

# Idiopathic pulmonary fibrosis (an update) and progressive pulmonary fibrosis in adults

Part II: Diagnosis and Treatment of PPF in Fibrotic ILD, Other than IPF

An official ATS/ERS/JRS/ALAT clinical practice guideline

April 2022

## Summary<sup>1</sup>



The guideline committee chose to adopt the term ‘progressive pulmonary fibrosis’ (PPF), a well-known, currently used term by both clinicians and patients



The ATS/ERS/JRS/ALAT clinical practice guideline:

- Defines PPF as at least two of three criteria (worsening symptoms, physiological progression and radiological progression) occurring within the past year with no alternative explanation, in a patient with an ILD other than IPF
- Acknowledges that PPF occurs in many different fibrotic lung diseases
- Clarifies that PPF is not a diagnosis but a common feature of progression in these ILDs



Nintedanib is recommended for the treatment of patients with PPF who have failed standard management\* and further research into pirfenidone is suggested

\* Standard management includes observation, immunosuppressive treatment and/or antigen remediation

## The definition of PPF<sup>1</sup>

In a patient with ILD of known or unknown etiology (other than IPF) who has radiological evidence of pulmonary fibrosis, PPF is defined as **at least two of the following three criteria** occurring **within the past year** with no alternative explanation:

01

**Worsening respiratory symptoms**

02

**Physiological evidence of disease progression**

Either of the following:

- Absolute decline in FVC  $\geq 5\%$  predicted within 1 year of follow-up
- Absolute decline in DLCO  $\geq 10\%$  (corrected for Hb) predicted within 1 year of follow-up

03

**Radiological evidence of disease progression**

One or more of the following:

- Increased extent or severity of traction bronchiectasis and bronchiolectasis
- New ground-glass opacity with traction bronchiectasis
- New fine reticulation
- Increased extent or increased coarseness of reticular abnormality
- New or increased honeycombing
- Increased lobar volume loss

The guideline provides evidence-based recommendations for the treatment of PPF (other than IPF)<sup>1</sup>



**Nintedanib:**  
*A conditional recommendation is made for nintedanib in PPF*

**Pirfenidone:**  
*Additional research into pirfenidone is recommended*

### Abbreviations:

ALAT, Asociación Latinoamericana del Tórax; ATS, American Thoracic Society; DLCO, diffusing capacity of the lungs for carbon monoxide; ERS, European Respiratory Society; FVC, forced vital capacity; ILD, interstitial lung disease; IPF, idiopathic pulmonary fibrosis; JRS, Japanese Respiratory Society; PPF, progressive pulmonary fibrosis

Reference: 1. Raghu G et al. Am J Respir Crit Care Med 2022;205:e18–47