Idiopathic Pulmonary Fibrosis — Diagnosis & Management

HRCT/biopsy diagnostic criteria, evidence-based pharmacotherapy recommendations, and supportive care for adults with IPF.

2011 ATS/ERS/JRS/ALAT Guideline
Idiopathic Pulmonary Fibrosis: Evidence-Based Guidelines for Diagnosis and Management
ATS/ERS/JRS/ALAT Committee · Am J Respir Crit Care Med 2011;183:788-824

GRADE — SF · Strong For | WF · Weak For | WA · Weak Against | SA · Strong Against

1 Diagnostic Criteria — HRCT & Biopsy

–	GUIDANCE
UIP on HRCT	Diagnostic of IPF without biopsy, once known causes of ILD are excluded and clinical context fits.
Combination patterns	UIP HRCT + probable/possible UIP or nonclassifiable fibrosis on biopsy = IPF; "inconsistent with UIP" HRCT + biopsy UIP = possible IPF, requires MDD.
MDD accuracy	Diagnostic accuracy increases with multidisciplinary discussion, especially when radiology and histopathology are discordant; refer to ILD experts.

2 Diagnostic Evaluation

WA **Transbronchial biopsy** — not for use in the majority; may be reasonable in a minority.

WF **Serologic testing** (RF, anti-CCP, ANA) in the majority to exclude connective tissue disease, even without CTD signs/symptoms.

SF **Multidisciplinary discussion** — pulmonology, radiology, and pathology discussion should be used in evaluation of suspected IPF.

3 Pharmacotherapy — Not Recommended

AGENT	GUIDANCE	GRADE
Corticosteroid monotherapy	No survival benefit shown; substantial treatment-related morbidity.	SA
Cyclosporine A	Discordant, very low-quality data.	SA
Interferon-γ1b	No mortality difference in a definitive RCT of 800+ patients (high-quality evidence).	SA
Bosentan	No benefit on mortality/disease progression.	SA
Etanercept	Insufficient evidence of benefit.	SA
Anticoagulation	Very low-quality data; risks/cost outweigh unproven benefit.	SA

4 Pharmacotherapy — Selective Use Only

AGENT	GUIDANCE	GRADE
Colchicine	No survival benefit shown; may suit a minority who accept low expected benefit.	WA
Acetylcysteine monotherapy	Low-quality supportive data only.	WA
Prednisone + azathioprine + NAC	Triple therapy; low-quality evidence, considerable committee debate.	WA
Pirfenidone	Reduced FVC decline in Japanese RCTs but inconsistent primary endpoints; GI, hepatic, and photosensitivity adverse effects.	WA

5 Supportive & Non-Pharmacologic Care

SF **Long-term oxygen** — clinically significant resting hypoxemia treated with LTOT, extrapolated from other chronic lung disease evidence.

SF **Lung transplantation** — appropriate candidates should undergo transplant; 5-yr survival 50–56%; no clear consensus on timing.

WA **Mechanical ventilation** — not for the majority with respiratory failure (hospital mortality 87–96% in cohort studies); may suit a minority, e.g. bridge to transplant.

WF **Pulmonary rehabilitation & GERD treatment** — rehabilitation for use; asymptomatic GERD treated medically in the majority.

6 Acute Exacerbation, Comorbidities & Progression

WF **Acute exacerbation** — corticosteroids for the majority despite absent controlled trials; supportive care remains the mainstay.

–	GUIDANCE
Pulmonary hypertension	Data on PH-specific therapy in IPF are limited; no routine recommendation.
Progression markers	≥10% absolute FVC decline or ≥15% DLCO decline (with/without the other) is a surrogate marker of mortality; smaller sustained changes (5–10%) may also reflect progression.

Raghu G, Collard HR, Egan JJ, 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.* 2011;183(6):788–824.