AT THE FOREFRONT
UChicago Medicine

Imaging cystic lung diseases: Diagnostic algorithm and imaging findings

Jonathan H. Chung, MD
Professor of Radiology
Chief Quality Officer, Radiology
Section Chief Thoracic Imaging

@jonherochung

Disclosures

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Goals/Objectives

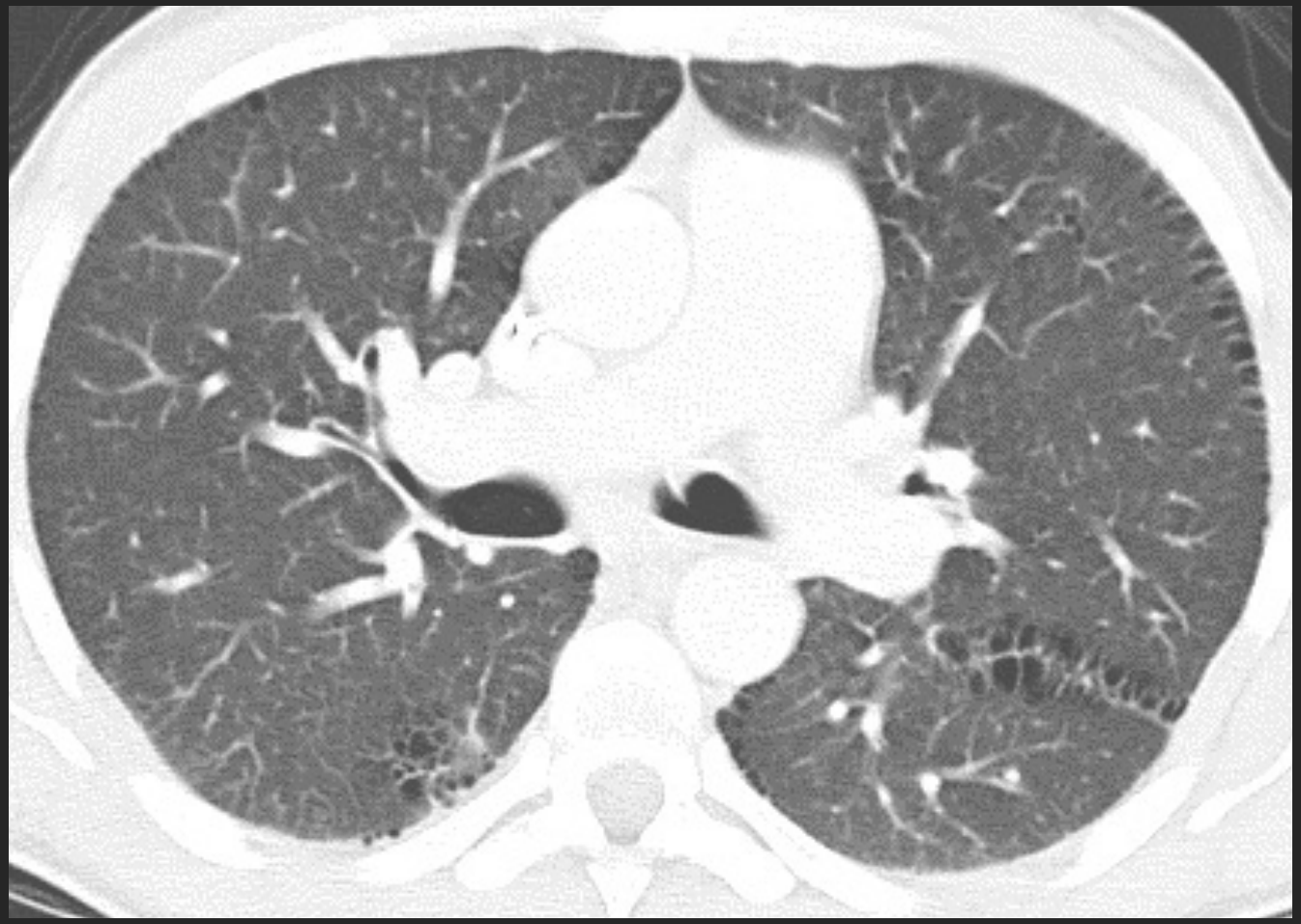
Be able to:

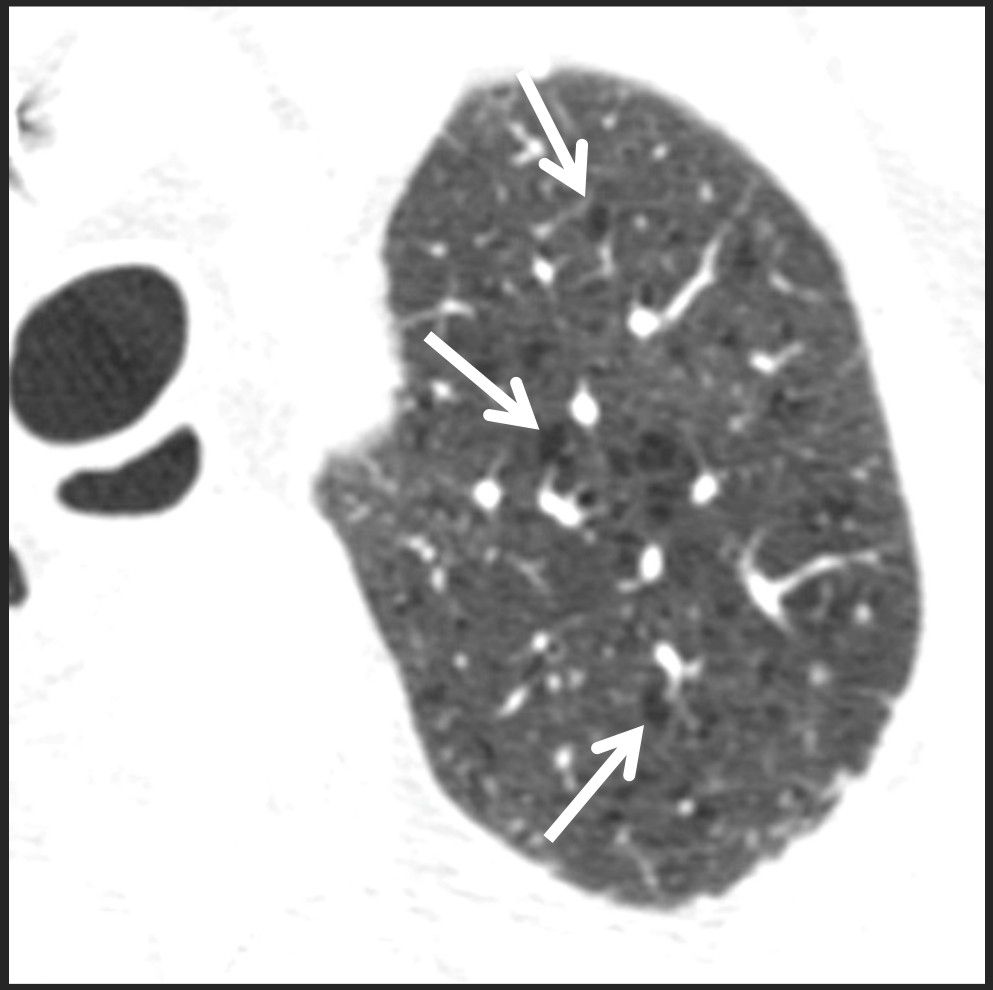
- List common causes of cystic lung disease
- Identify the presence of cystic lung disease and a general diagnostic approach
- Discuss specific findings more suggestive of BHD as opposed to LIP

Diffuse Cystic Lung Disease: DDx

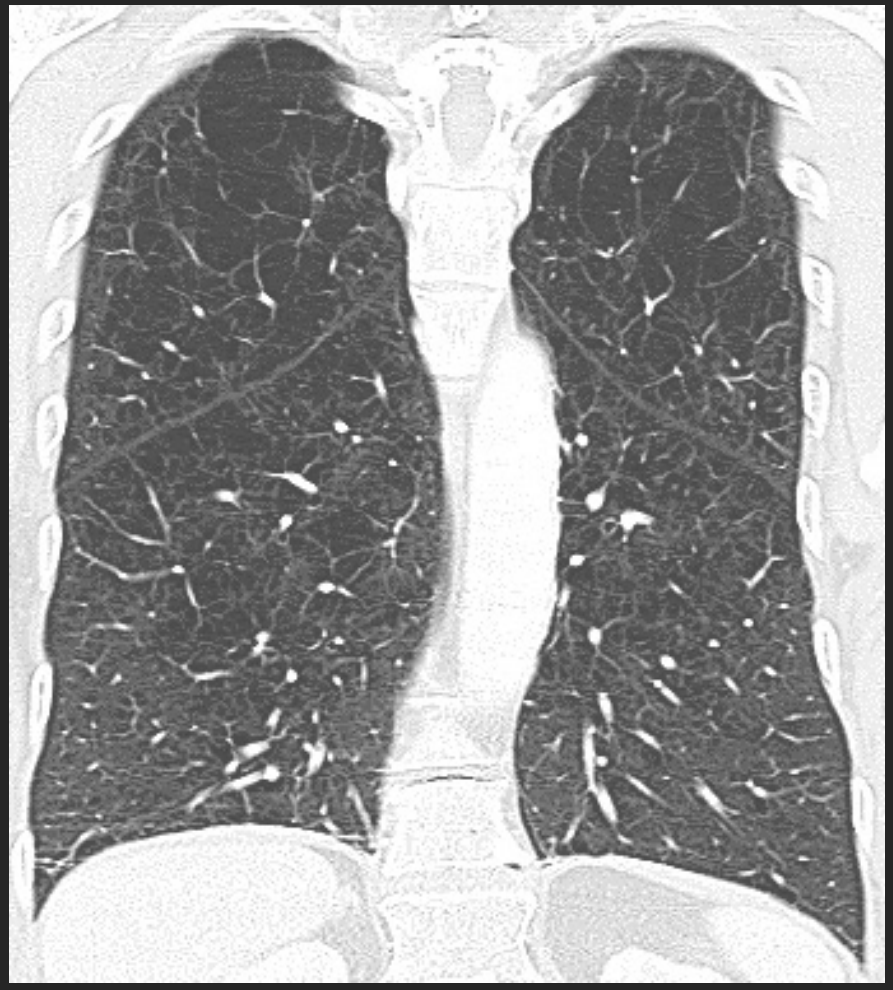
- Langerhans cell histiocytosis
- Lymphangioleiomyomatosis
- Lymphocytic interstitial pneumonitis
- Birt-Hogg-Dube syndrome (rare)
- Light chain deposition disease
- Desquamative interstitial pneumonitis (rare)
- Emphysema (mimic)
- Pneumatoceles (mimic)

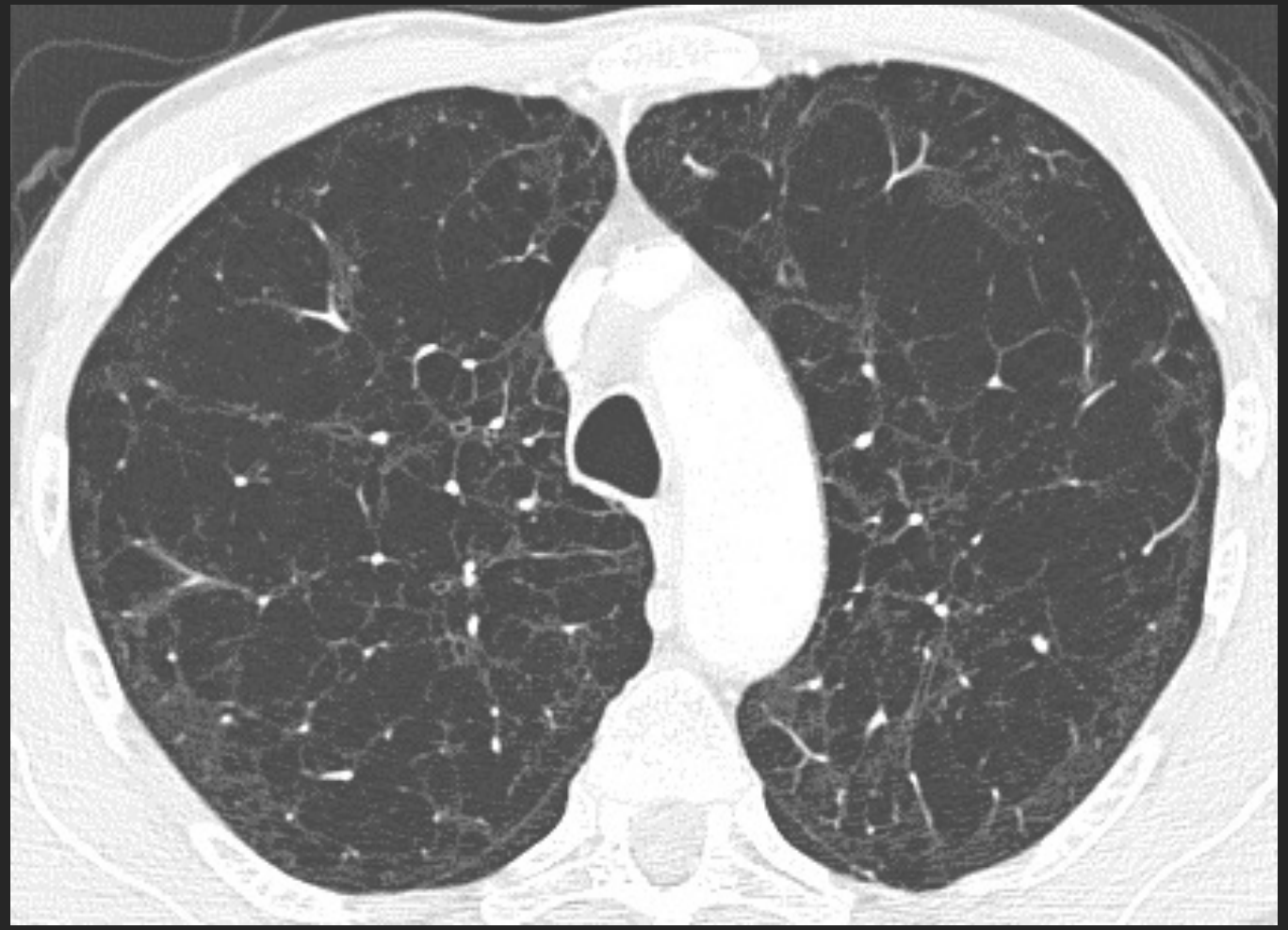
Emphysema





Emphysema

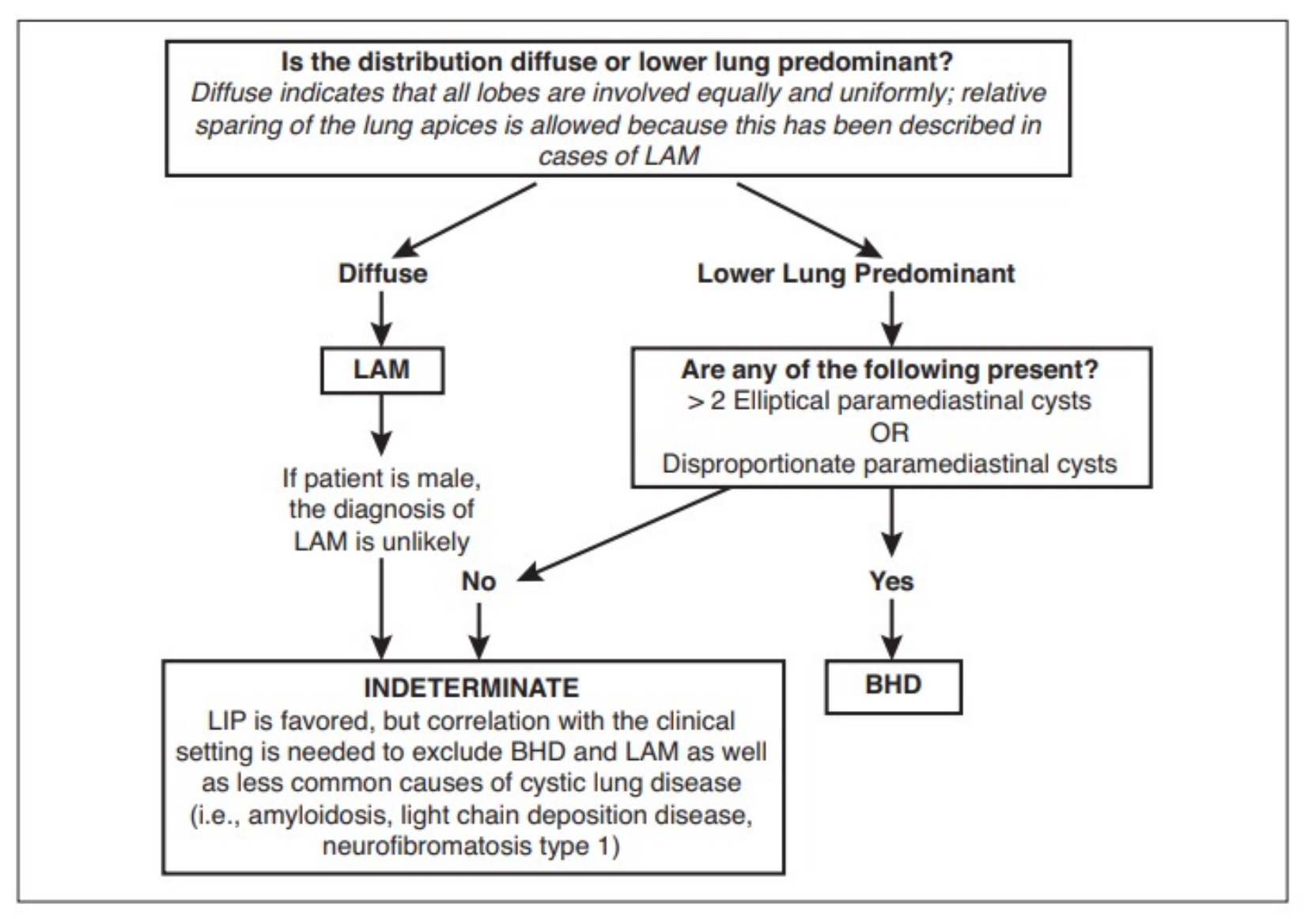




Diffuse Cysts: Approach

- Make sure it's not CLO and/or PS emphysema and that there are "enough" cysts to exclude post-infectious or traumatic pneumatoceles
  - DDX: LCH vs LAM, then consider LIP and BHD
  - LCH usually very characteristic

Diffuse Cysts Approach: LAM vs LIP vs BHD



Escalon JG,..., Lynch DA. AJR Am J Roentgenol. 2019 Mar 19;1-5.

TABLE 4: Diagnostic Accuracy of Readers Using Cystic Lung Disease Algorithm

Accuracy	Patients With BHD Syndrome (*n* = 16)		Patients With LIP (*n* = 14)		Patients With LAM (*n* = 17)	
	Reader 1	Reader 2	Reader 1	Reader 2	Reader 1	Reader 2
Correct diagnosis, no. (%) of patients	12 (75)	13 (81)	9 (64)	12 (86)	16 (94)	15 (88)
Incorrect diagnosis, no. of patients						
Indeterminate	4	3	—	—	0	2
BHD syndrome			2	2	1	
LAM	0	0	3	0	—	—

Note—The interreader agreement for diagnosis had a kappa value of 0.809 (95% CI, 0.668–0.950). BHD = Birt-Hogg-Dubé, LIP = lymphocytic interstitial pneumonia, LAM = lymphangioleiomyomatosis. Dash [—] incidates correct diagnosis.

Escalon JG,..., Lynch DA. AJR Am J Roentgenol. 2019 Mar 19;1-5.

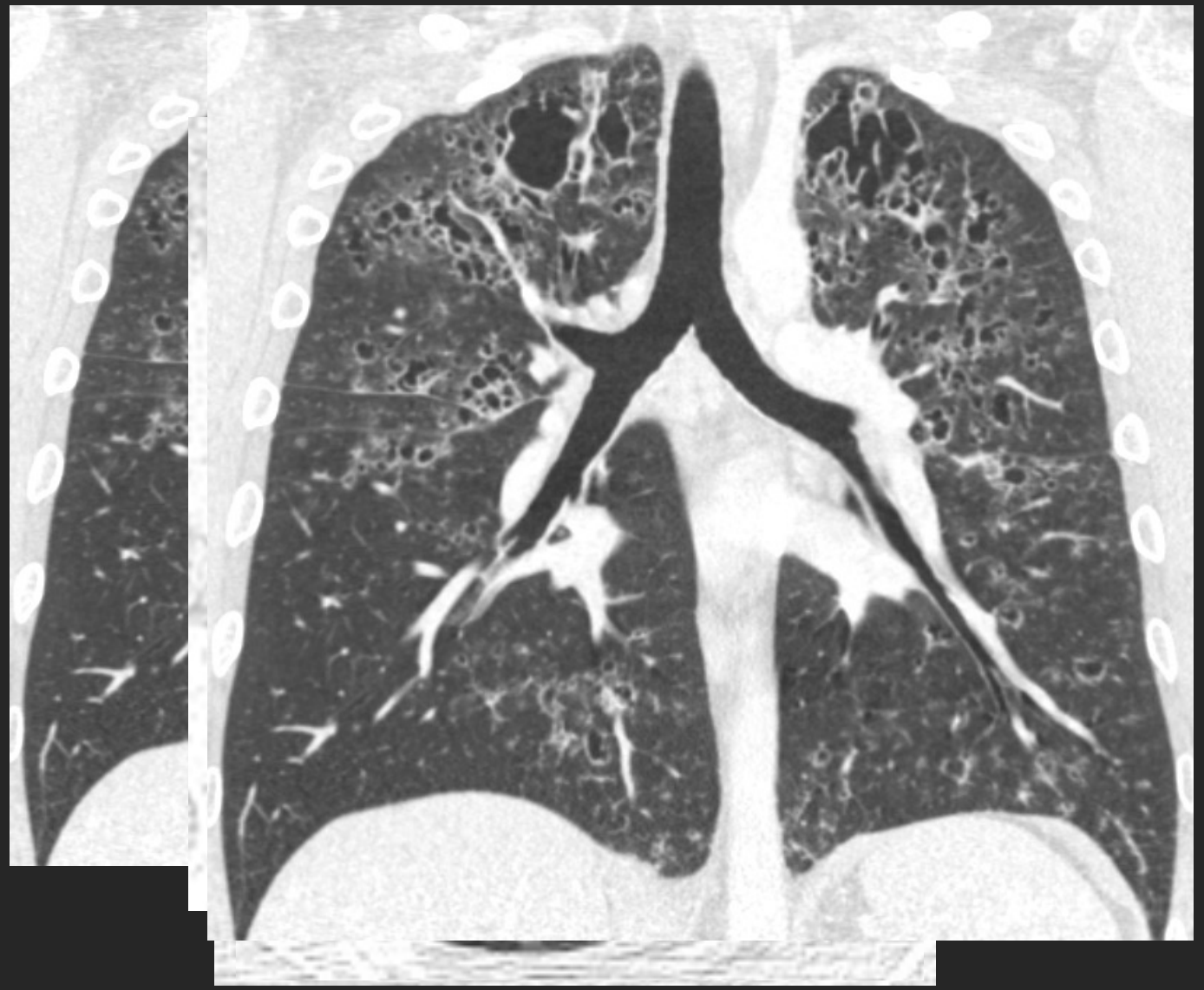
Pulmonary Langerhans Cell Histiocytosis (PLCH)

Chest CT

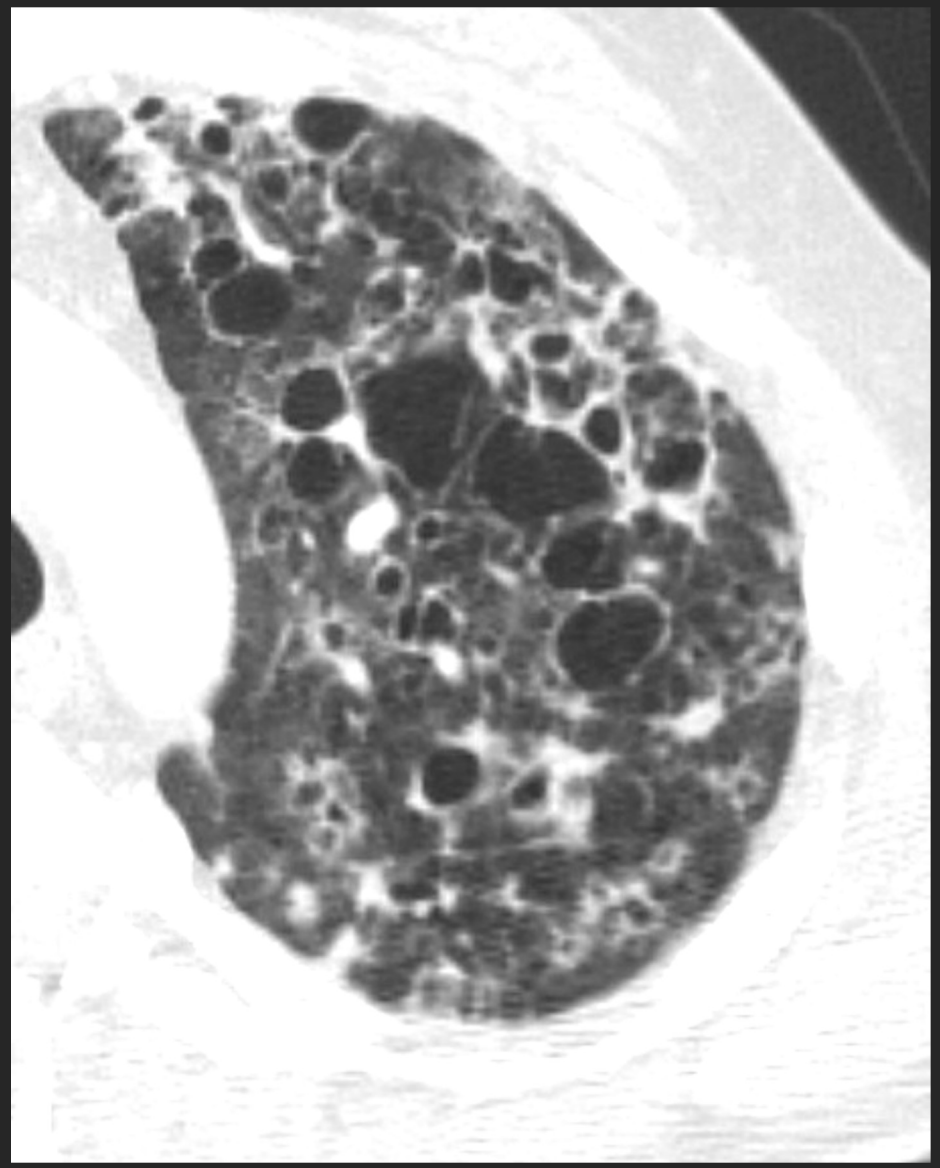
- Cysts: thin- or thick-walled; combination of round and bizarre shaped cysts
- Nodules: poorly- or well-defined; some may show cavitation
- Nodules predominate in early disease and cysts in advanced disease
- Upper lobe zone distribution with sparing of the costophrenic sulci
- Cysts may decrease in size over time

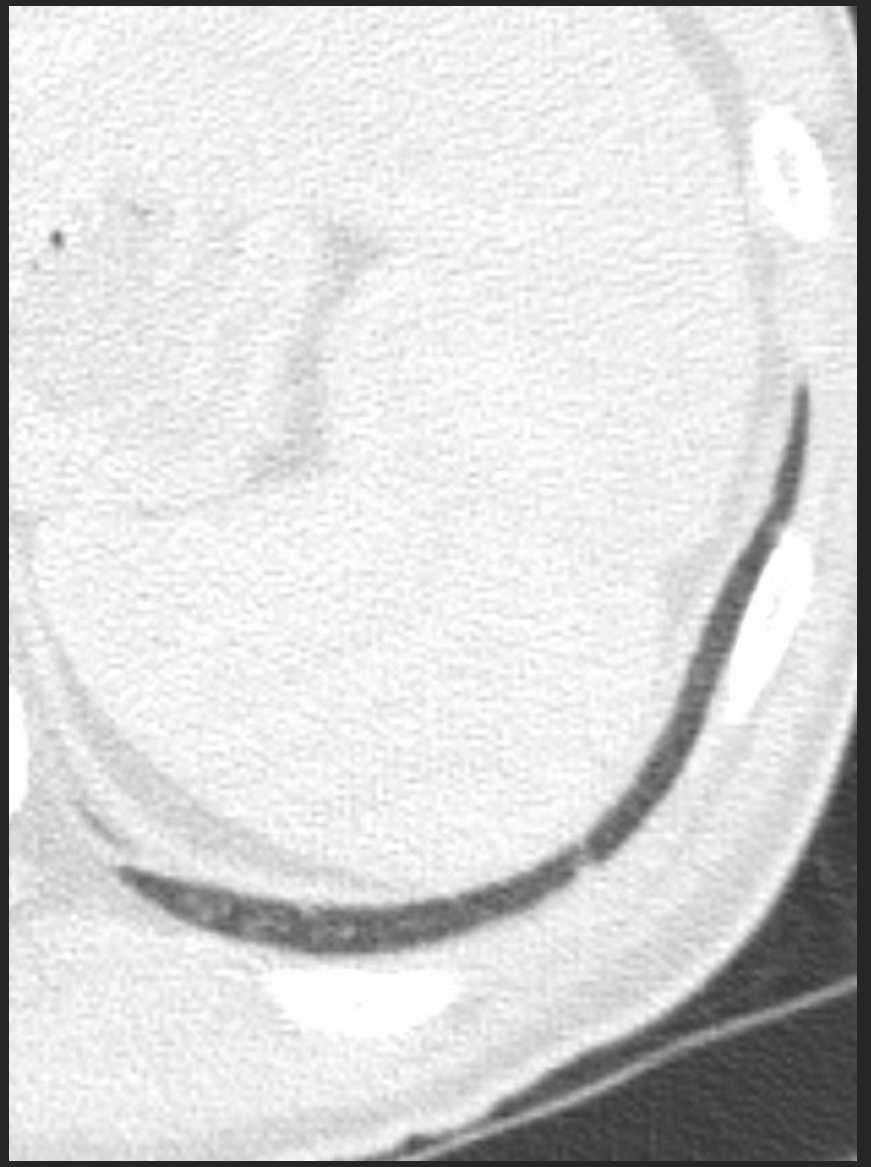
Other Imaging Findings

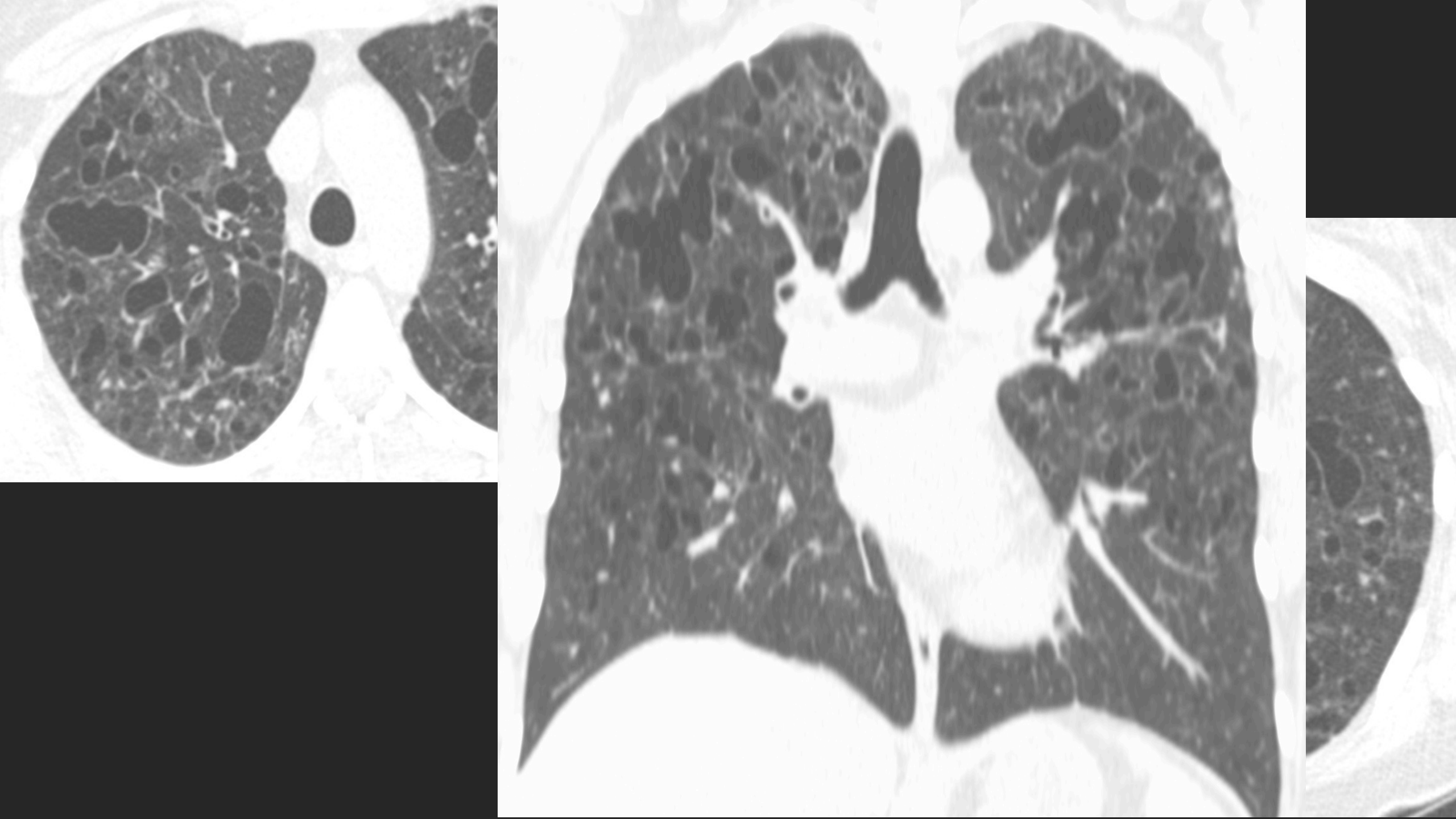
- Pneumothorax



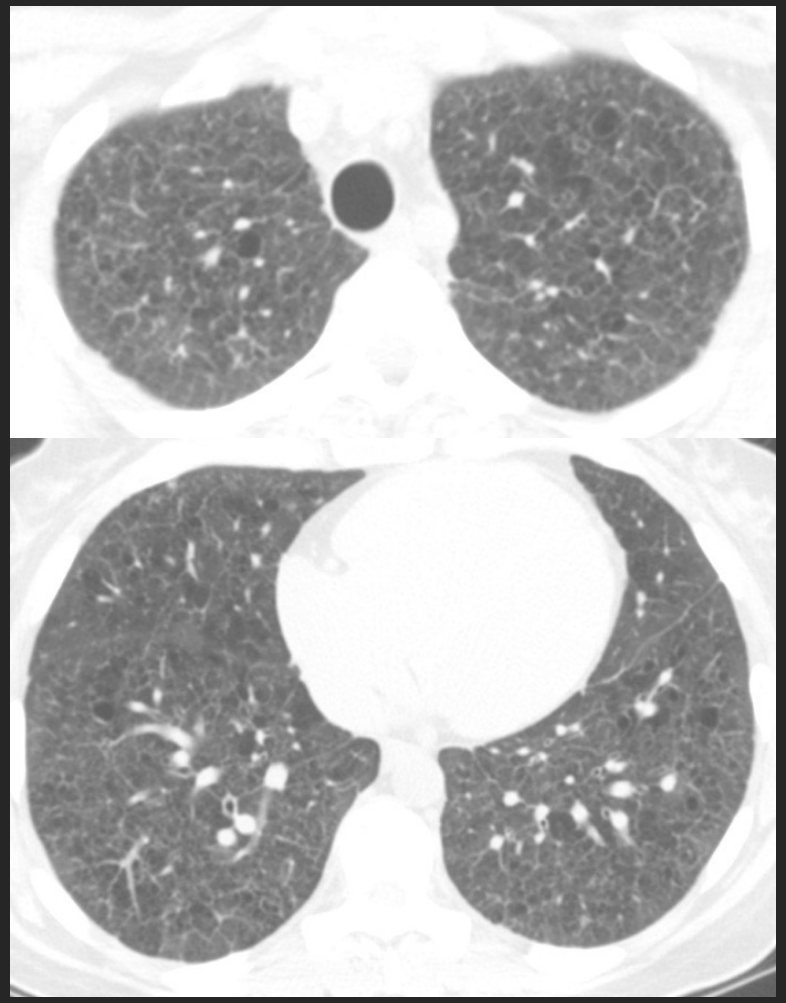
Langerhans cell histiocytosis

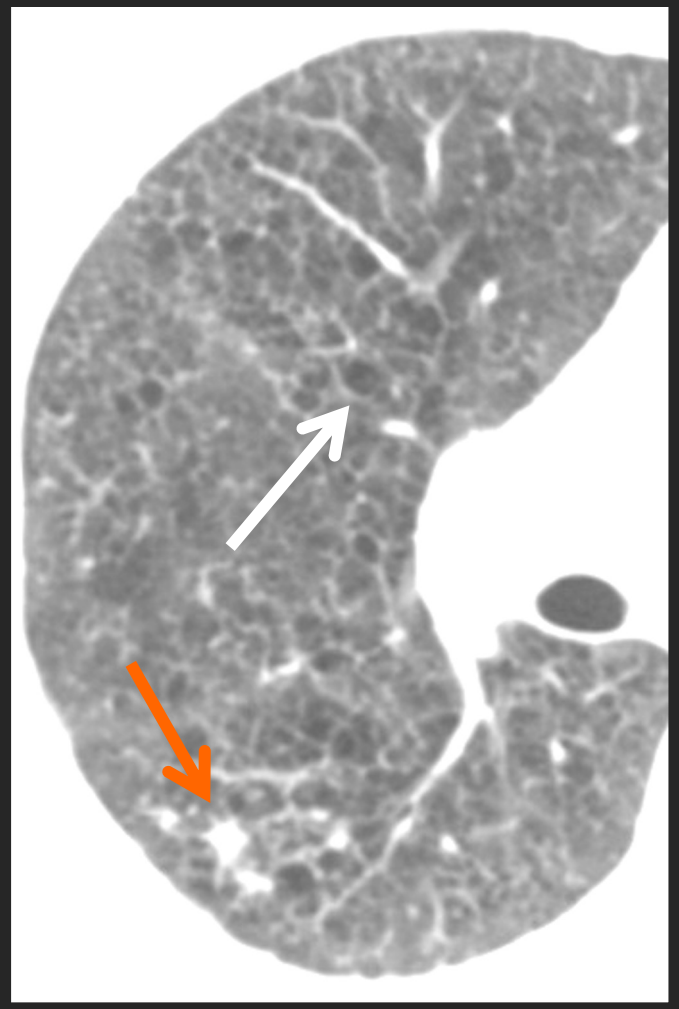






Langerhans cell histiocytosis

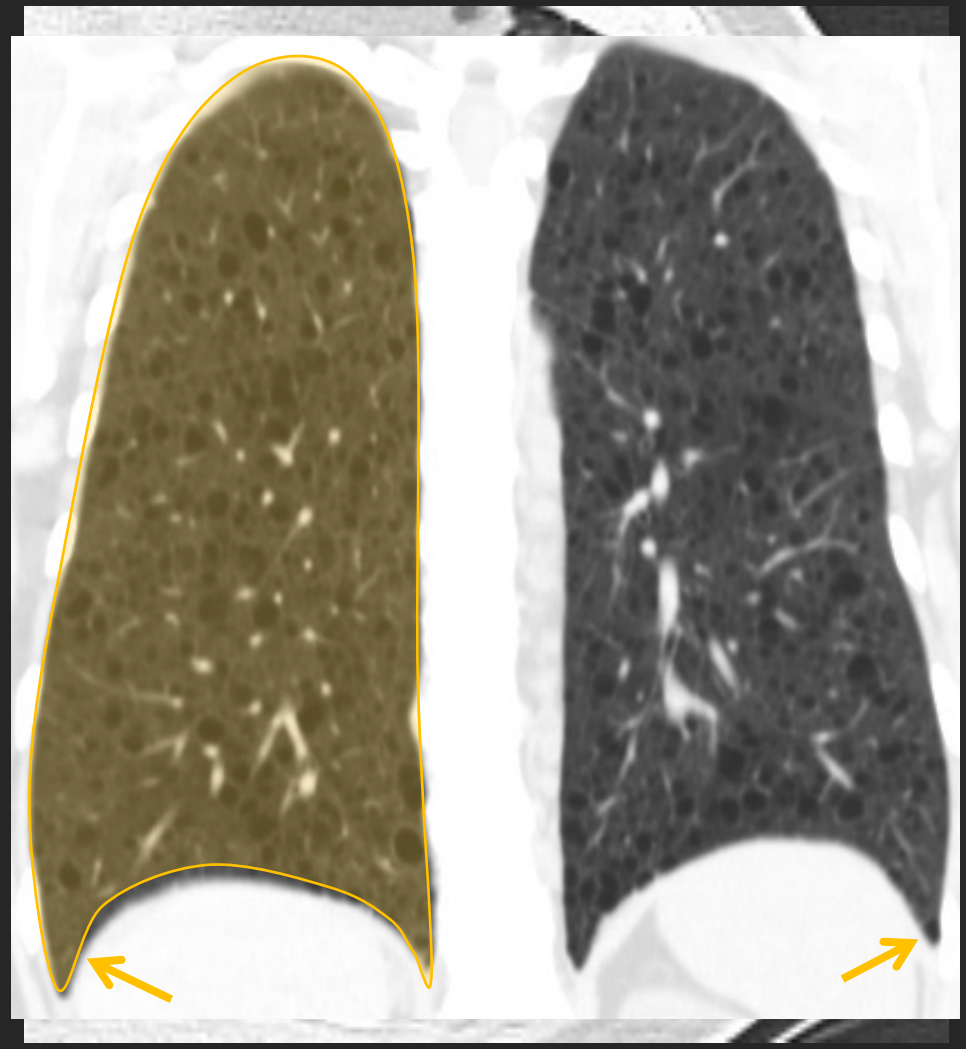




Lymphangioleiomyomatosis (LAM)

Chest CT

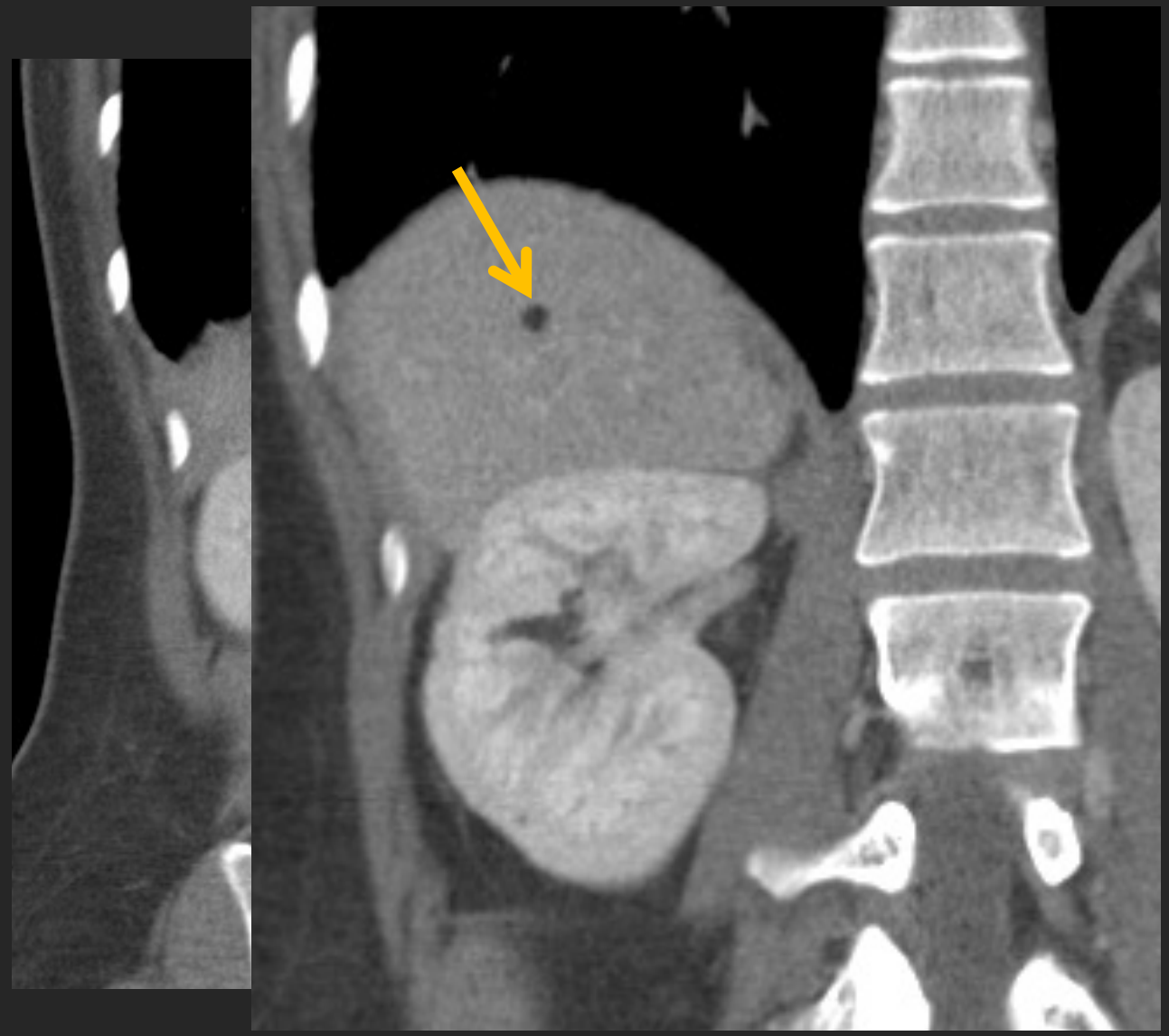
- **Cysts**:
  - Thin-walled (1 – 2 mm)
  - Round or oval shaped generally uniform in size (5 – 15 mm)
  - Diffuse distribution
  - Involve costophrenic sulci (in contrast to LCH)
  - Normal appearing intervening lung



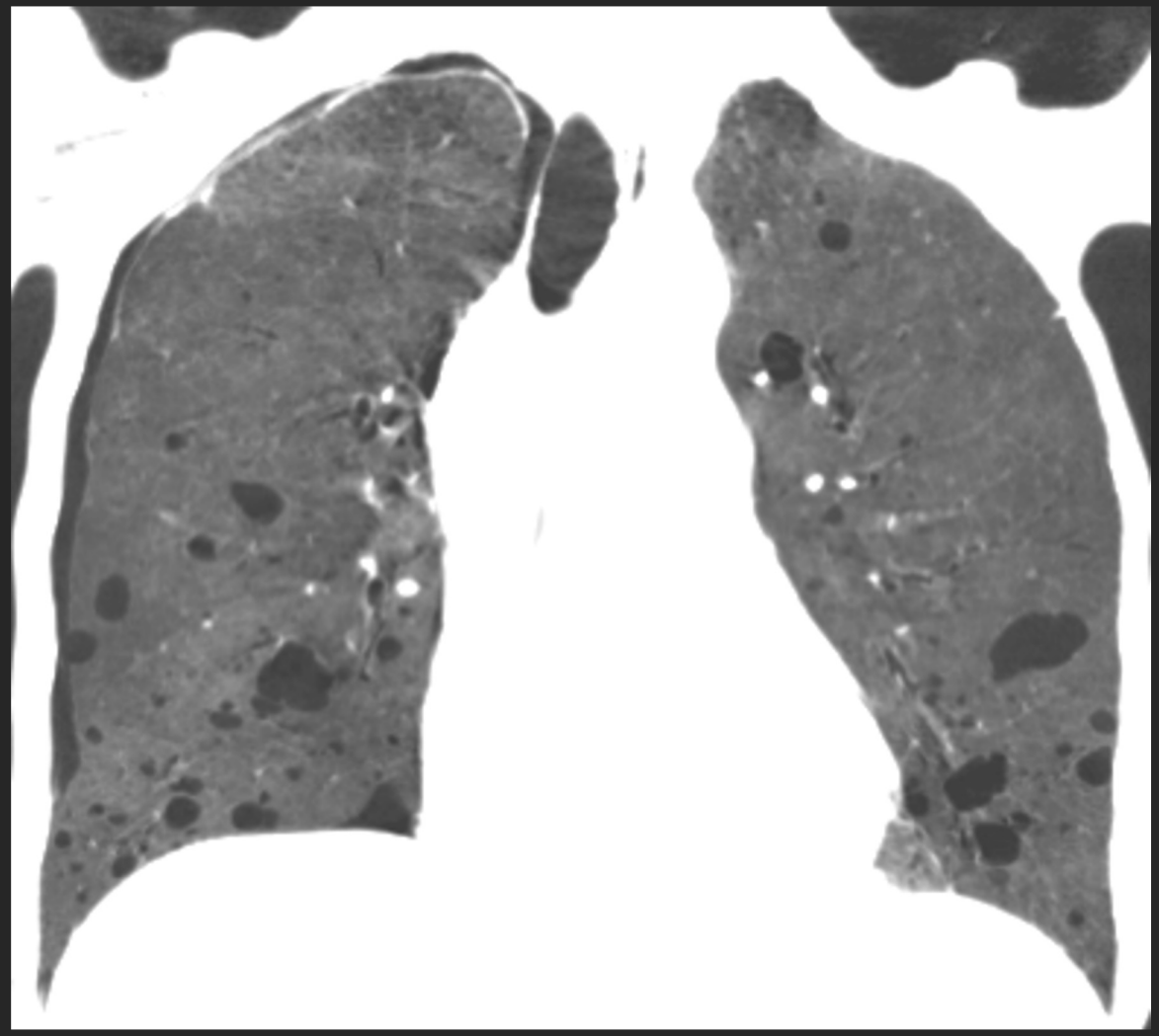
Lymphangioleiomyomatosis (LAM)

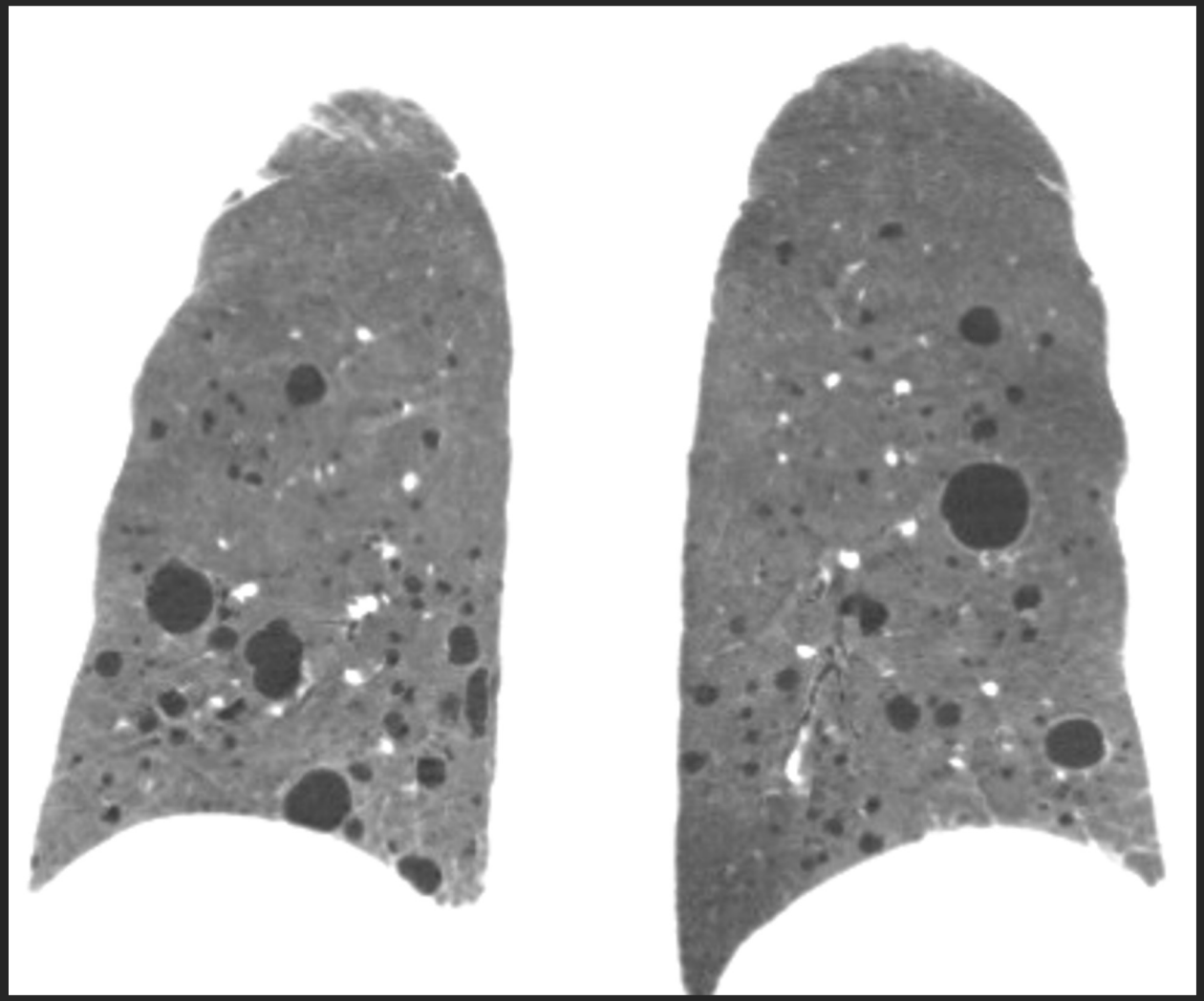
Extrathoracic Findings

- Renal angiomyolipomas
- Hepatic angiomyolipomas
- Lymphangiomyomas
- Retroperitoneal lymphadenopathy
- Chylous ascites

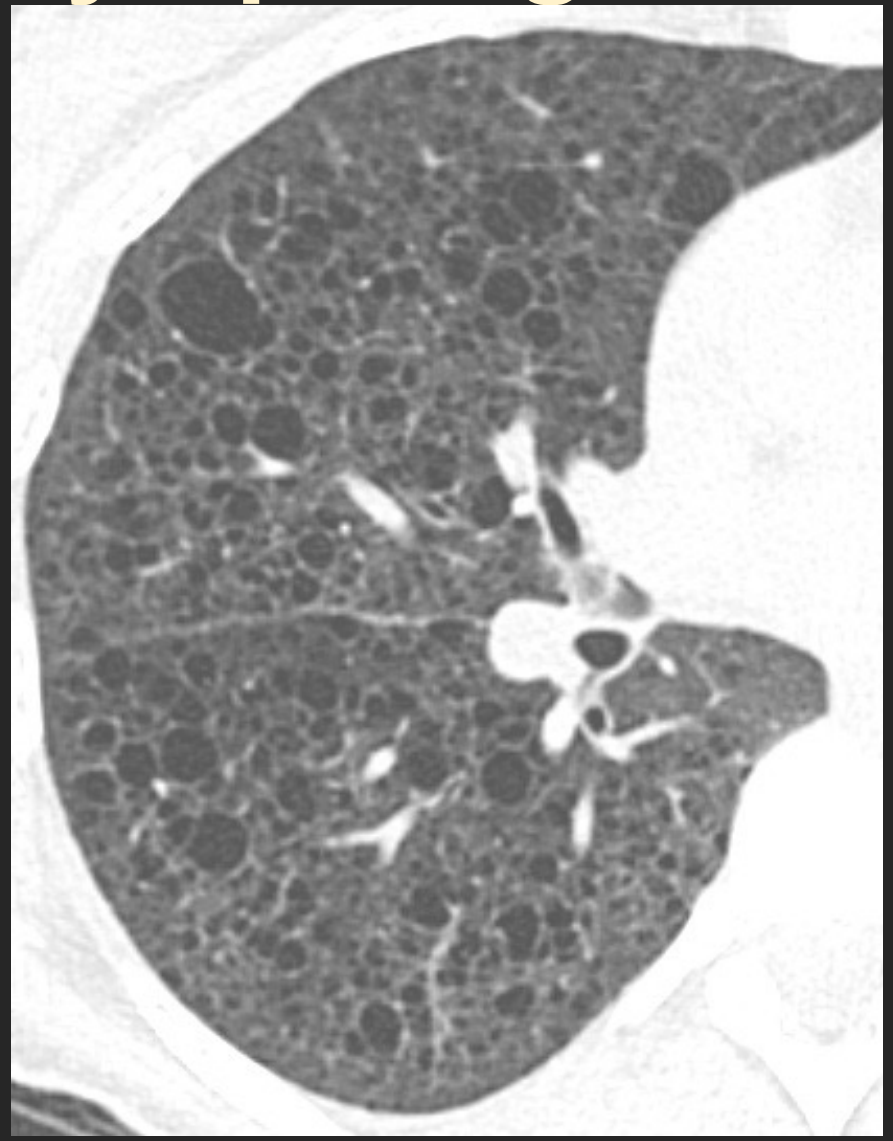


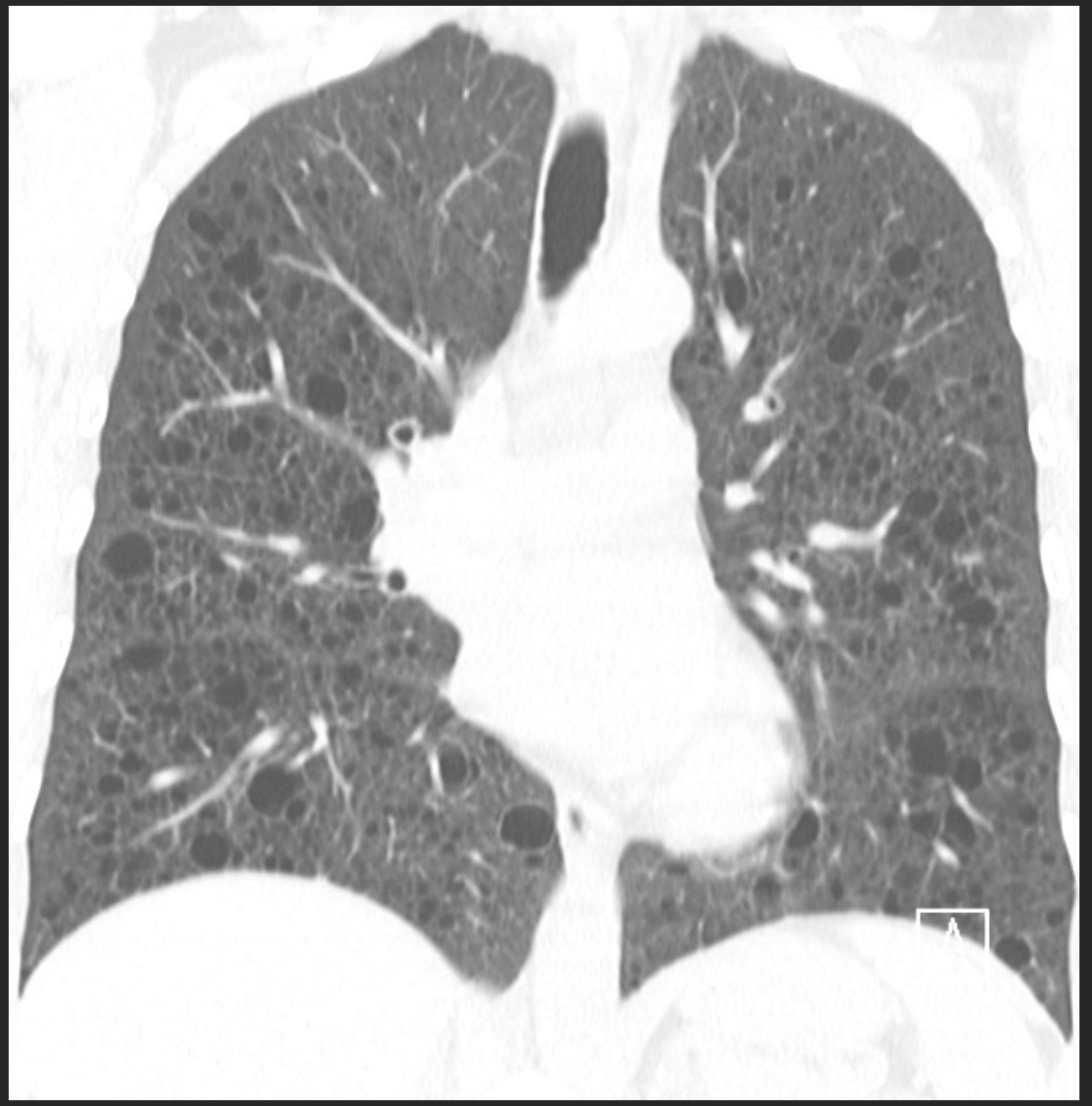
Lymphangioleiomyomatosis





Lymphangioleiomyomatosis





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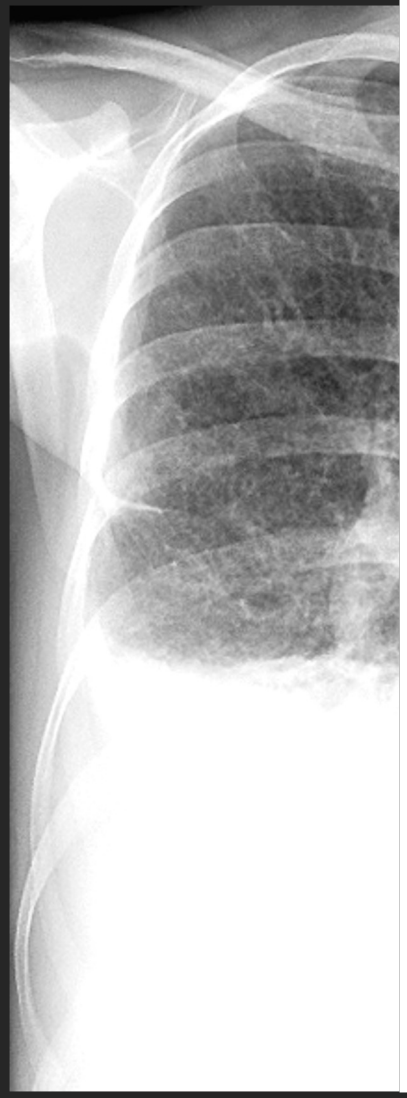
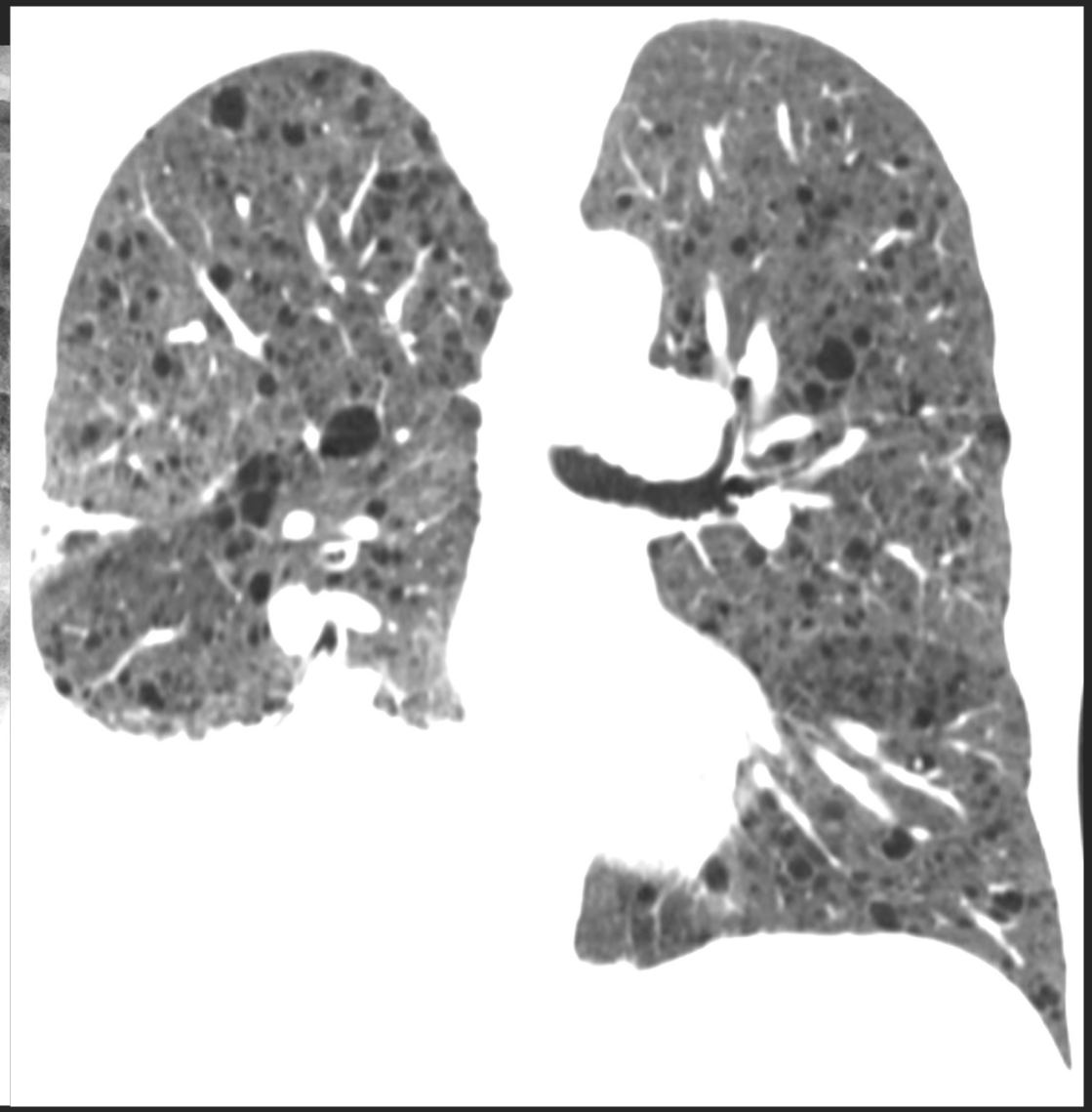
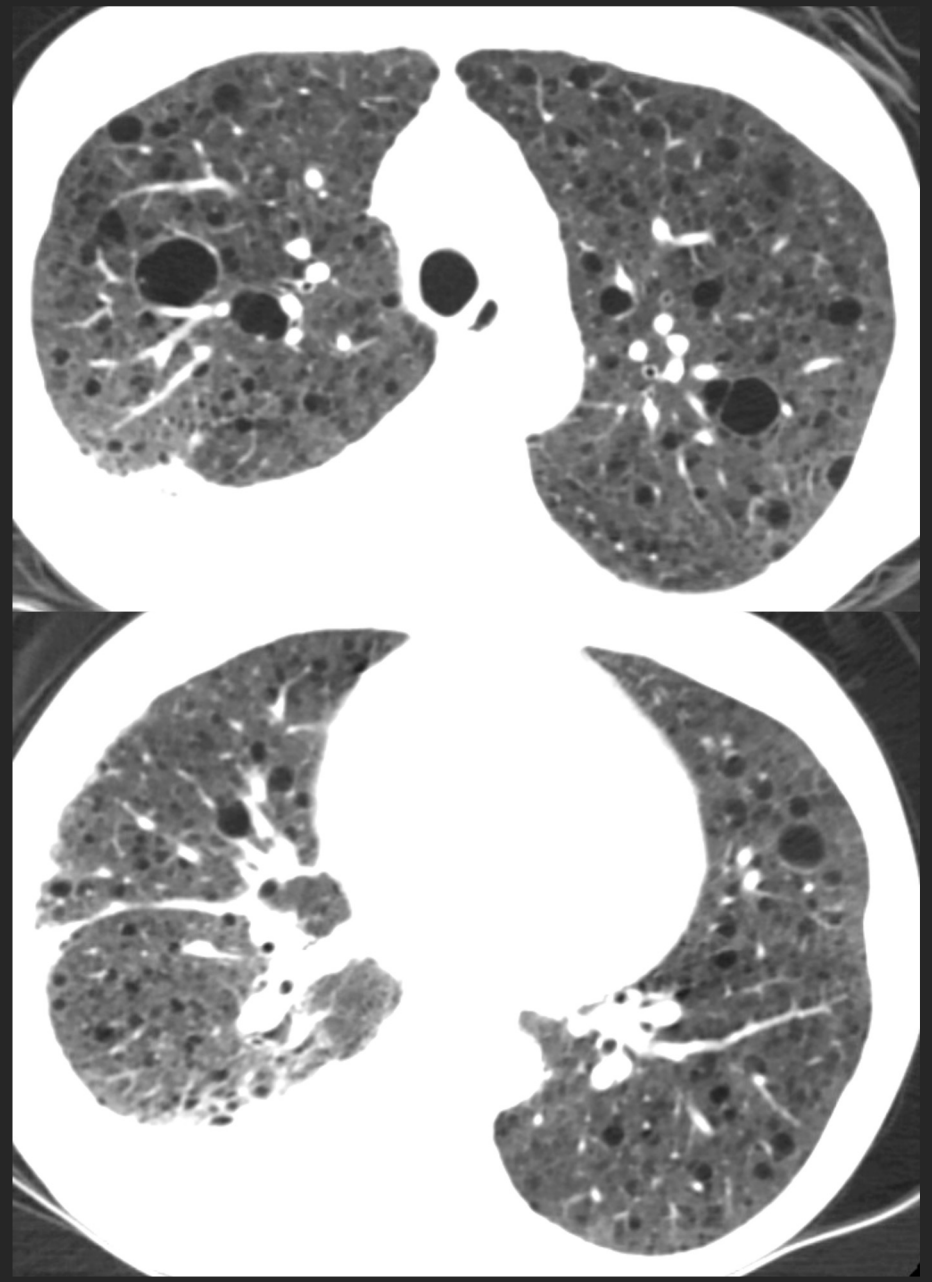
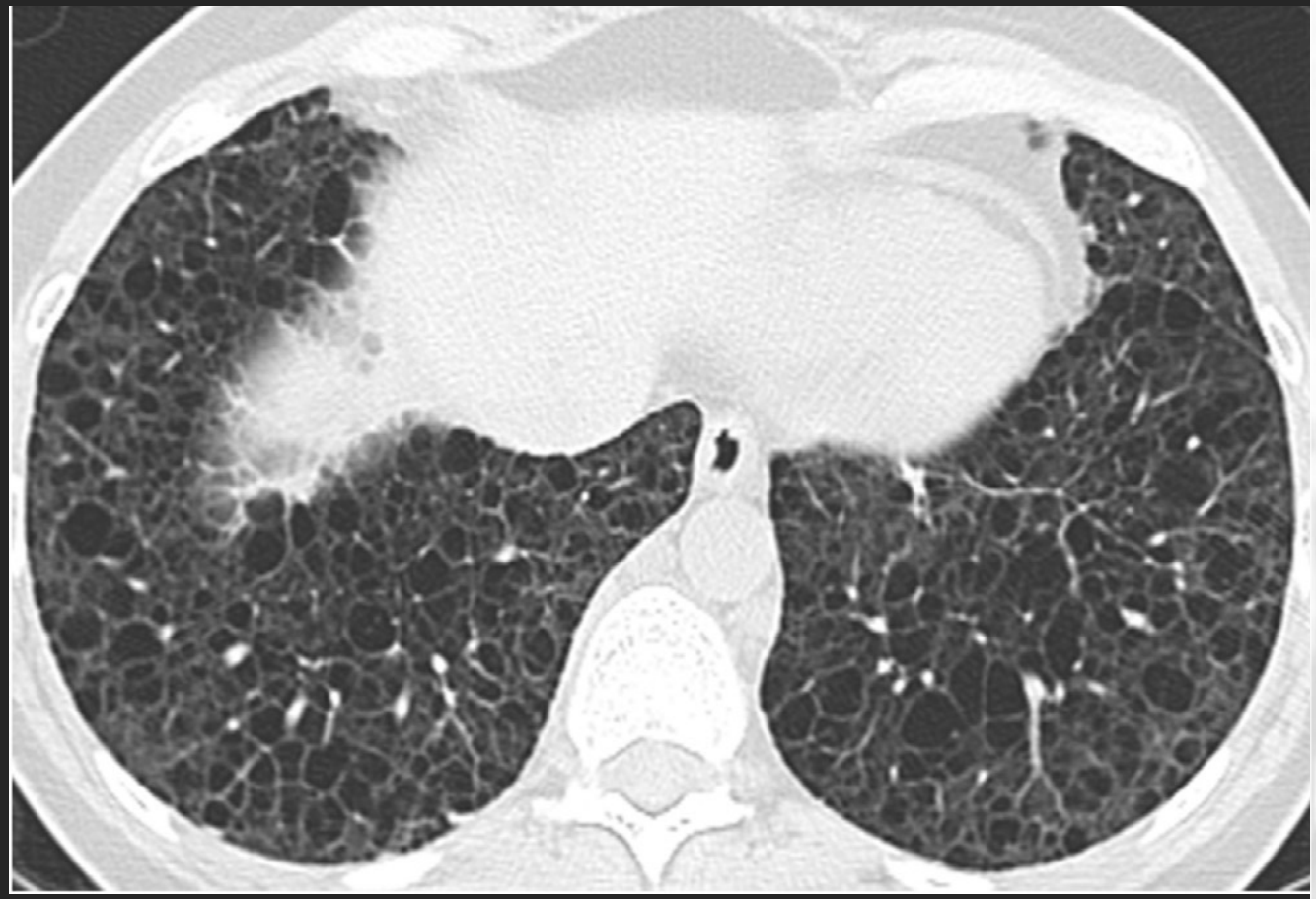
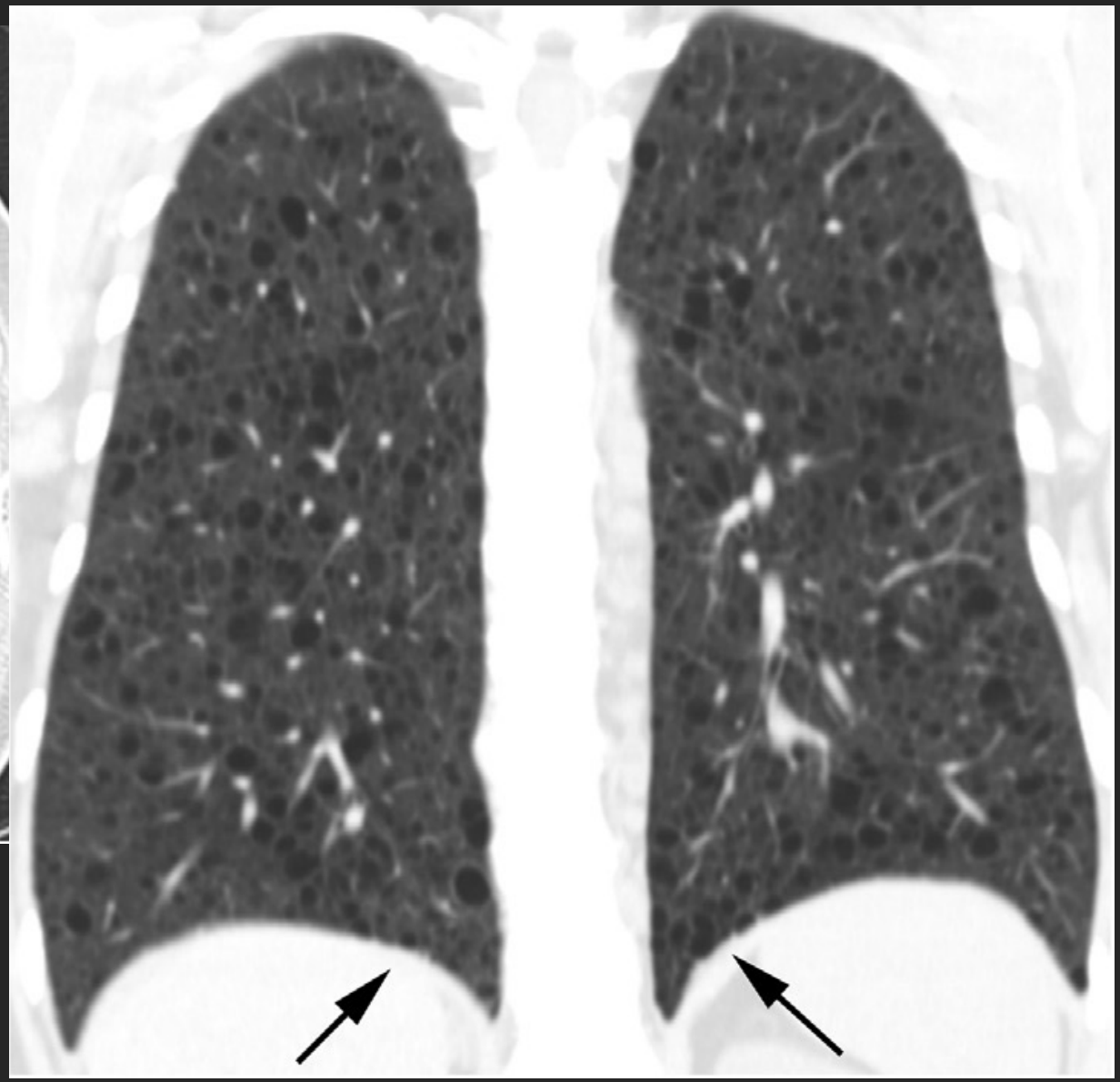


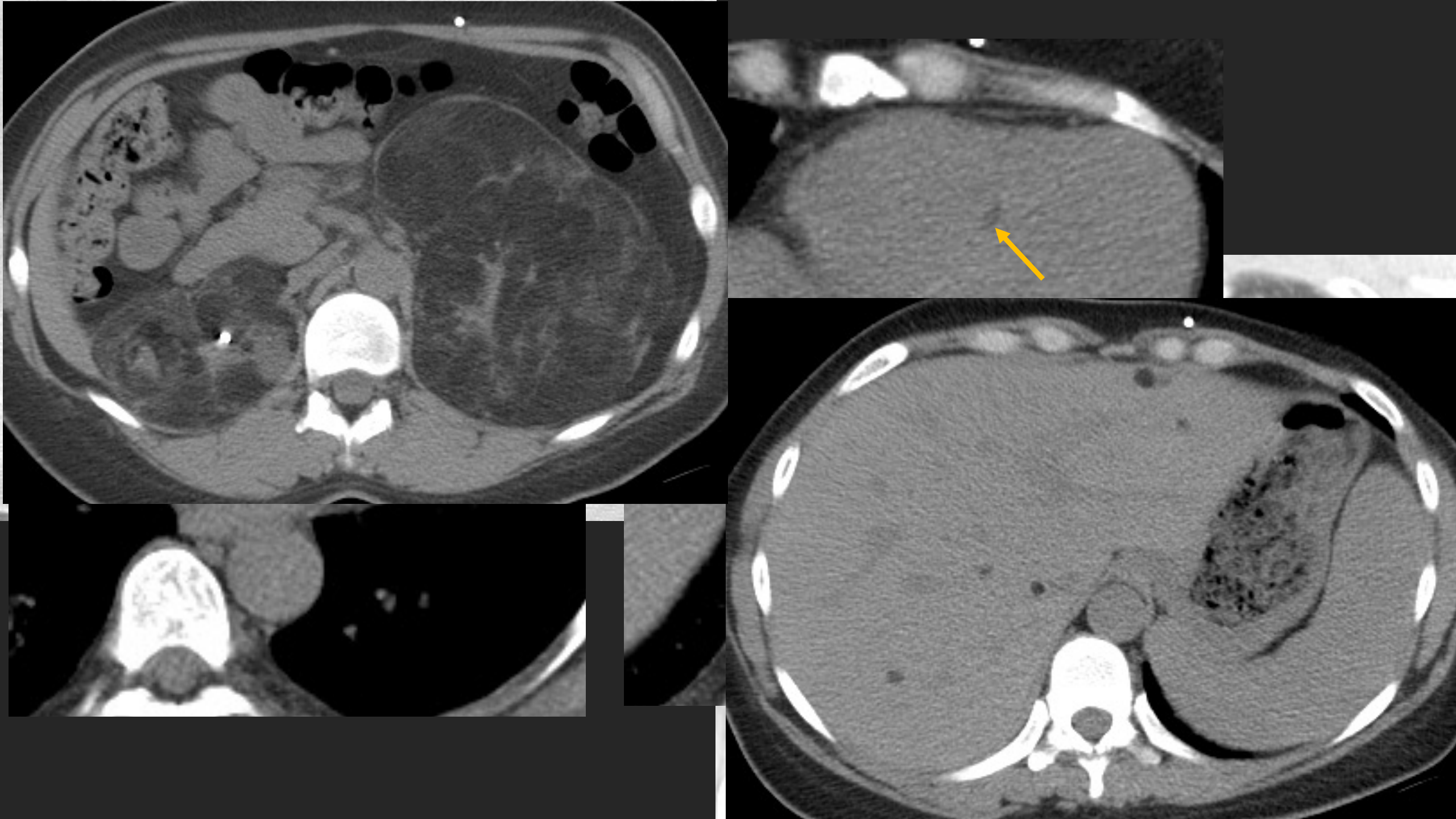




Lymphangioleiomyomatosis



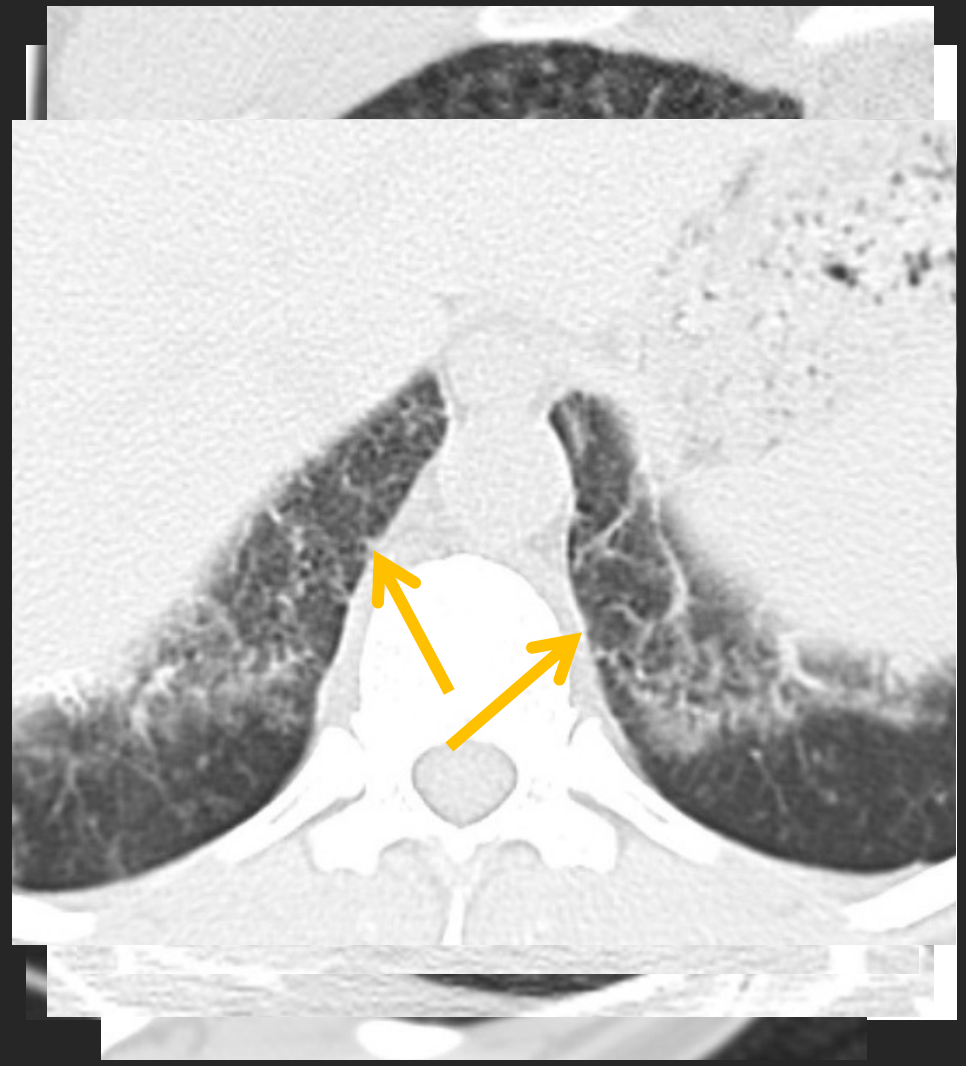




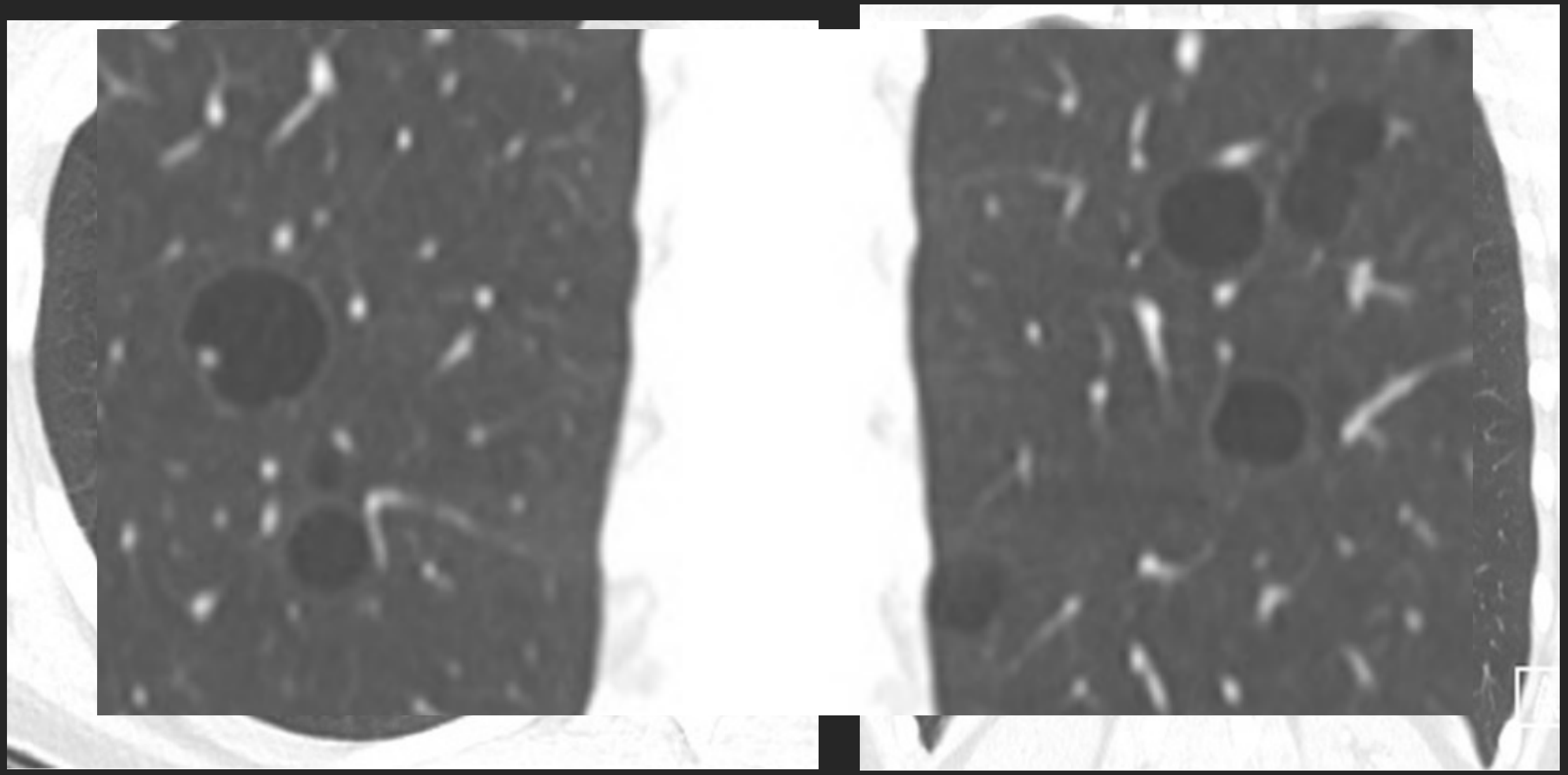
Lymphocytic Interstitial Pneumonia (LIP)

Chest CT

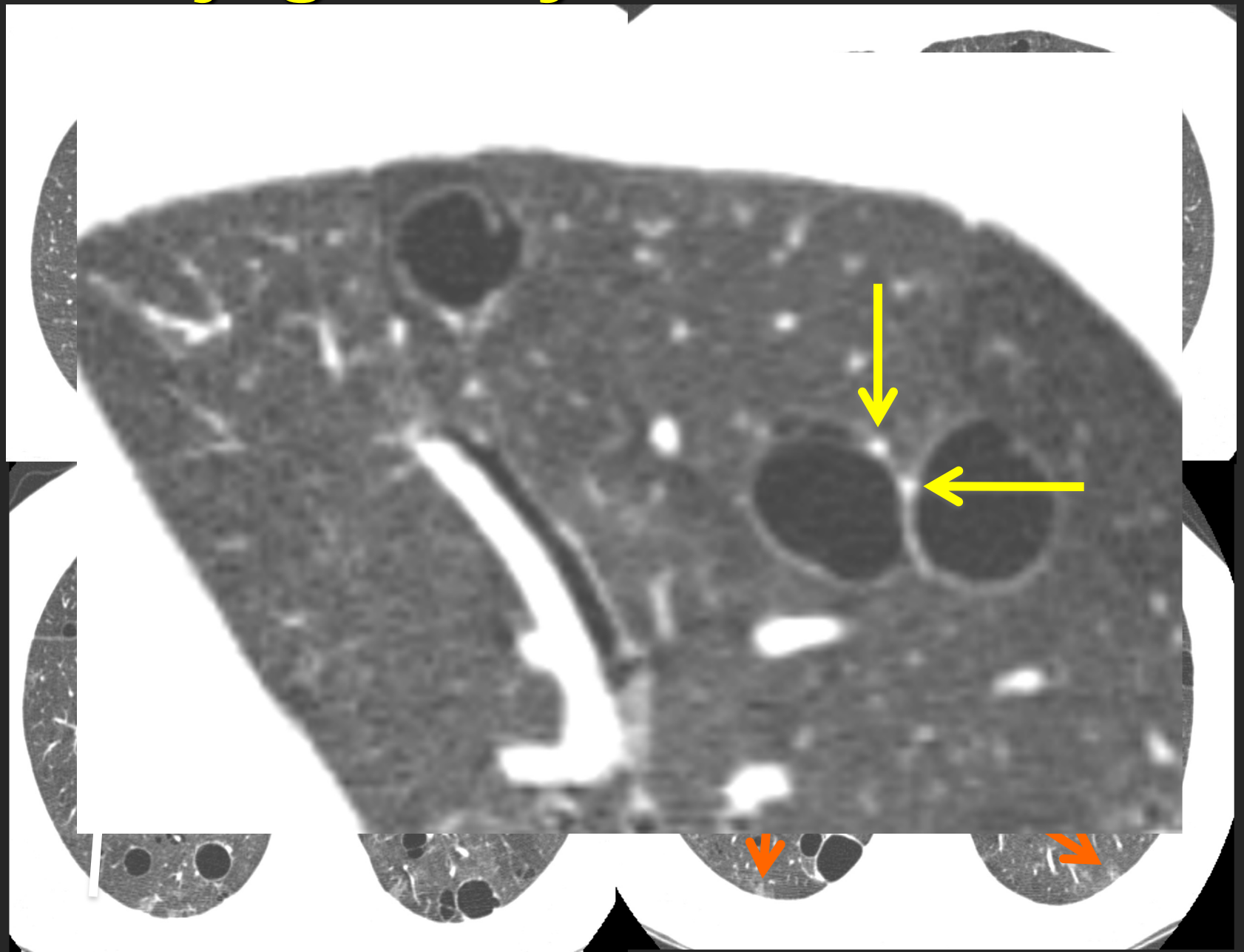
- Ground-glass opacities
  - Dominant feature, often bilateral
- Cysts:
  - Variable in number
  - Size generally < 3 cm
  - Perivascular or subpleural cysts common; basilar
  - Typically form in areas of previous centrilobular nodules
  - Common in areas of ground-glass opacity
- Centrilobular nodules
- Reticulation
- Distribution:
  - Diffuse or lower lung predominant



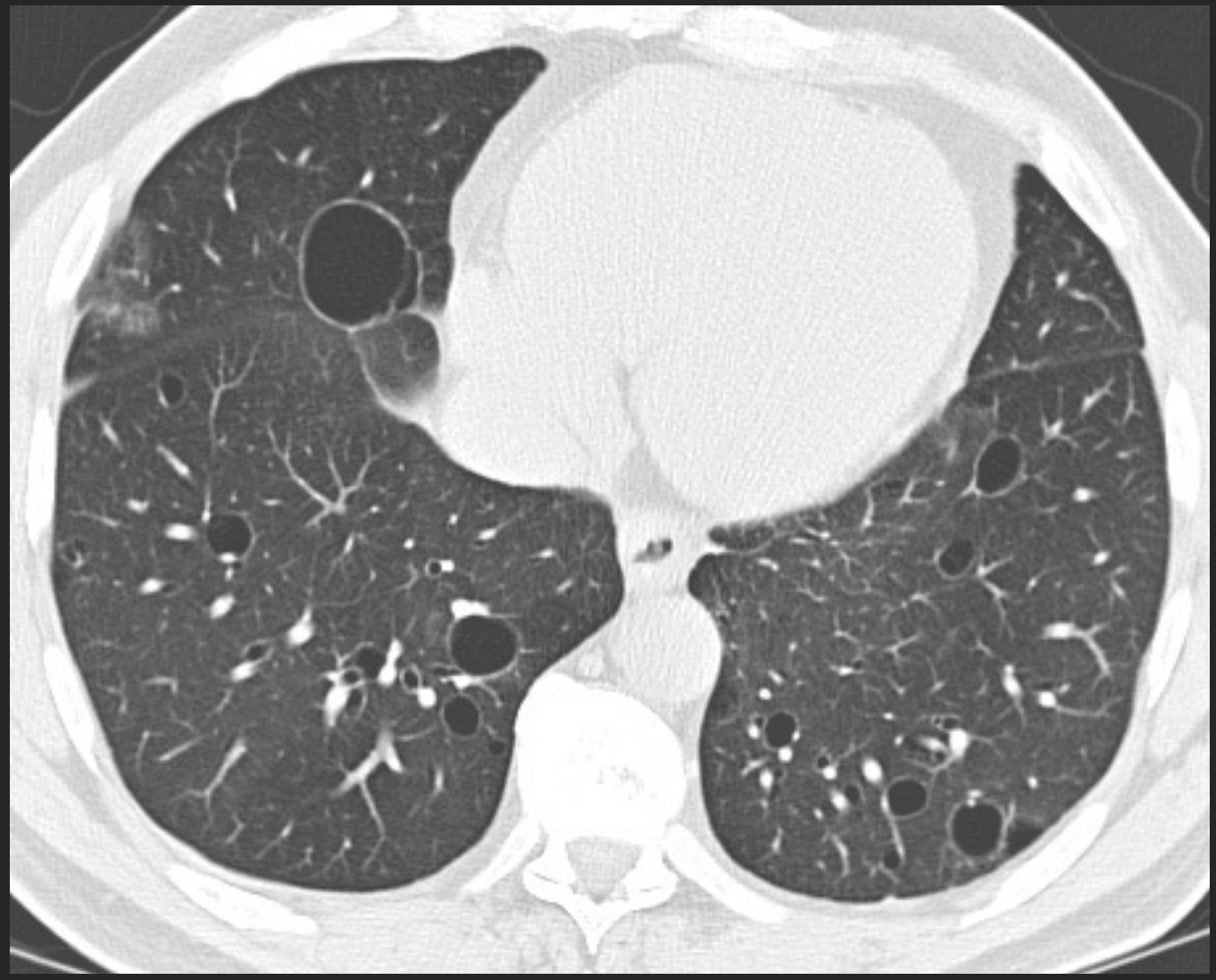
Sjögren syndrome: LIP



Sjögren syndrome: LIP



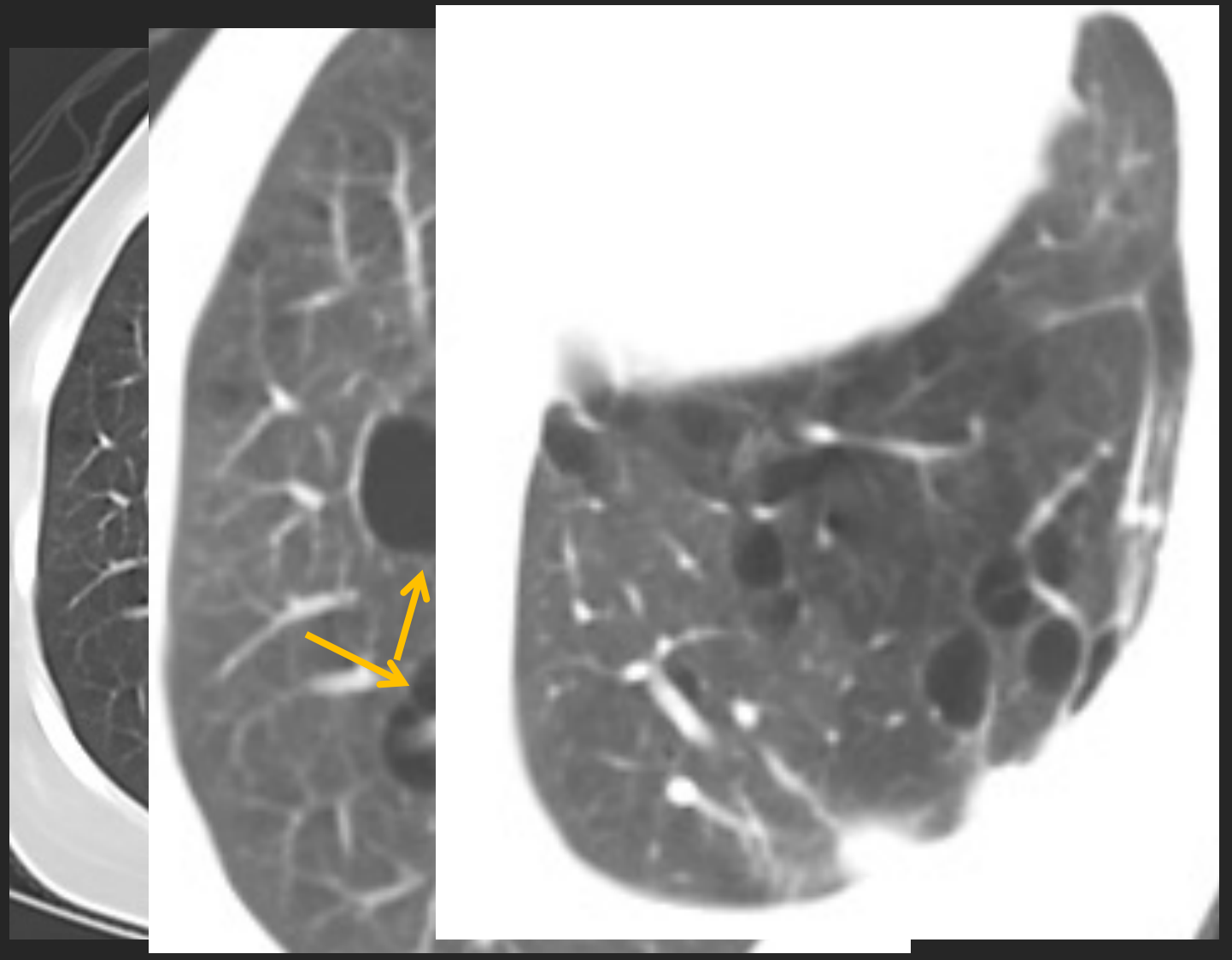
LIP and Sjögren syndrome



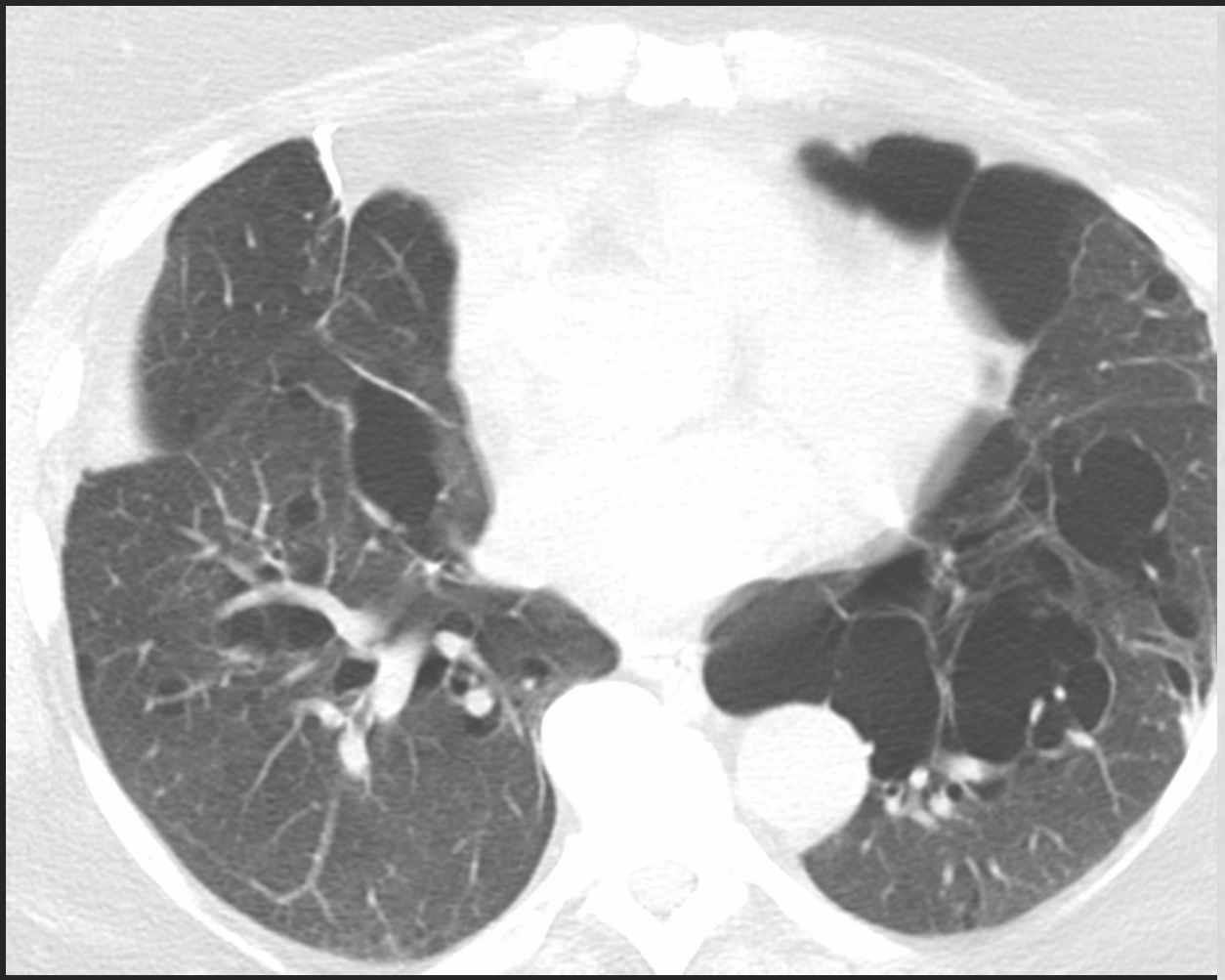
Birt-Hogg-Dubé Syndrome

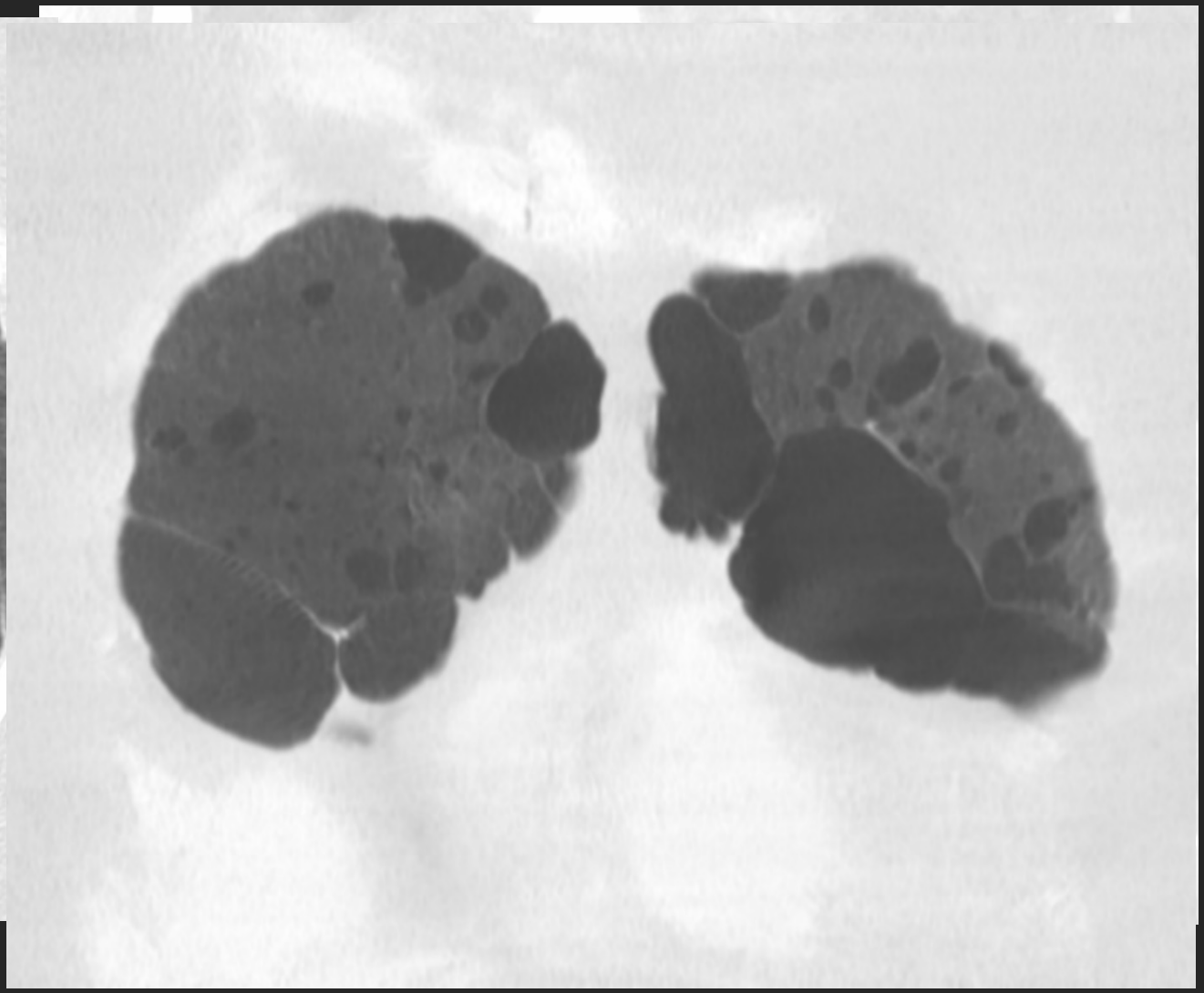
Cysts on CT

- Extent:
  - Sparse to innumerable; variable size (3mm to 8 cm)
  - Often remain relatively stable on follow-up CT scans
- Distribution:
  - More concentrated and larger in the lower and medial lung regions
  - Subpleural or paramediastinal cysts
  - Cysts along the lower lobe proximal pulmonary arteries and veins
- Morphology:
  - Thin uniform wall
  - Uniformly round or combination of round-, ovoid-or lentiform-shape
  - Large, multi-septated, air cuff sign

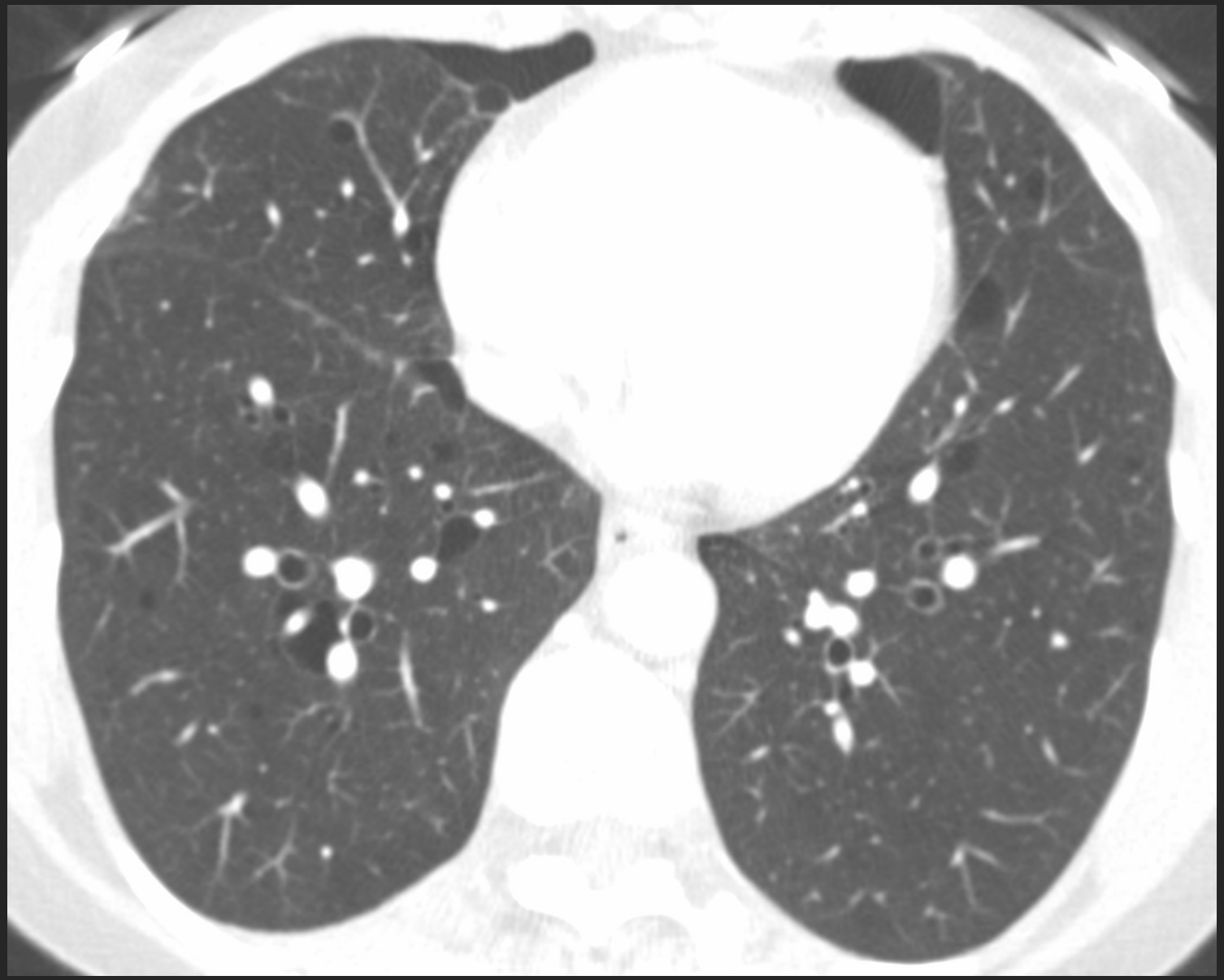


Birt Hogg Dube





Birt Hogg Dube



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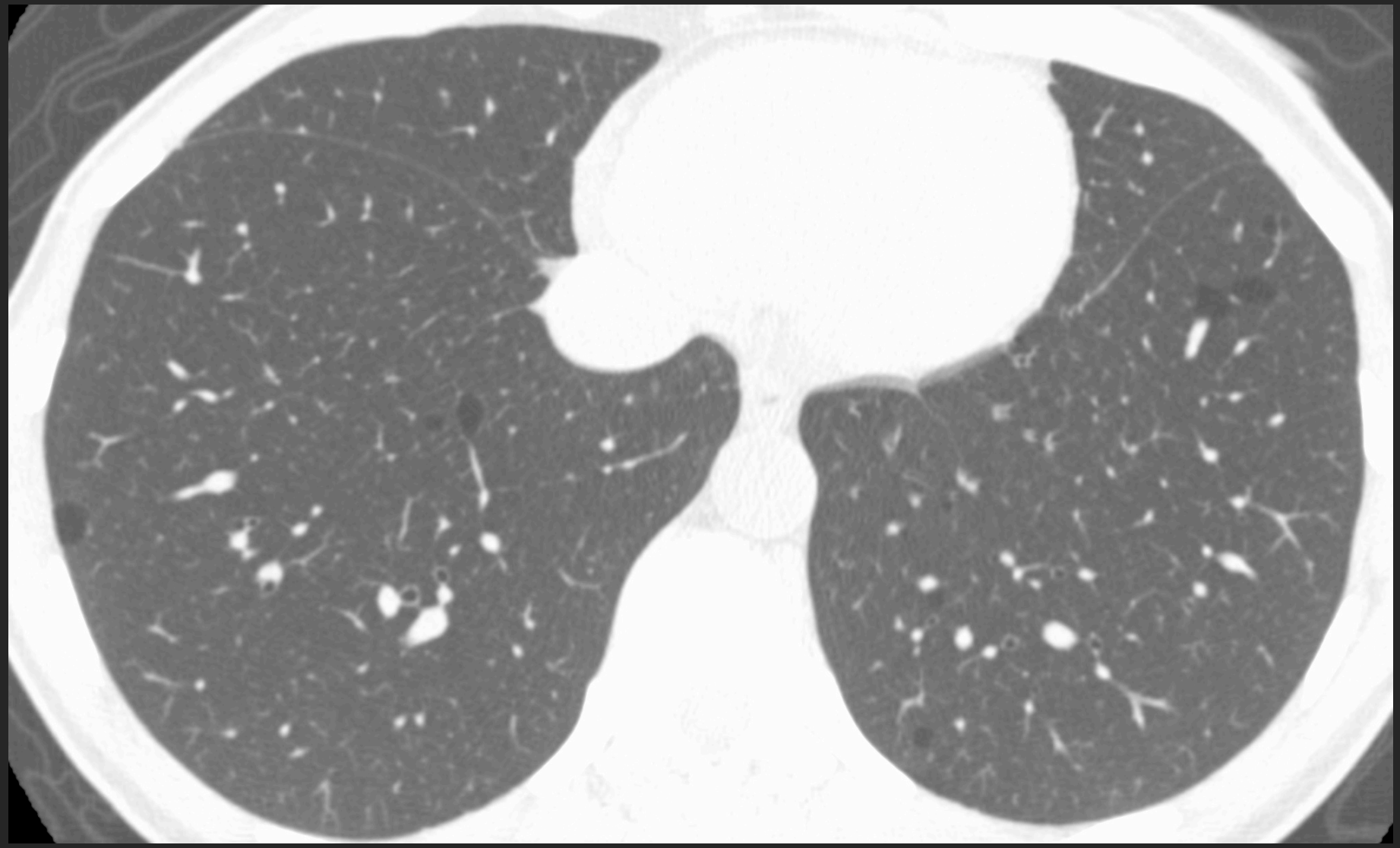
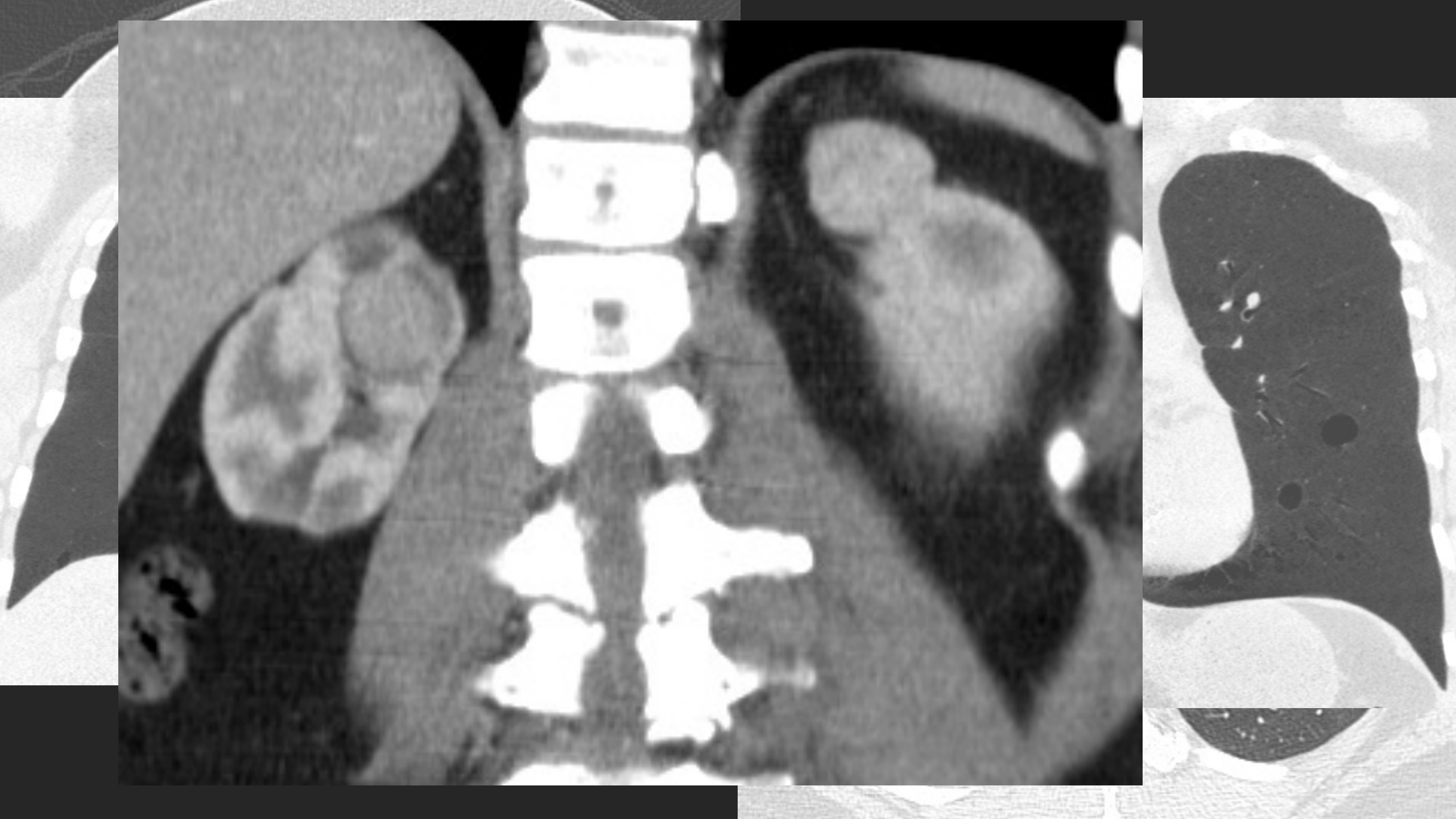
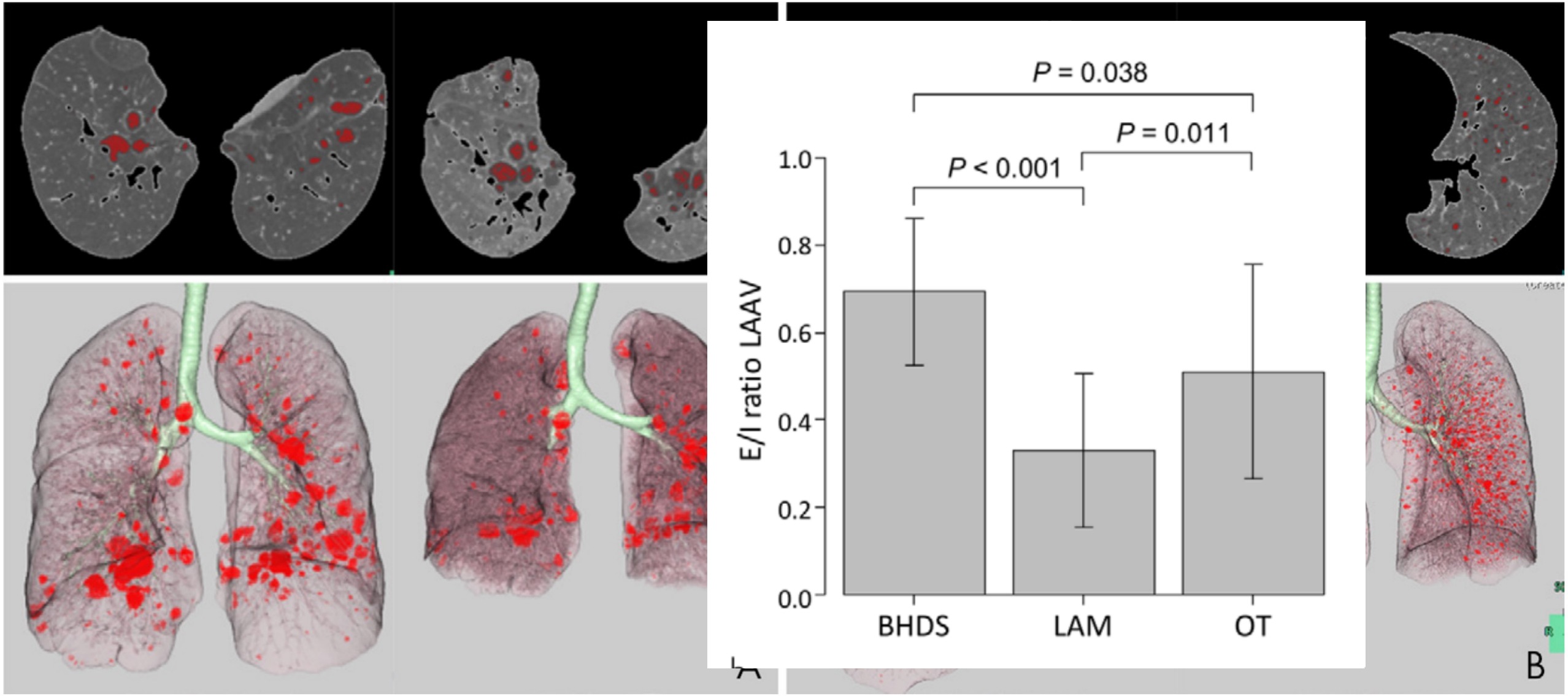


Courtesy of Nishant Gupta, MD



Birt Hogg Dube



Suzuki K, et al. **Radiol Cardiothorac Imaging** 2020;2(2):e190097.

Cystic Disease	Cyst Wall	Cyst Distribution	Cyst Shape and Size	Associated HRCT Findings
LAM	Thin	Diffuse	Round or oval	Usually normal intervening lung; TS-LAM: Multifocal micronodular pneumocyte hyperplasia
PLCH	Thin and thick	Upper and middle lung zones; spares costophrenic angles	Bizarre shaped, round (often heterogeneous)	Nodules (well - or poorly-defined) often with central lucencies (cheerio-like appearance)
LIP	Thin	Lower lung zone; perivascular and subpleural cysts	Round or oval	Ground-glass opacities, reticulation, centrilobular nodules
BHD	Thin	Lower lung; subpleural and perivascular; peramediastinal cysts	Round-, ovoid-or lentiform-shaped; air cuff sign, multi-septated cysts, less exp collapse	Renal tumors (RCC>oncocytomas)

Thank You

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Dr. Nishant Gupta

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