Idiopathic Interstitial Pneumonias

Multidisciplinary classification of the IIPs — major, rare, and unclassifiable entities — plus a clinical disease-behavior framework for treatment and monitoring.

2013 ATS/ERS Statement
Update of the International Multidisciplinary Classification of the Idiopathic Interstitial Pneumonias
ATS/ERS Committee on IIPs · Am J Respir Crit Care Med 2013;188:733–748

FORMAT *This is a classification scheme, not a graded recommendation set. Diagnosis requires multidisciplinary discussion (MDD) integrating clinical, radiologic, and pathologic data.*

1 Major IIPs — Chronic Fibrosing

ENTITY	CLINICAL–RADIOLOGIC–PATHOLOGIC CORRELATE
IPF UIP	Requires exclusion of known ILD causes plus UIP on HRCT (no biopsy needed) or specific HRCT+biopsy combinations; atypical HRCT (ground-glass, nodules, mosaic attenuation) may still = IPF after MDD.
Idiopathic NSIP NSIP	Bilateral ground-glass opacity, traction bronchiectasis, subpleural sparing; predominantly fibrotic subtype most common; now a distinct entity, "provisional" dropped.

2 Major IIPs — Smoking-Related

ENTITY	CLINICAL–RADIOLOGIC–PATHOLOGIC CORRELATE
RB-ILD RB	Always present in current smokers. Ground-glass + centrilobular nodules on HRCT; a minority progress despite cessation.
DIP DIP	Macrophage accumulation; can occur in nonsmokers (SP gene mutations); ~70% 10-year survival, resistant to treatment in a minority.

3 Major IIPs — Acute/Subacute

ENTITY	CLINICAL–RADIOLOGIC–PATHOLOGIC CORRELATE
COP OP	Subacute illness under 3 mo; patchy/migratory consolidation, reversed halo sign; most recover with corticosteroids but relapse is common.
AIP DAD	Fulminant respiratory failure; histologically indistinguishable from ARDS of known cause.

4 Rare IIPs

ENTITY	CLINICAL–RADIOLOGIC–PATHOLOGIC CORRELATE
Idiopathic LIP LIP	Dense lymphoid interstitial infiltrate; rare, often overlaps with Sjögren's/autoimmune disease workup.
Idiopathic PPFE PPFE	Dense subpleural fibroelastosis, upper-lobe predominant; recurrent pneumothorax common; progresses in 60%, death from disease in 40%.

5 Unclassifiable IIP — Causes

CAUSE	DESCRIPTION
Inadequate data	Insufficient clinical, radiologic, or pathologic information to reach a confident MDD diagnosis.
Altered findings	Prior therapy substantially alters radiologic/histologic findings (e.g., biopsy of DIP after steroids showing only residual NSIP).
Novel/discordant pattern	New entity or unusual variant not characterized by current classification; multiple discordant HRCT and/or pathologic patterns present simultaneously.

6 Disease Behavior Classification

BEHAVIOR	TREATMENT GOAL · MONITORING
Reversible, self-limited	e.g. many RB-ILD. Remove possible cause; short-term (3–6 mo) observation to confirm regression.
Reversible, risk of progression	e.g. cellular/some fibrotic NSIP, DIP, COP. Achieve response then rationalize longer-term therapy; short-term then long-term observation to confirm gains preserved.
Stable, residual disease	e.g. some fibrotic NSIP. Maintain status; long-term observation to assess disease course.
Progressive, potentially stabilizes	e.g. some fibrotic NSIP. Stabilize; long-term observation to assess course.
Progressive despite therapy	e.g. IPF, some fibrotic NSIP. Slow progression; long-term observation for need for transplant or effective palliation.

7 Diagnostic Principles

MDD — the dynamic, integrated multidisciplinary discussion, not histology alone, is the diagnostic gold standard. **Category correlation:** chronic fibrosing = UIP/NSIP pattern; smoking-related = RB/DIP pattern; acute/subacute = OP/DAD pattern. **Biomarkers:** elevated SP-A/SP-D, KL-6, MMP-7, and CCL18 associate with worse IPF survival — investigational, not yet routine practice.

Travis WD, Costabel U, Hansell DM, 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.* 2013;188(6):733–748.