New Classification of Interstitial Pneumonias

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Disclosures

- No current or past financial conflict of interests

- AI was used to help with editing and graphs but not to generate content
- I have more slides that allowed by time, I will skip a few

Learning Objectives

By the end of this session, participants will be able to:

- Describe the major changes in the **2025 ERS/ATS classification of interstitial pneumonias**
- Explain the shift from an **idiopathic-only framework to one integrating idiopathic and secondary causes**
- Recognize the new **bronchiolocentric interstitial pneumonia (BIP)** pattern and its key radiologic and histologic features
- Review updated terminology, including **alveolar macrophage pneumonia (AMP)** and **idiopathic diffuse alveolar damage (DAD)**
- Apply **diagnostic confidence** and the updated classification framework to clinical evaluation of patients with interstitial pneumonia

The Document

- 49 pages in PDF format
- Available for Free
- Both ERS/ATS
- First since 2013

EUROPEAN RESPIRATORY JOURNAL
ERS OFFICIAL DOCUMENT
C.J. RYERSON ET AL.

Update of the international multidisciplinary classification of the interstitial pneumonias: an ERS/ATS statement

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How the Statement Was Developed

1

Committee formation

Five co-chairs assembled 32 experts across pulmonology, radiology, pathology, and molecular medicine, plus 2 patient representatives

2

Structured drafting

Full-committee video meetings, then subgroups drafted individual sections in parallel

3

Consensus, not systematic review

Literature was selected by expert consensus rather than systematic search, given the breadth of the topic

4

Voting when needed

Decisions were made by discussion; when consensus was unclear, ≥70% agreement was required by vote

Guarding against bias

The co-chairs deliberately assembled a committee with a range of perspectives, and every member was encouraged to select and discuss literature — reducing the risk that evidence was cherry-picked to fit predetermined views.

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Ryerson CJ, Adegunsoye A, Piciucchi S, et al. Eur Respir J 2025; 66: 2500158

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Outlines

- Expansion beyond idiopathic IP to include secondary causes of IP
- Identification of new subcategories and updated terminology:
  - Addition of bronchiolocentric IP (BIP) as a major pattern
  - Changing acute interstitial pneumonia (AIP) to idiopathic diffuse alveolar damage (DAD)
  - Changing desquamative interstitial pneumonia (DIP) to alveolar macrophage pneumonia (AMP)
- Consideration of diagnostic confidence in patient evaluation and management
- Providing a comprehensive update on potential molecular tools and future research priorities

The 2013 Classification, Briefly

- **Six major idiopathic patterns** plus two rare idiopathic patterns, and a category of unclassifiable disease.
- **Pattern-based:** patients were categorised primarily by histological pattern.
- **Sub classified by behaviour:** chronic fibrosing, smoking-related, and acute/subacute subtypes.
- **Known-cause disease set aside:** important differentials such as fibrotic hypersensitivity pneumonitis and CTD-ILD were discussed, but excluded from the idiopathic category by definition.
- **Disease behaviour framework:** a complementary approach based on anticipated clinical trajectory, with monitoring and treatment implications.

2013 MAJOR IIPs

- IPF
- Idiopathic NSIP
- Acute interstitial pneumonia (AIP)
- Cryptogenic organising pneumonia (COP)
- RB-ILD
- Desquamative interstitial pneumonia (DIP)

Plus rare IIPs (iPPFE, iLIP) and unclassifiable IIP

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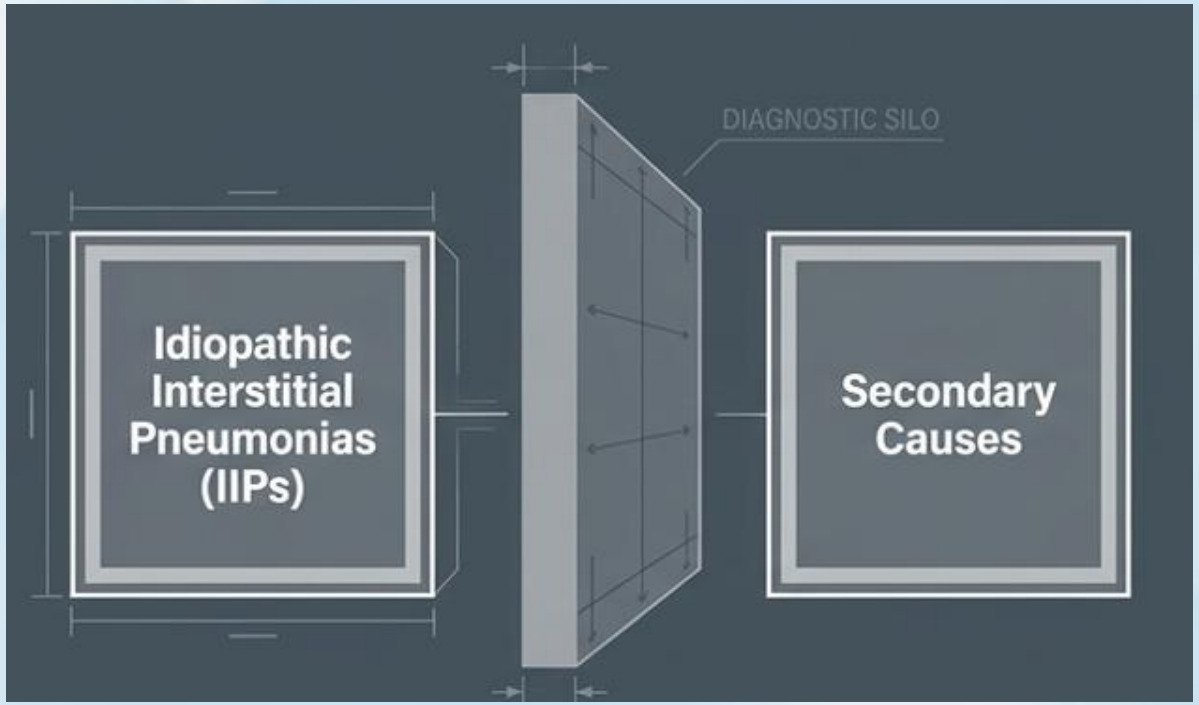
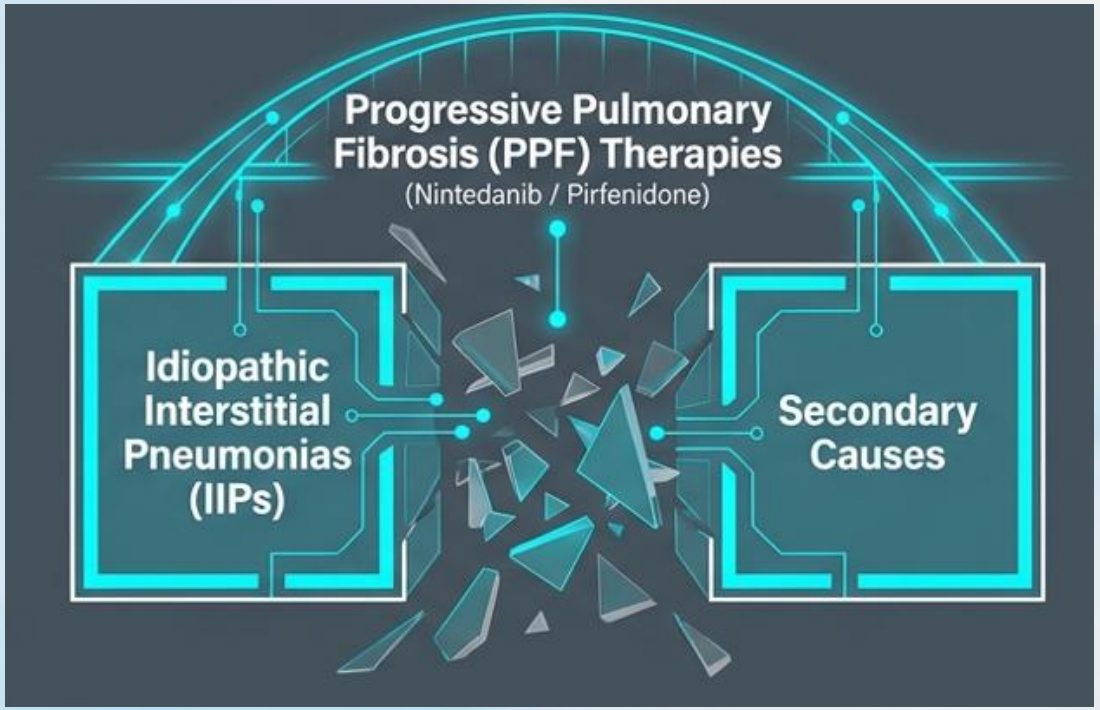
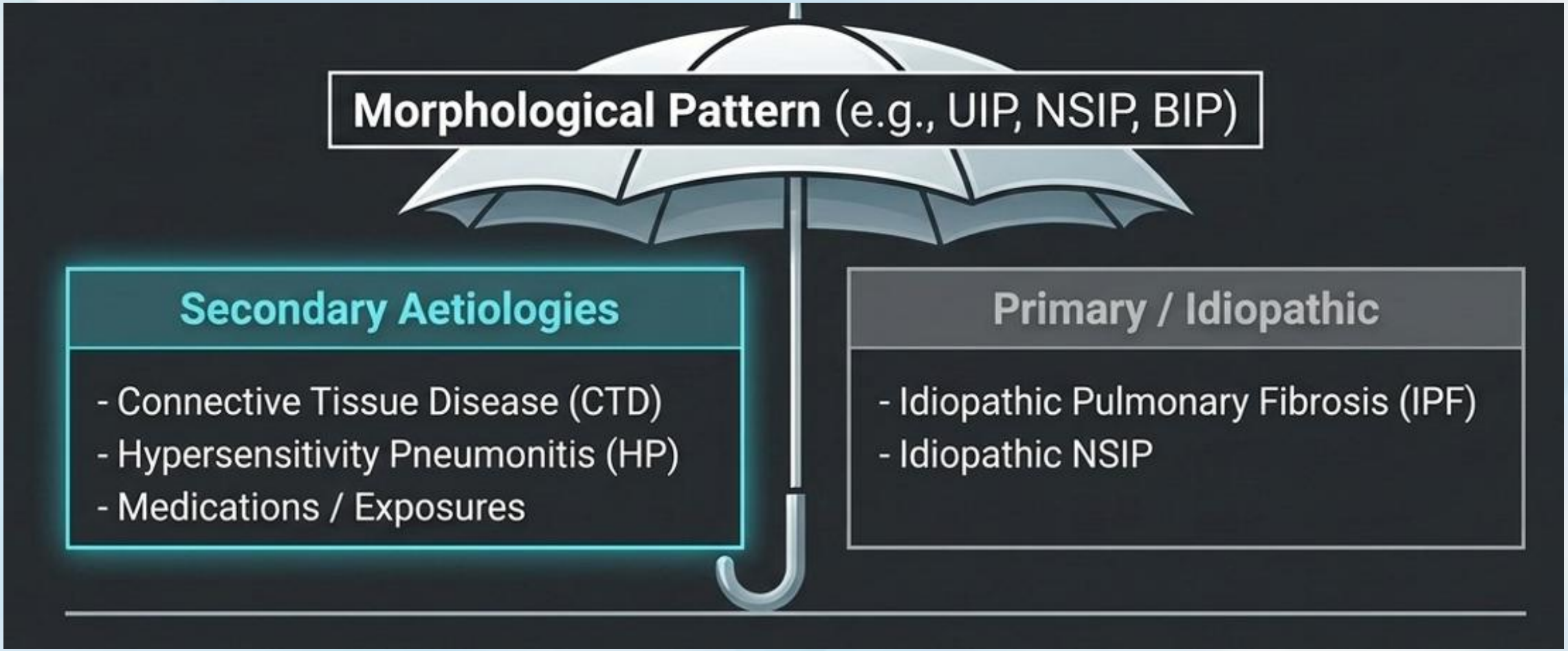
2013 Framework



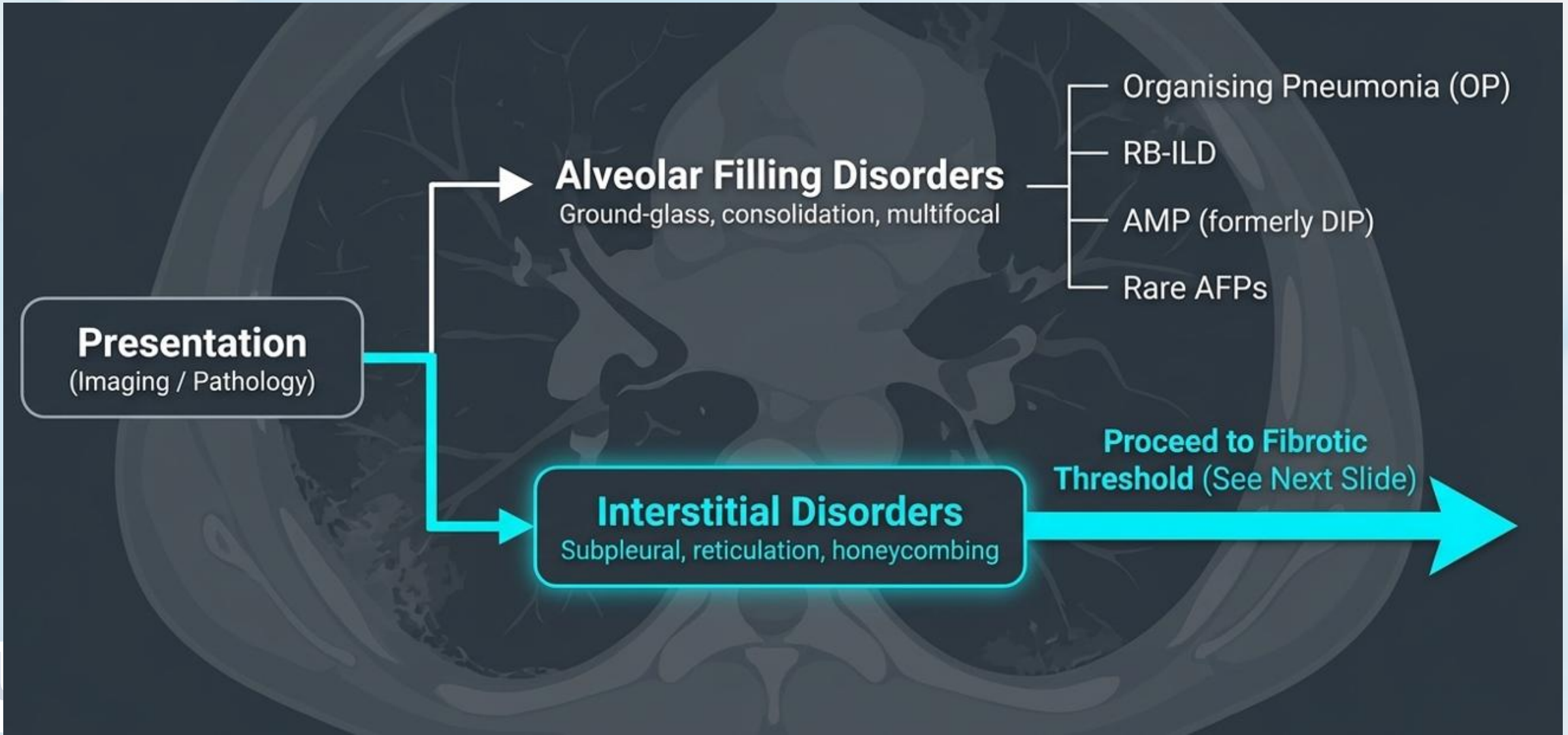
2025 Reality



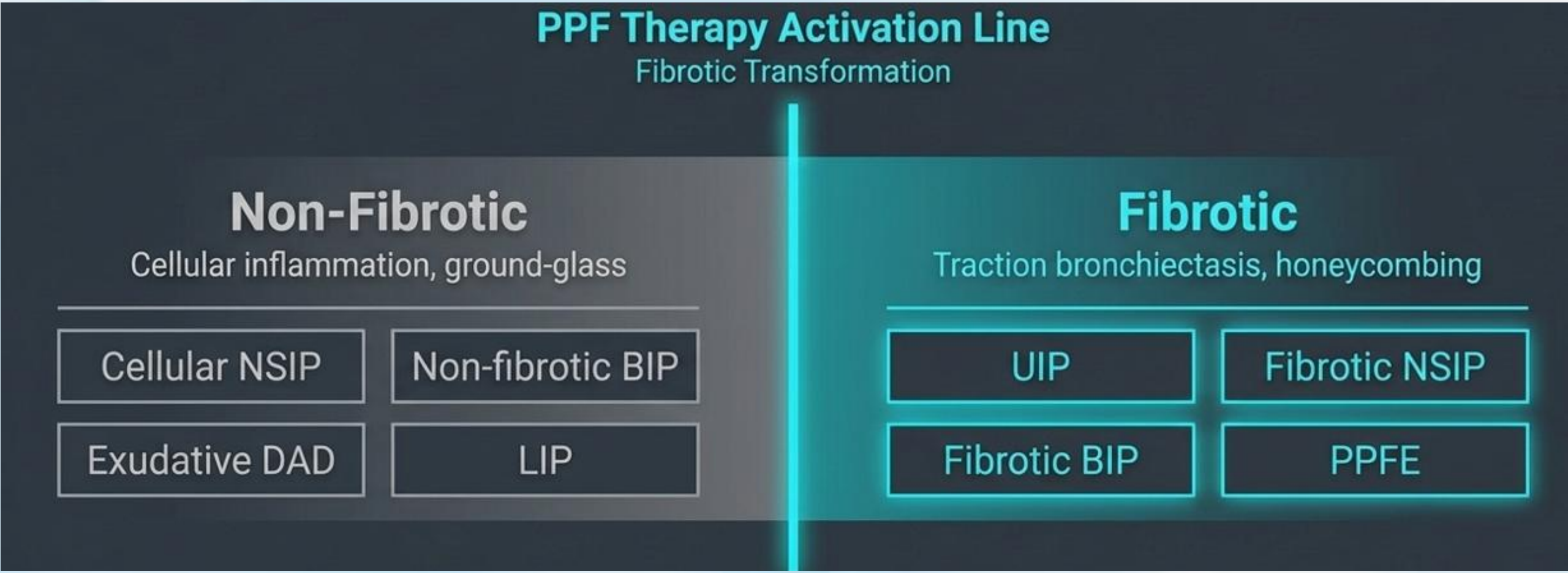
Expansion: The Umbrella Concept



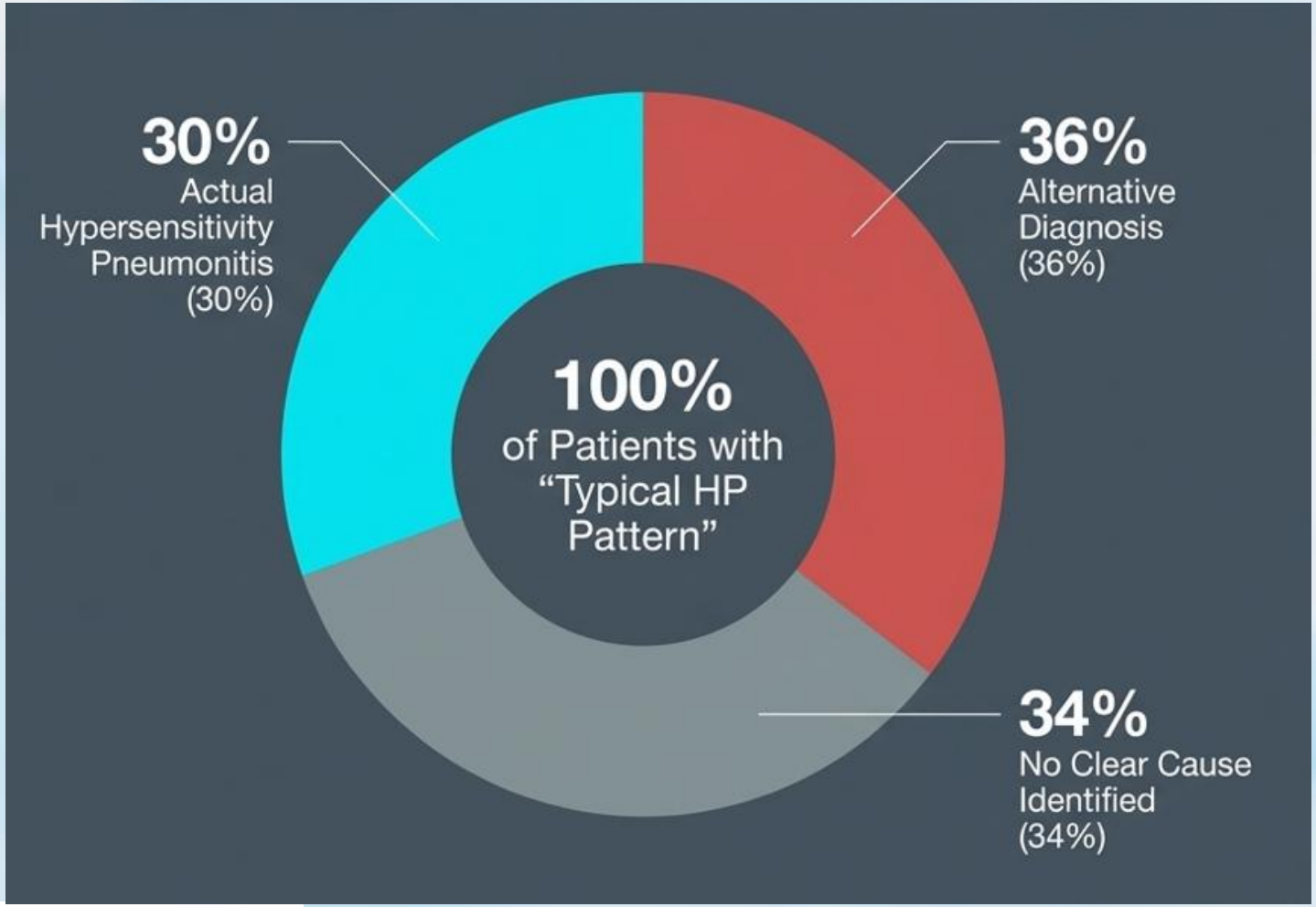
Subclassification: The Diagnostic Route



Subclassification: The Fibrotic Threshold

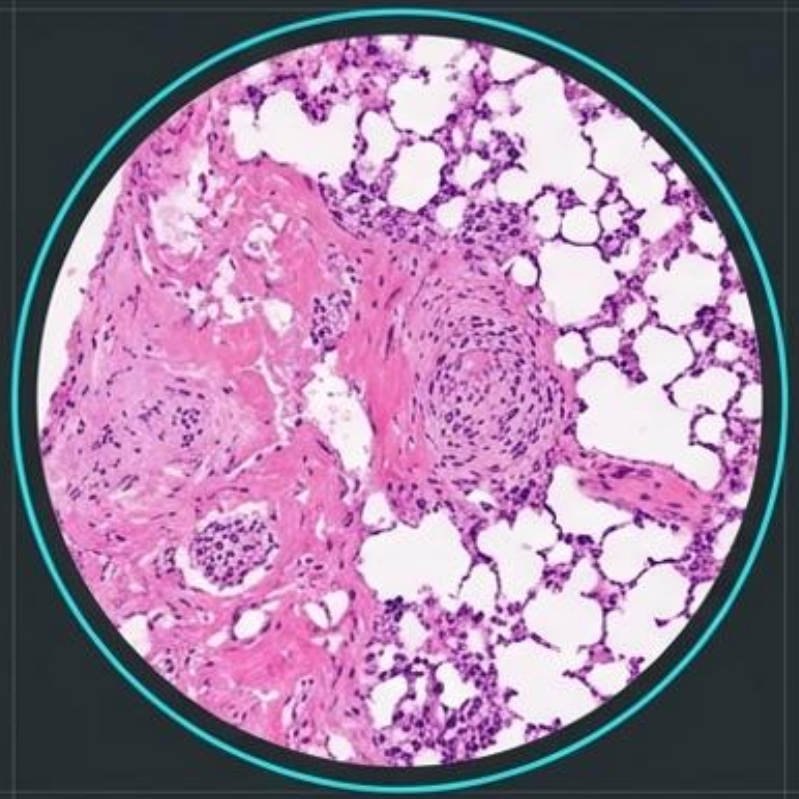


The Death of "HP Pattern"



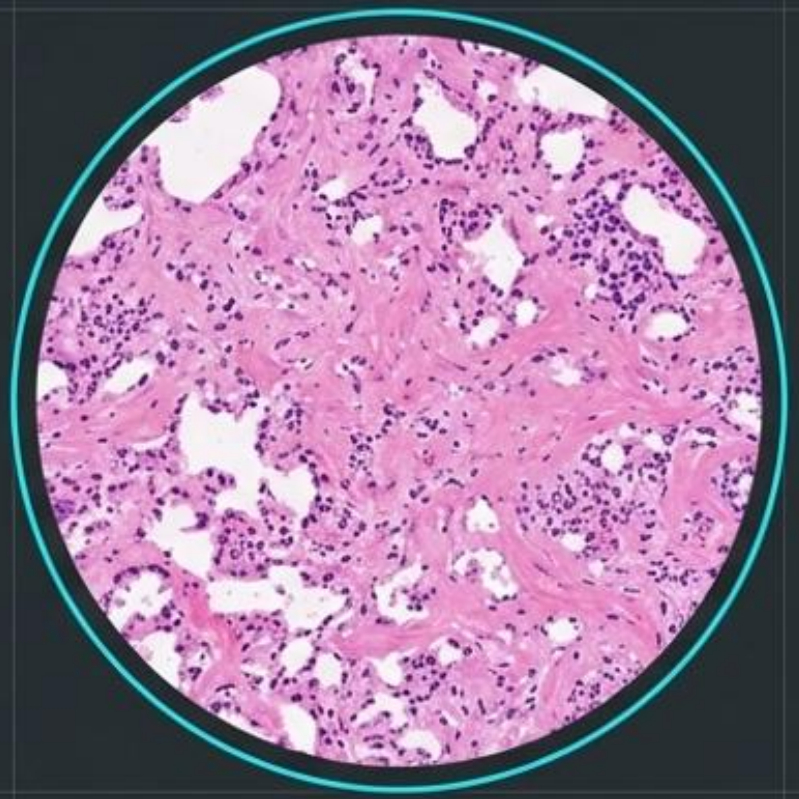
The Microscopic Big Three

distiguishing the major interstitial patterns at the cellular level



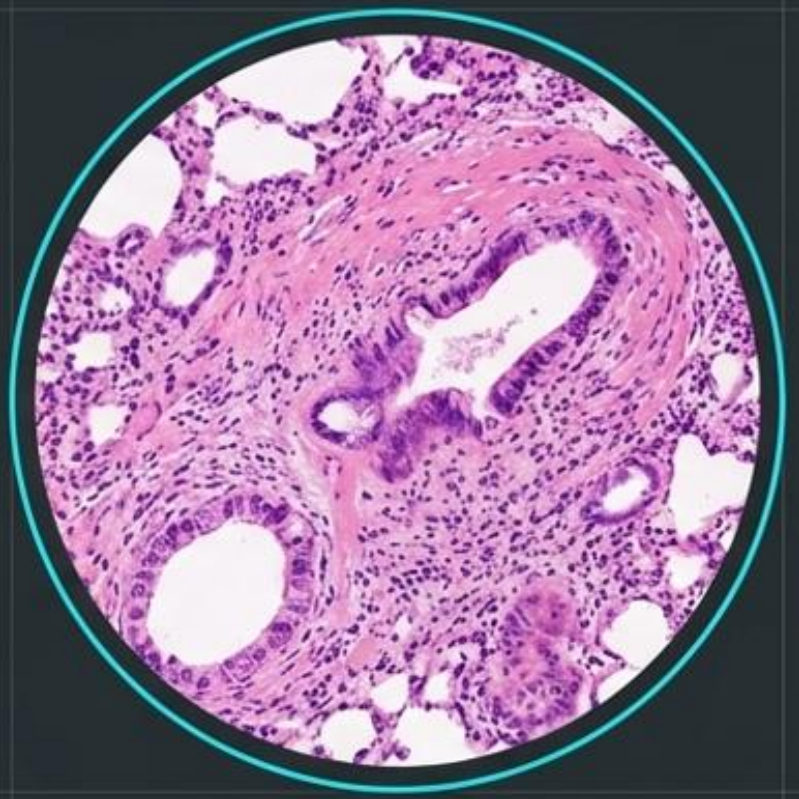
UIP Pattern

Peripheral/subpleural distribution.
Temporal heterogeneity with fibroblast foci.



NSIP Pattern

Uniform/diffuse distribution.
Diffuse thickening without fibroblastic foci.



BIP Pattern

Airway-centered distribution.
Bronchiolocentric fibrosis, peribronchiolar metaplasia.

Confidence: formalization Diagnostic Threshold



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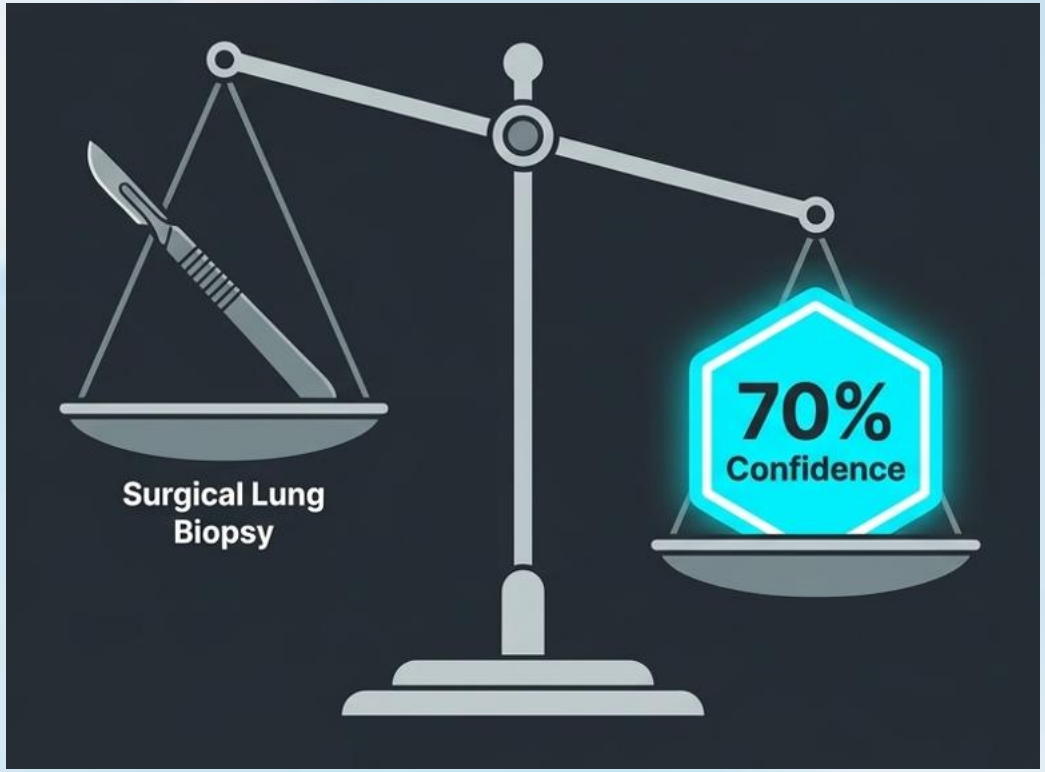
The Bedside Impact of Confidence Dials



The 70% Rule:

studies suggest a 70% confidence threshold in a clinical-radiologist diagnosis of IPF is sufficient to support immediate management decisions such as prescribing anti-fibrotics

The Clinical payoff:

This structured probabilistic framework empowers clinicians to implicitly agree on a “provisional diagnosis” at 70% certainty, entirely bypassing surgical lung biopsies in older, high risk or fragile patients

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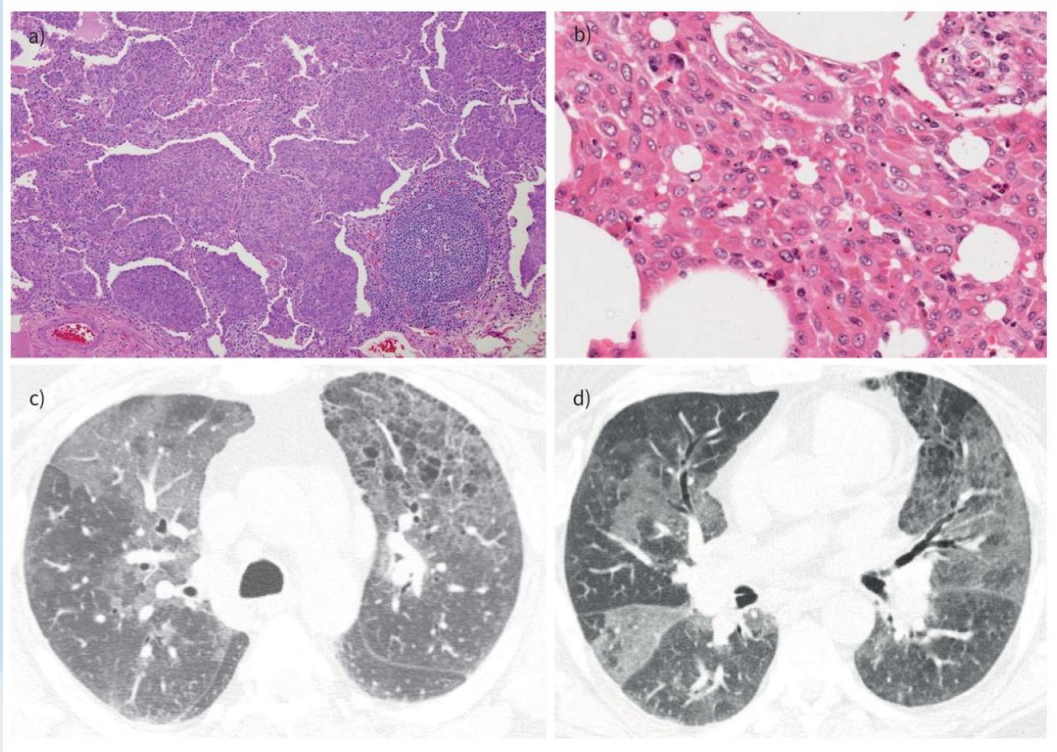
Alveolar Macrophage Pneumonia (AMP)

- Alveolar Macrophage Pneumonia (AMP) to replace prior inaccurate terminology of desquamative interstitial pneumonia (DIP) to better reflect the underlying pathology of the disease
- Disease of alveolar filling with macrophages, not desquamation of pneumocytes.



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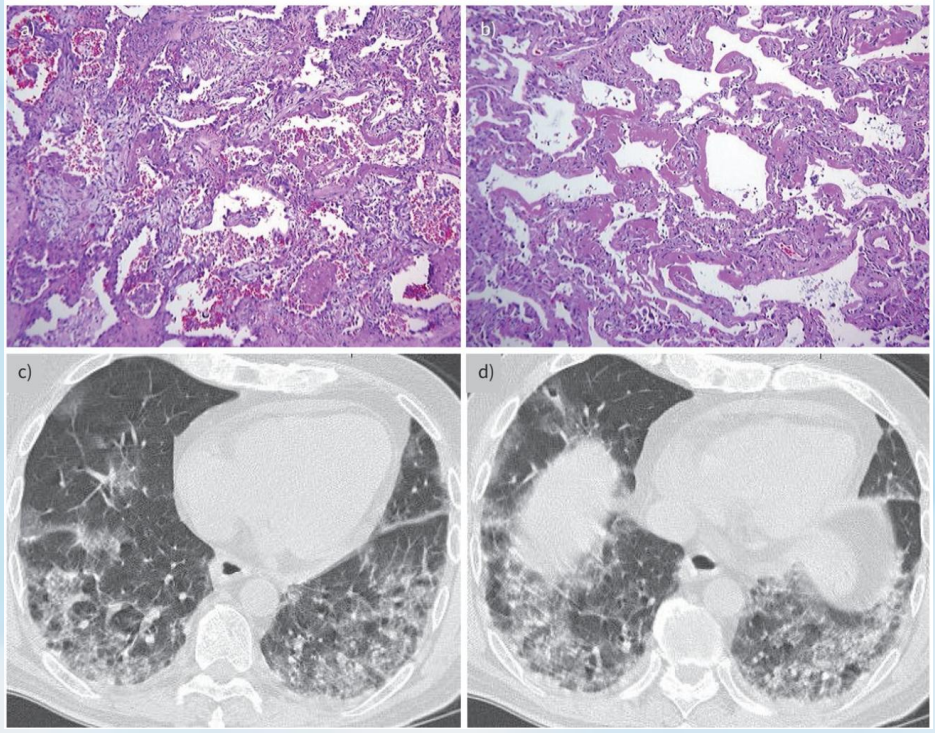
Idiopathic Diffuse Alveolar Damage

- Idiopathic diffuse alveolar damage (DAD) in introduced to replace acute interstitial pneumonia (AIP)
- AIP was considered an imprecise term given the potential for acute presentations of other interstitial pneumonitis



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Bronchiolocentric Interstitial Pneumonia (BIP)

- Rationale: not all HPs
- A substantial proportion of patients have bronchiolocentric disease on pathology and/or CT but may not necessarily have hypersensitivity pneumonitis (i.e., CTD-ILD, familial ILD, aspiration, drug reaction)
- The field was missing a description to cover bronchiolocentric patterns, thus BIP was brought to the table to decouple the pattern (BIP) from etiology (e.g, HP, CTD-ILD, etc)
- Pattern triad of UIP, NSIP or BIP (or mixed)
- Etiologies for any pattern(s) can be idiopathic or secondary

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Bronchiolocentric Intersititial Pneumonia (BIP)

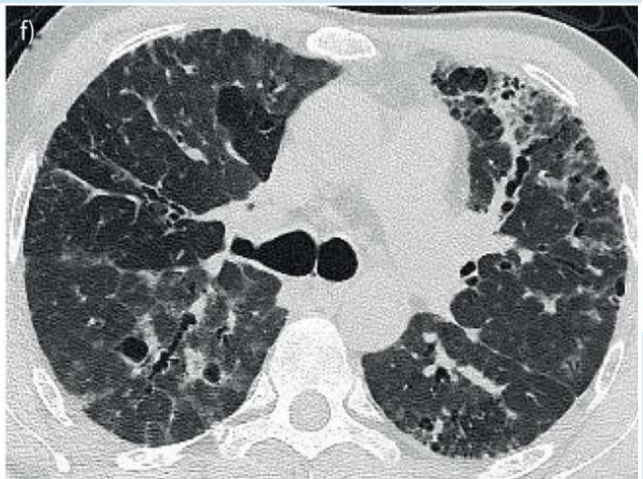
CT features

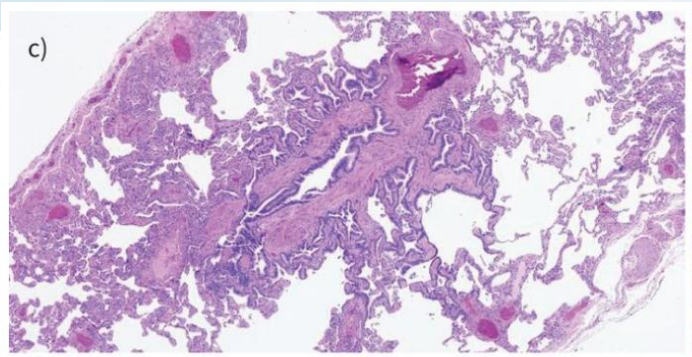
- non-fibrotic BIP: centrilobular ground-glass nodule, mosaic attenuation, and/or lobular air trapping
- Fibrotic BIP: peribronchovascular ground-glass traction bronchiectasis, mosaic attenuation, lobular air trapping, and three-density sign. Features can overlap with non-fibrotic BIP

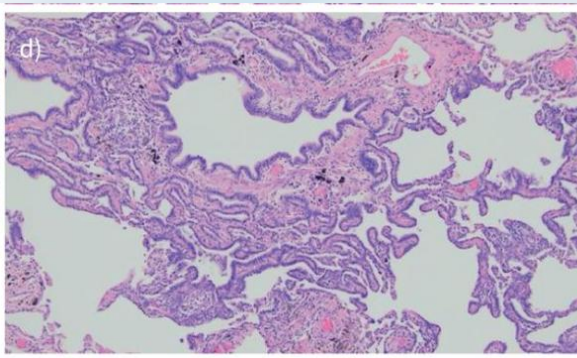
Histologic features

- Non-fibrotic BIP: bronchiolocentric chronic inflammation± non-necrotizing granulomatous inflammation
- Fibrotic BIP: bronchiolocentric fibrosis often with peribronchiolar metaplasia ± brochiolocentric inflammation









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A Diagnostic Blueprint Build not Just To Classify but to Treat: 1, 2, 3

1. Look for behavior

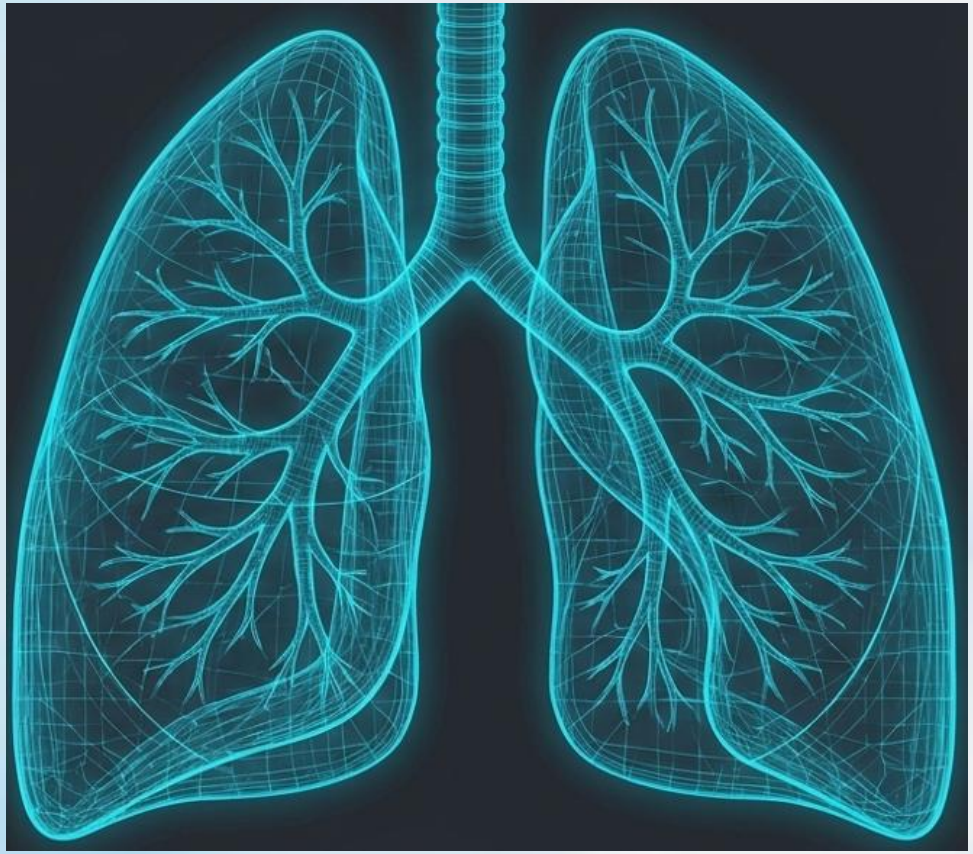
Subclassify fibrotic vs non-fibrotic early to flag PPF

2. Separate pattern from disease

adopt BIP and AMP. Retire the historical terms HP pattern, AIP and DIP

3. Use the dials

Embrace probabilistic diagnosis (51-89%) to spare patient unnecessary invasive biopsies while accelerating access to care



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Where the Field Is Headed

Genetics

MUC5B promoter variant (rs35705950) raises pulmonary fibrosis risk up to 20x with two copies; telomere-related genes drive familial disease

Telomere length

Critically short peripheral-blood telomeres predict worse survival across IPF, fibrotic HP, and unclassifiable ILD

Transcriptomics

The Envisia genomic classifier aids UIP recognition (88% specificity) but cannot yet distinguish among non-UIP patterns

Protein biomarkers

MMP-7, KL-6, and CA-125 trends help track prognosis and treatment response — clinical utility still being defined

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Ryerson CJ, Adegunsoye A, Piciucchi S, et al. Eur Respir J 2025; 66: 2500158 · Endorsed by ERS (23 Jul 2025) and ATS (10 Jul 2025)

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Summary Take-Home Message

One framework: idiopathic and secondary interstitial pneumonias are now classified together
Pattern ≠ etiology: UIP, NSIP, and **BIP** patterns may be idiopathic or secondary
BIP is new: a bronchiolocentric pattern is formally recognized, separating morphology from a specific diagnosis such as hypersensitivity pneumonitis
Terminology matters: DIP → **alveolar macrophage pneumonia (AMP)**; AIP → **idiopathic diffuse alveolar damage (DAD)**
State diagnostic confidence: classify diagnoses as **confident, provisional, or unclassifiable**
The future is molecular: genetics, telomere biology, transcriptomics, and biomarkers are increasingly complementing radiology and pathology

Thank You!

Questions or Comments are welcome

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