P8867

Scientific content on demand
To request a copy of this poster:
Scan QR code
via a barcode reader application

Variability of Criteria Used to Confirm Progressive Pulmonary Fibrosis: Results from a Real-World Survey

Pooja Chopra[1], Rebecca Hardy-Woods[2], Mark Small[2], Lisa Lancaster[3]

[1] Bristol Myers Squibb, Princeton, NJ, [2] Adelphi Real World, Bollington, UK, [3] Division of Allergy, Pulmonary and Critical Care Medicine, Department of Medicine, Vanderbilt University Medical Center*, Nashville, Tennessee, USA

INTRODUCTION

- Progressive Pulmonary Fibrosis (PPF), formerly referred to as progressive-fibrosing interstitial lung disease (PF-ILD) [1], refers to a subset of ILDs with continued fibrosis of the lungs.
- Prior to the ATS/ERS/JRS/ALAT terminology and guideline update in 2022 for PPF, definitions of progression in ILD patients varied widely in literature and were largely shaped by criteria used in clinical trials. Although variation was noted, progression was defined by evidence of worsening of symptoms, decline in forced vital capacity (FVC) and/or increased fibrosis as measured by high-resolution computed topography (HRCT) in clinical trials [2].
- In 2020, a new ICD-10 code (J84.170) was introduced for ILD with a progressive fibrotic phenotype in diseases classified elsewhere. Additionally, ICD-10 code J84.10 is available for pulmonary fibrosis (unspecified).
- The ATS/ERS/JRS/ALAT guideline update [3] provided a recommended PPF definition, related to evidence of at least two of the following criteria occurring within the past 12 months with no alternative explanation: worsening of respiratory symptoms, radiological progression or physiological evidence of progression. Following the guideline update, and to our knowledge, no other studies have assessed real-world guideline application for PPF and use of ICD-codes within a PPF sample of patients.

OBJECTIVES

- The aim of the study was to describe the variability in guideline definition criteria used to confirm PPF in a real-world setting.

METHODS

DESIGN

Adelphi Real World PPF-ILD Disease Specific Programme™ [4,5]

Real-world data in five European countries*, the USA and Japan

*France, Germany, Italy, Spain, United Kingdom

Cross-sectional with elements of retrospective data collection

Physicians completed patient record forms for up to 5 consecutively consulting patients with PPF

INCLUSION CRITERIA

Physician:

Pulmonologists / Internal medicine specialists who saw at least 4 types of ILDs in a typical month

Rheumatologists who saw at least 4 autoimmune (AI) associated ILD in a typical month

Qualifying ILD types: Non-Specific Interstitial Pneumonia (iNSIP), Chronic / Fibrotic Hypersensitivity Pneumonitis (cHP/fHP), Unclassifiable Interstitial lung disease (uILD), **AI-ILD:** Scleroderma-Associated Interstitial lung disease (SSc-ILD), **AI-ILD**: Rheumatoid arthritis-associated interstitial lung disease (RA-ILD), **AI-ILD:** Polymyositis / Dermatomyositis Interstitial lung disease (PM/DM-ILD), **AI-ILD:** Sjögren's syndrome Interstitial lung disease (SS-ILD).

Patient:

Physician confirmed diagnosis of ILD with PPF

Adult patients, not involved in a clinical trial

DATA COLLECTION & ANALYSIS

- Data was collected between February 2024 – October 2024, based on available medical history to the physician
- Physicians provided demographic, diagnostic (including most recent ICD-10 code) and treatment data for consulting PPF patients
- Data was analysed descriptively by country and by type of ILD

CONCLUSION

- **The current application of guideline criteria used by physicians to confirm PPF in a real-world setting differs between country and type of ILD. The relatively high proportion of patients defined as PPF using a single criterion indicates a need for closer adherence to guidelines.**
- **Of patients where two or more criteria were used to confirm PPF, a considerable proportion of patients were not treated with an antifibrotic.**
- **Furthermore, although ICD-10 codes are approved for progressive pulmonary fibrosis, findings from this study highlight a low proportion of patients categorized as pulmonary fibrosis. This highlights the difficulties in identifying PPF-ILD patients in existing datasets based on standardized diagnostic classifications vs. real-world classification and management, also indicating potential underdiagnosis and undertreatment.**

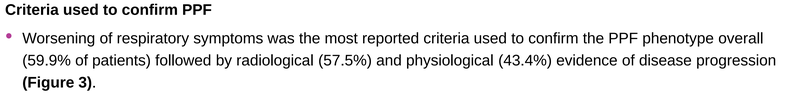
RESULTS

- Overall, 376 physicians, of which 198 were based in an ILD specialist centre (**Table 1**) provided data on 1761 patients with PPF (**Figure 1**).

Table 1. Number of physicians included (predefined splits) by country

	France (n=38)	Germany (n=50)	Italy (n=56)	Spain (n=43)	UK (n=50)	USA (n=76)	Japan (n=63)
Pulmonologist / Internal Medicine specialist (IMs)	17 / 12	35	40	32	35	46	43
Rheumatologist	9	15	16	11	15	30	20

Figure 1. Breakdown of PPFs (predefined splits) (n=number of patients)



- Mean (SD) age was 63.0 (12.4) years, 46.3% were biologically male, and 42.6% were retired. Patient demographics by country and type of ILD shown in **Table 2 and 3**, respectively.

Table 2. Patient demographics stratified by country

	France (n=221)	Germany (n=262)	Italy (n=222)	Spain (n=205)	UK (n=208)	USA (n=334)	Japan (n=309)
Mean (SD) age - survey date	61.9 (11.0)	55.7 (12.2)	63.8 (12.3)	66.3 (10.2)	63.4 (11.9)	61.2 (12.6)	69.1 (11.3)
Biological sex - Male, %	40.7	47.7	52.3	42.9	45.2	42.2	52.1
Employment – retired, %	48.0	33.2	52.7	56.6	51.0	36.5	31.1
Ethnicity - White, %	*	95.8	98.2	95.1	77.4	72.8	*

*Question relating to ethnicity is not answered in France and Japan

Table 3. Patient demographics stratified by ILD type

	iNSIP (n=418)	cHP/fHP (n=306)	uILD (n=167)	SSc-ILD (n=246)	RA-ILD (n=389)	PM/DM-ILD (n=137)	SS-ILD (n=98)
Mean (SD) age - survey date	65.7 (10.8)	65.4 (12.5)	67.6 (11.3)	57.8 (12.5)	62.6 (11.4)	58.0 (13.0)	58.8 (14.4)
Biological sex - Male, %	57.7	62.8	58.7	17.5	43.2	36.5	23.5
Employment – retired, %	49.1	48.7	58.7	30.5	42.9	19.7	29.6
Ethnicity - White, %	88.4	89.9	84.8	83.6	87.5	79.3	88.0

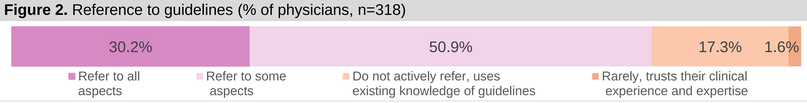
Top three tests/assessments performed to confirm diagnosis of ILD

- Overall (n=1759), FVC was the most reported test/assessment performed to confirm the diagnosis of ILD (83.3%) in patients, followed by DLco (79.1%) and HRCT scan (76.1%).

Physician-reported use of guidelines for ILD with PPF

- Of the 376 physicians, the majority (84.6%) reported referring to guidelines to diagnose, monitor or treat ILD with PPF patients. Over three quarters of physicians (n=318) reported referring to some / all aspects of guidelines when making decisions **(Figure 2).**

Figure 2. Reference to guidelines (% of physicians, n=318)



- A range of ICD-10 codes were assigned to patients: 79.7% of SSc-ILD patients, 78.8% of PM/DM ILD patients, 78.7% of RA-ILD patient, 75.1% of iNSIP patients, and 76.6% of uILD patients were assigned an ICD-10 code specific to the underlying CTD or ILD aetiology. For cHP/fHP, 76.6% of patients were assigned a non-specific ICD code. Similarly, for SS-ILD, 64.3% of patients were assigned a non-specific ILD code. Overall, 8.1% of all patients were assigned code J84.10 and 0.2% were assigned code J84.170.

Criteria used to confirm PPF

- Worsening of respiratory symptoms was the most reported criteria used to confirm the PPF phenotype overall (59.9% of patients) followed by radiological (57.5%) and physiological (43.4%) evidence of disease progression **(Figure 3)**.
  - For worsening of symptoms, dyspnea on exertion was most frequently reported (78.6% of patients).
  - For physiological evidence of progression, decline in FVC was measured for 92.5% of patients. Mean decline in absolute FVC percent predicted was 14.5%.
  - For radiological evidence of progression, new ground-glass opacity with traction bronchiectasis (43.8% of patients), increase extent or increase coarseness of reticular abnormality (40.8%), new fine reticulation (40.2%) was reported by physicians.

RESULTS

Criteria used to confirm PPF [continued]

- For 25.0% of patients, worsening of symptoms only was used to confirm PPF (**Figure 3**), ranging from 16.7% in France to 35.9% in Germany (**Figure 4a**), and 19.5% for SSc-ILD to 28.7% for uILD (**Figure 4b**).
- Two criteria were used to confirm PPF for 22.9% of patients (**Figure 3**). This ranged by country and type of ILD (**Figure 4a and 4b**).
- All three criteria were used to confirm PPF in 18.9% of patients (**Figure 3**), ranging from 11.8% in Germany to 23.9% in Spain (**Figure 4a**), and 14.3% SS- ILD to 21.1% for SSc-ILD (**Figure 4b**).

Figure 3. Overview of criteria used to confirm PPF (% of patients, n=1761)

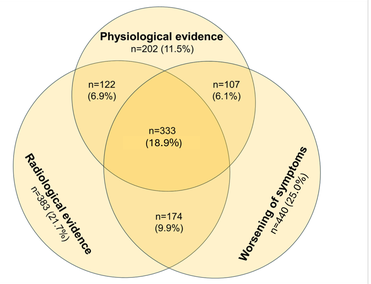
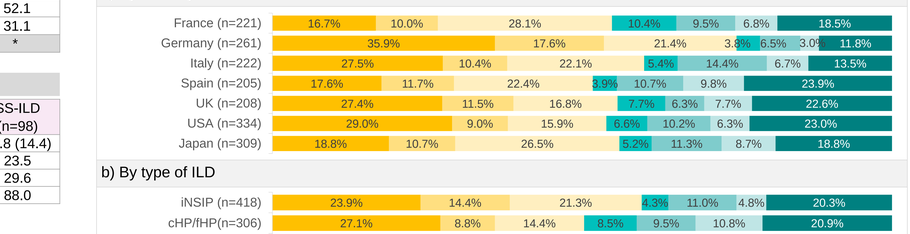
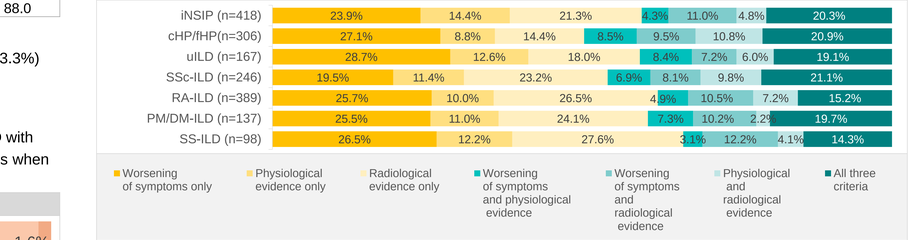


Figure 4. Use of each criteria to confirm PPF Split by a) Country and b) type of ILD (% of patients)

a) By Country



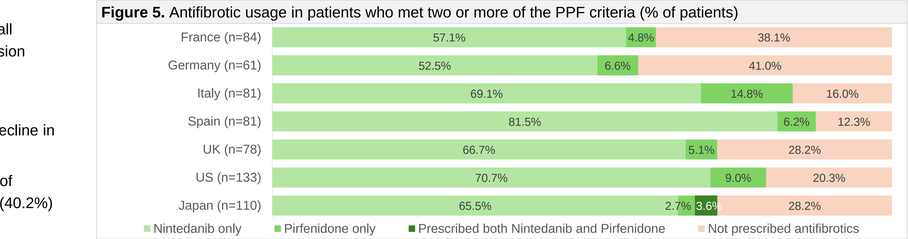
b) By type of ILD



Antifibrotic prescribing patterns for patients with confirmed PPF

- At survey date, 80.0% of patients were prescribed treatment for their PPF, whilst 14.7% had never received treatment for their PPF.
- Of patients currently prescribed PPF treatment where one criterion was used to confirm PPF, worsening symptoms (n=346), radiological (n=265) or physiological evidence (n=170), 62.1%, 67.1% and 58.9% were prescribed an antifibrotic, respectively.
- Of patients currently prescribed PPF treatment where two or more criteria were used to confirm PPF (n=628), 74.5% were prescribed an antifibrotic, most commonly nintedanib (90.6%). Data shown by country in **Figure 5**.

Figure 5. Antifibrotic usage in patients who met two or more of the PPF criteria (% of patients)



American Thoracic Society (ATS), May 16th-21st 2025, San Francisco, CA, USA

Disclosures: Pooja Chopra is an employee of Bristol Myers Squibb. Mark Small and Rebecca-Hardy Woods are employees of Adelphi Real World. Lisa Lancaster has received grant/research support from Bellerophon, Biogen, Boehringer Ingelheim, Celgene, Galactic, Galapagos NV, Genentech, Novartis, Bristol Myers Squibb, Fibrogen and Pliant; has worked as a paid consultant for AstraZeneca, Bristol Myers Squibb, DevPro Biopharma, Galapagos NV, Genentech, United Therapeutics and Veracyte; and has been paid as a speaker (disease state education) by Boehringer Ingelheim, Genentech, United Therapeutics and Veracyte. Lisa Lancaster is an employee of Vanderbilt University Medical Center (*employer at the time the study was conducted). She was appointed as Chief Medical Officer at Endeavor BioMedicines in April 2025

Funding: Data collection was undertaken by Adelphi Real World as part of an independent survey, entitled the Adelphi Real World PPF-ILD DSP. The DSP is a wholly owned Adelphi Real World product. Bristol Myers Squibb is one of multiple subscribers to the DSP.

References:

[1] W. A. Wong, C. J. Ryerson and S. A. Guler, "Progression of fibrosing interstitial lung disease," Respiratory Research, vol. 21, no. 1, 2020.
[2] K. R. Flaherty, K. K. Brown, W. U. Athol, E. Clerisme-Beaty and H. R. Collard, "Design of the PF-ILD trial: a double-blind, randomised, placebo-controlled phase III trial of nintedanib in patients with progressive fibrosing interstitial lung disease," BMJ Open Respiratory Research, vol. 17, no. 4, pp. e000212, 2017.
[3] G. Raghu, M. Remy-Jardin, L. Richeldi and C. C. Thomson, "Idiopathic Pulmonary Fibrosis (an Update) and Progressive Pulmonary Fibrosis in Adults: An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline," AJRCCM, vol. 205, no. 9, 2022.
[4] P. Anderson, J. Courcy and V. Higgins, "real-world evidence generation from patients, their caregivers and physicians supporting clinical, regulatory and guideline decisions: an update on Disease Specific Programmes," Current Medical Research Opinions, vol. 39, no. 12, pp. 1707-1715, 2023.
[5] P. Anderson, M. Karavali, N. Harris, M. Benford and J. Piercy, "Real-world physician and patient behaviour across countries: Disease-Specific Programmes - a means to understand," Current Medical Research Opinions, vol. 24, no. 11, pp. 3063-72, 2008.