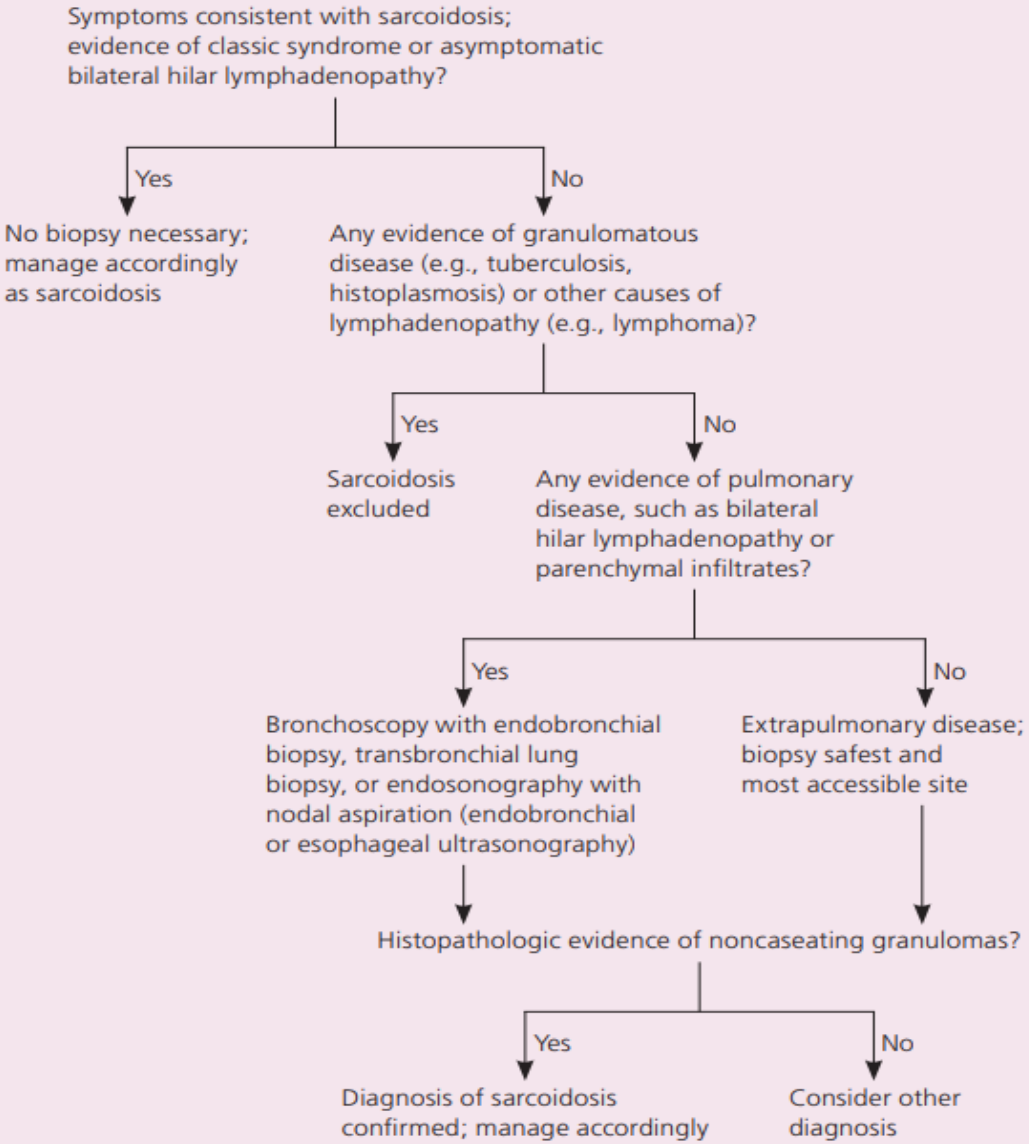
- V/Q scanning is the screening test of choice at most centers for CTEPH .
- Confirmatory imaging should be done on patients with unmatched perfusion defects on V/Q.
- If you are not sure about the imaging, ask for help.
- Patients with CTEPH should be evaluated for operability

Sarcoidosis Epidemiology

- Affects people of all racial and ethnic groups
- > 80% of cases occur in adults 20-50 years of age
- Children rarely affected
- 4-10% of patients have a first degree relative with sarcoidosis

Iannuzzi MC, et al. *NEJM*. 2007;357:2153-2165.
Soto-Gomez N, et al. *Am Fam Physician*. 2016;93:840-848.

Diagnosis of Sarcoidosis



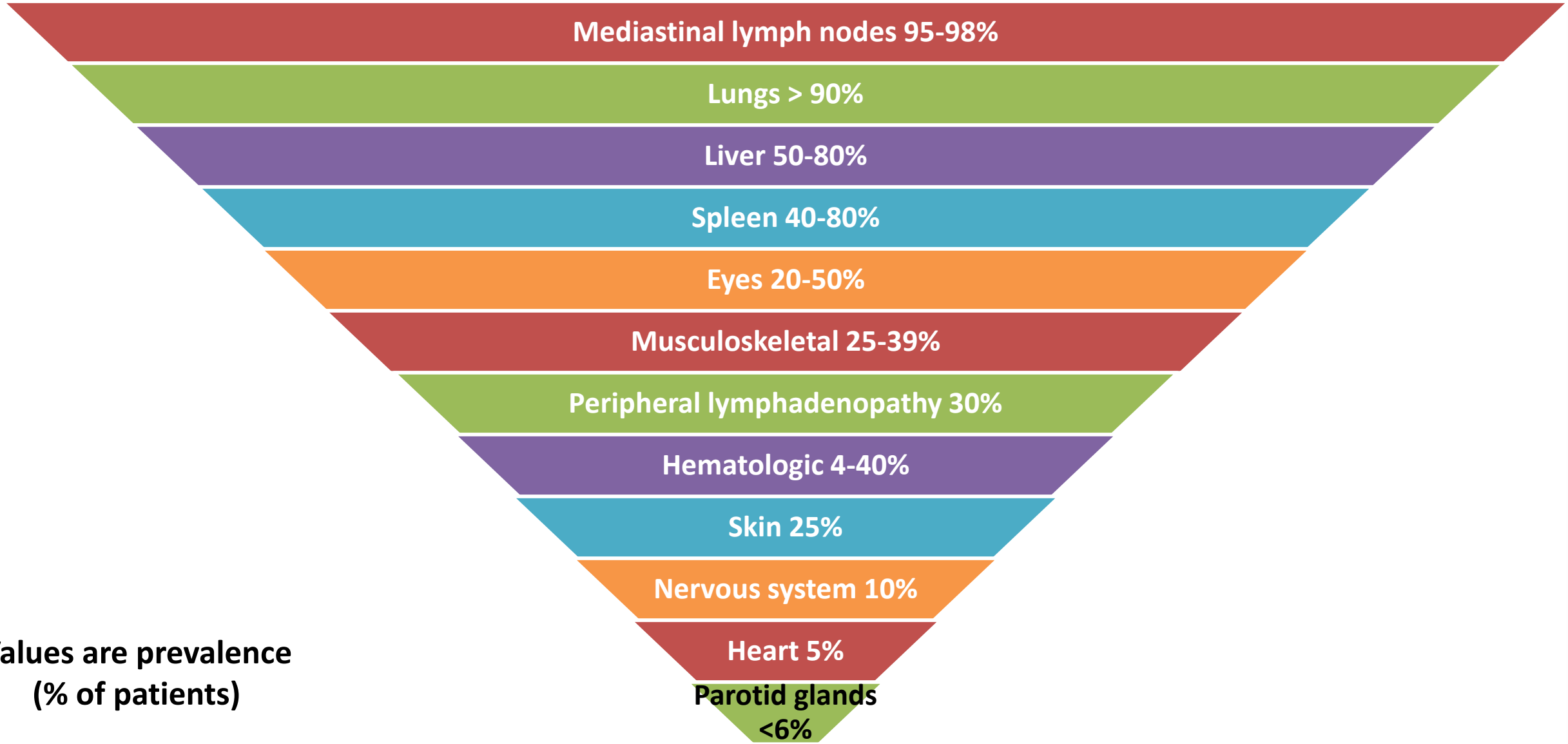
Soto-Gomez N, et al. *Am Fam Physician*. 2016;93:840-848.

Role for Different Types of Imaging

Study	Findings
Chest CT	Useful for differential diagnosis of diffuse interstitial changes in lung parenchyma and pulmonary fibrosis
CXR	Bilateral hilar lymphadenopathy and interstitial changes, necessary for staging
^{18}F-fluorodeoxy-glucose PET	Useful for finding areas to biopsy; May aid in the diagnosis of cardiac sarcoidosis May correlate with active inflammation and disease activity
MRI	**CNS**: useful for identification of lesions **Cardiac MRI**: Findings include focal intramyocardial zones of increased signal intensity due to edema and inflammation Delayed gadolinium enhancement is a predictor of ventricular arrhythmias and poor outcomes

Soto-Gomez N, et al. *Am Fam Physician*. 2016;93:840-848.

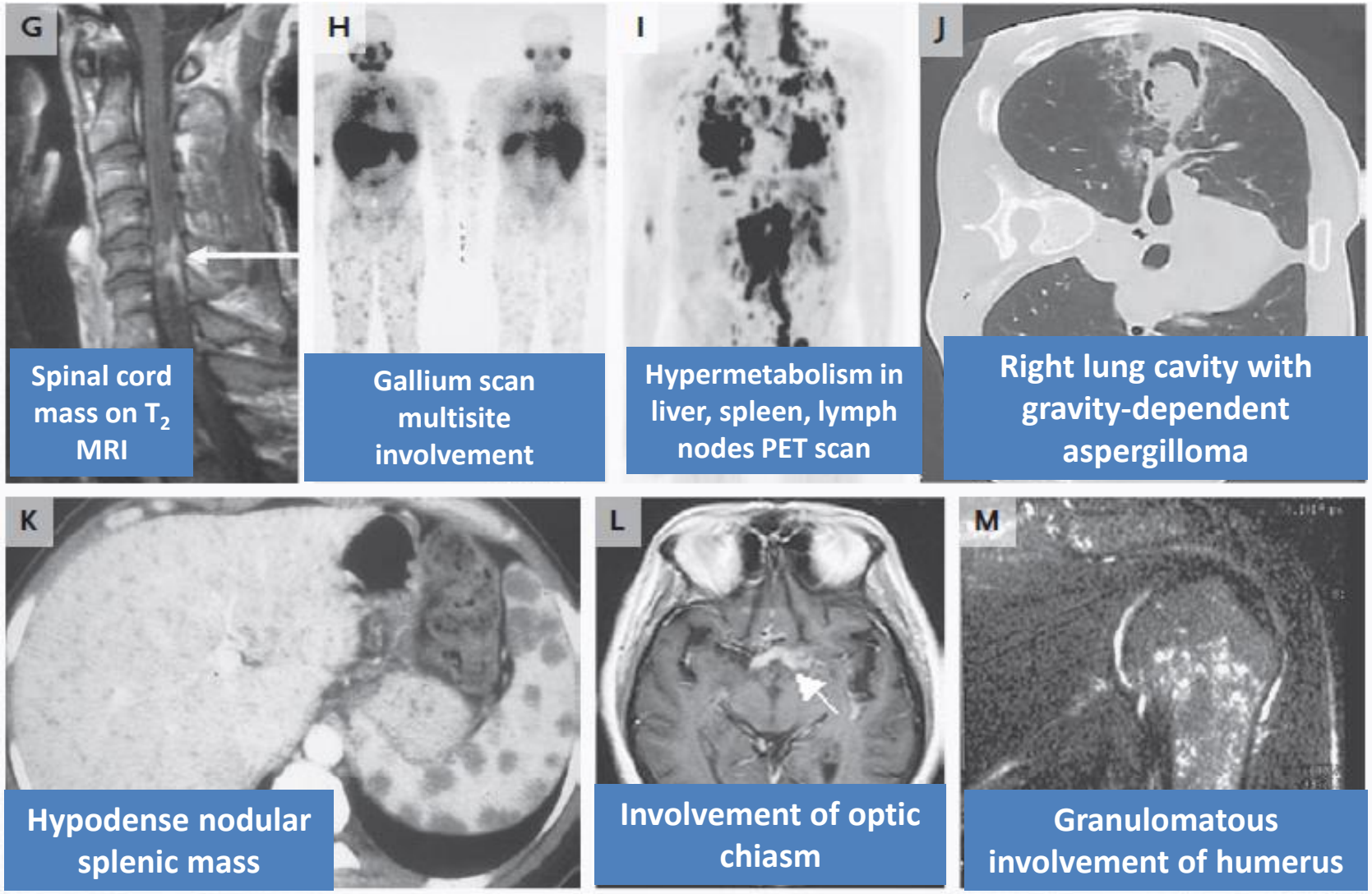
Organ Involvement in Sarcoidosis



Values are prevalence (% of patients)

Soto-Gomez N, et al. *Am Fam Physician*. 2016;93:840-848.

Clinical Features of Sarcoidosis



Pulmonary Involvement
Clinical Manifestations

- Cough, dyspnea
- Hilar and mediastinal lymphadenopathy
- Pulmonary hypertension
- Interstitial lung disease and pulmonary fibrosis

Sarcoid: Lymphadenopathy (hilar and mediastinal)

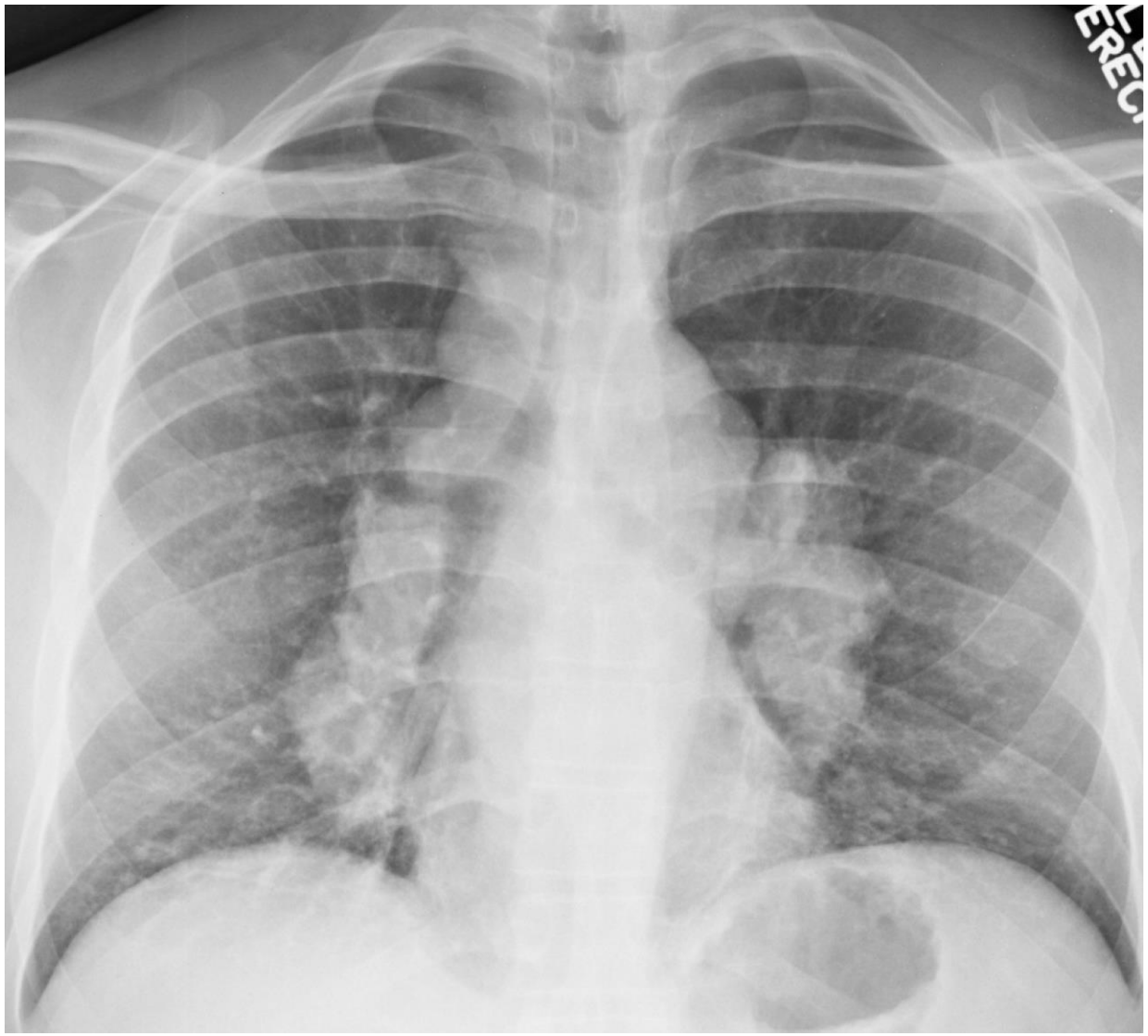


Image courtesy of L. Heyneman, MD

Sarcoid: Lymphadenopathy and Parenchyma

Subtle upper lobe nodules

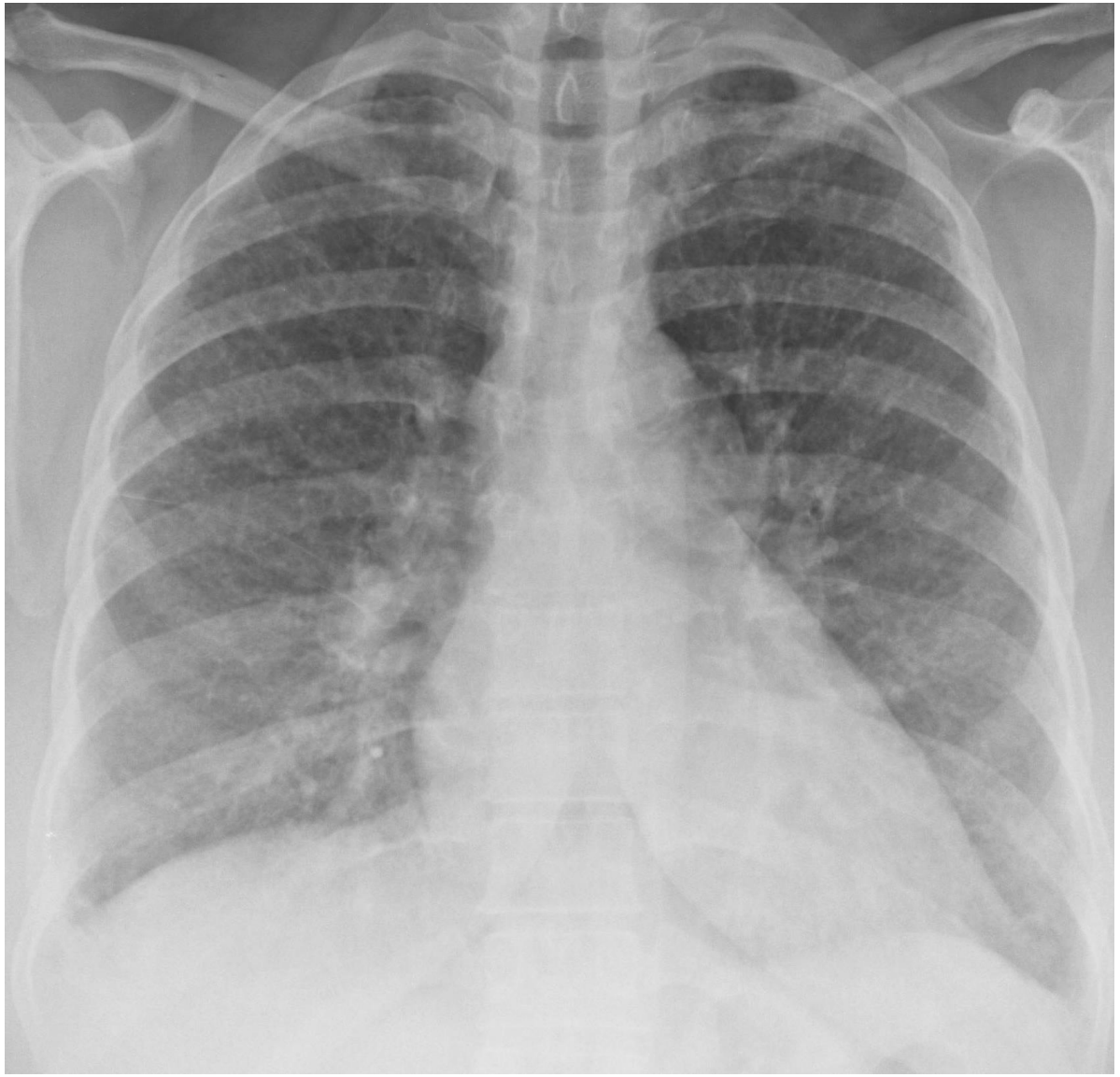
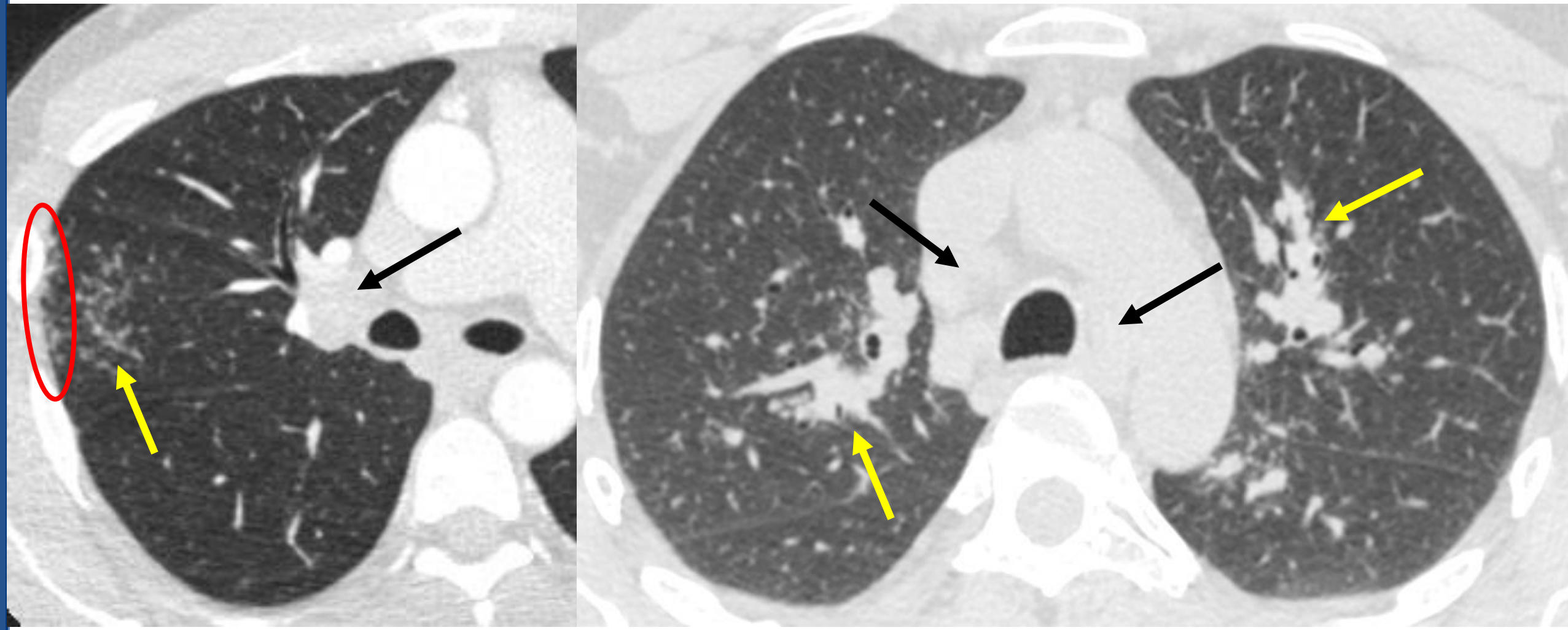


Image courtesy of L. Heyneman, MD

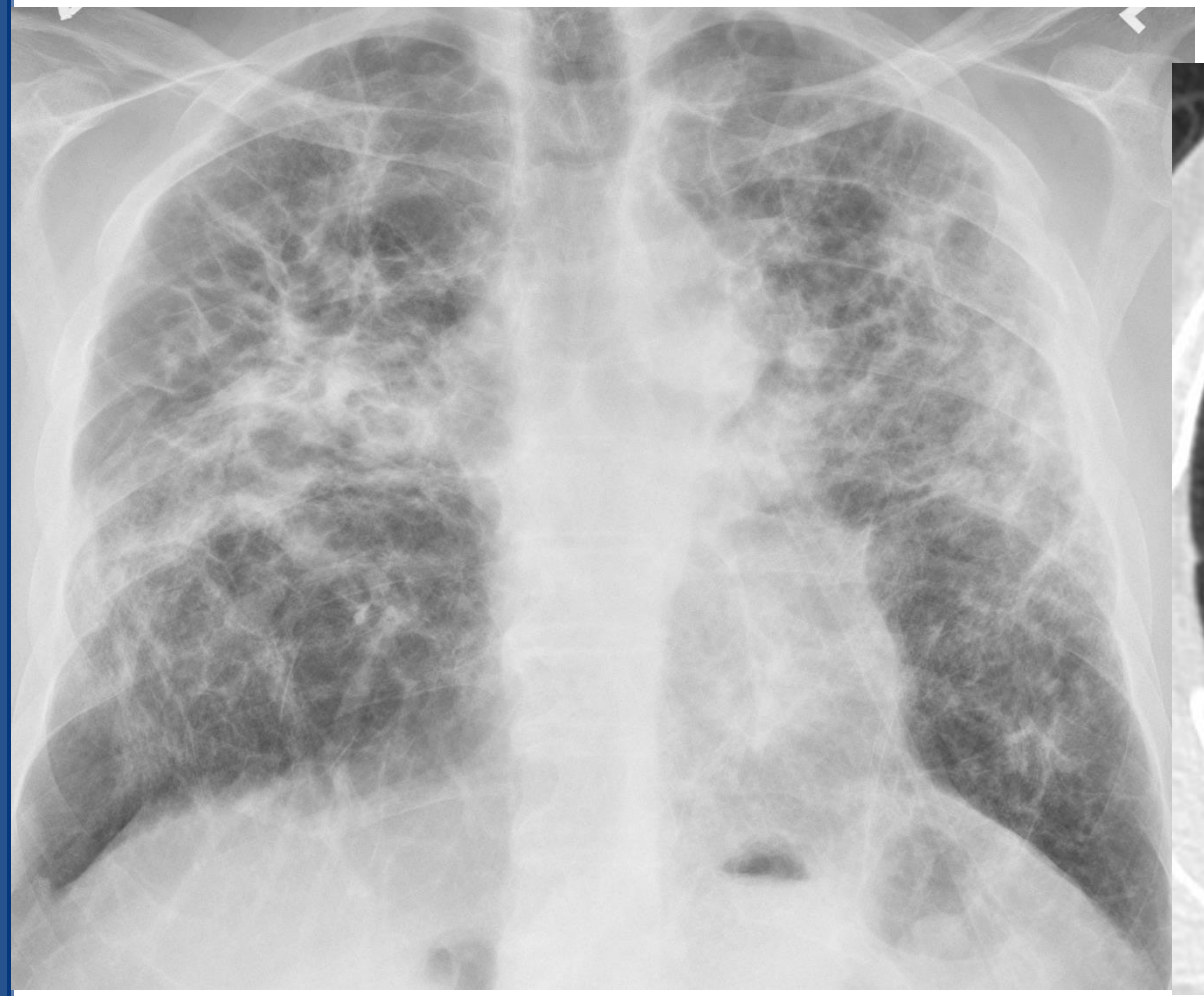
Lymphadenopathy (↑) and Pulmonary Parenchyma

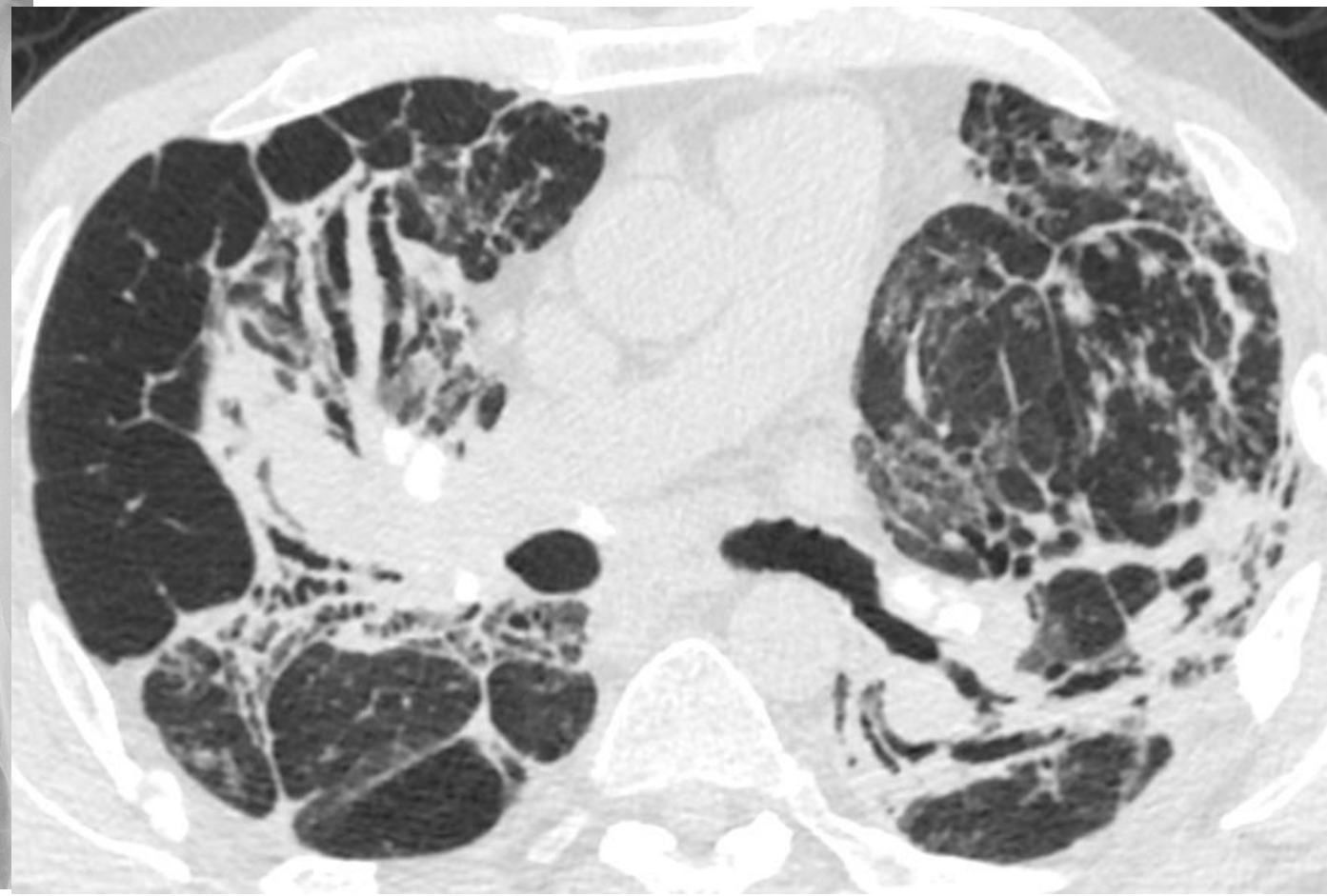
Peribronchovascular (↓) + subpleural (○) nodularity



Images courtesy of L. Heyneman, MD

Sarcoid: End-Stage Fibrosis





Images courtesy of L. Heyneman, MD

Summary

- Sarcoidosis is a systemic inflammatory disease with a predilection for the respiratory system.
- Diagnosis relies on 3 criteria: compatible clinical and radiologic presentation; pathologic evidence of noncaseating granulomas; exclusion of other diseases
- Up to 20% develop fibrotic lung disease (granulomatous inflammation evolves to pulmonary fibrosis).
  - Morbidity and mortality are increased for these patients.
- Immunosuppressive therapy may be beneficial in patients with active inflammation.