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Fibrosis Pulmonar Idiopática: qué cambió en 2018?

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Estructura:

- Neumonias intersticiales idiopáticas / FPI
- Guía de práctica clínica FPI
  - Principales diferencias 2011 / 2018
- Resultados de nuestro estudio
- Conclusiones

Neumonías Intersticiales idiopáticas / FPI

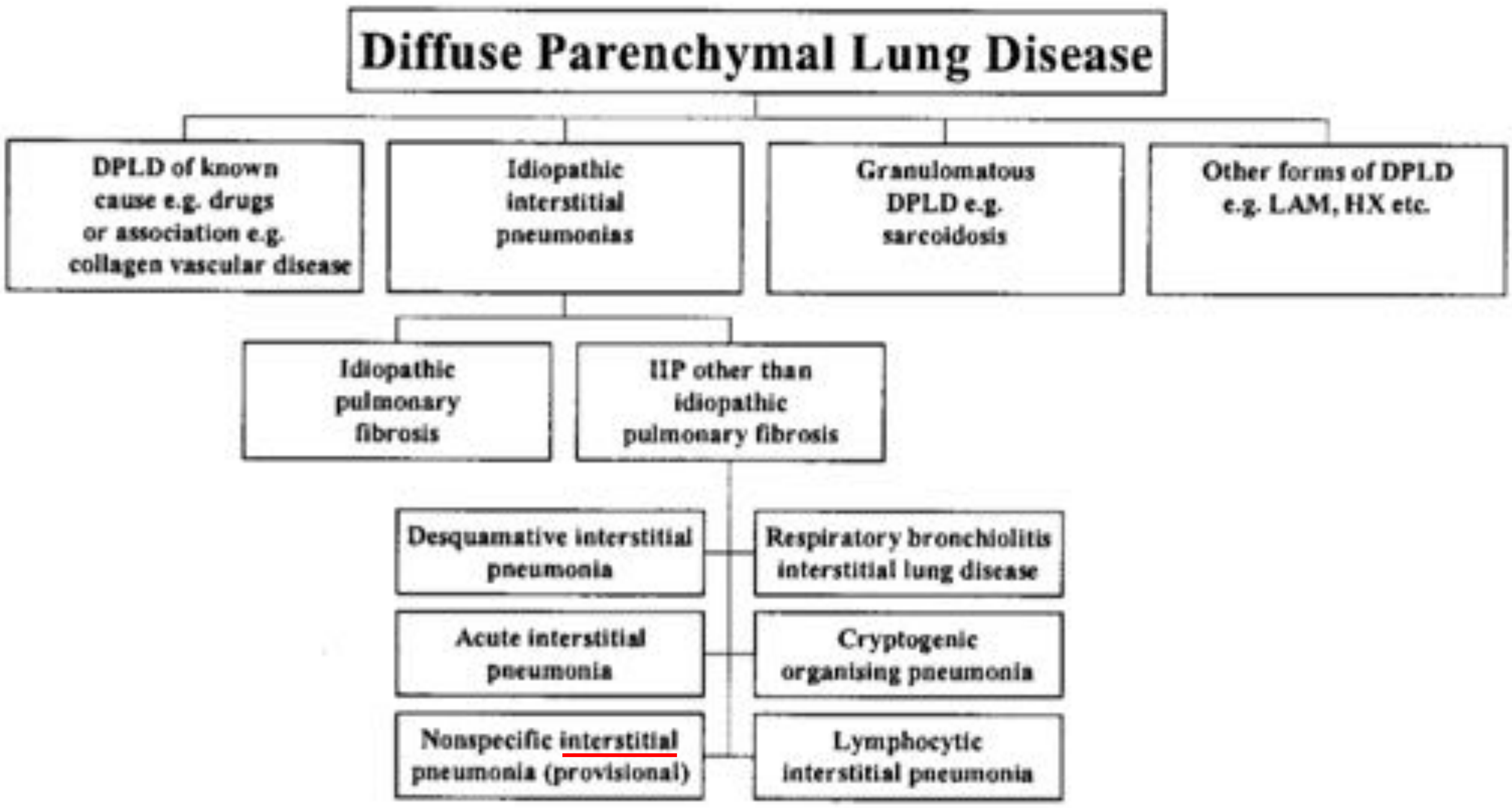
Grupo de enfermedades del parenquima pulmonar, sin etiología conocida, que comparten muchas características pero que son lo suficientemente diferentes para que las separemos como distintas enfermedades:

- NIU, la más común (50-60%).

- No NIU (NINE, Bronquiolitis-intersticial respiratoria, Neumonia intersticial descamativa, Neumonia organizada criptogenética, Neumonia intersticial aguda, Neumonia intersticial linfoide, Fibroelastosis pleuroparenquimatosa idiopatica)

Excluir: enfermades del colágeno, neumonitis por hipersensibilidad, toxicidad farmacológica, enfermedades profesionales.

Neumonías Intersticiales idiopáticas / FPI



ATS, 2002

Neumonías Intersticiales idiopáticas / FPI

TABLE 1. REVISED AMERICAN THORACIC SOCIETY/EUROPEAN RESPIRATORY SOCIETY CLASSIFICATION OF IDIOPATHIC INTERSTITIAL PNEUMONIAS: MULTIDISCIPLINARY DIAGNOSES

Major idiopathic interstitial pneumonias	
Idiopathic pulmonary fibrosis	**Fibróticas crónicas**
Idiopathic nonspecific interstitial pneumonia	
Respiratory bronchiolitis–interstitial lung disease	**Relacionadas con tabaquismo**
Desquamative interstitial pneumonia	
Cryptogenic organizing pneumonia	**Agudas / subagudas**
Acute interstitial pneumonia	
Rare idiopathic interstitial pneumonias	
Idiopathic lymphoid interstitial pneumonia	
Idiopathic pleuroparenchymal fibroelastosis	
Unclassifiable idiopathic interstitial pneumonias*	

ATS, 2013

Fibrosis Pulmonar Idiopática (FPI):

- Síndrome clínico asociado a NIU. Todas las FPI tienen histologia NIU pero no todas las NIU son FPI.

- 60 / 1.000.000 casos/hab. Hombres >50 años, fumadores, >6meses de disnea y tos no productiva, crepitantes en auscultacion, patrón restrictivo en espirometria.

- Esperanza de vida 2,5-3,5 años.

- 20% de los casos son familiares (gen Muc5B, acortamiento de telomerasas).

- Factores de riesgo - reflujo gastroesofagico, infecciones viricas cronicas (EBV, VHC).

Criterios Radiológicos:

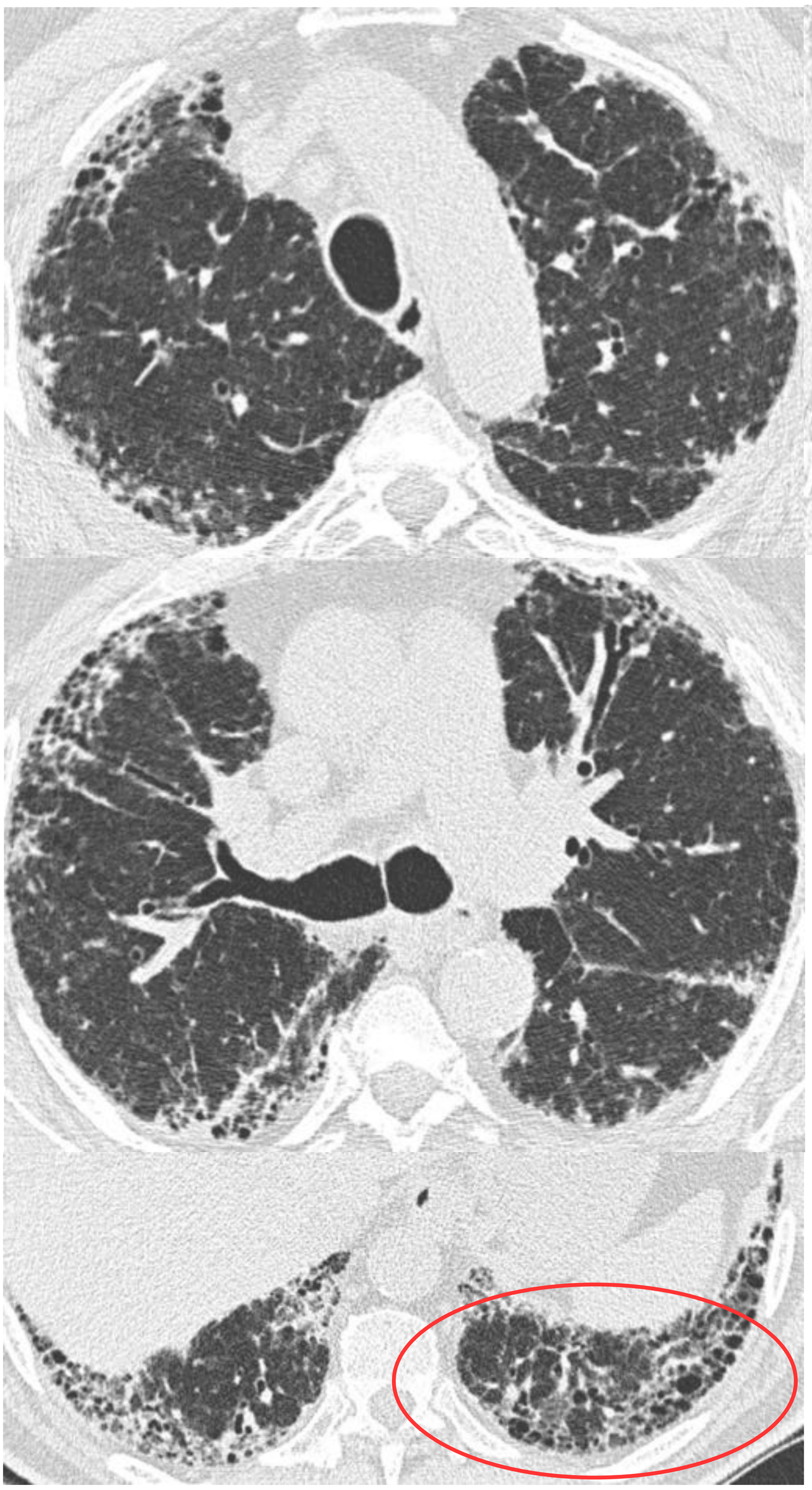
2011

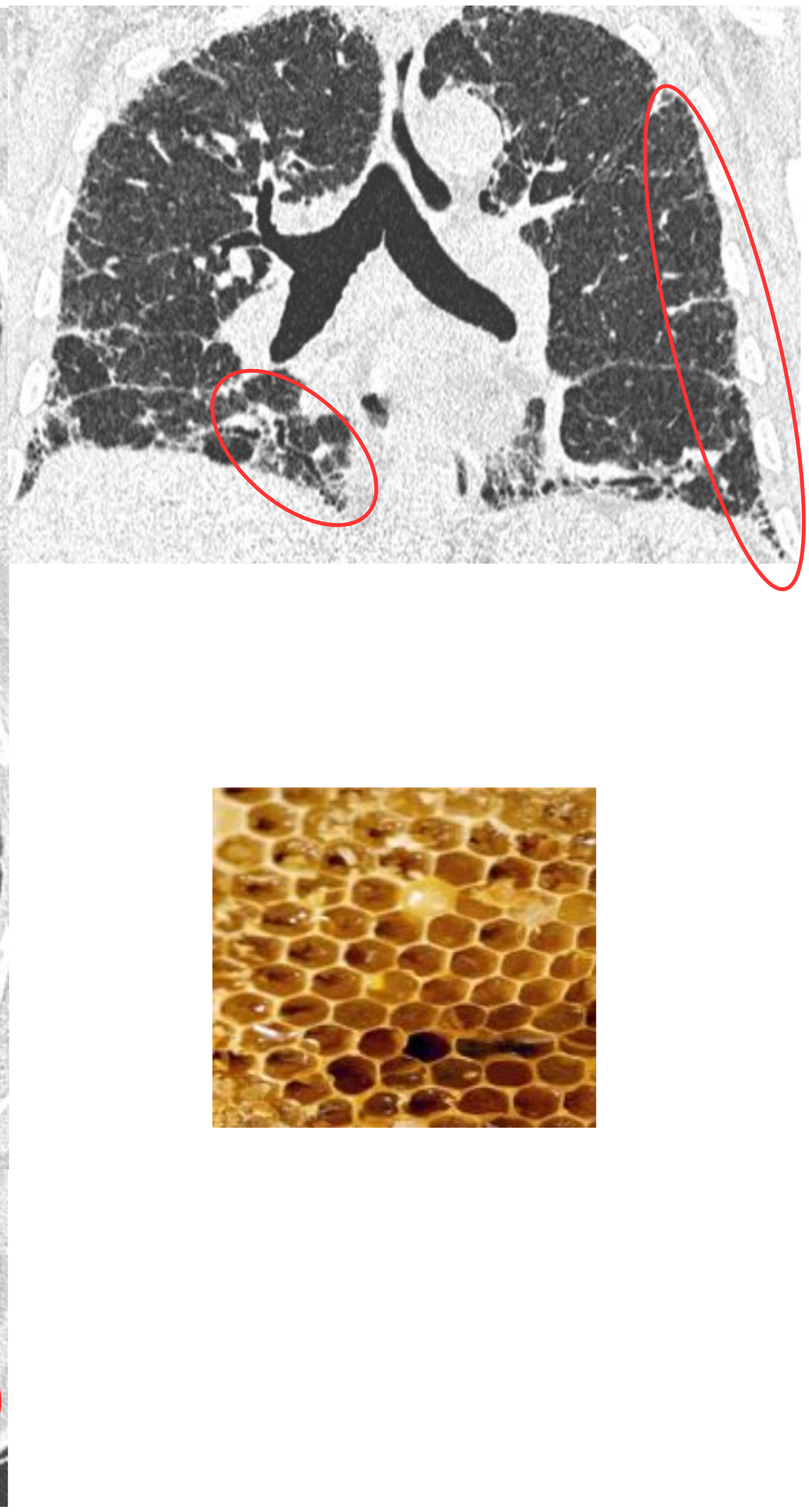
UIP Pattern (All Four Features)	Possible UIP Pattern (All Three Features)	Inconsistent with UIP Pattern (Any of the Seven Features)
• Subpleural, basal predominance • Reticular abnormality • Honeycombing with or without traction bronchiectasis • Absence of features listed as inconsistent with UIP pattern (*see* third column)	• Subpleural, basal predominance • Reticular abnormality • Absence of features listed as inconsistent with UIP pattern (*see* third column)	• Upper or mid-lung predominance • Peribronchovascular predominance • Extensive ground glass abnormality (extent > reticular abnormality) • Profuse micronodules (bilateral, predominantly upper lobes) • Discrete cysts (multiple, bilateral, away from areas of honeycombing) • Diffuse mosaic attenuation/air-trapping (bilateral, in three or more lobes) • Consolidation in bronchopulmonary segment(s)/lobe(s)

2018

UIP	Probable UIP	Indeterminate for UIP	Alternative Diagnosis
Subpleural and basal predominant; distribution is often heterogeneous* Honeycombing with or without peripheral traction bronchiectasis or bronchiolectasis†	Subpleural and basal predominant; distribution is often heterogeneous Reticular pattern with peripheral traction bronchiectasis or bronchiolectasis May have mild GGO	Subpleural and basal predominant 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")	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)

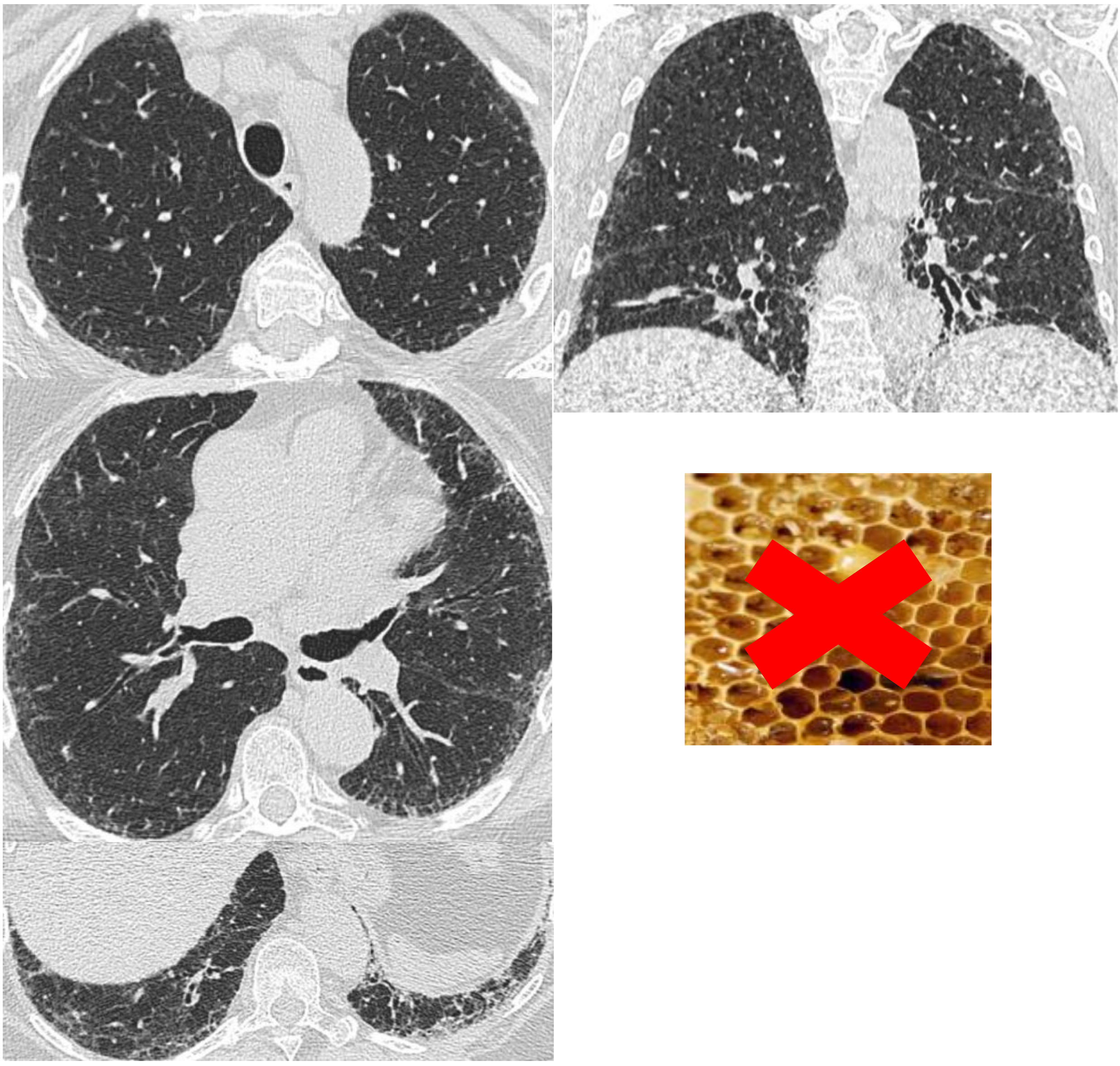
Criterios Radiológicos:





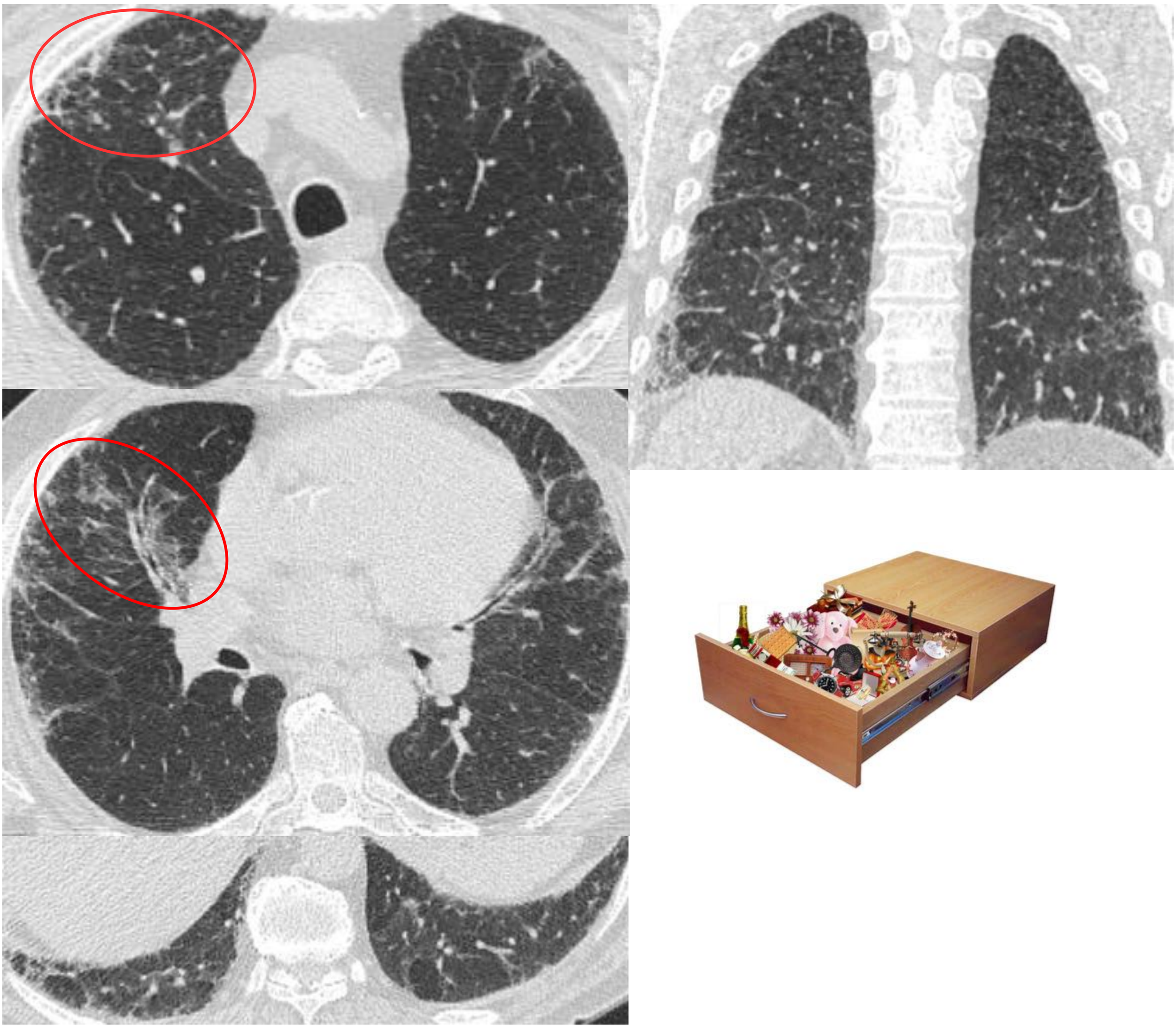
NIU

Criterios Radiológicos:



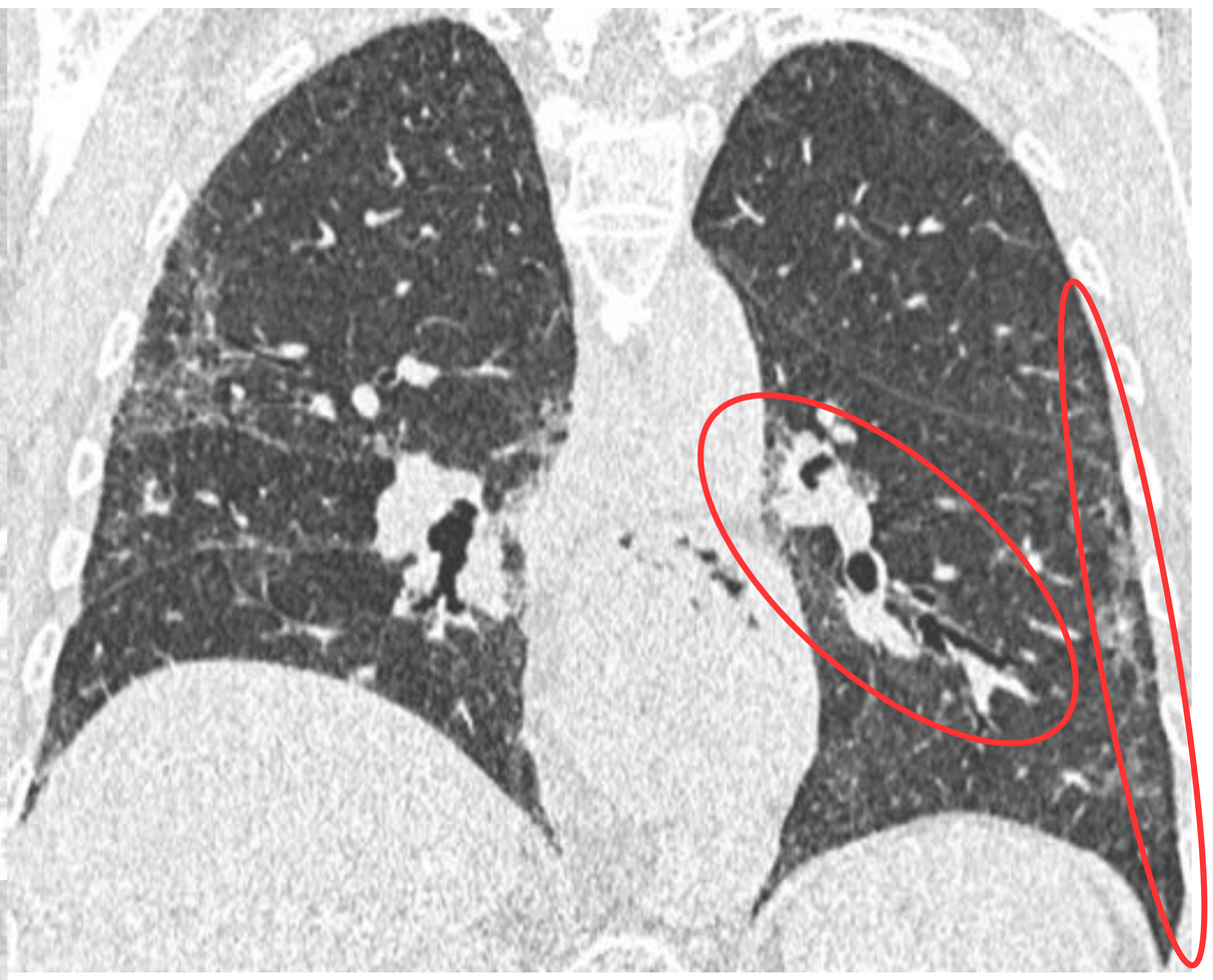
PROBABLE NIU

Criterios Radiológicos:



INDETERMINADA

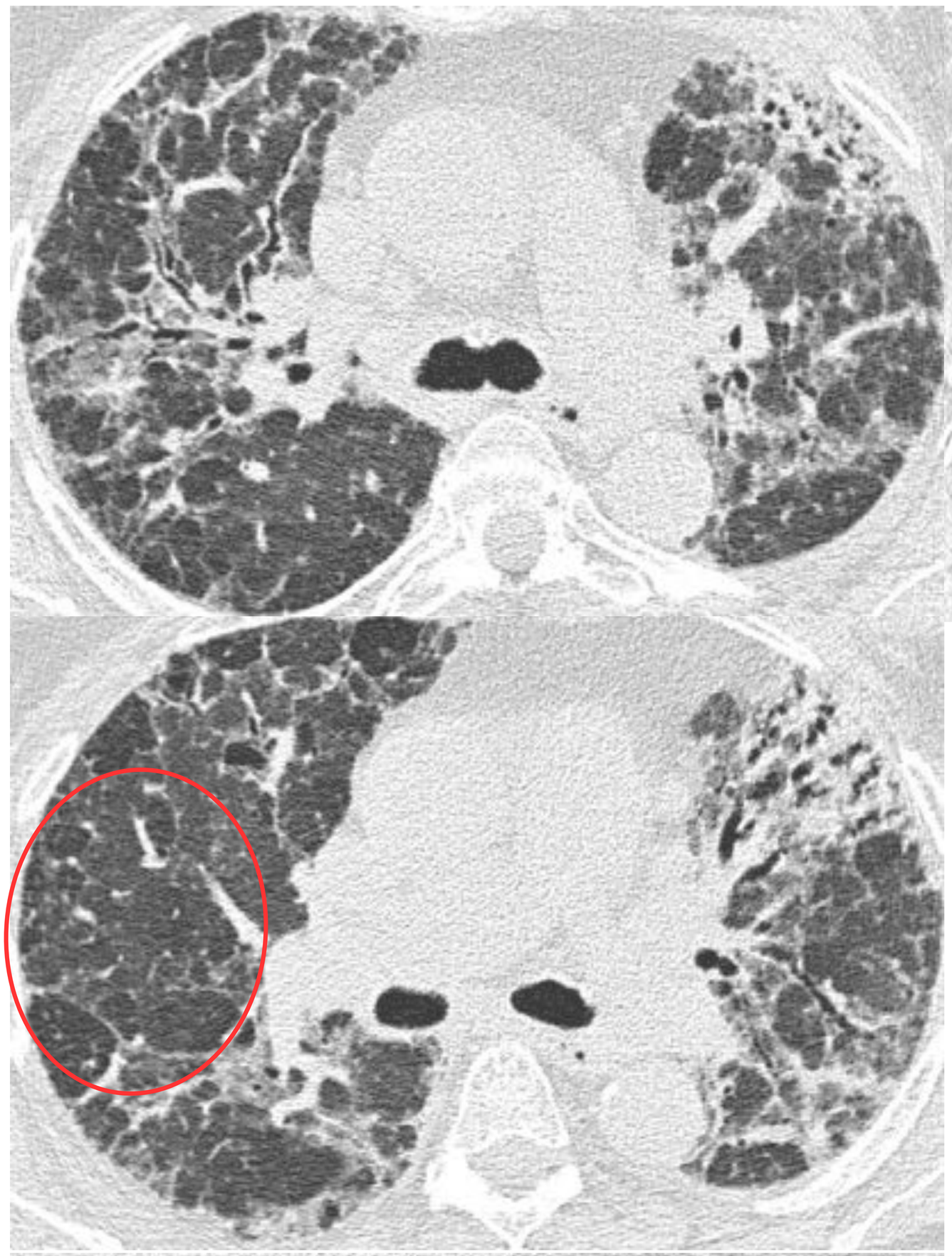
Criterios Radiológicos:

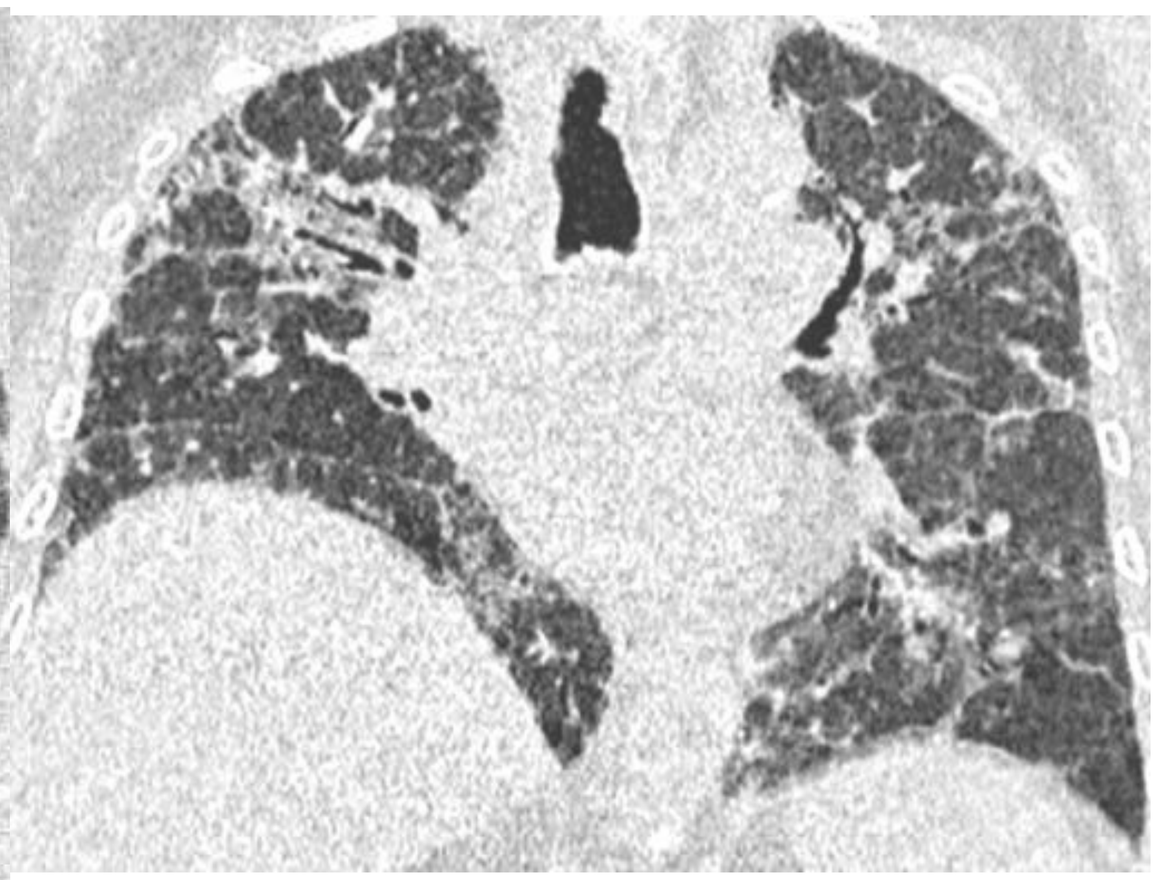


Paciente con NINE

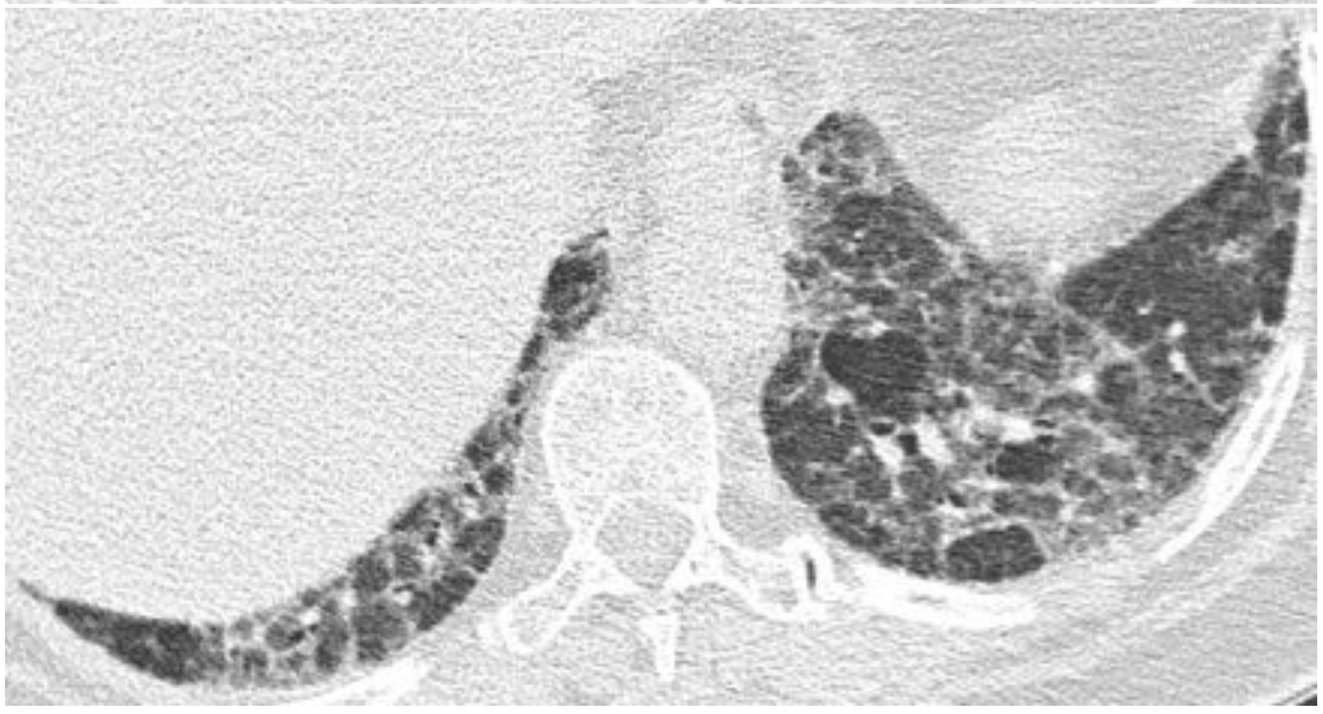
ALTERNATIVO

Criterios Radiológicos:





Paciente con NHc



ALTERNATIVO

Criterios histológicos:

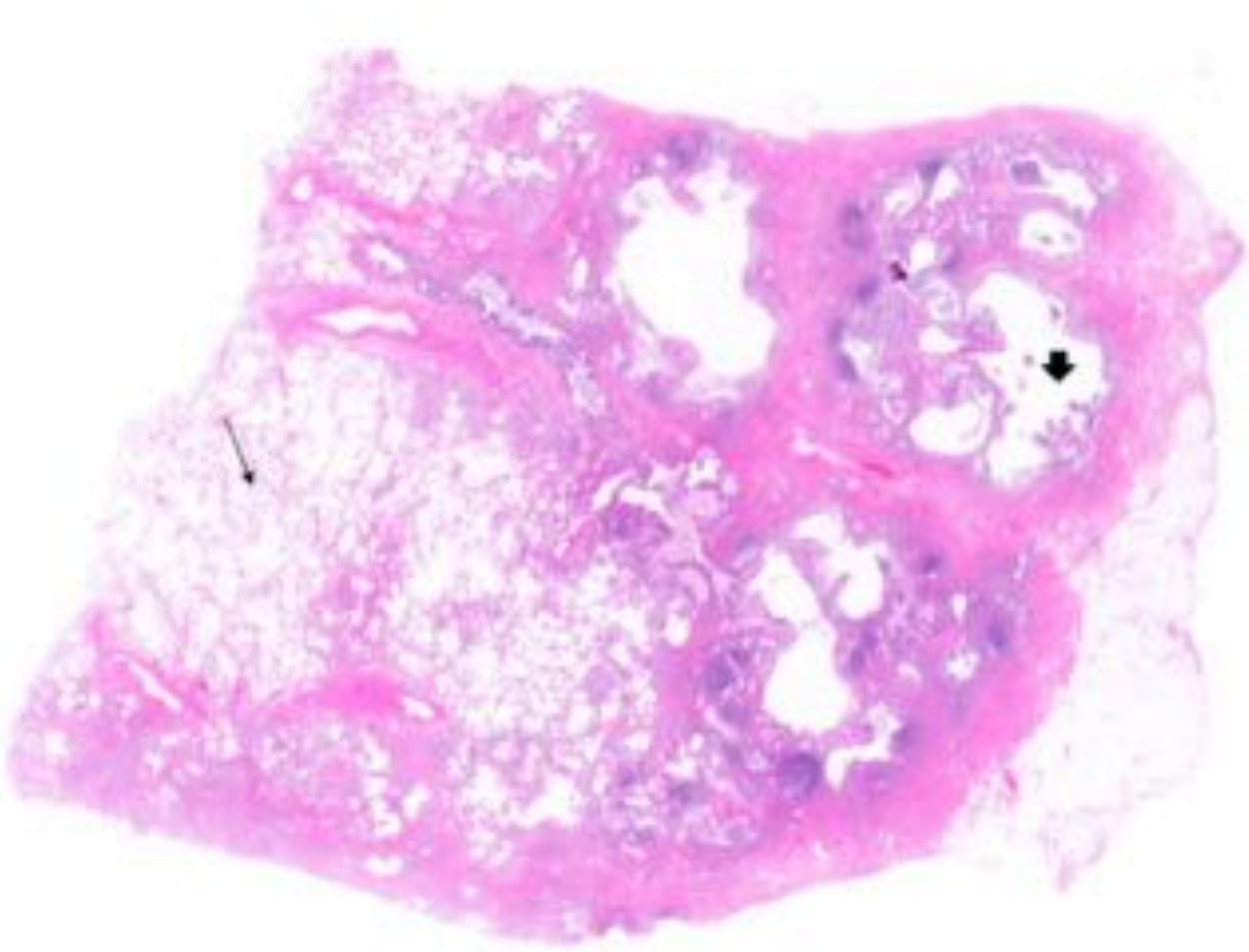
2011

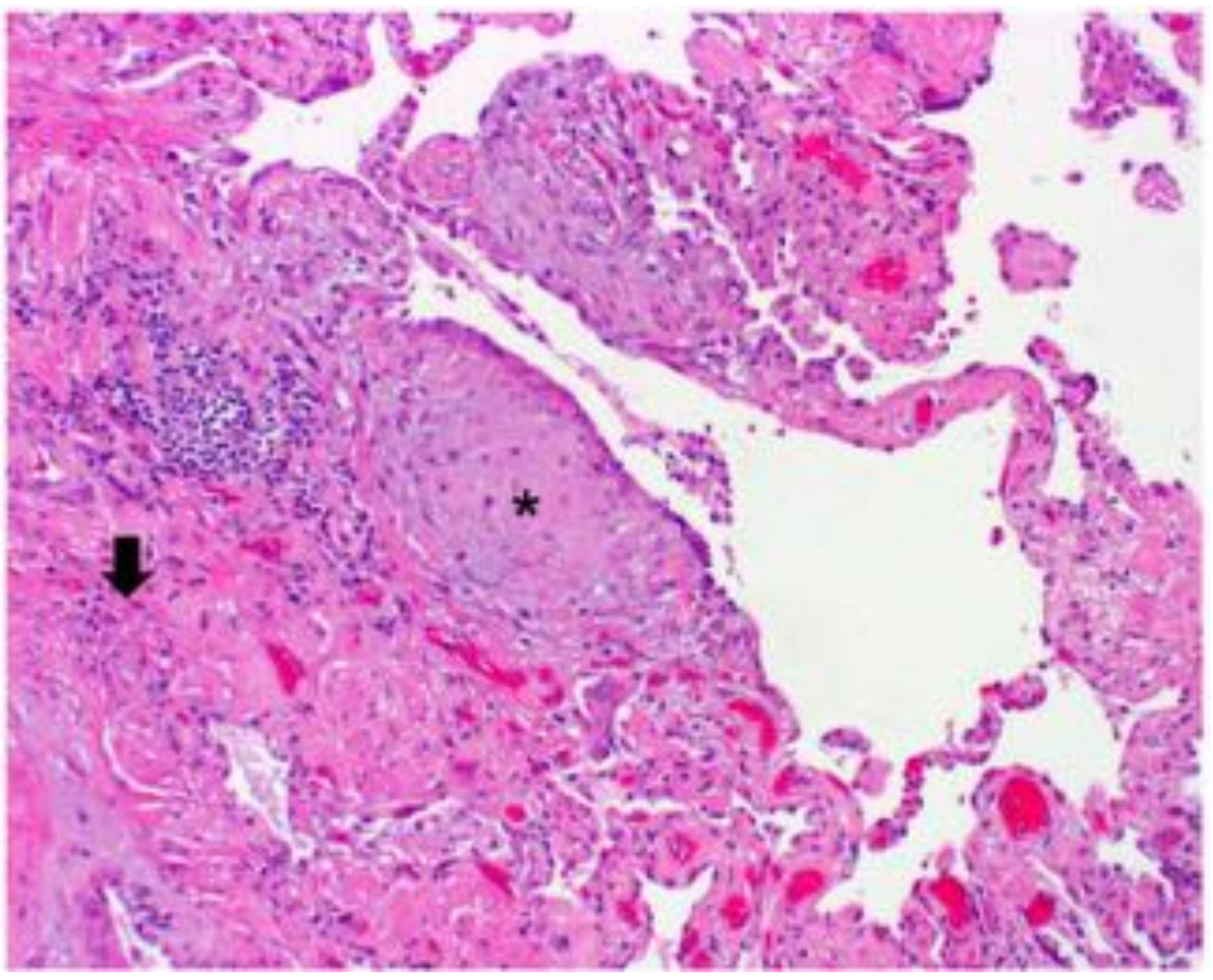
UIP Pattern (All Four Criteria)	Probable UIP Pattern	Possible UIP Pattern (All Three Criteria)	Not UIP Pattern (Any of the Six Criteria)
• Evidence of marked fibrosis/ architectural distortion, ± honeycombing in a predominantly subpleural/ paraseptal distribution • Presence of patchy involvement of lung parenchyma by fibrosis • Presence of fibroblast foci • Absence of features against a diagnosis of UIP suggesting an alternate diagnosis (see fourth column)	• Evidence of marked fibrosis / architectural distortion, ± honeycombing • Absence of either patchy involvement or fibroblastic foci, but not both • Absence of features against a diagnosis of UIP suggesting an alternate diagnosis (see fourth column) OR • Honeycomb changes only[a]	• Patchy or diffuse involvement of lung parenchyma by fibrosis, with or without interstitial inflammation • Absence of other criteria for UIP (see UIP Pattern column) • Absence of features against a diagnosis of UIP suggesting an alternate diagnosis (see fourth column)	• Hyaline membranes* • Organizing pneumonia*† • Granulomas‡ • Marked interstitial inflammatory cell infiltrate away from honeycombing • Predominant airway centered changes • Other features suggestive of an alternate diagnosis

2018

UIP	Probable UIP	Indeterminate for UIP	Alternative Diagnosis
• Dense fibrosis with architectural distortion (i.e., destructive scarring and/or honeycombing) • Predominant subpleural and/or paraseptal distribution of fibrosis • Patchy involvement of lung parenchyma by fibrosis • Fibroblast foci • Absence of features to suggest an alternate diagnosis	• Some histologic features from column 1 are present but to an extent that precludes a definite diagnosis of UIP/IPF *And* • Absence of features to suggest an alternative diagnosis *Or* • Honeycombing only	• Fibrosis with or without architectural distortion, with features favoring either a pattern other than UIP or features favoring UIP secondary to another cause* • Some histologic features from column 1, but with other features suggesting an alternative diagnosis†	• Features of other histologic patterns of IIPs (e.g., absence of fibroblast foci or loose fibrosis) in all biopsies • Histologic findings indicative of other diseases (e.g., hypersensitivity pneumonitis, Langerhans cell histiocytosis, sarcoidosis, LAM)

Criterios histológicos:





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Criterios diagnósticos:

2011

HRCT Pattern*	Surgical Lung Biopsy Pattern* (When Performed)	Diagnosis of IPF?*
UIP	**UIP** **Probable UIP** **Possible UIP** **Nonclassifiable fibrosis**[‡]	**YES**
	Not UIP	No
Possible UIP	**UIP** **Probable UIP**	**YES**
	Possible UIP Nonclassifiable fibrosis	Probable[‡]
	Not UIP	No
Inconsistent with UIP	UIP	Possible[‡]
	Probable UIP Possible UIP Nonclassifiable fibrosis Not UIP	No

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	IPF	IPF (Likely)**	Indeterminate***	Non-IPF dx
	Alternative diagnosis	IPF (Likely)** /non-IPF dx	Non-IPF dx	Non-IPF dx	Non-IPF dx

Resultados del estudio:

- 63 pacientes diagnosticados de FPI tratados con fármacos anti-fibróticos desde noviembre del 2014 hasta la actualidad.

- 3 Pacientes derivados de otros centros y sin pruebas radiológicas previas fueron excluidos, quedando una población de 60 pacientes.

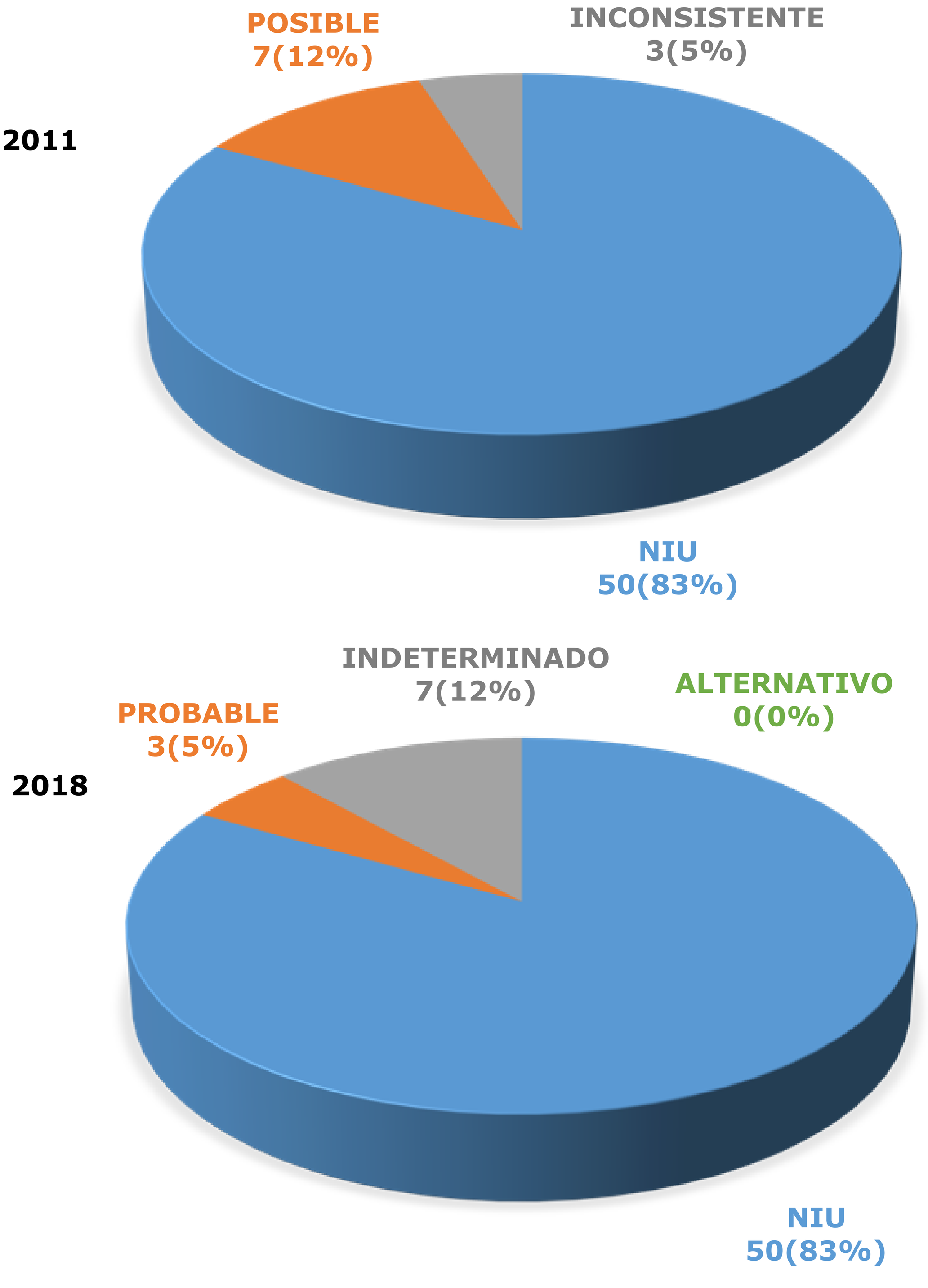
- 40 Hombres (67%), 20 mujeres (33%)

- Reclasificamos todas las TC diagnósticas de estos pacientes aplicando los nuevos critérios ATS para FPI buscando:

  - Cambios en su diagnóstico radiológico

  - Potencial impacto en su manejo clínico-terapéutico

Resultados del estudio:



Conclusiones:

- La aplicación de los nuevos criterios ATS/ERS no supondría un cambio relevante en el manejo clínico de los pacientes hasta su diagnostico, pero si tendría implicaciones en su tratamiento:

1. Mayor impacto si se prescindiese de la biopsia en pacientes con "Probable NIU“ como propuso la sociedad Fleischner.

2. Pacientes "no NIU“ que pasarían a "indeterminado para NIU“ tienen más posibilidades de ser tratados con anti-fibróticos.

3. La histología “manda” pero la radiología gana protagonismo.

1. **Mayor impacto si se prescindiese de la biopsia en pacientes con "Probable NIU“ como propuso la sociedad Fleischner.**

UIP Pattern (All Four Features)	Possible UIP Pattern (All Three Features)	Inconsistent with UIP Pattern (Any of the Seven Features)
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2011

BIOPSIA

UIP	Probable UIP	Indeterminate for UIP	Alternative Diagnosis
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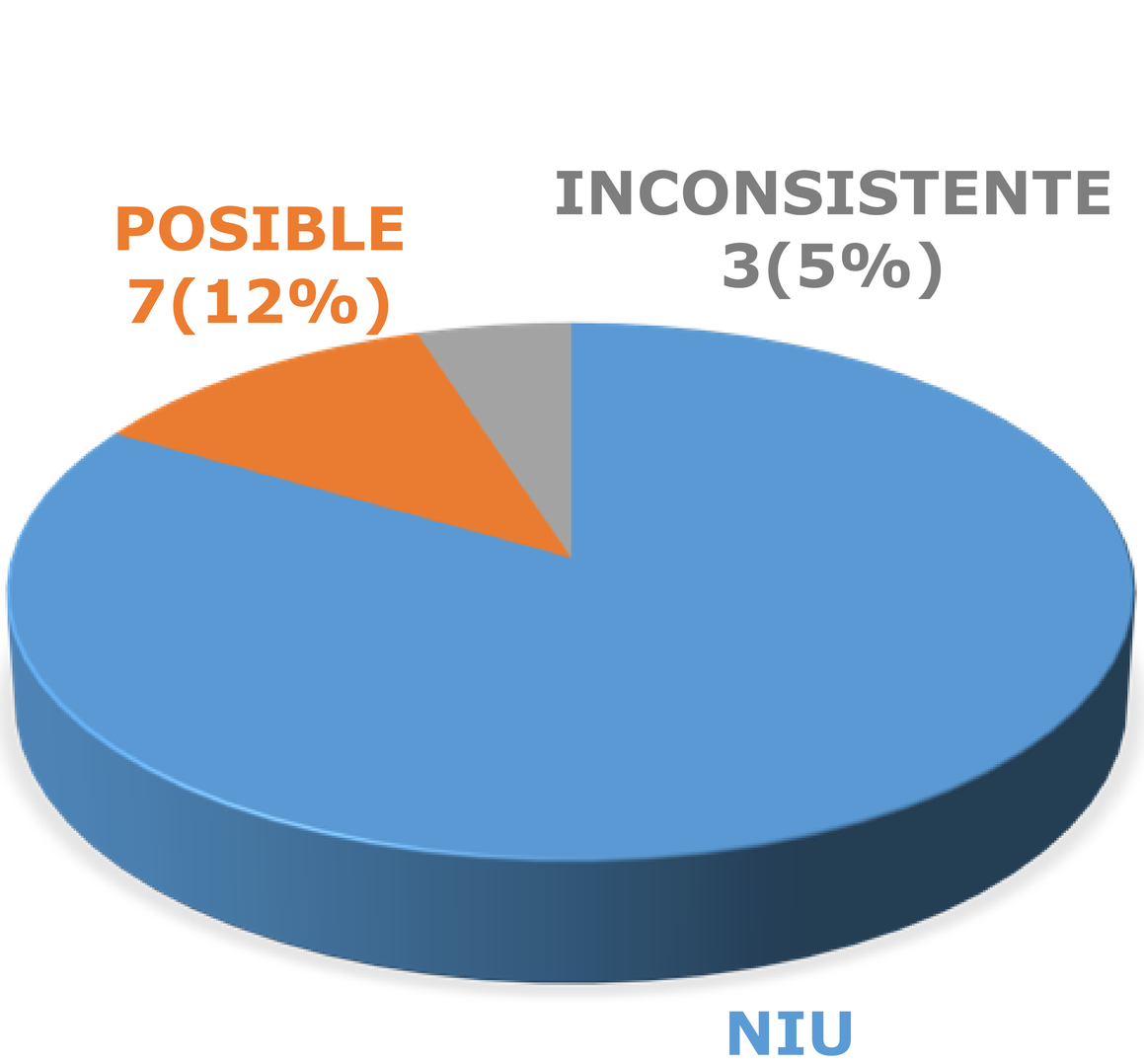
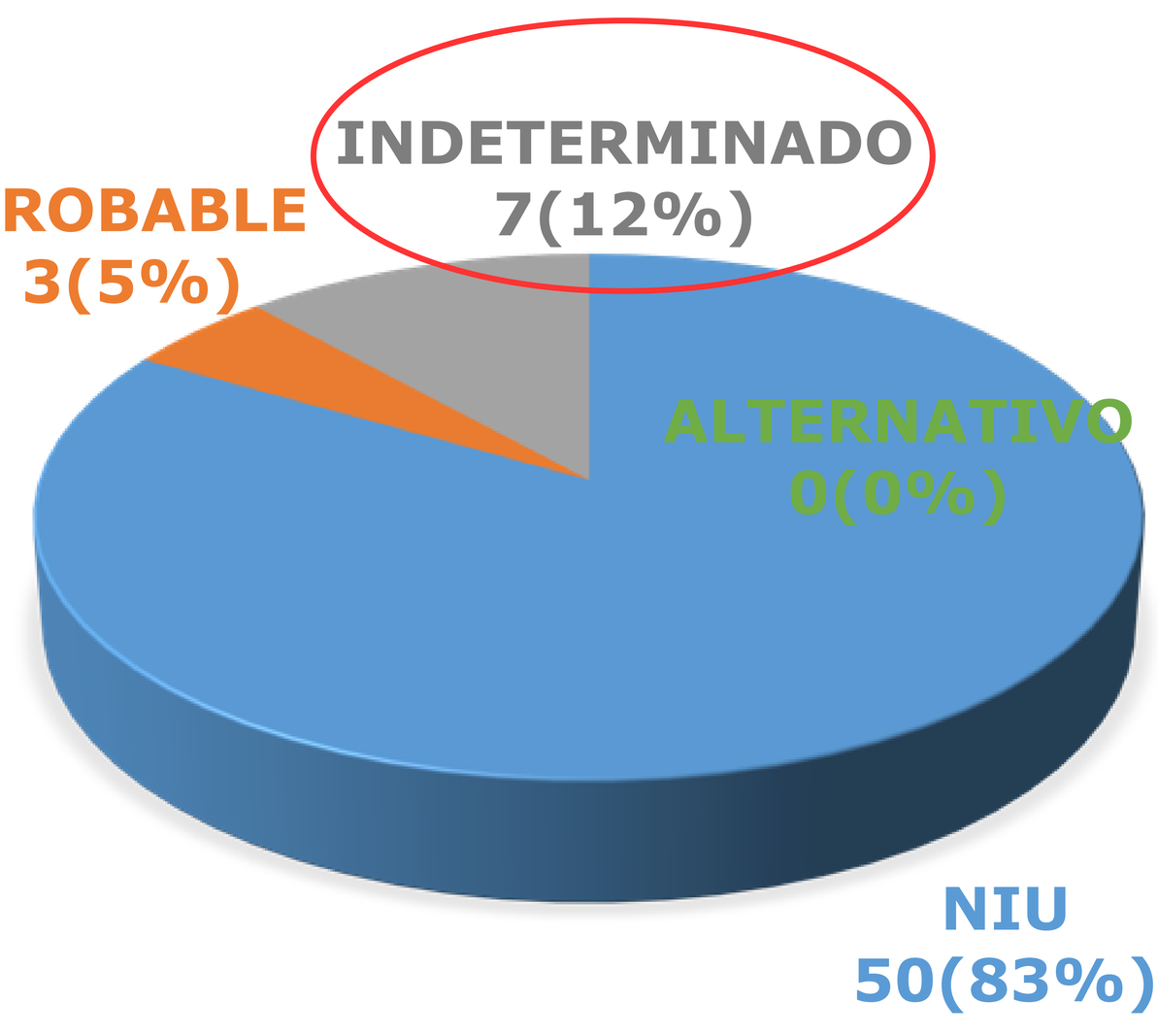
2018

2. Pacientes “no NIU” que pasarían a “indeterminados para NIU” tendrían más posibilidades de ser tratados con anti-fibróticos.





3. La histología “manda” pero la radiología gana protagonismo.

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

Tendencias:

- Prescindir de la biopsia

- Cuantificar vidrio esmerilado/fibrosis con fines de diagnóstico diferencial y pronóstico

- Patrones histológicos comparados por IA (BAL, SLB, criobiopsia, TBB)

- Marcadores genéticos/serológicos.

Bibliografía:

- American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias. Am J Respir Crit Care Med Vol 165. pp 277–304, 2002

- William D. Travis, Ulrich Costabel, David M. Hansell, et al. An Official American Thoracic Society/European Respiratory Society Statement: Update of the International Multidisciplinary Classification of the Idiopathic Interstitial Pneumonias. Am J Respir Crit Care Med Vol 188, Iss. 6, pp 733–748, Sep 15, 2013

- Ganesh Raghu, Harold R. Collard, Jim J. Egan, et al. An Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management. Am J Respir Crit Care Med Vol 183. pp 788–824, 2011

- Ganesh Raghu, Martine Remy-Jardin, Jeffrey L. Myers, et al. Diagnosis of Idiopathic Pulmonary Fibrosis An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline. Am J Respir Crit Care Med Vol 198, Iss 5, pp e44–e68, Sep 1, 2018

- Takeshi Johkoh. Nonspecific Interstitial Pneumonia and Usual Intertitial Pneumonia: Is Differentiation Posible by High-Resolution Computed Tomography? Elevier Inc Semin Ultrasound CT MRI 35:24-28, 2014.