10° International Meeting on Pulmonary Rare Diseases and Orphan Drugs

Milan 3-4.03.2023

IMAGING IN ILD - CTD

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Disclosure

Nothing to disclose

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Background

- Lung diseases in CTDs cause morbidity and mortality
- Patterns of ILD mirrors those seen in the IIPs
- Lung involvement in CTDs no limited to ILDs

  Airways disease

  Pleural involvement

  Cardiovascular disease

Spagnolo P et al, Eur Respir J 2016; 47: 1535-1558 *Fischer A and du Bois RM, Lancet 2012; 380:689–698* *Kim EA et al, Radiographics 2002; 22:S151–S165*

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What is the role of imaging?

Suggest diagnosis

Prognostication

Follow-up

- Progression
- Complications

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What is the role of imaging?

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Complications

- *HRCT Pattern*
- *"Peculiar" features of the pattern*
- *Additional findings*

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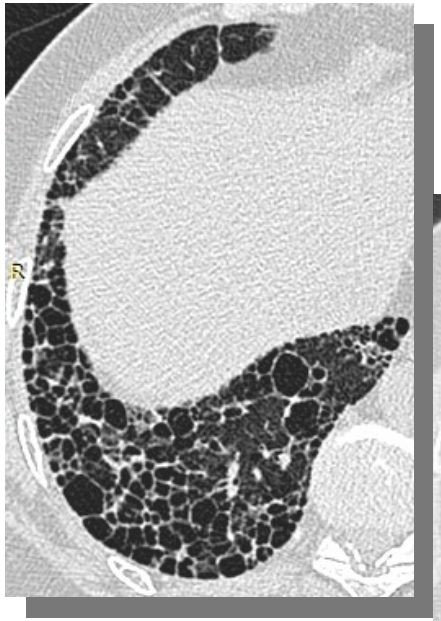
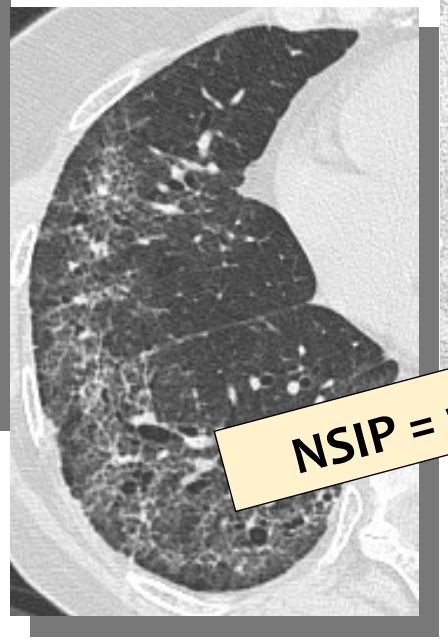
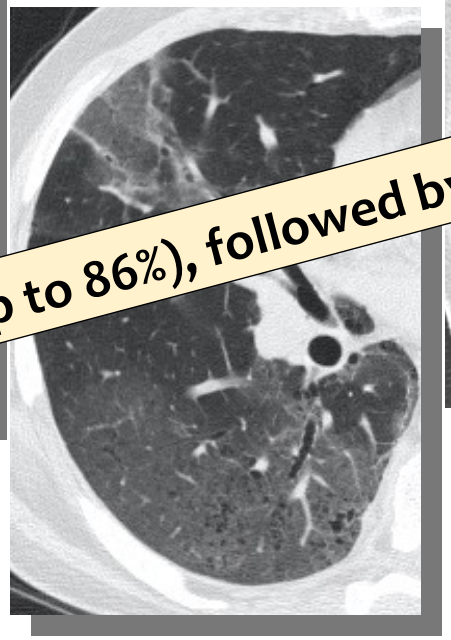


UIP

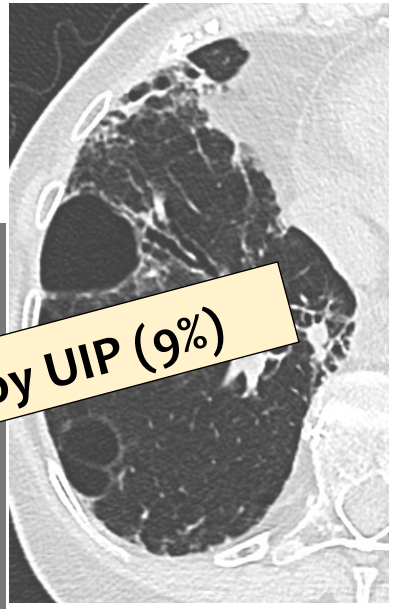


NSIP

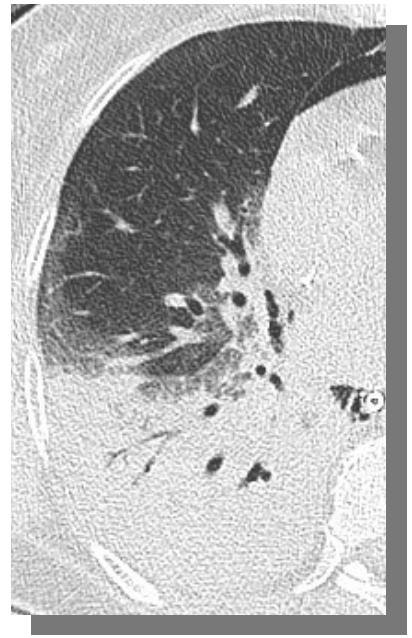
OP



DIP



LIP



DAD

NSIP = most common (up to 86%), followed by UIP (9%)

Kim EA et al, Radiographics 2002; 22:S151–S165

Walsh SL et al, Thorax 2014; 69:216-22

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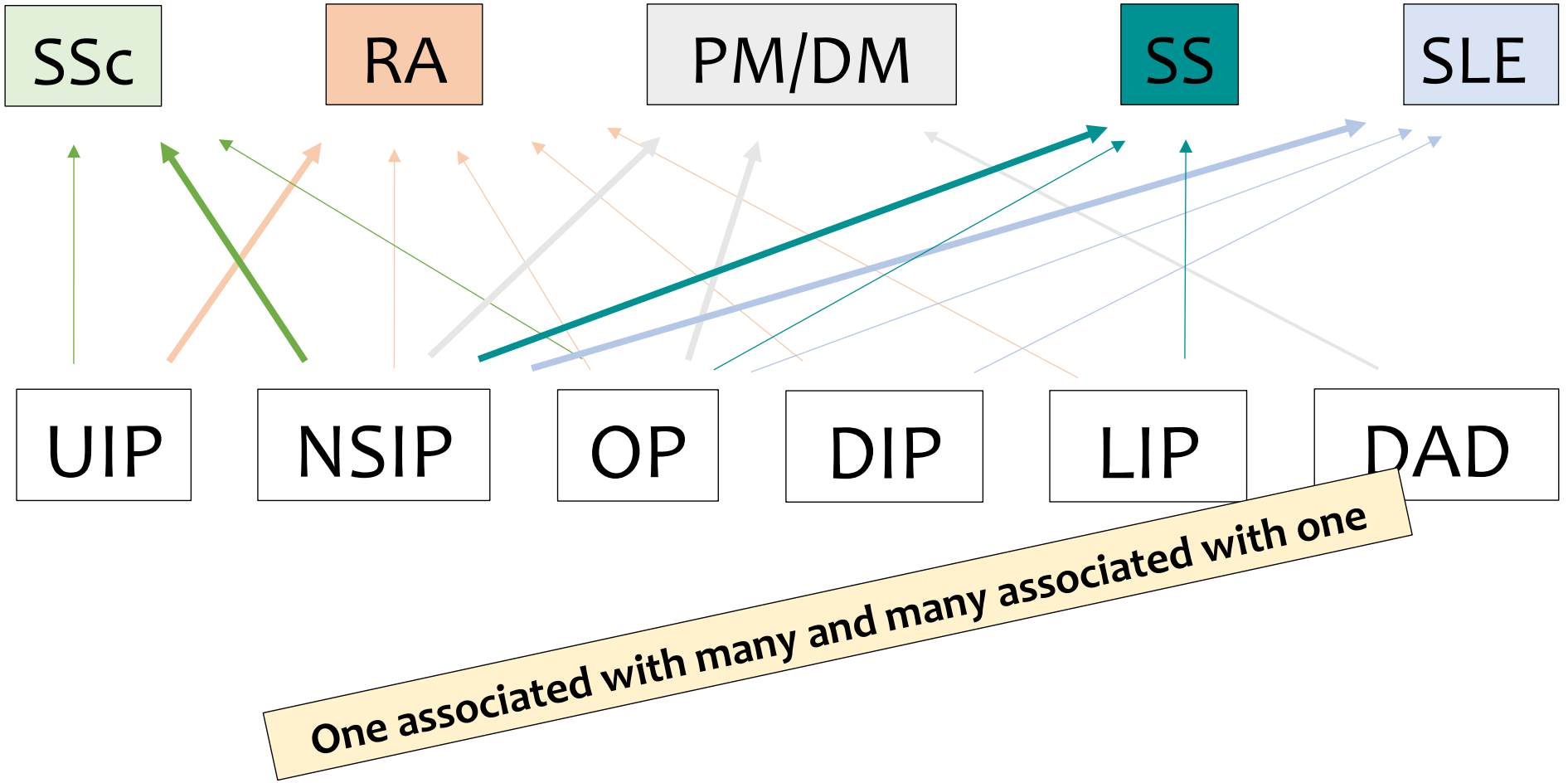

Palmucci S et al, Insights Into Imaging 2022; 13:108

Kim EA et al, Radiographics 2002; 22:S151–S165

Walsh SL et al, Thorax 2014; 69:216-22

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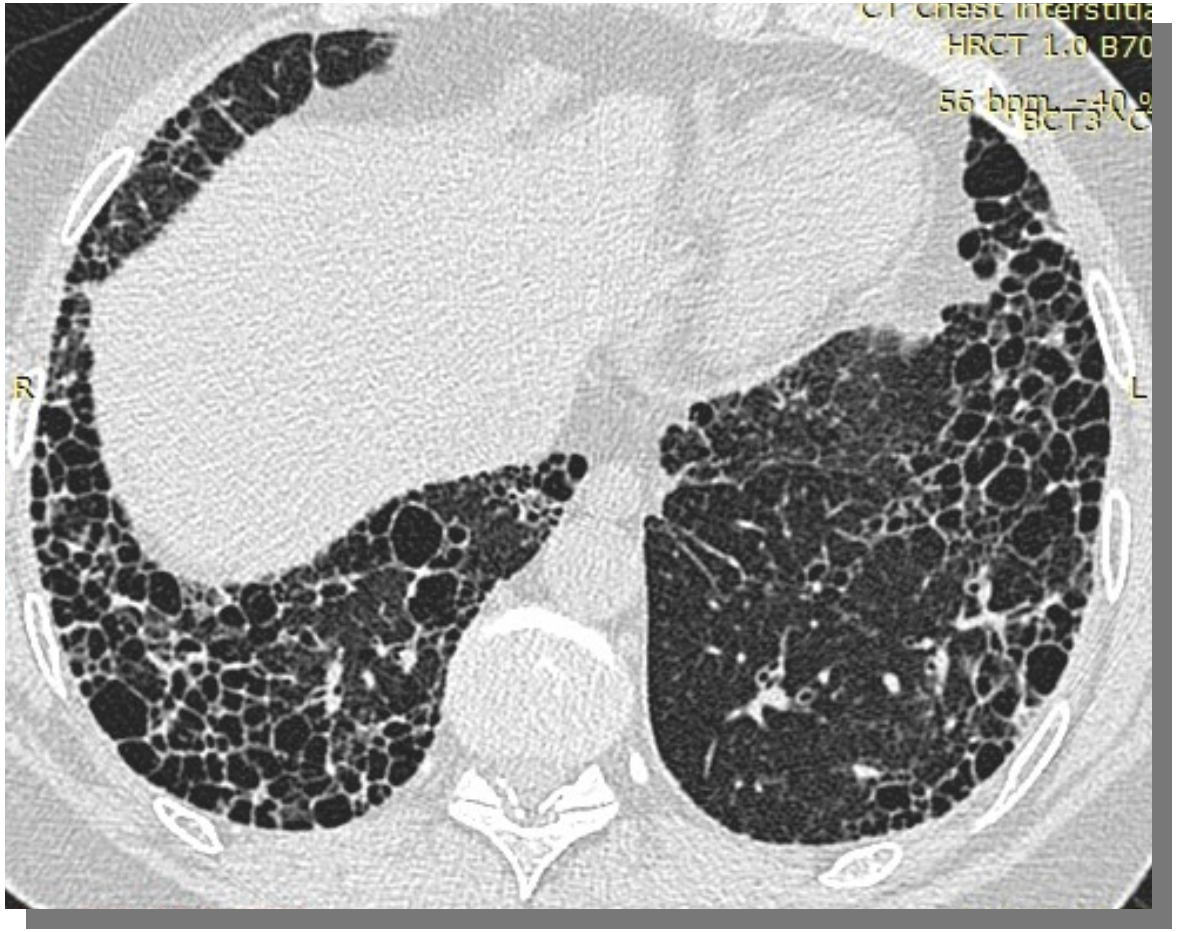
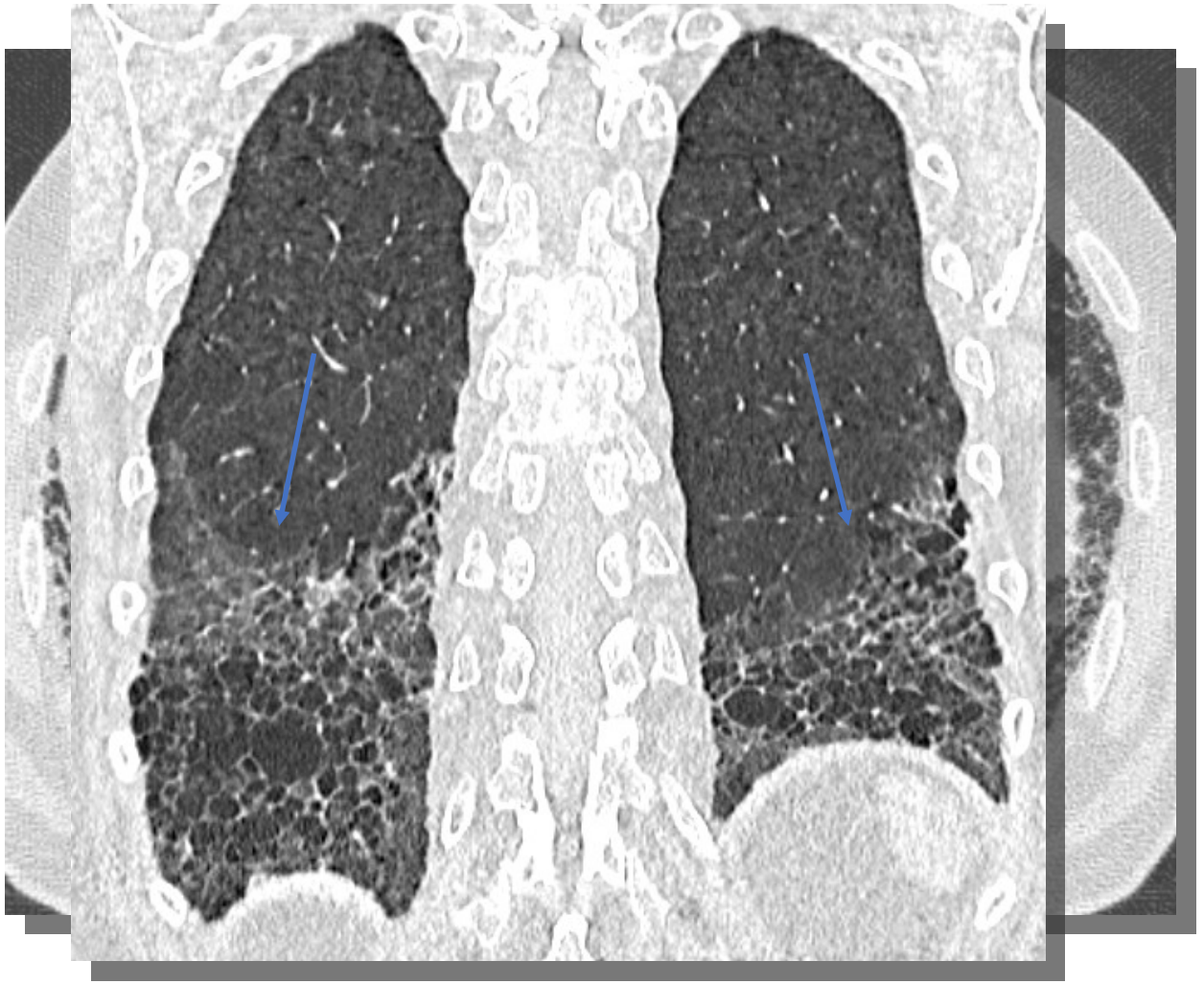


UIP in RA

Exuberant honeycombing



UIP in SSc

Straight-edge sign

Chung JH et al, AJR 2017; 210:307–313

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	SSc	RA	PM/DM	SS	SLE
Lung parenchyma					
NSIP	+++	++	++	++	+
UIP	+	++	+	+	+
LIP		+		++	
OP		++	++	+	
DAD		+	+		+
Hemorrhage					++
Nodules		++			
Pleura		+++			+++
Airways		++		++	
Vessels	+++				++

Kim EA et al, Radiographics 2002; 22:S151–S165

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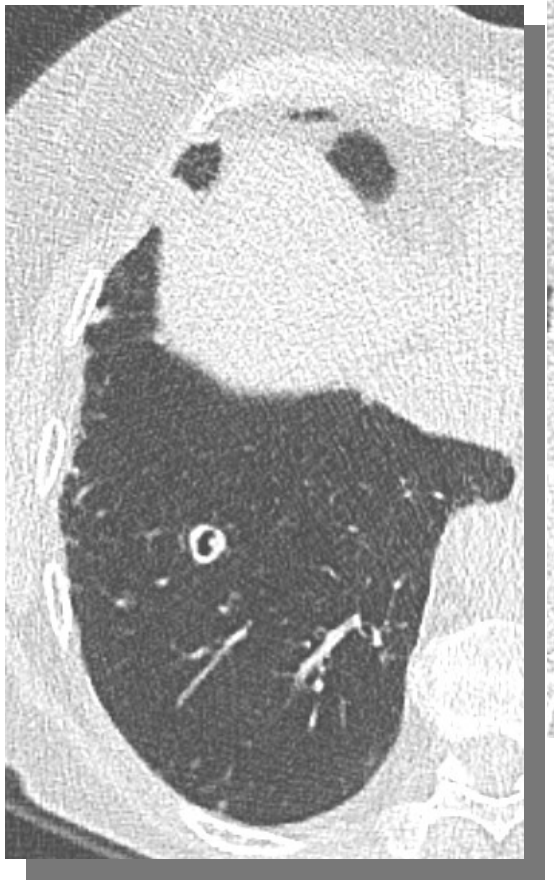

Nodules in RA

- *Longstanding disease*
- *Subcutaneous nodules*
- *Interlobular septa or subpleural*
- *Single or multiple*
- *From few mm to several cm*
- *Cavitation and rupture can occur*

Shaw M et al, Eur Respir Rev 2015; 24: 1–16

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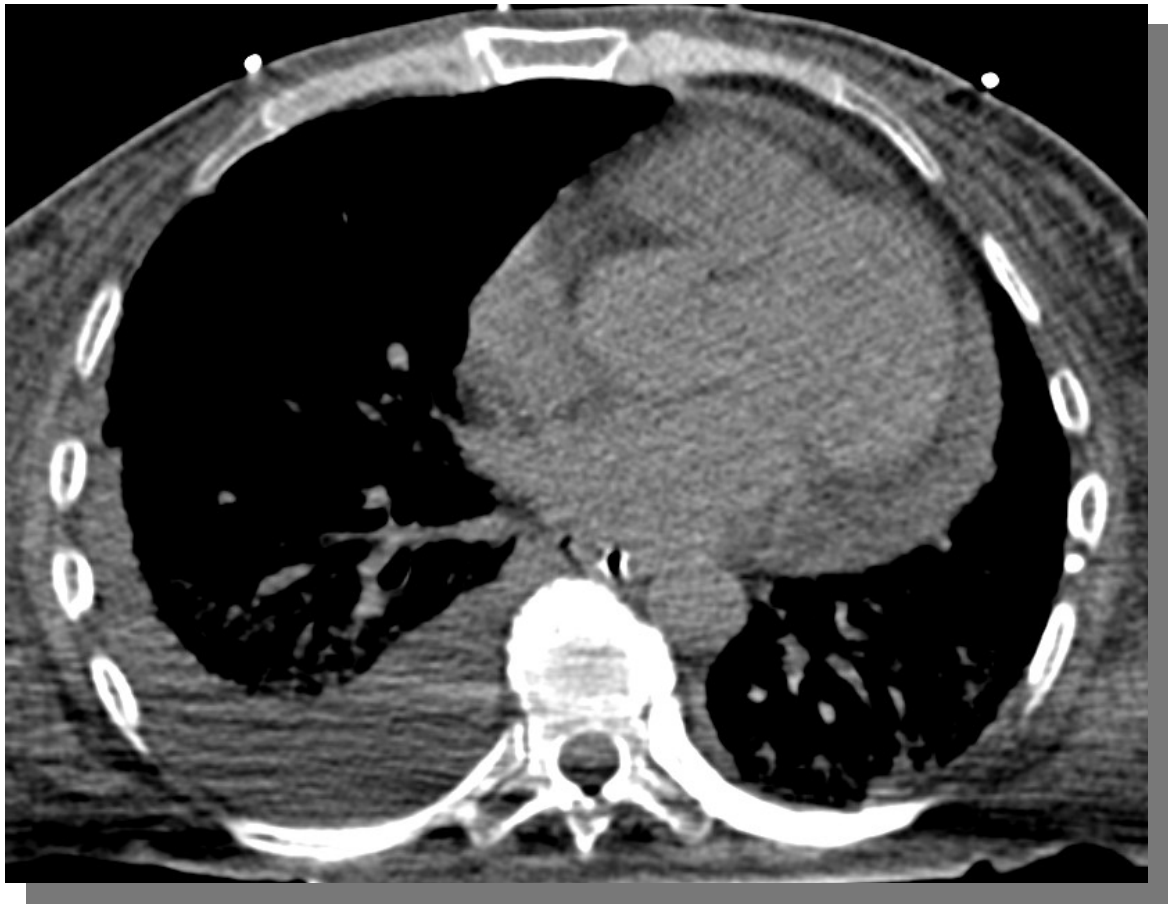
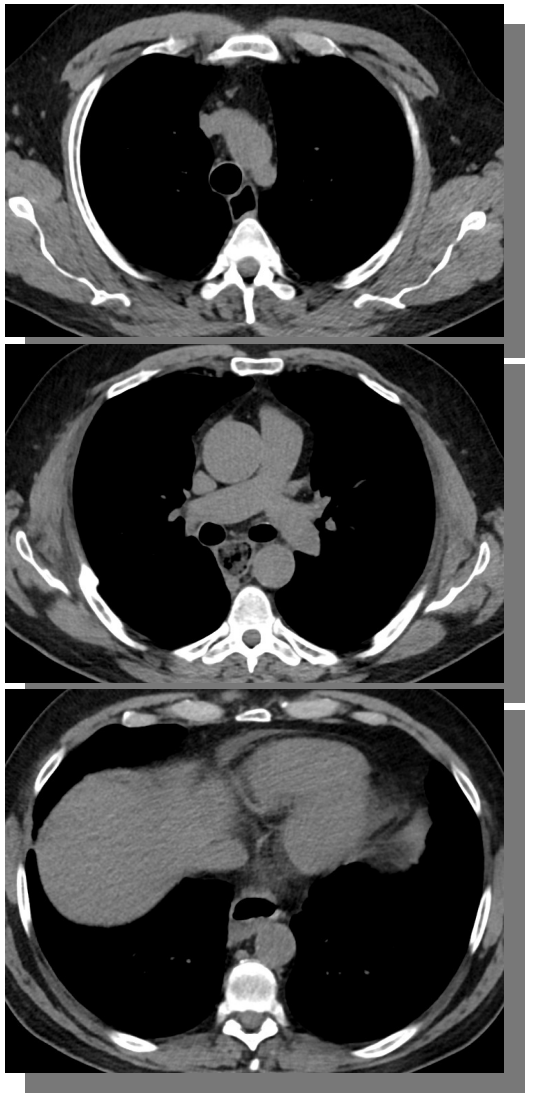


Pleural and pericardial effusion in SLE



Esophageal dilatation in SSc

Palmucci S et al, Insights Into Imaging 2022; 13:108

Lee MH et al, RadioGraphics 2019; 39:1411–1434

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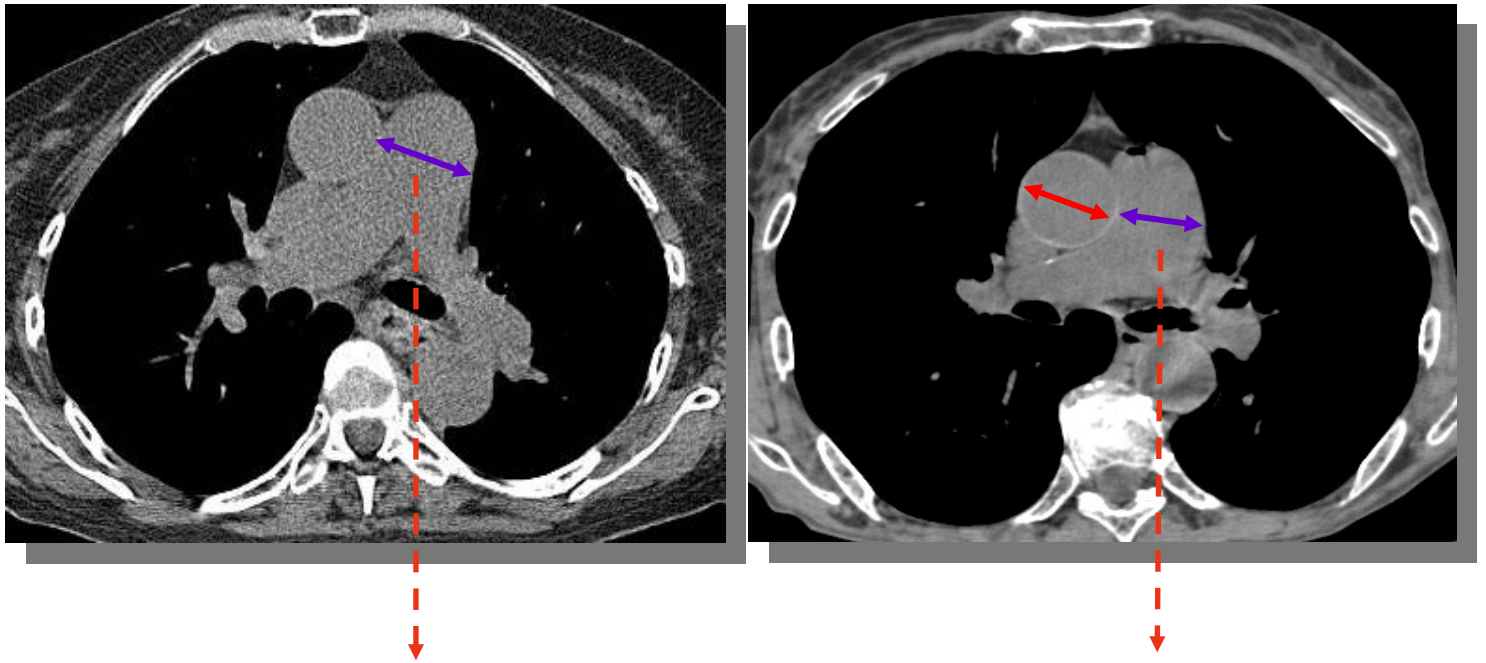
SSc and pulmonary hypertension

CT findings:

- *Enlarged main pulmonary artery +++*
- *Enlarged intralobar arteries ++*
- *Pericardial effusion +++*
- *Enlarged right cardiac chambers ++*
- *Contrast reflux in IVC +*



≥ 29mm

AP/Ao > 1

Haque A et al, Eur Respir Rev 2021; 30: 210053 *Remy-Jardin M et al, Radiology 2021; 298:531–549*

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What is the role of imaging?

Suggest diagnosis

Prognostication

- *HRCT Pattern*
- *Extent*

Follow-up

Progression

Complications

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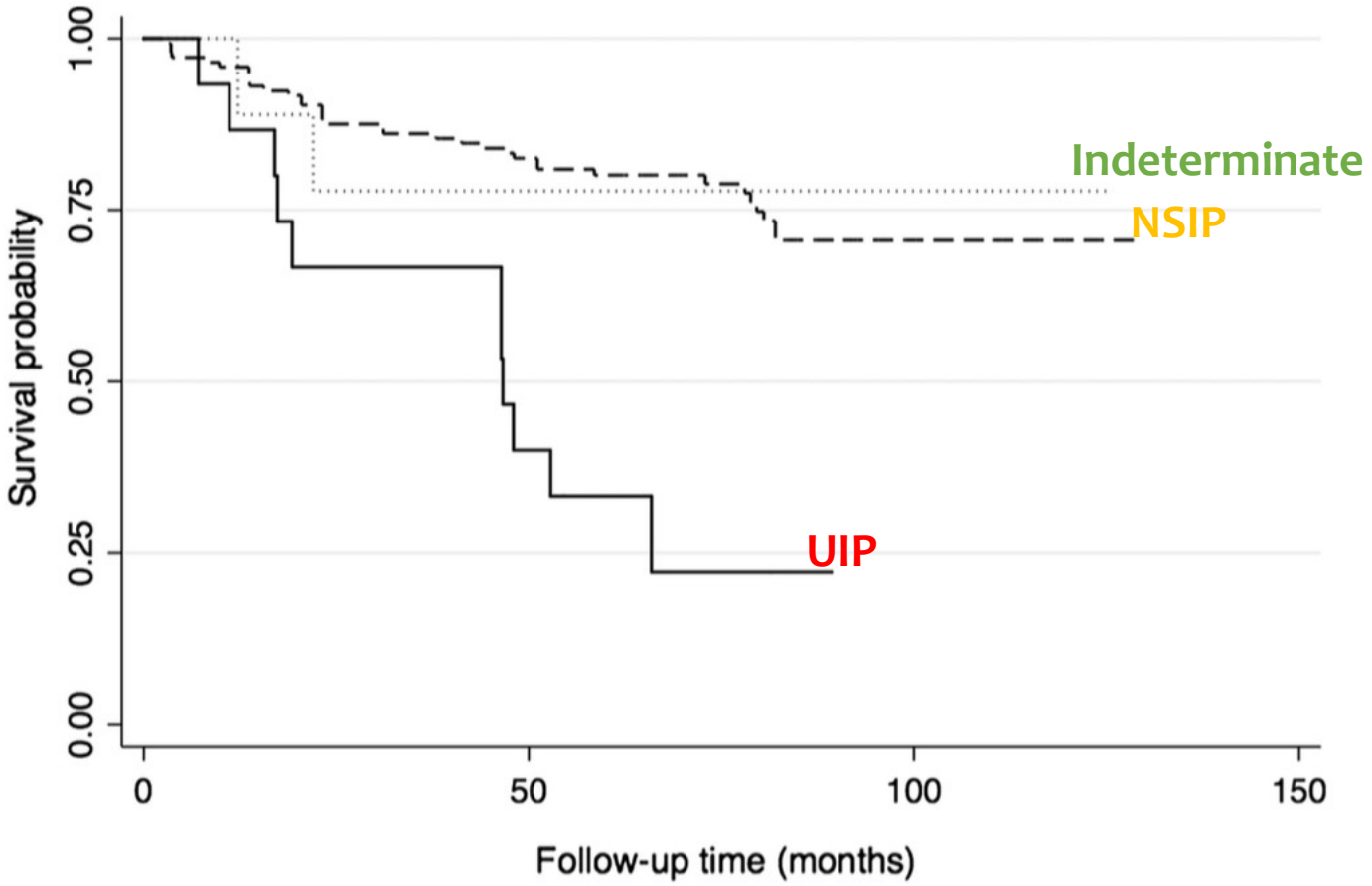
Prognostication | HRCT Pattern



Walsh SL et al, Thorax 2014; 69:216-22

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Prognostication | RA-ILD

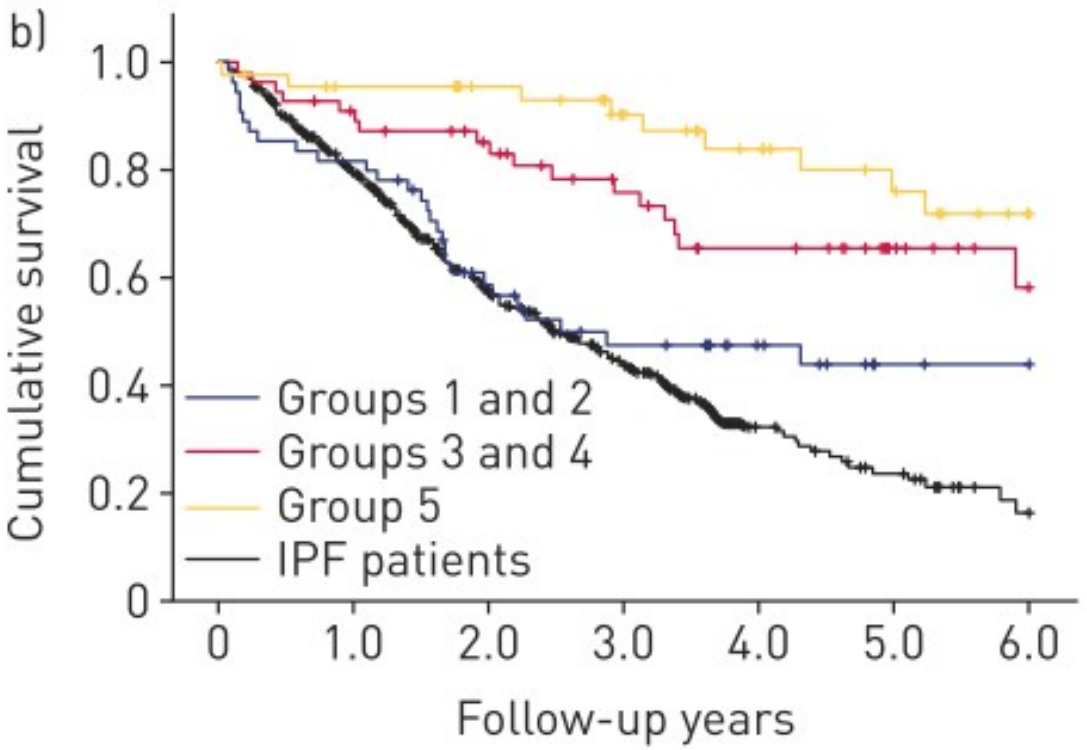
Group 1: definite UIP pattern in an IPF distribution

Group 2: definite UIP pattern not in an IPF distribution

Group 3: probable UIP in an IPF distribution

Group 4: probable UIP pattern not in an IPF distribution

Group 5: features inconsistent with UIP (excluding disease distribution and a mosaic attenuation pattern)



Jacob J et al, Eur Respir J 2019; 53: 1800869

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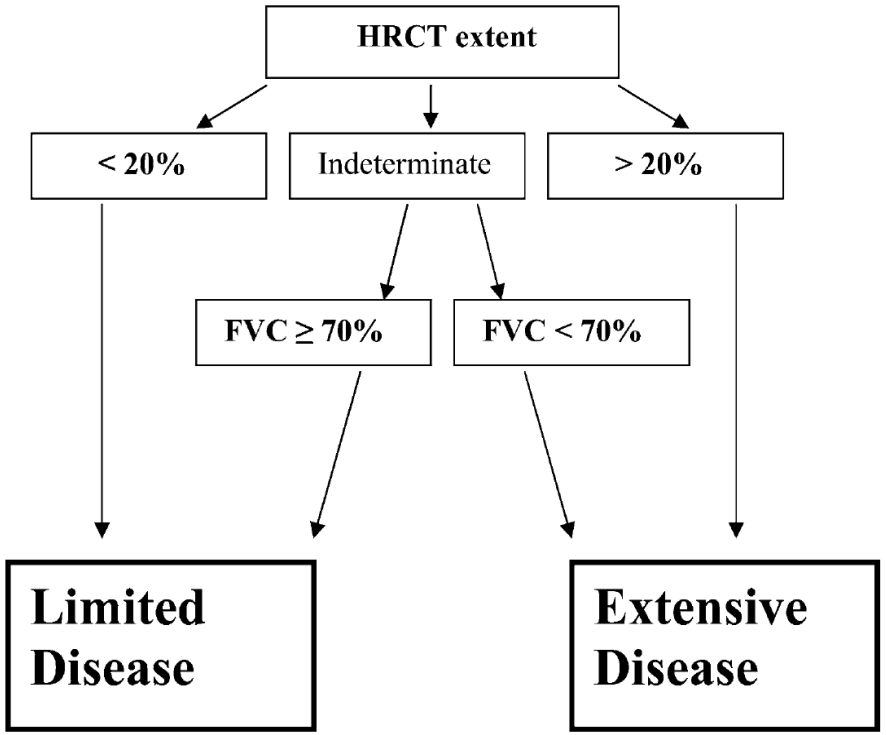
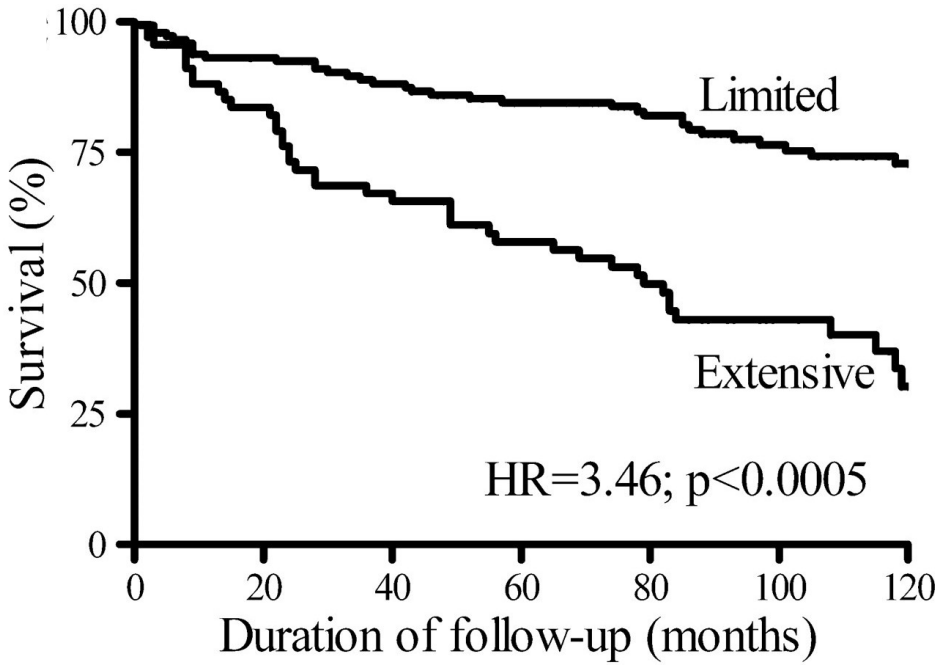
Prognostication | SSc-ILD





Goh NSL et al, Am J Respir Crit Care Med 2008; 177: 1248–1254

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What is the role of imaging?

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Prognostication

Follow-up

- Progression
- Complications

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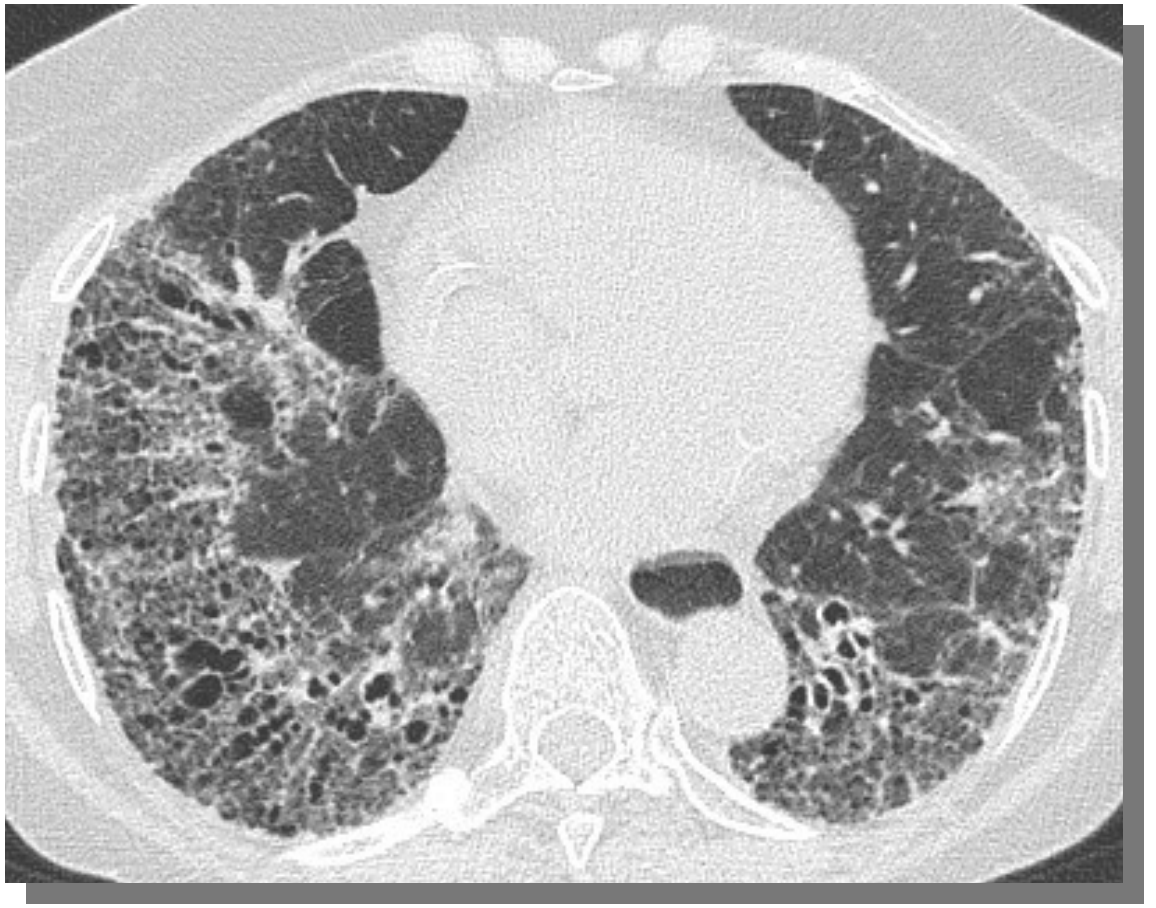
Progression

Inc

2018



2021

NSIP in SSc

Devaraj A, Eur Respir Rev 2014; Jun;23(132):215-9

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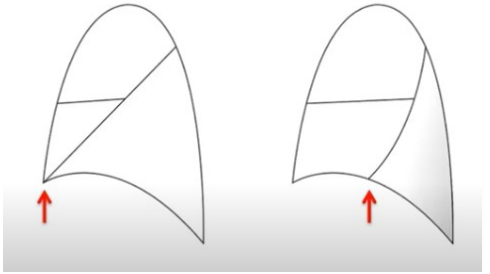
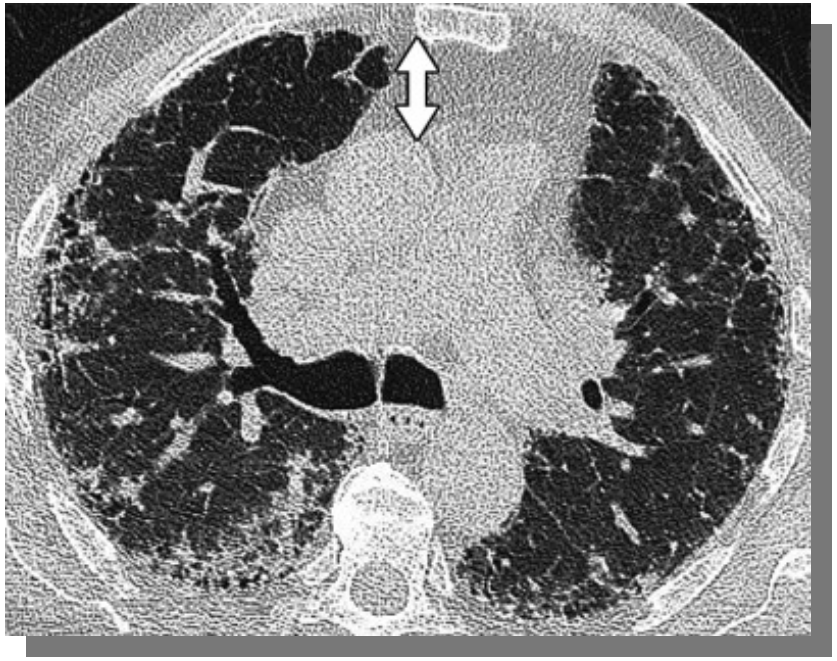
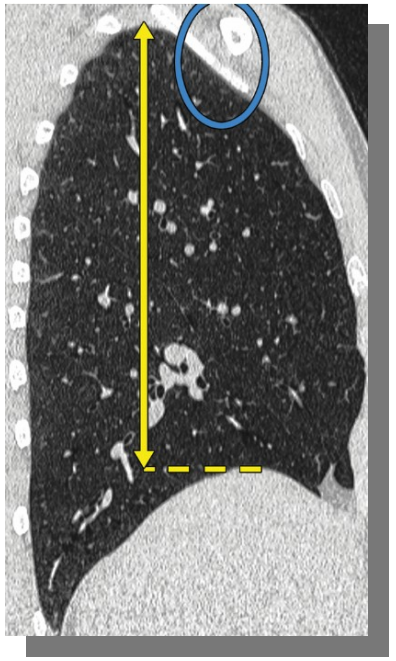
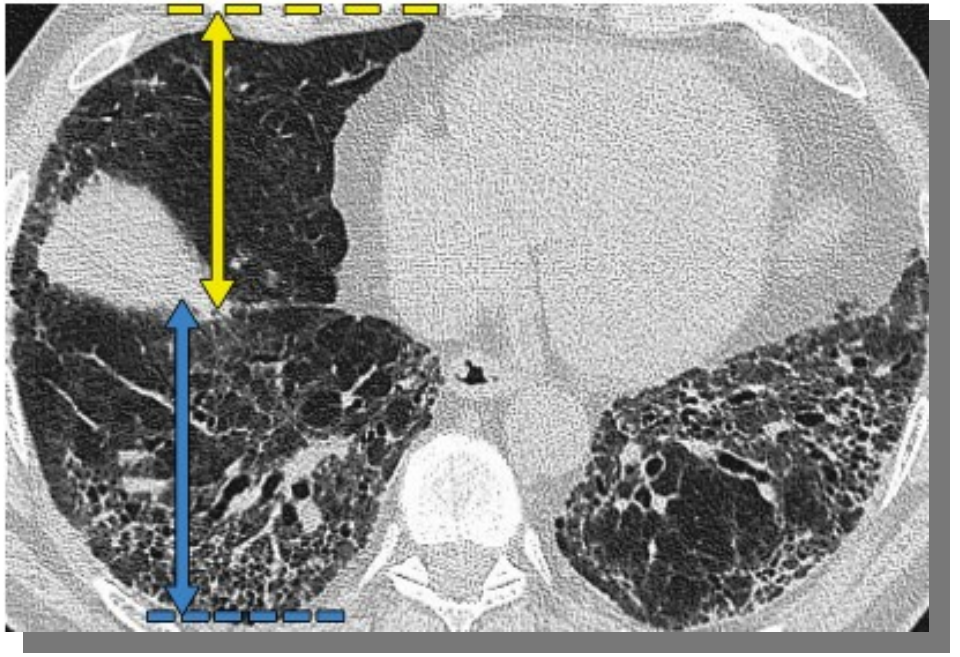




Aorto-sternal distance



Lung height



Oblique fissure retraction distance (ratio)

Robbie H et al, AJR 2019;213(2):318-24

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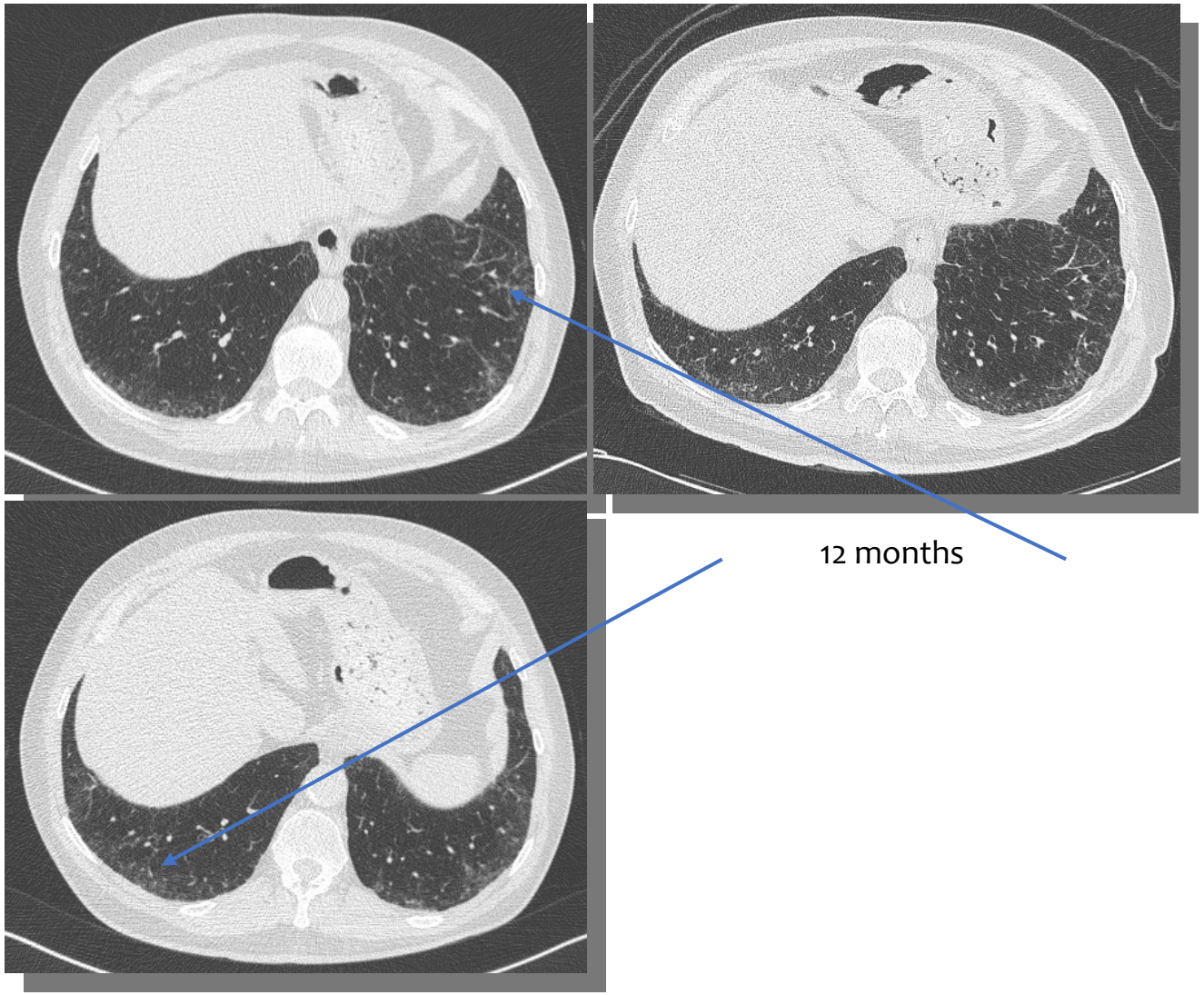
Visual score



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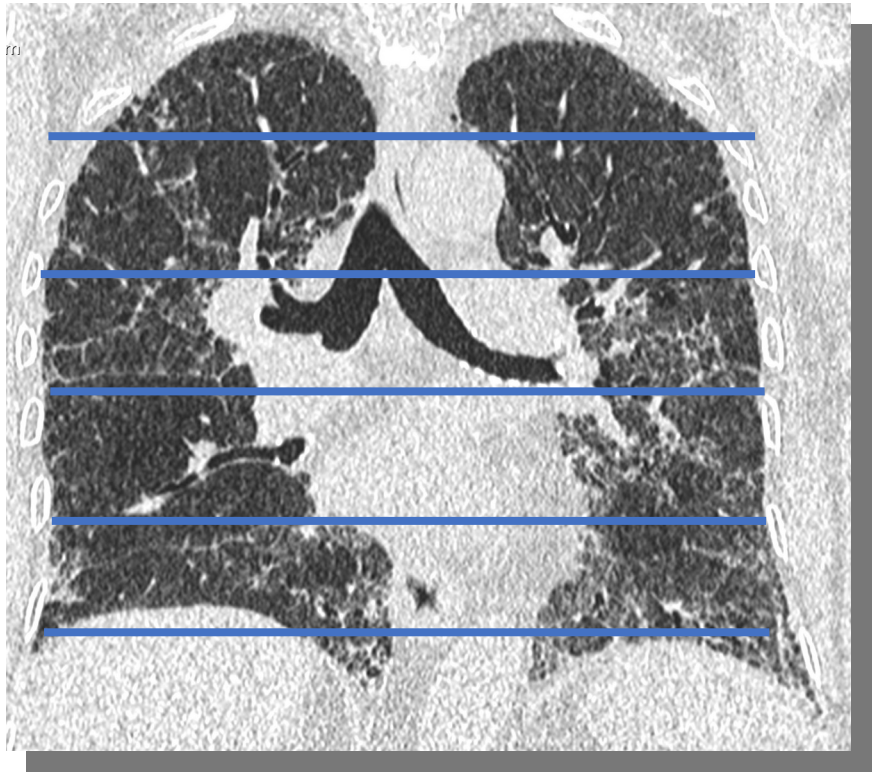
Semi-quantitative score

Nearest 5% extent of each HRCT finding (ground-glass opacity, reticulation, honeycombing, and emphysema)

3-6 predefined **sample slices** deemed representative for the entire lung:

- Supra-aortic vessels
- Carina
- Right pulmonary vein
- Diaphragmatic dome



Best AC et al, Radiology 2008;246(3): 935–940

Jacob J et al, J Thoracic Imaging 2016;31:304–311

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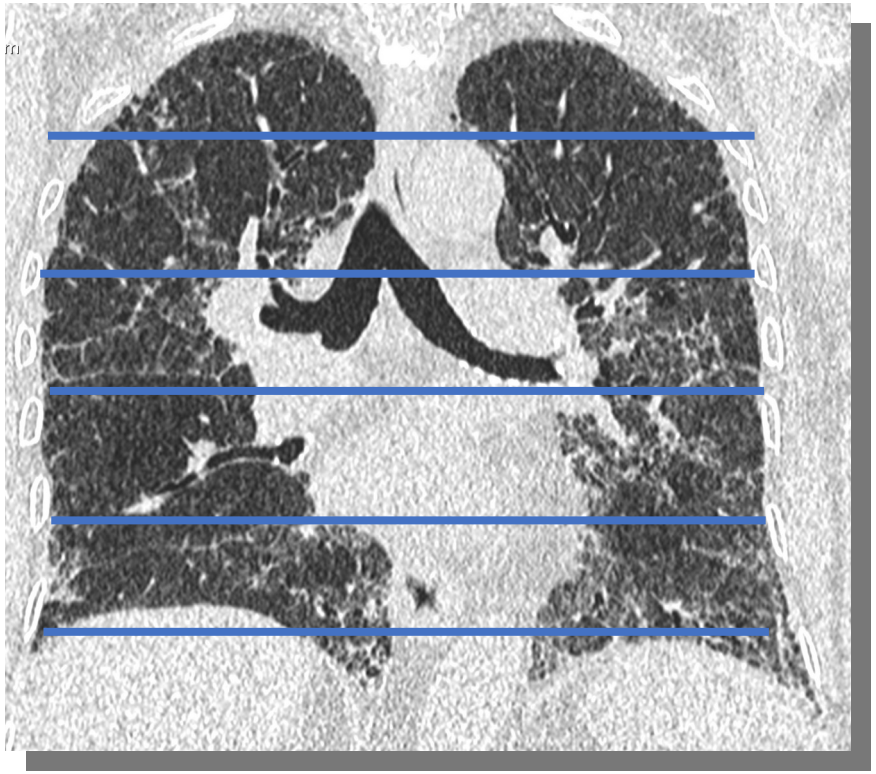
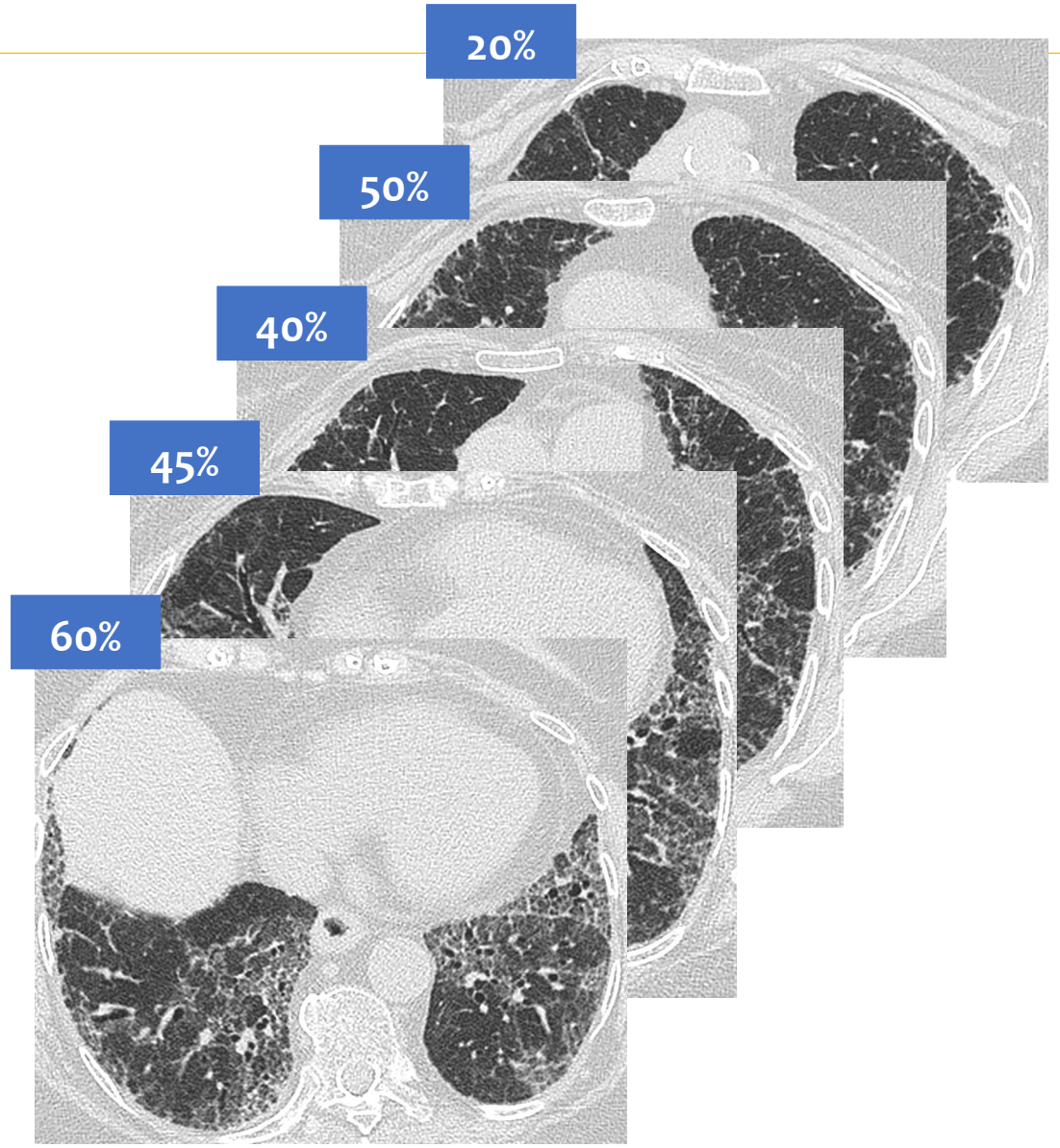
Semi-quantitative score





(20 + 50 + 40 + 45 + 60)/ 5 = 43%

Best AC et al, Radiology 2008;246(3): 935–940

Jacob J et al, J Thoracic Imaging 2016;31:304–311

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One or more of:

A. Increased extent or severity of traction bronchiectasis and bronchiolectasis
B. New ground-glass opacity with traction bronchiectasis
C. New fine reticulation
D. Increased extent or increased coarseness of reticular abnormality
E. New or increased honeycombing
F. Increased lobar volume loss

1 Worsening respiratory symptoms

2 Physiological evidence of disease progression

3 Radiological evidence of disease progression

Raghu G et al. Am J Respir Crit Care Med 2022;205:e18–47

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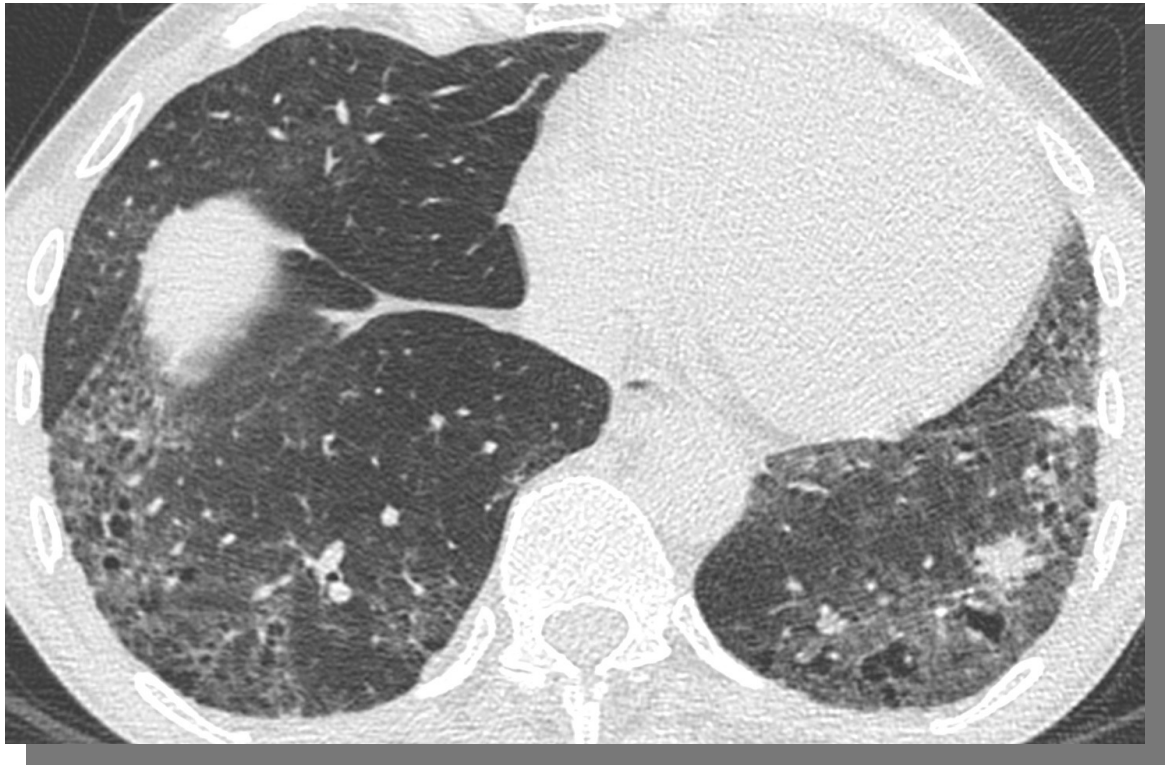
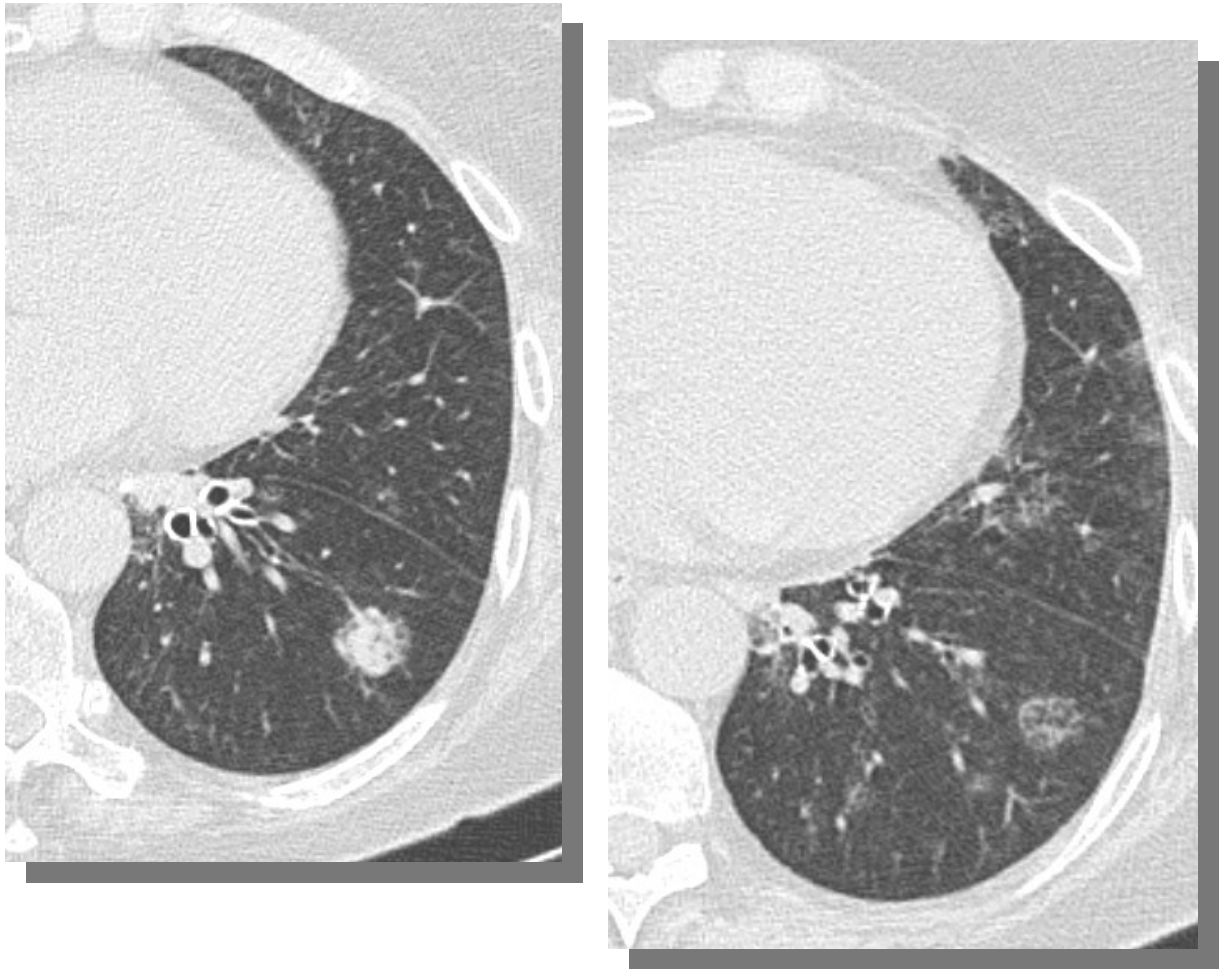
Complications



Adenocarcinoma in SSc



LNH in SS

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Key points

- To become familiar with patterns/abnormalities associated with individual CTD
- Different patterns of lung disease contribute to morbidity/mortality in CTDs
- Disease behavior is difficult to predict and interpret
- Multidisciplinary approach is key

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