MEETING 16. 01. 2019

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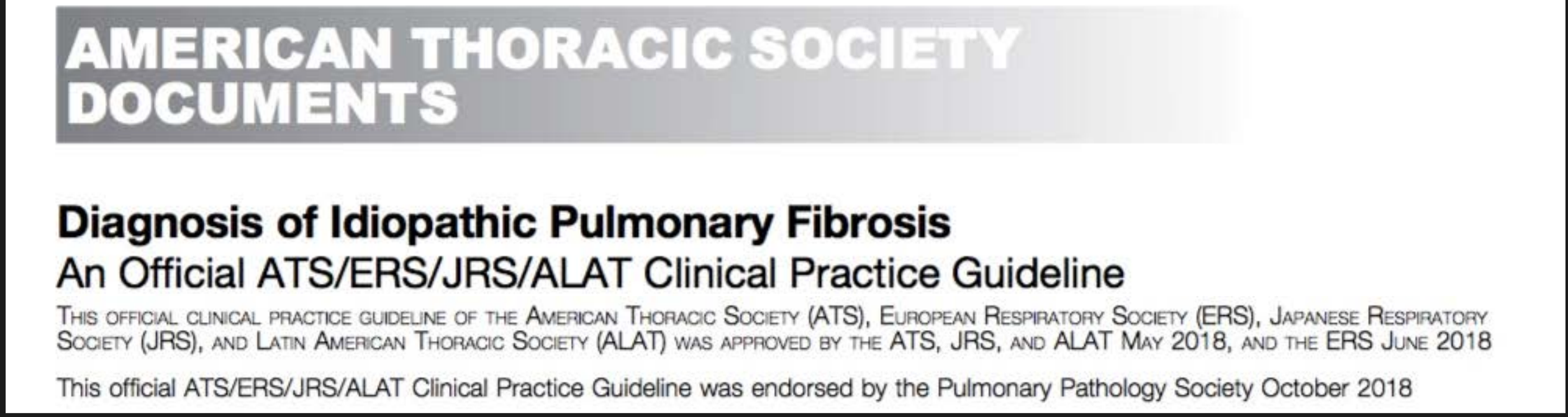
IPF: FIBROSI POLMONARE IDIOPATICA. LE NUOVE LINEE GUIDA



- Aggiornamento sulla base di un confronto ATS 2011 e Linee Guida Fleischner 2018.

- Forte bisogno di MDD
- Malattia emergente in cui la diagnosi radiologica è fondamentale
- Necessaria integrazione CLINICA-RADIOLOGICA

INQUADRAMENTO CLINICO

- Specifica forma CRONICA, PROGRESSIVA di pneumopatia fibrosante da causa sconosciuta
- Uomo adulto > 60 aa
- Dispnea insidiosa ed ingravescente, tosse, clubbing

- Prognosi infausta a causa di diagnosi clinica errata (COPD, asma, polmonite)

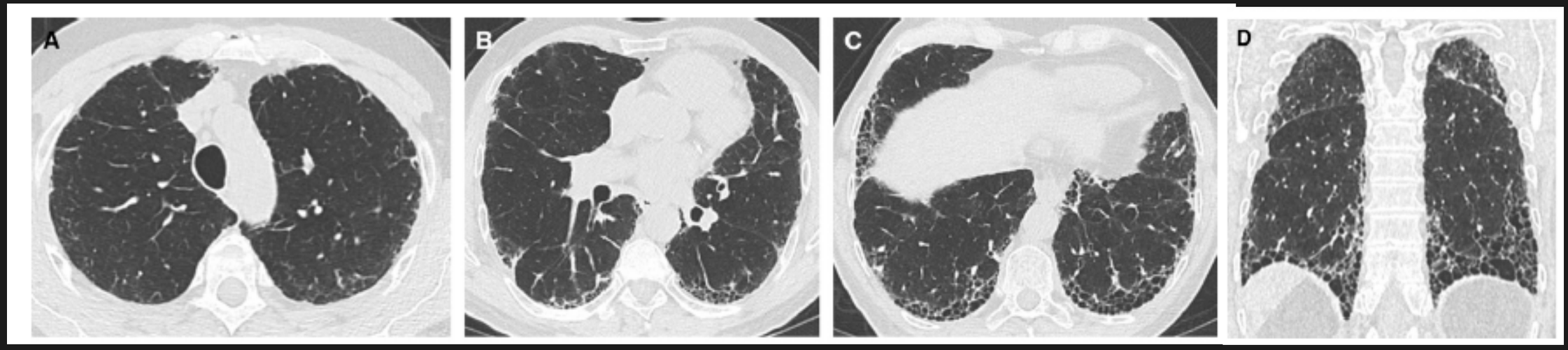
- Escludere altre cause di IPF
- Anamnesi lavorativa-occupazionale
- Storia di fumo
- Parametri di laboratorio per escludere malattie reumatologiche
- Anamnesi farmacologica
- Analisi genetiche

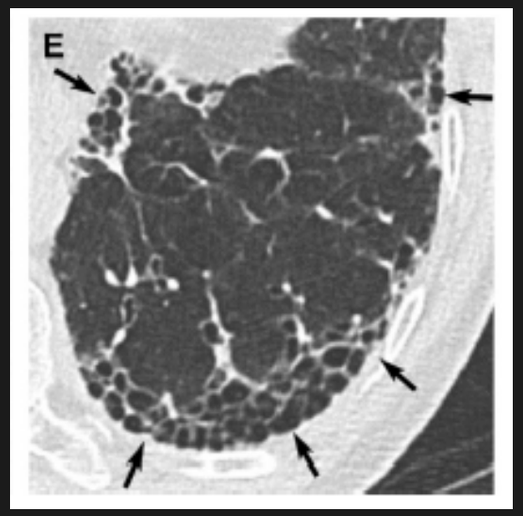
INQUADRAMENTO RADIOLOGICO: HRTC

- 4 pattern radiologici
- 90-100% VPP

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")	

HRTC: UIP PATTERN



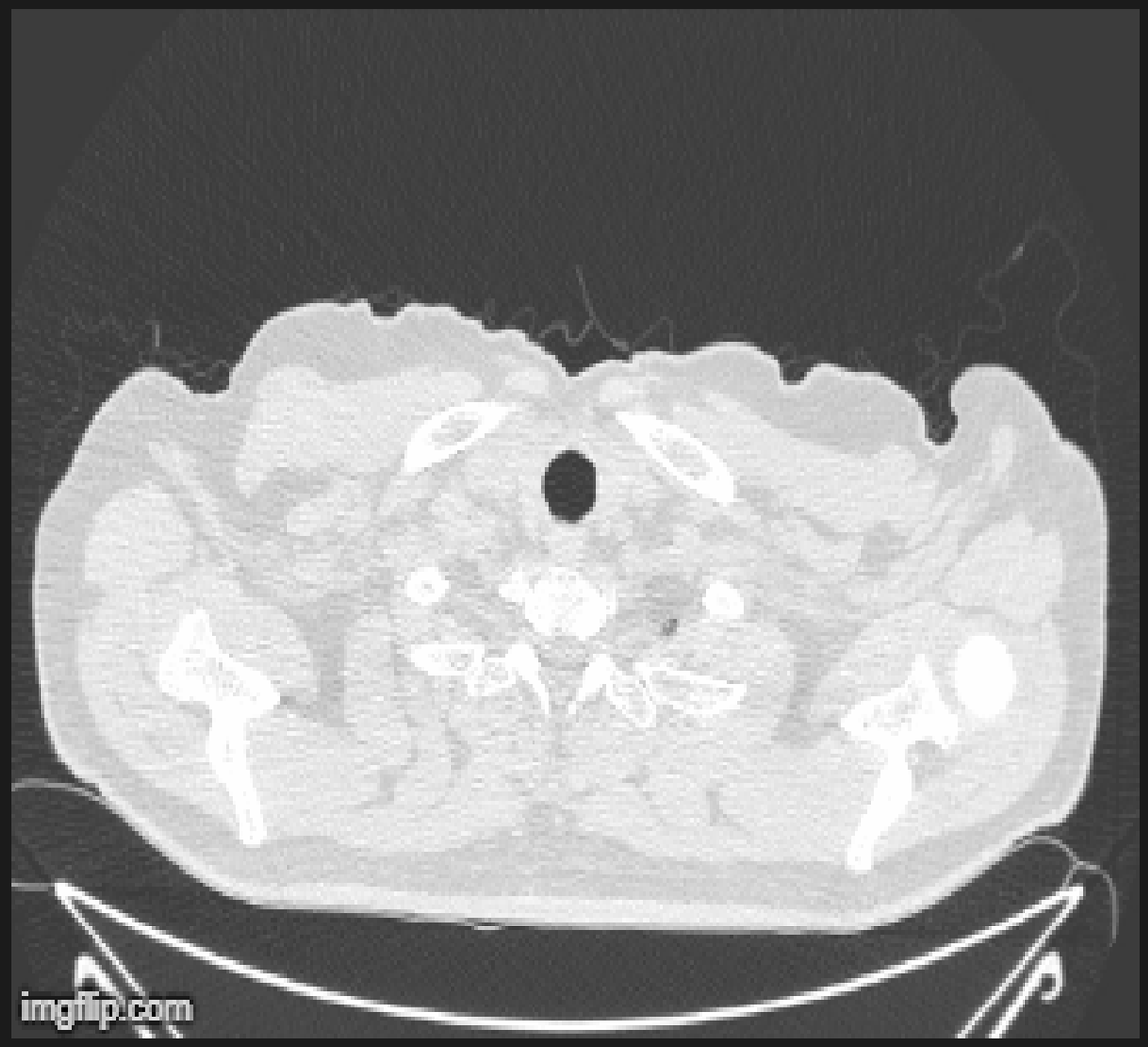


HONEYCOMBING: Cisti da bronchiolo-ostruzione, generalmente disposte in più strati, con parete spessa e ben definita. DDF: bronchiectasie da trazione.

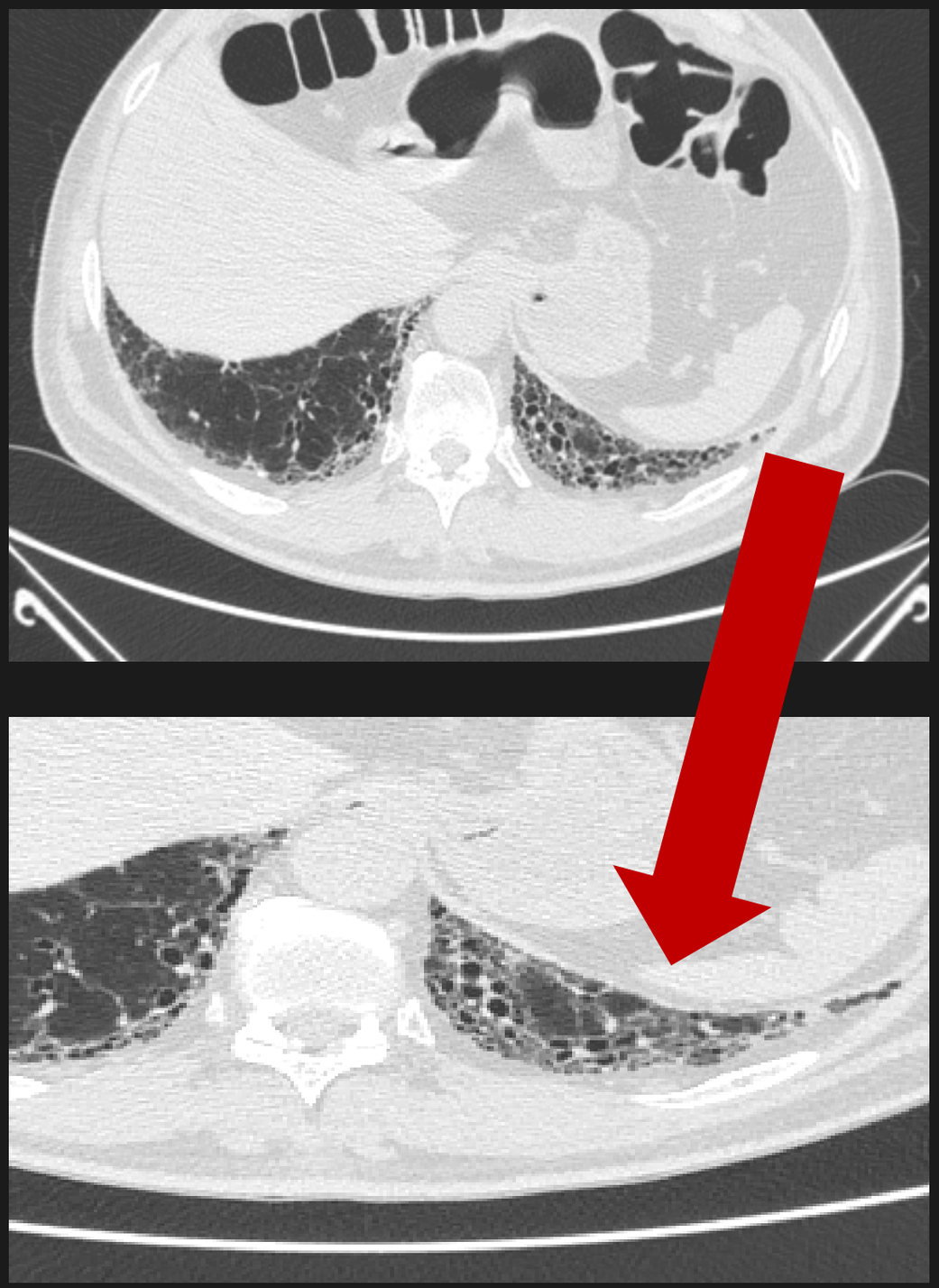
- Predominantly subpleural and basal
- Distribution is often heterogeneous and occasionally diffuse or asymmetrical
- Honeycombing, with or without peripheral traction bronchiectasis or bronchiolectasis, must be present for a definite HRCT diagnosis of UIP
- Mild GGO associated with reticular pattern. Pulmonary ossification may be present

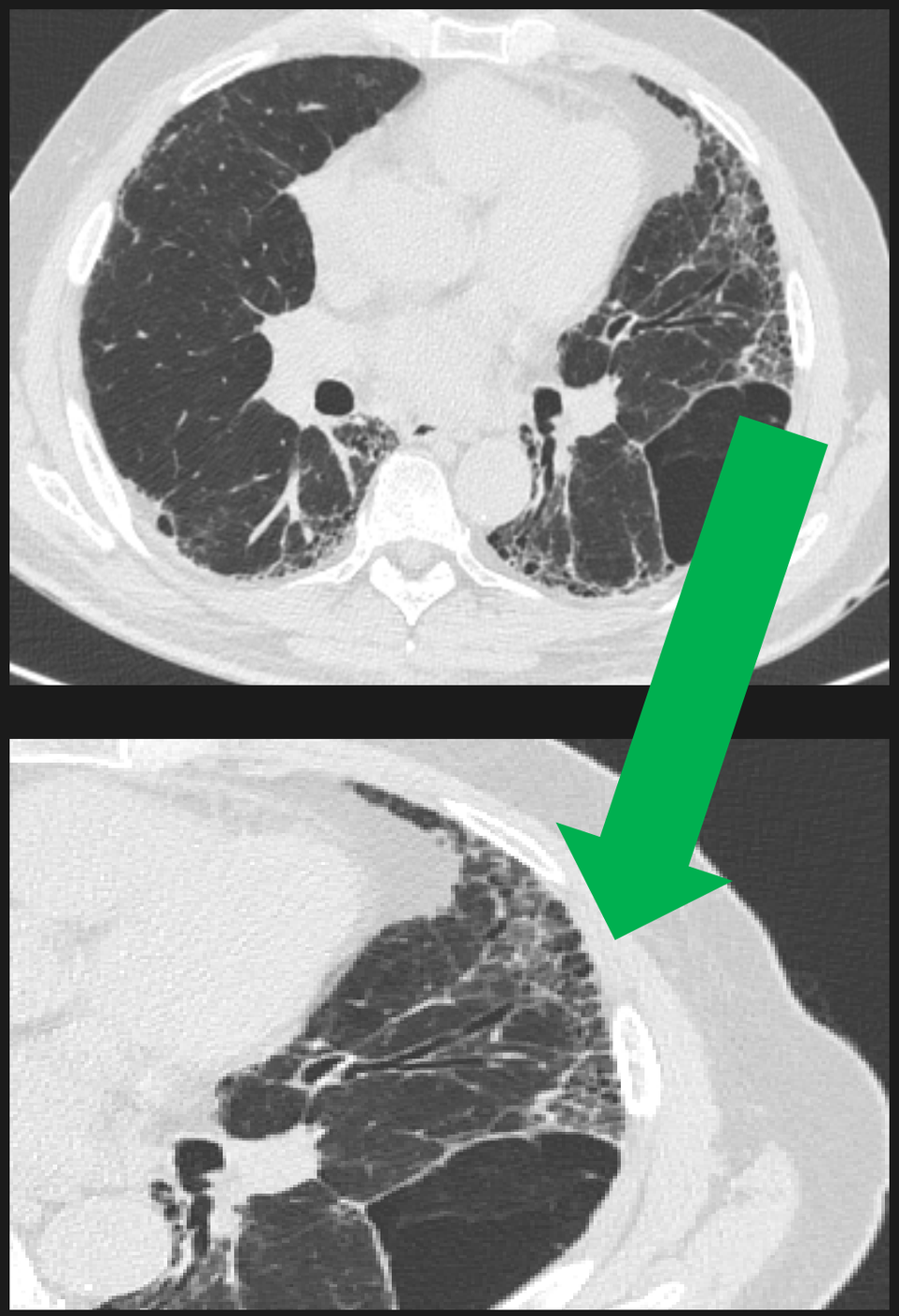
UIP PATTERN

- Uomo, 65anni
- Ex fumatore (1pack/die x 25 yrs)
- Ex vigile del fuoco, nega esposizione a farmaci ed allergeni
- Nega familiarità
- Autoimmunità negativa
- Esordio con dispnea da sforzo

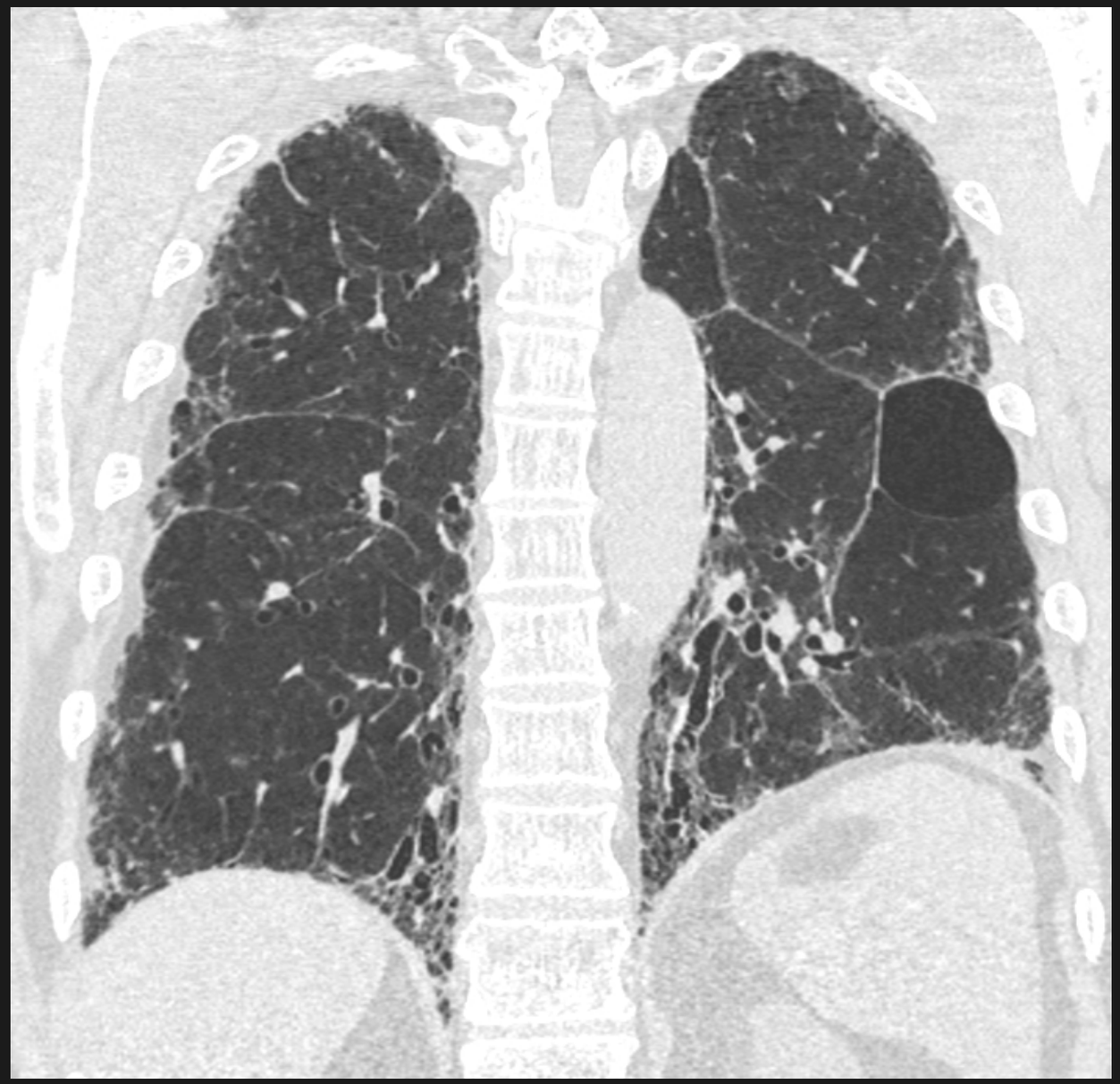


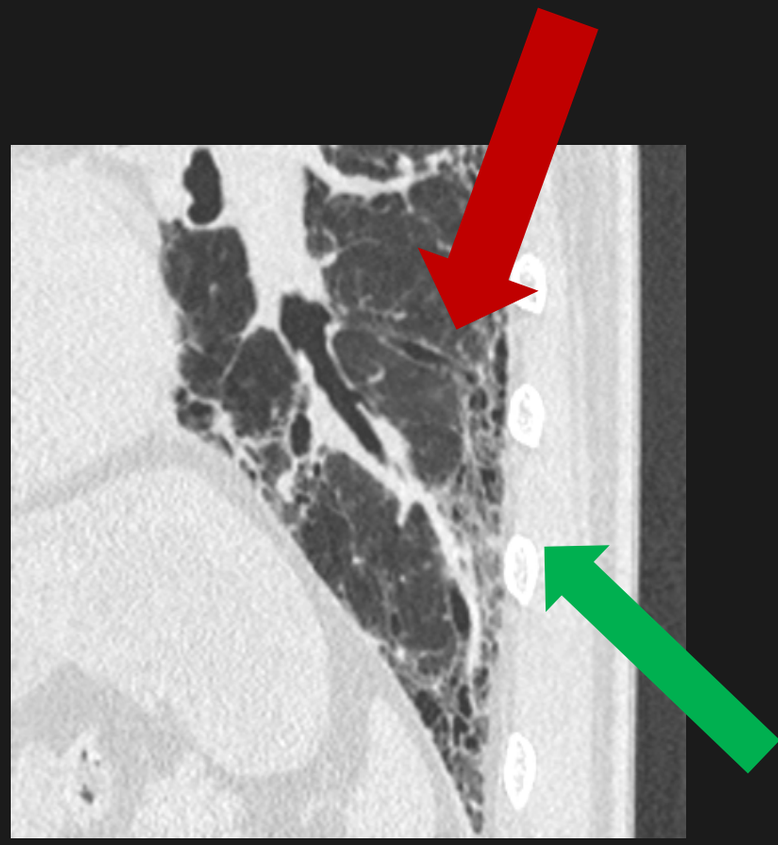
UIP PATTERN

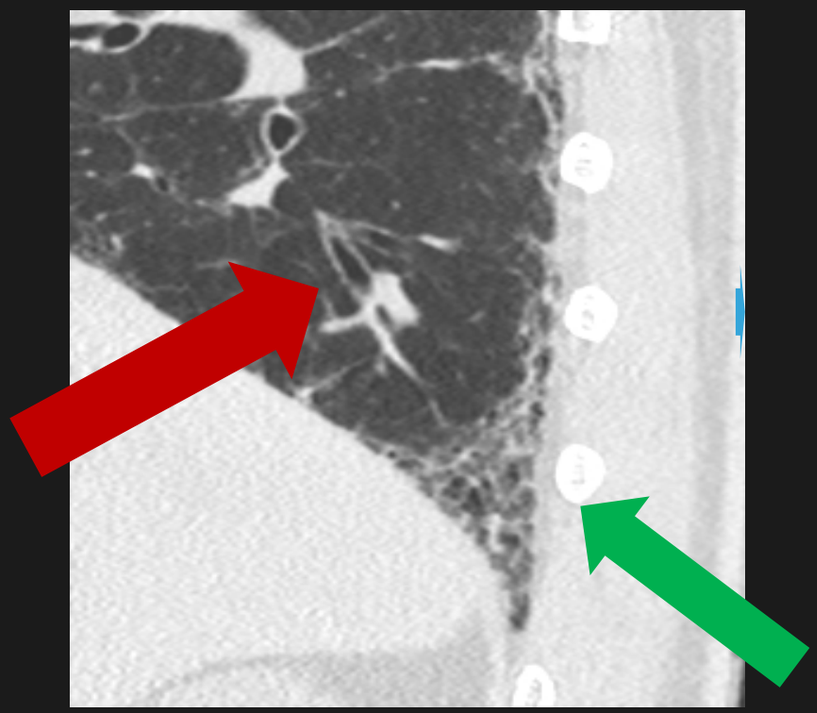




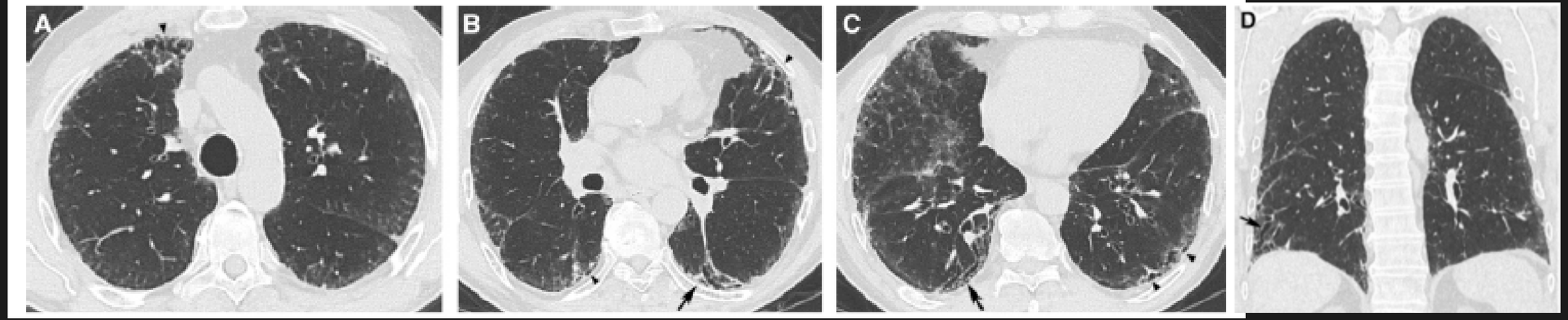
UIP PATTERN

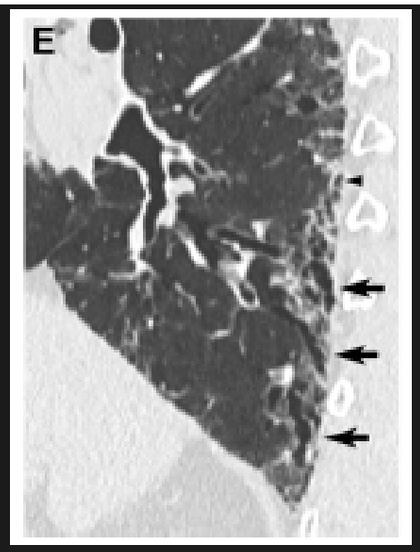






HRTC: UIP PROBABLE





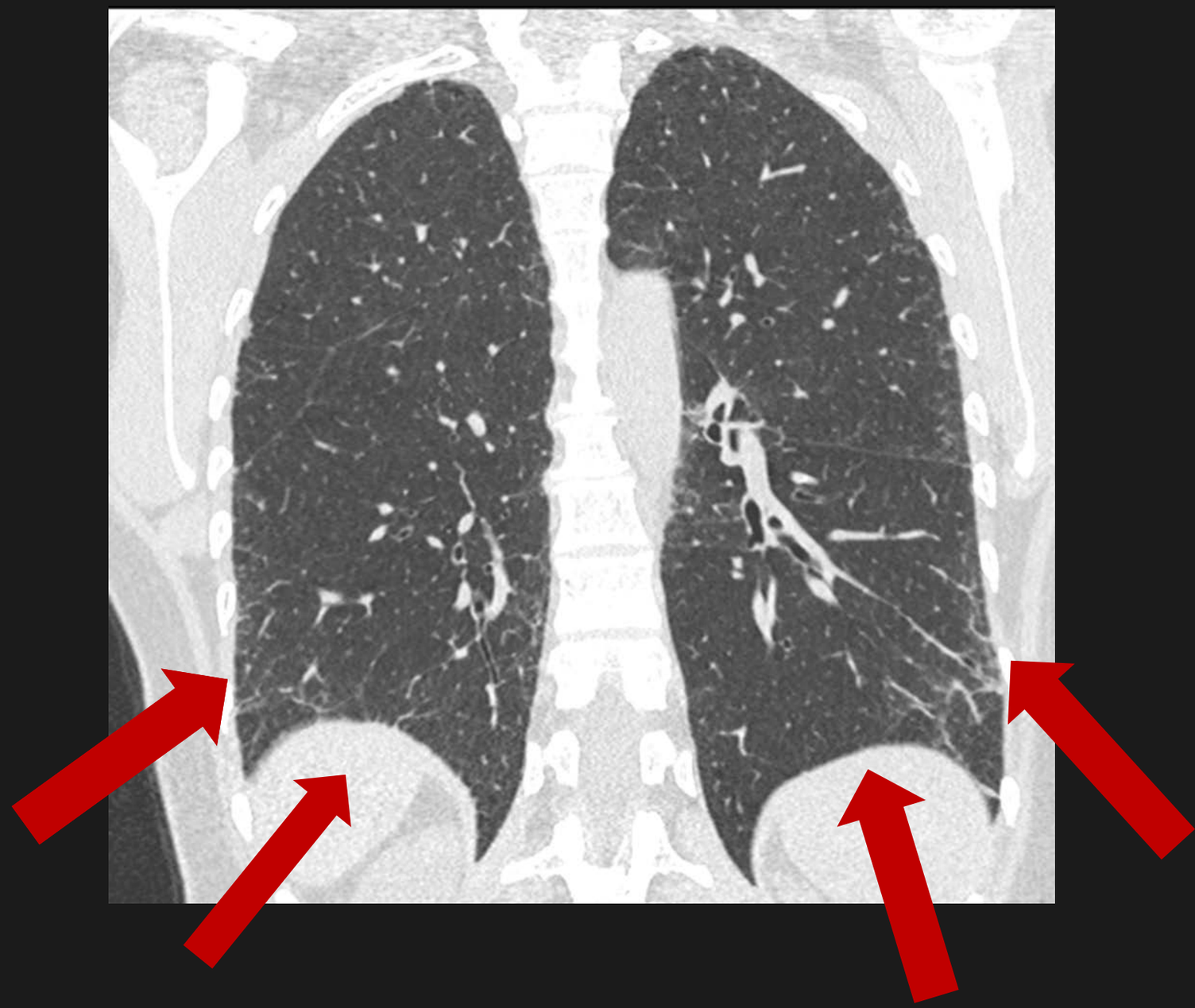
Reticular pattern: ispessimento dei setti, nel caso della fibrosi è fine ed irregolare, a differenza delle quello delle malattie interstiziali non fibrotiche in cui i setti sono più spessi e regolari

Traction bronchiectasis: dilatazione irregolare dei bronchi o dei bronchioli causata da retrazione fibrotica del parenchima.

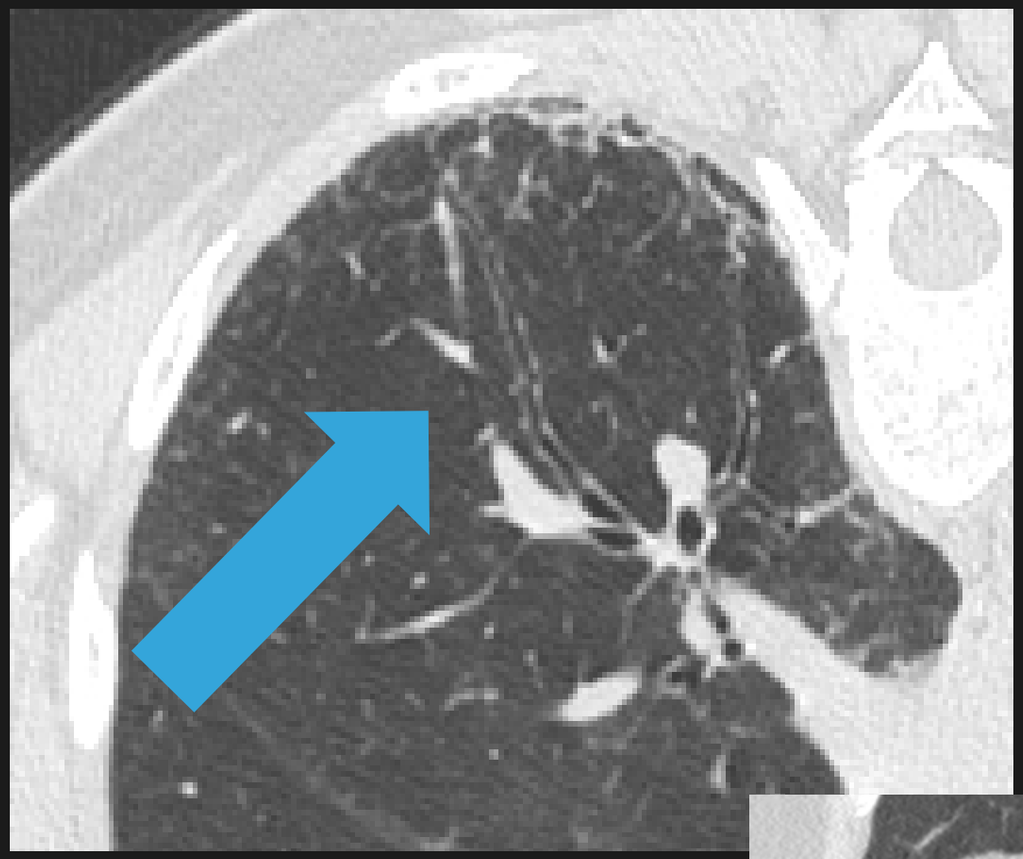
- Predominantly subpleural and basal
- Often heterogeneous distribution
- Reticular pattern with peripheral traction bronchiectasis or bronchiolectasis
- May have mild GGO

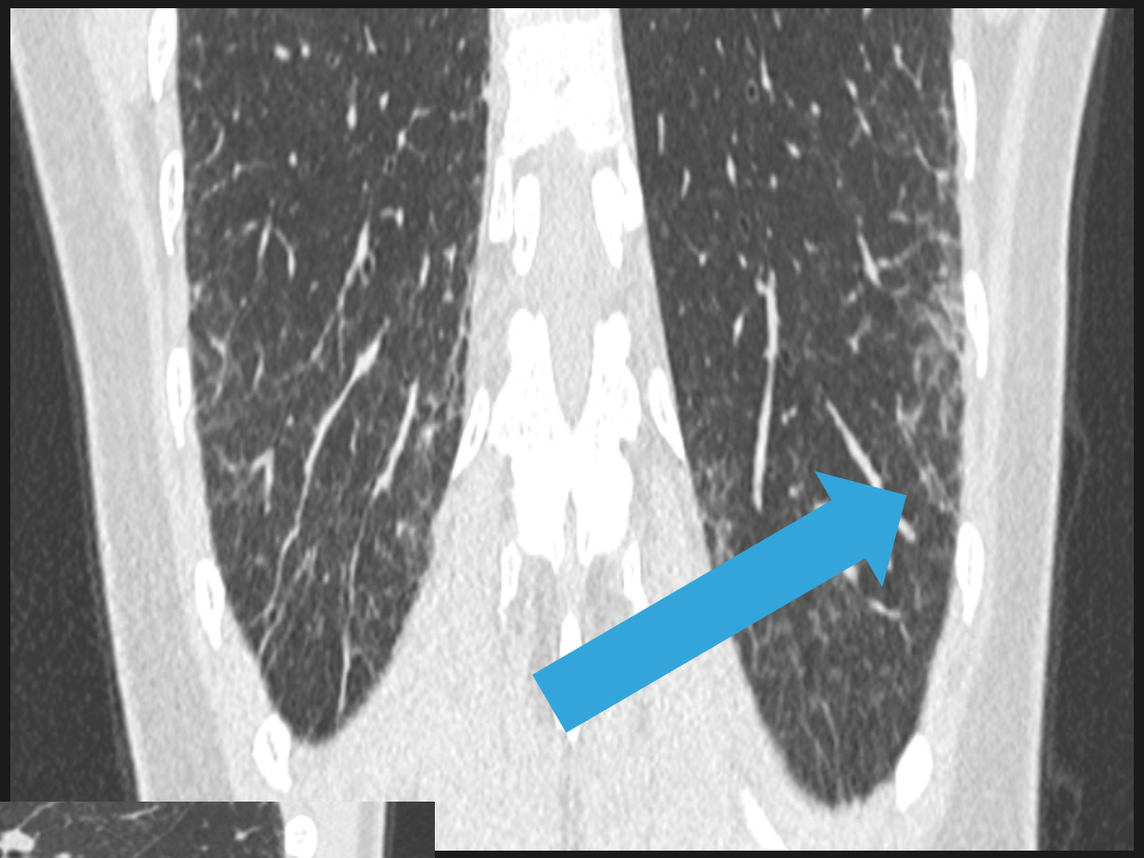
UIP PROBABLE

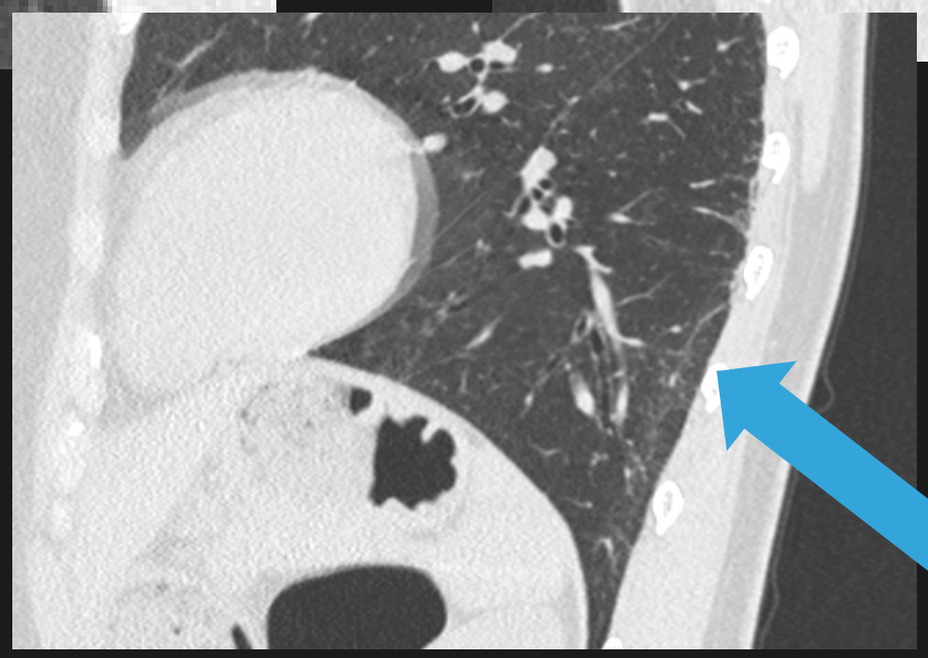
- Donna, 52 anni
- Familiarità per IPF
- Infermiera. Nega esposizione a sostanze tossiche, allergeni, farmaci, nega fumo
- Autoimmunità negativa
- Asintomatica



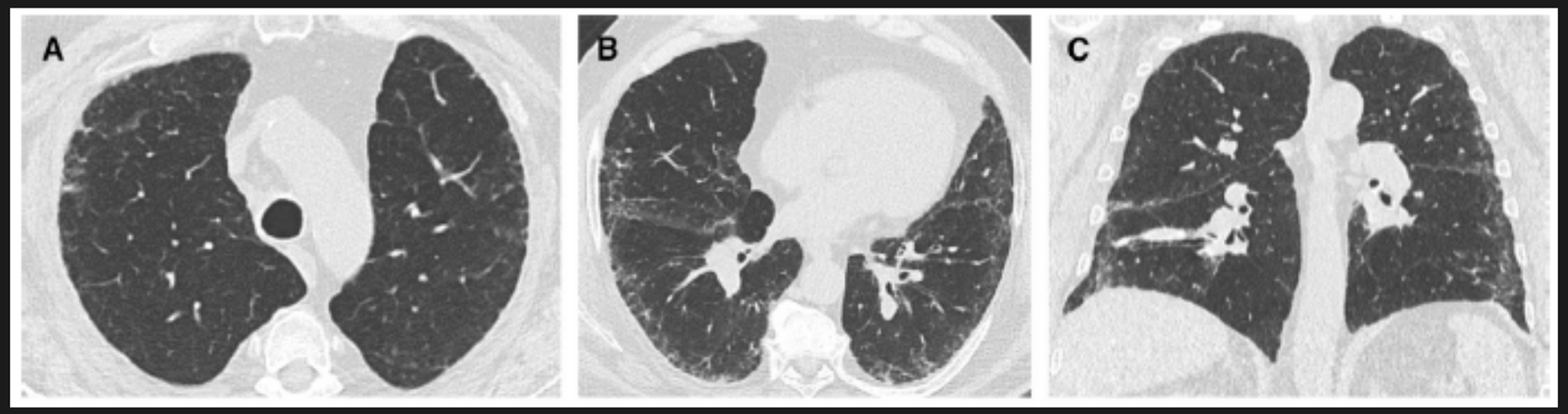
UIP PROBABLE

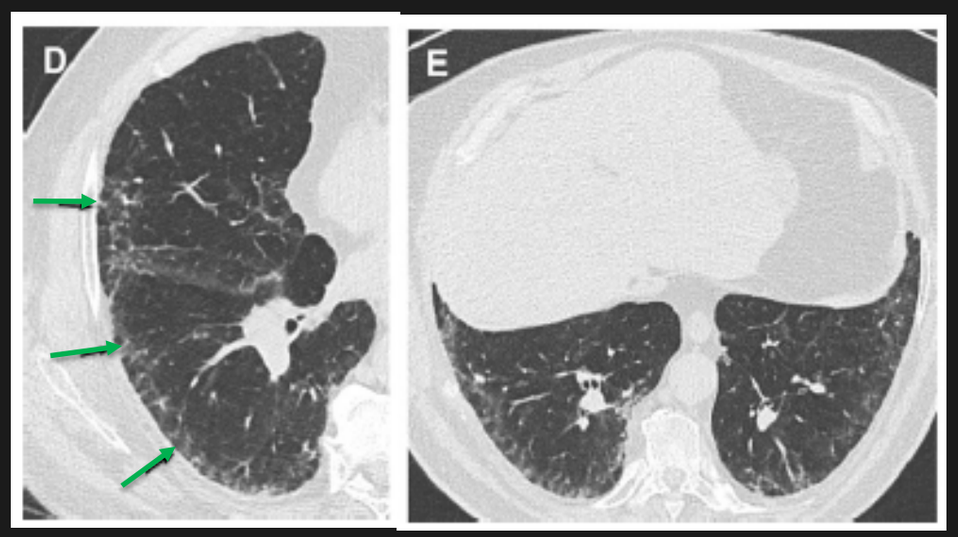






HRTC: UIP INDETERMINATE

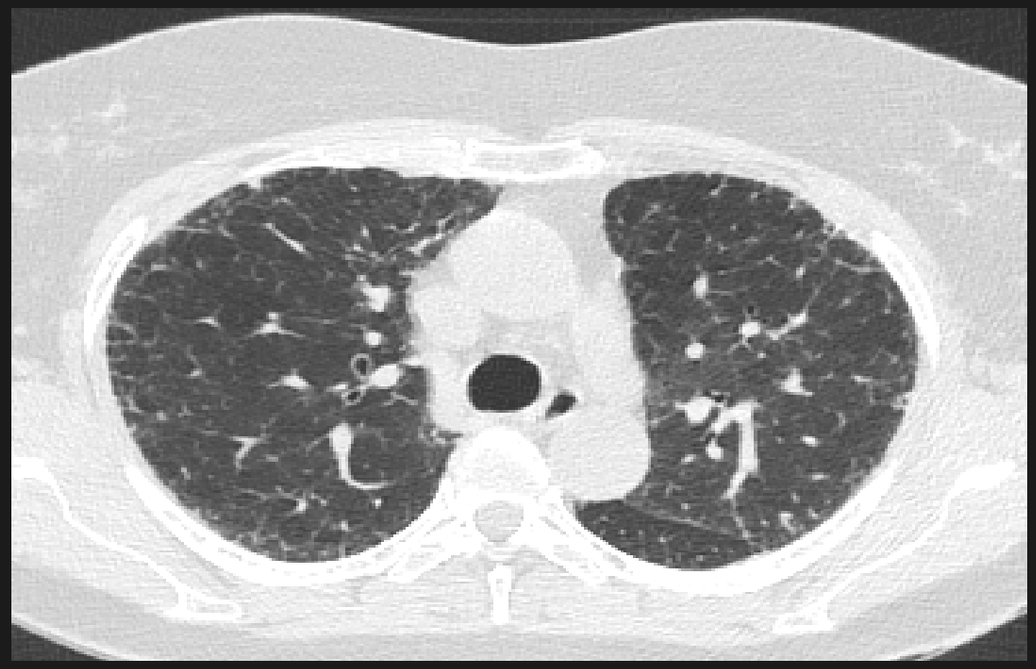


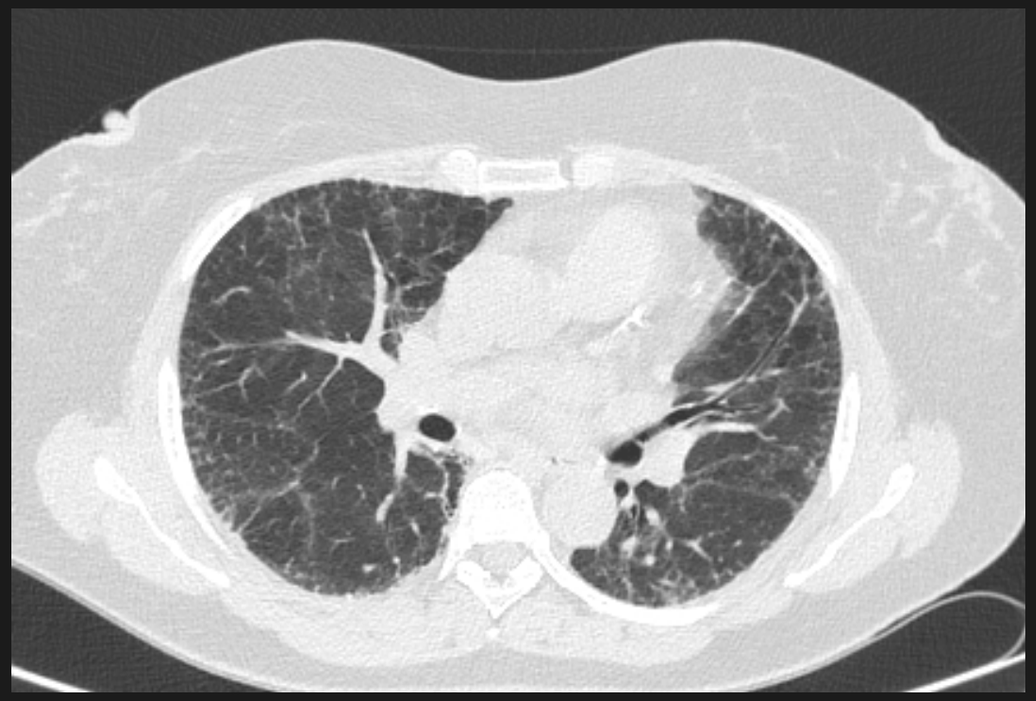


- Predominantly subpleural and basal
- Subtle reticulation
- Mild GGO or distortion may be present
- The CT features and/or the distribution of lung fibrosis do not suggest any specific etiology (‘truly indeterminate’)

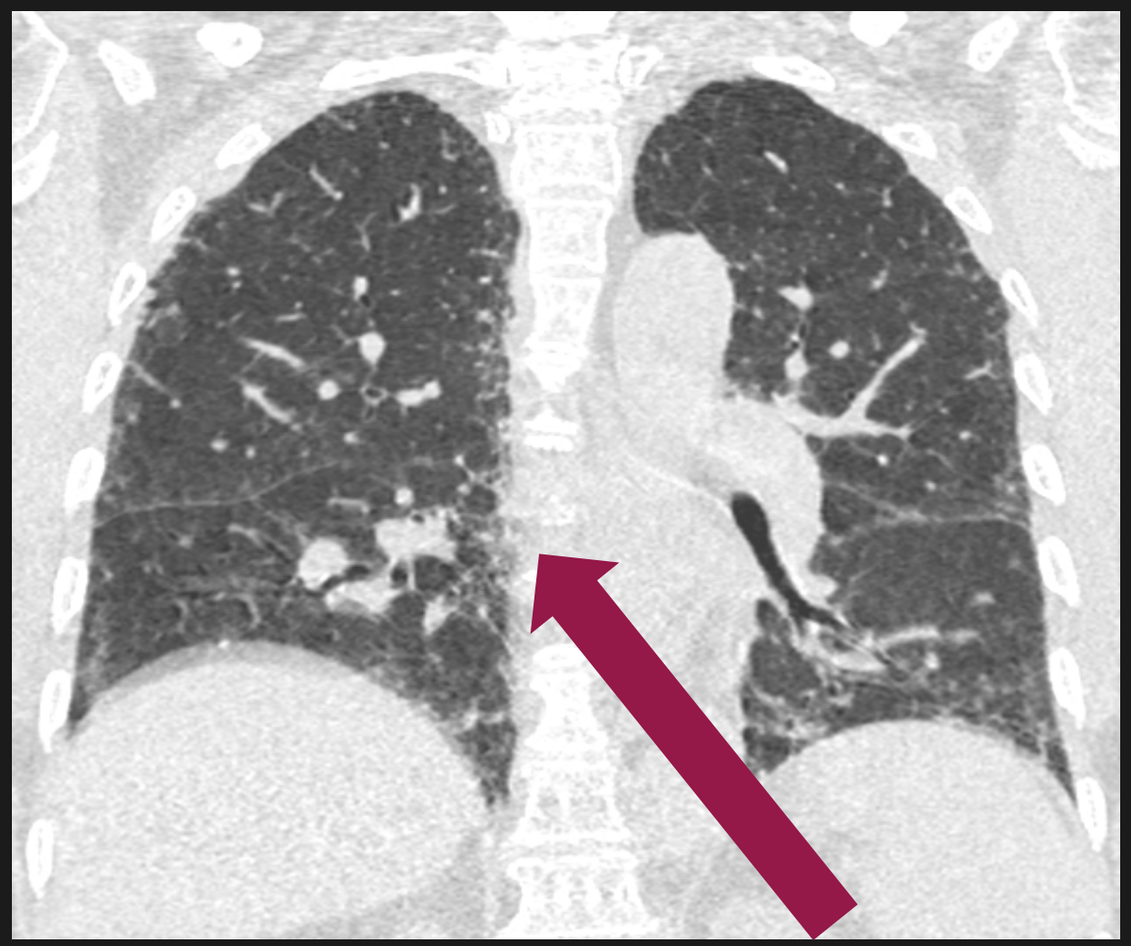
UIP INDETERMINATE

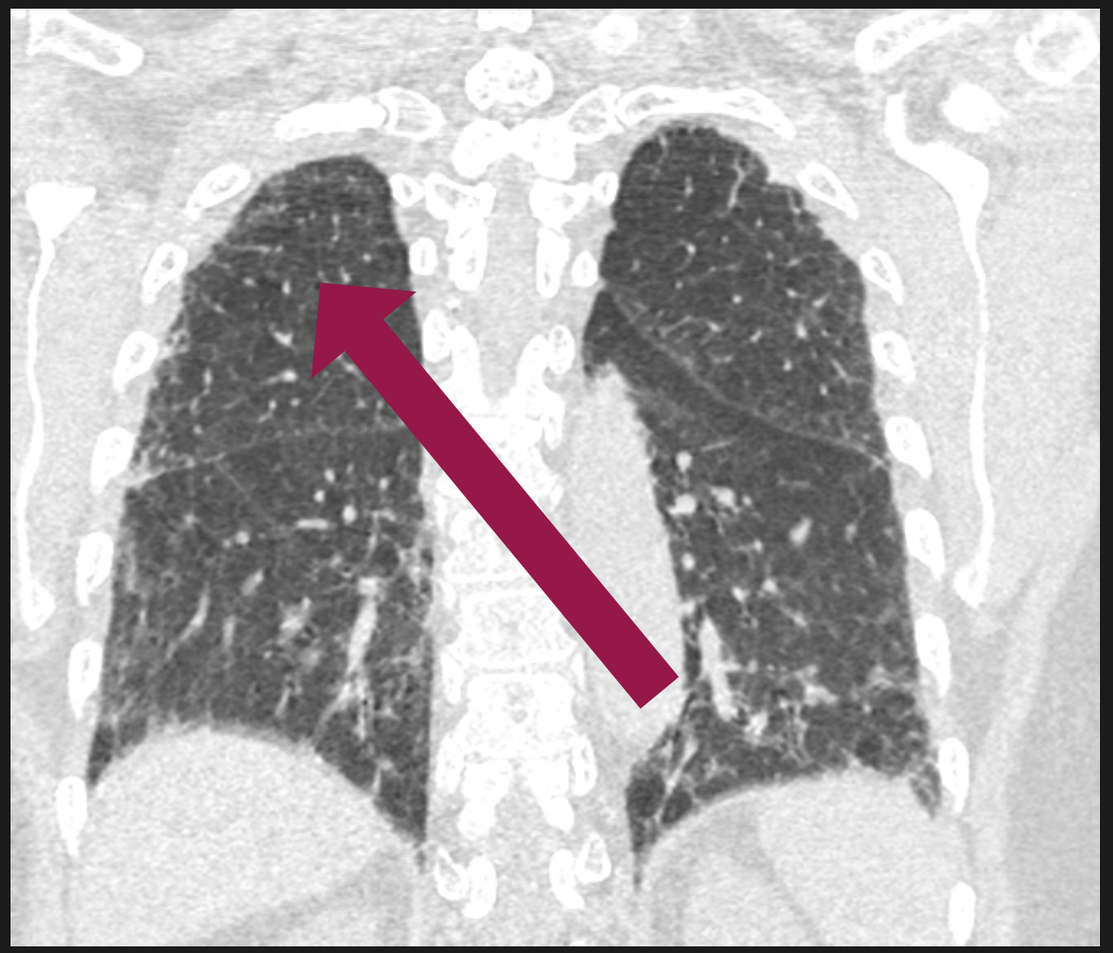
- Donna, 72 anni
- Casalinga, nega esposizione a sostanze tossiche, allergeni, farmaci, nega fumo
- Nega familiarità
- Autoimmunità negativa ad eccezione di fattore reumatoide debolmente positivo
- Esordio con febbricola e tosse resistente a terapia antibiotica



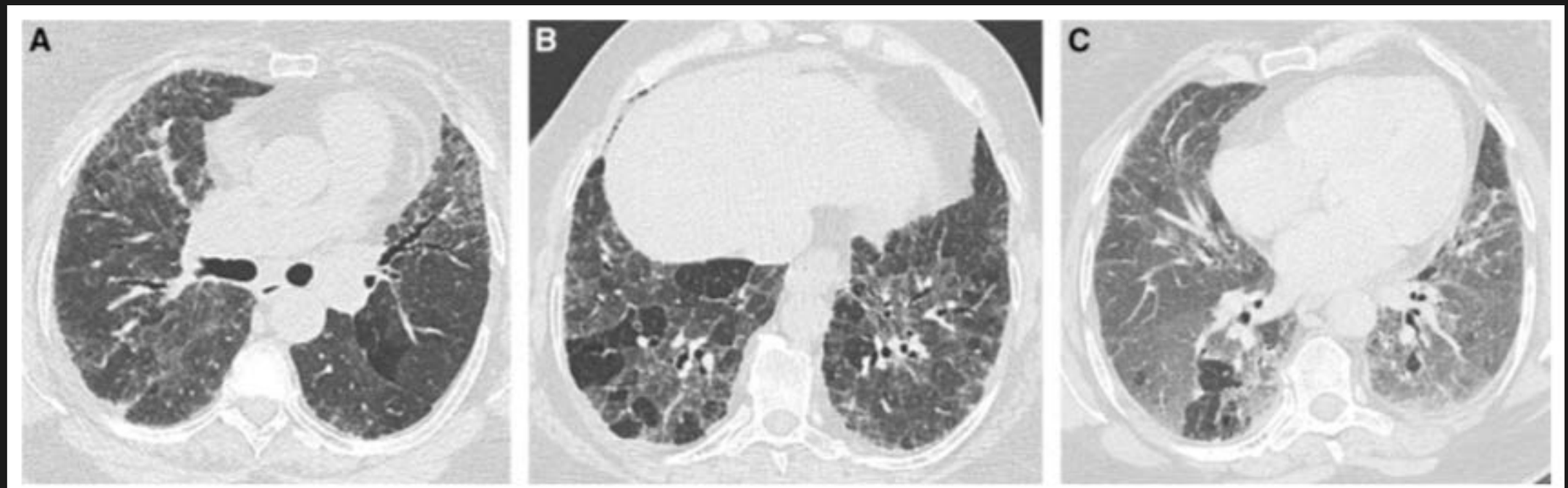


UIP INDETERMINATE





HRTC: NON UIP/ ALTERNATIVE DIAGNOSIS



CT features:

- Cysts
- Marked mosaic attenuation
- Predominant GGO
- Profuse micronodules
- Centrilobular nodules
- Nodules
- Consolidation

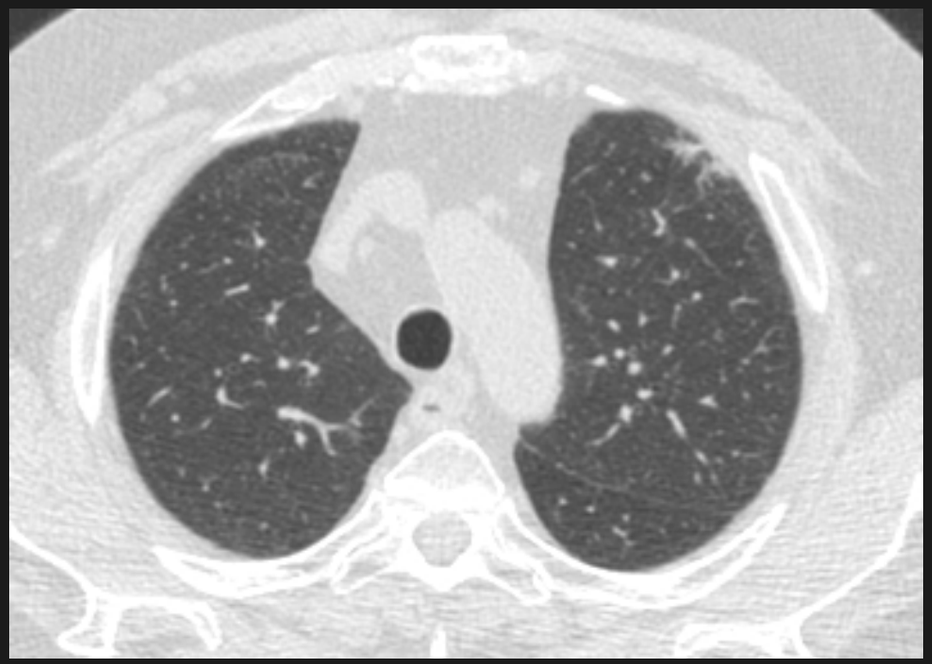
Predominant distribution:

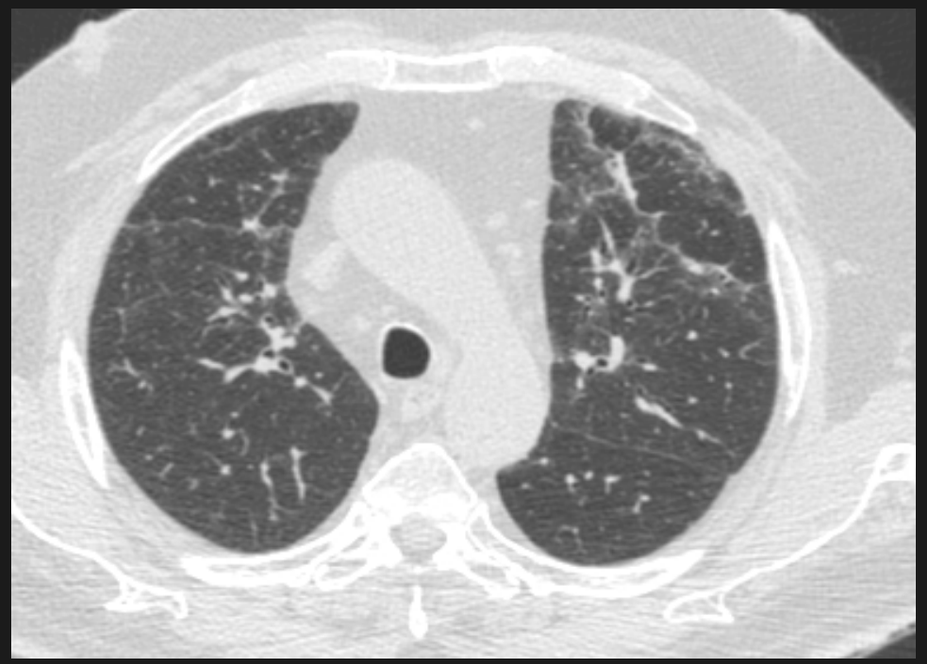
- Peribronchovascular
- Perilymphatic
- Upper or mid lung

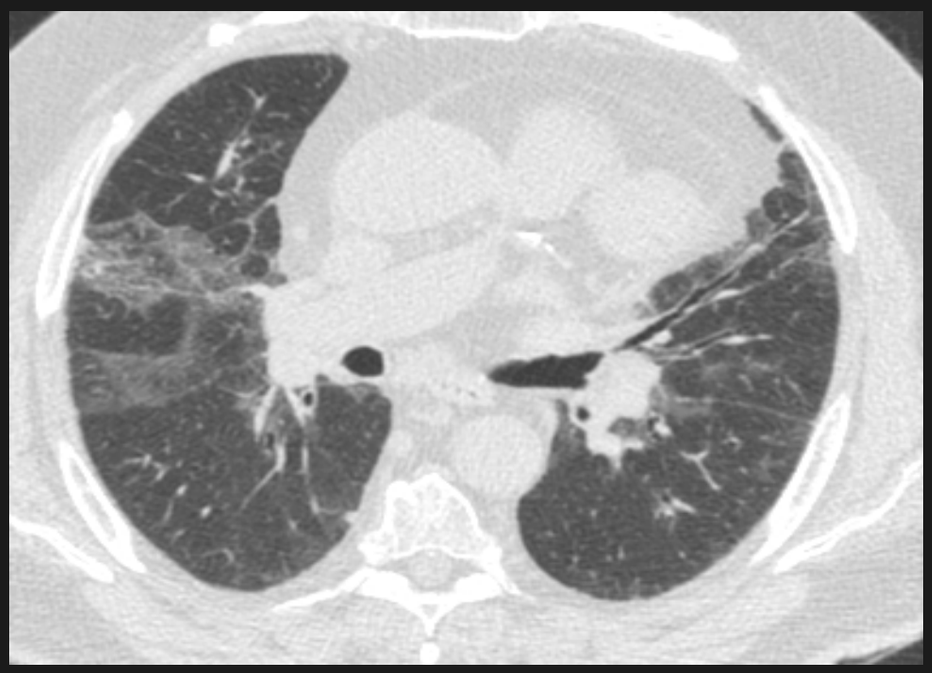
Other:

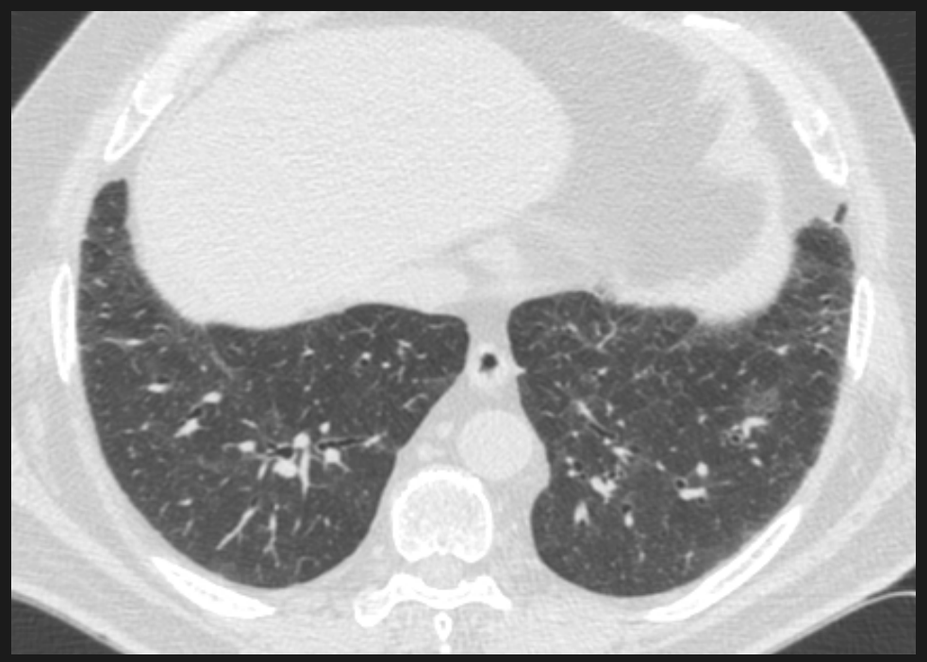
- Pleural plaques (consider asbestosis)
- Dilated esophagus (consider CTD)
- Distal clavicular erosions (consider RA)
- Extensive lymph node enlargement (consider other etiologies)
- Pleural effusions, pleural thickening (consider CTD/drugs)

ALTERNATIVE DIAGNOSIS







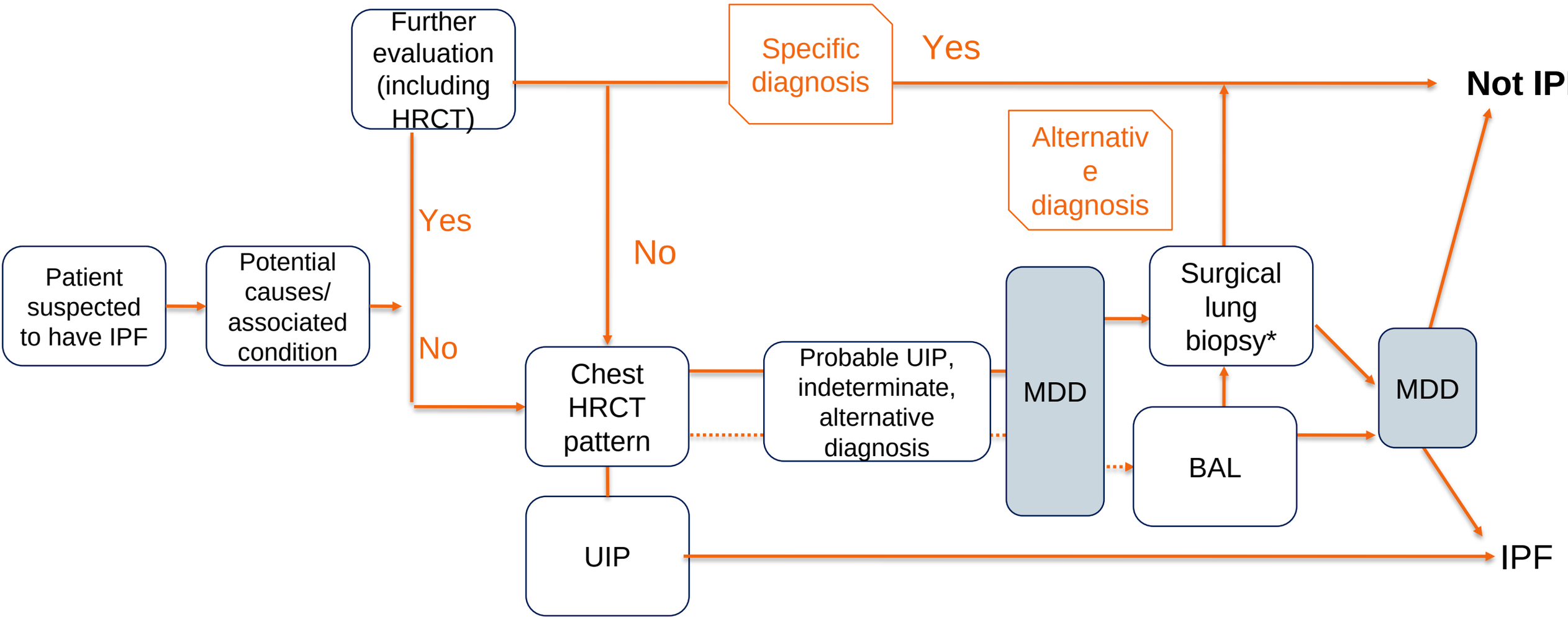


Diagnostic algorithm for IPF

Excluding other causes of ILD

HRCT patterns

Patients categorized as probable UIP, indeterminate for UIP or alternative diagnosis at HRCT



TERAPIE

Agent	2015 guidelines
Nintedanib	Conditional recommendation **for** use*
Pirfenidone	Conditional recommendation **for** use*
Anticoagulation (warfarin)	Strong recommendation **against** use*
Combination prednisone + azathioprine + N-acetylcysteine	Strong recommendation **against** use†
Selective endothelin receptor antagonist (ambrisentan)	Strong recommendation **against** use†
Imatinib, a tyrosine kinase inhibitor with one target	Strong recommendation **against** use*
Dual endothelin receptor antagonists (macitentan, bosentan)	Conditional recommendation **against** use†
Phosphodiesterase-5 inhibitor (sildenafil)	Conditional recommendation **against** use*
Antiacid therapy	Conditional recommendation **for** use‡
N-acetylcysteine monotherapy	Conditional recommendation **against** use†
Antipulmonary hypertension therapy for idiopathic pulmonary fibrosis-associated pulmonary hypertension	Reassessment of the previous recommendation was deferred
Lung transplantation: single vs bilateral lung transplantation	Formulation of a recommendation for single vs bilateral lung transplantation was deferred

Importanza di conoscere quali sono i farmaci fortemente SCONSIGLIATI, come Prednisone, Azatioprina, N-acetilcisteina.

TAKE HOME MESSAGE

SCHEDA RACCOLTA DATI PER PATOLOGIE INFILTRATIVE DIFFUSE POLMONARI

Nome_____Cognome_____

Data di nascita: _____ Sesso: F ☐ M ☐

ANAMNESI

Fumatore ☐ N° pacchetti al giorno: Da (età inizio):_____

Ex fumatore (>3 aa) ☐ Da (età di interruzione): _____

Non fumatore ☐

Attività lavorativa (specificare mansione) _____

Esposizione a (specificare): Silice ☐ Asbesto ☐ Metalli duri _____

Sostanze inorganiche _____Sostanze organiche _____

Assunzione farmaci (specificare quali): _____

Assunzione sostanze di abuso (specificare quali) _____

Infezioni respiratorie: In atto ☐ Da quanto tempo: _____

Pregresse ☐

Diatesi allergica: No ☐

Si ☐ Specificare: _____

Malattie del connettivo e/o autoimmuni note: No ☐

Si ☐ Specificare: _____

Altre malattie note: No ☐

Si ☐ Specificare: _____

Sintomatologia attuale: No ☐

Si ☐ Specificare: _____

Familiarità: No☐

Si☐ Specificare _____

Eventuali diagnosi istologiche precedenti: No ☐

Si ☐ Specificare: _____

FUNZIONALITA' RESPIRATORIA

Normale ☐

Alterata: ☐ Ostruttiva ☐ Restrittiva ☐ Mista ☐

Specificare: TLC (% pred):_____ DLCO (%):_____ PaO2 (mmHg):_____

FVC (%pred): _____ FEV1 (%pred): _____ FEV1/FVC (%): _____

BIBLIOGRAFIA

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 L. F. 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

This official ATS/ERS/JRS/ALAT Clinical Practice Guideline was endorsed by the Pulmonary Pathology Society October 2018

Review

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