Diagnosis of Idiopathic Pulmonary Fibrosis

HRCT-pattern-based diagnostic testing recommendations and multidisciplinary decision-making for newly detected ILD of unknown cause with suspected IPF.

2018 ATS/ERS/JRS/ALAT Guideline
Diagnosis of Idiopathic Pulmonary Fibrosis
ATS/ERS/JRS/ALAT Committee · Am J Respir Crit Care Med 2018;198:e44-e68

GRADE — Recommend (Strong) — Suggest (Conditional) — Strong Against

1 When to Suspect IPF

Typical presentation	Unexplained bilateral fibrosis on chest imaging, bibasilar inspiratory crackles, age typically >60 yr.
Atypical presentation	Middle-aged adults (40–60 yr), especially with familial pulmonary fibrosis risk, may present the same way.

2 Initial Screening — All Suspected IPF

- Rec. — **Exposure history** — detailed history of medication use and environmental exposures at home, work, and elsewhere.
- Rec. — **Serologic testing** — test to exclude connective tissue disease as a potential cause.

3 Testing by HRCT Pattern

HRCT PATTERN — UIP

- Suggest — **BAL cellular analysis** — against performing.
- Strong — **Surgical lung biopsy** — against performing.
- Strong — **Transbronchial biopsy** — against performing.
- Strong — **Lung cryobiopsy** — against performing.

HRCT — PROBABLE/INDETERMINATE OR ALTERNATIVE

- Suggest — **BAL cellular analysis** — for performing.
- Suggest — **Surgical lung biopsy** — for performing.

Transbronchial biopsy	No recommendation for or against.
Lung cryobiopsy	No recommendation for or against.

4 Multidisciplinary Discussion & Biomarkers

- Suggest — **MDD** — multidisciplinary discussion suggested for diagnostic decision-making.
- Strong — **Serum biomarkers** — against measuring MMP-7, SP-D, CCL-18, or KL-6 to distinguish IPF from other ILDs.

Format note: "we recommend" = strong; "we suggest" = conditional/weak; most recommendations rest on very-low-quality evidence. Condensed from the Summary of Recommendations.

Raghu G, Remy-Jardin M, Myers JL, et al. Diagnosis of Idiopathic Pulmonary Fibrosis: An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline. *Am J Respir Crit Care Med.* 2018;198(5):e44-e68.