Patient journey in progressive pulmonary fibrosis in Europe, USA, and Japan: real-world survey findings

PA4005

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*At the time study was conducted

Aims

- Interstitial lung diseases (ILDs) are a heterogeneous group of diffuse parenchymal lung disorders with varying degrees of inflammation and fibrosis[1]
  - Idiopathic pulmonary fibrosis (IPF) is the archetypal ILD, with progressive fibrosis manifesting as increased coughing and dyspnea and with a poor prognosis if left untreated[1,2]
- Of patients with fibrosing ILDs other than IPF, approximately a third will have progressive pulmonary fibrosis (PPF)[1]
- PPF is characterized by a decline in lung function, radiographic progression, early mortality, and a heterogeneous disease course.[1] Due to the high symptom and economic burden and poor prognosis of PPF, a robust multidisciplinary assessment is needed for diagnosis and monitoring[3]
- Real-world data from first symptom to current management are urgently needed to inform the best approach to timely diagnosis, treatment, and optimal management of PPF[3]
- Here, we aim to describe the diagnostic journey, treatment patterns, and burden of patients with PPF in Europe, the USA, and Japan

Methods

DESIGN

- Data drawn from the Adelphi Real World PPF-ILD Disease Specific Programme™ (DSP)[1,4,5]
- Real-world data in 5 European countries,[a] the USA, and Japan
  - [a]France, Germany, Italy, Spain, United Kingdom.
- Cross-sectional survey of pulmonologists, rheumatologists, or internal medicine (IM) specialists (France only)
- Physicians completed patient record forms for up to 5 consecutively consulted patients with PPF

INCLUSION CRITERIA

Physicians included:

- Pulmonologists/IM specialists seeing ≥ 4 types of ILDs[a] with PPF or IPF in a typical month
- Rheumatologists seeing ≥ 2 autoimmune-associated ILDs* in a typical month
  - [a]Qualifying ILD types: idiopathic nonspecific interstitial pneumonia (iNSIP), chronic hypersensitivity pneumonitis (cHP), unclassifiable ILD (uILD), rheumatoid arthritis-ILD (RA-ILD)*, systemic sclerosis-ILD (SSc-ILD)*, polymyositis/dermatomyositis-ILD (PM/DM-ILD)*, or Sjögren's syndrome-ILD (SS-ILD)*

Patients included:

- Physician-confirmed diagnosis of ILD with PPF
- Adult patients, not involved in a clinical trial

ANALYSIS

- Data were collected between February and October 2024
- Physicians reported data on patient demographics, treatment history, and diagnostic journey
- Patients reported treatment satisfaction and completed the Work Productivity and Activity Impairment (WPAI), EQ-5D visual analog scale (VAS), and King's Brief Interstitial Lung Disease (KBILD) questionnaires
- Analyses were descriptive. Patients with IPF were excluded

Results

Patient demographic and clinical characteristics (physician-reported)

- Overall, 376 physicians (248 pulmonologists, 116 rheumatologists, and 12 IMs) participated in the survey and completed patient record forms for 1771 patients with a physician-confirmed ILD diagnosis with PPF
- Mean (standard deviation [SD]) patient age was 63.1 (12.4) years (n = 1766) and 46.3% were male (**Table 1**); 42.7% of 1771 patients were retired from employment
- In the total sample, the types of ILD with PPF were iNSIP 23.8%, RA-ILD 22.0%, cHP 17.3%, SSc-ILD 13.9%, uILD 9.5%, PM/DM-ILD 7.9%, and SS-ILD 5.6%
- Almost half of all patients with PPF (N = 1771) had rheumatologic disease (46.8%). Other common comorbidities included anxiety (17.4%), gastroesophageal reflux disease (GERD; 17.3%), depression (15.1%), and diabetes without chronic complications (10.7%)

Table 1. Patient demographics

	Country							
	Total (N = 1771)	France (n = 221)	Germany (n = 266)	Italy (n = 223)	Spain (n = 210)	UK (n = 208)	USA (n = 334)	Japan (n = 309)
Patient age at survey, mean (SD), years[a]	(N = 1766) 63.1 (12.4)	(n = 221) 61.9 (11.0)	(n = 266) 55.8 (12.2)	(n = 223) 63.8 (12.3)	(n = 210) 66.3 (10.3)	(n = 208) 63.4 (11.9)	(n = 334) 61.2 (12.6	(n = 304) 69.1 (11.1)
Patient sex, n (%)								
Male	819 (46.3)	90 (40.7)	127 (47.7)	116 (52.0)	90 (42.9)	94 (45.2)	141 (42.2)	161 (52.1)
Female	950 (53.6)	131(59.3)	139 (52.3)	107 (48.0)	120 (57.1)	114 (54.8)	191 (57.2)	148 (47.9)
Intersex	2 (0.1)	0	0	0	0	0	2 (0.6)	0
BMI, mean (SD), kg/m²	25.1 (4.5)	25.0 (4.1)	25.4 (4.1)	24.8 (3.1)	26.0 (3.9)	26.6 (5.1)	26.7 (4.8)	21.8 (3.6)
Smoking status, n (%)								
Current smoker	95 (5.4)	13 (5.9)	21 (7.9)	24 (10.8)	6 (2.8)	11 (5.3)	10 (3.0)	10 (3.2)
Ex-smoker	792 (44.7)	87 (39.4)	136 (51.1)	116 (52.0)	80 (38.1)	85 (40.8)	133 (39.8)	155 (50.2)
Never smoked	861 (48.6)	120 (54.3)	103 (38.7)	79 (35.4)	123 (58.6)	111 (53.4)	188 (56.3)	137 (44.3)
Don't know	23 (1.3)	1 (0.4)	6 (2.3)	4 (1.8)	1 (0.5)	1 (0.5)	3 (0.9)	7 (2.3)

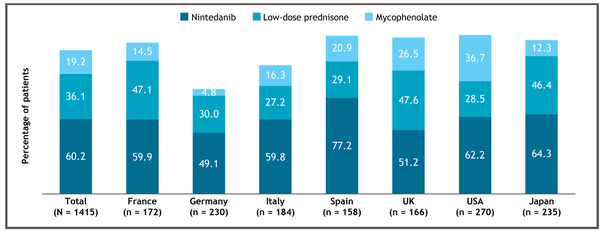
[a]Patients aged 90+ years were not included in the mean (SD) age.
BMI, body mass index.

Treatment landscape (physician- and patient-reported)

Currently prescribed treatment

- At survey date, 14.8% of 1771 patients were untreated for PPF (ranging from 10.8% in Italy to 18.4% in Japan), 79.9% were currently prescribed treatment for their PPF (ranging from 75.2% of patients in Spain to 86.5% in Germany), and 5.3% were not currently prescribed treatment for their PPF, but had been previously (ie, discontinued treatment, ranging from 1.5% in Germany to 6.7% in Italy and Spain)
- The top reasons for never prescribing treatment for PPF (n = 262) included a manageable profile without treatment (38.2%), patient concerns about side effects (28.2%), symptoms not being severe enough to warrant treatment (19.8%), and diagnosed too recently (17.9%)
- Among patients who were prescribed treatment (n = 1415), the most common treatments received were nintedanib (60.2%), low-dose prednisone (36.1%), and mycophenolate (19.2%) (**Figure 1**)
  - Across all countries, nintedanib was the most commonly prescribed treatment (ranging from 49.1% of patients in Germany to 77.2% in Spain)

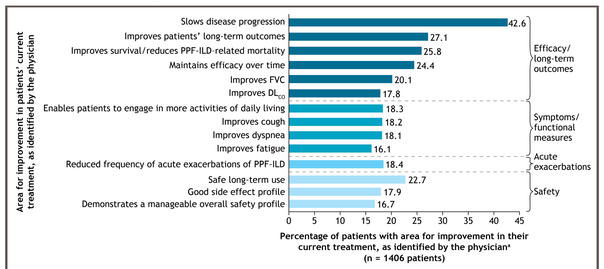
Figure 1. Top 3 prescribed treatments at survey date in the total sample and by country (physician-reported)



Satisfaction with current treatment

- Physicians reported being not very satisfied with patients' current treatment for PPF in 72.1% of cases (n = 1415; not at all, 6.2%; slightly, 28.1%; moderately, 37.8%)
  - Similarly, patients reported being not very satisfied with their current treatment for PPF in 61.1% of cases (n = 543; not at all, 4.6%; slightly, 17.1%; moderately, 39.4%)
- Physicians identified several areas in which patients' current treatment could be improved, spanning efficacy/long-term outcomes, safety profile, symptoms and functional measures, and acute exacerbations (**Figure 2**)

Figure 2. Most common physician-reported areas for improvement in the current treatments patients were receiving

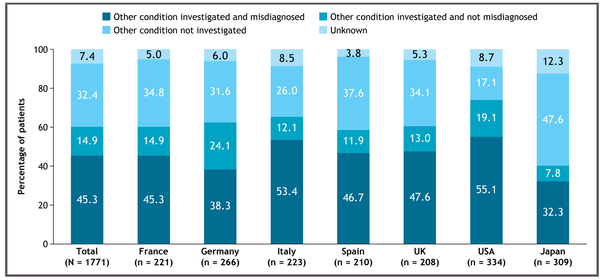


The most common areas for improvement in patients' current treatment are shown (ie, areas of improvement reported in ≥ 15% of patients).
[a]More than 1 area for improvement may have been reported for each patient.
DL_{CO}, diffusion capacity of lung for carbon monoxide; FVC, forced vital capacity.

Misdiagnosis (physician-reported)

- Almost half (45.3%) of 1771 patients were misdiagnosed with a condition other than PPF before receiving their PPF diagnosis, and 14.9% of patients had another condition investigated but were not misdiagnosed
  - Across all countries, the proportion of patients misdiagnosed with another condition ranged from 32.3% in Japan to 55.1% in the USA (**Figure 3**)
- Among all patients with a misdiagnosis (n = 802), the conditions most commonly misdiagnosed before a correct diagnosis of PPF were GERD (20.0%), chronic obstructive pulmonary disease (17.2%), and anxiety (16.6%)

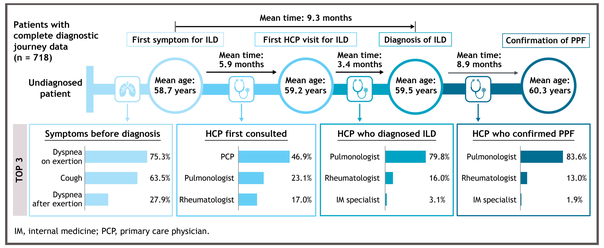
Figure 3. Patients who had undergone investigation for other conditions or misdiagnosed before ILD diagnosis, in the total sample and by country (physician-reported)



Patient journey (physician-reported)

- In total, 718 patients had complete diagnostic journey data (**Figure 4**). The mean (SD) age at first symptom of ILD was 58.7 (12.5) years
- The mean (SD) age at first healthcare practitioner (HCP) consultation for ILD was 59.2 (12.6) years. There was a mean (SD) delay of 5.9 (10.0) months between first ILD symptom and first consultation with an HCP
  - The mean (SD) delay by country ranged from 3.3 (5.5) months (Germany) to 9.3 (16.6) months (Spain)
- There was a further delay of 3.4 (5.8) months between the first HCP consultation and ILD diagnosis
  - The mean (SD) delay by country ranged from 2.2 (France, 2.9; Germany, 2.8; Japan, 8.2) months to 7.0 (7.0) months (Spain)
- The mean (SD) time from confirmed ILD diagnosis to PPF confirmation was 8.9 (17.5) months
  - The mean (SD) delay by country ranged from 1.3 (5.2) months (Germany) to 22.3 (27.5) months (Spain)
- Overall, the mean (SD) delay from first symptom to ILD diagnosis was 9.3 (12.0) months
  - The mean (SD) delay by country ranged from 5.5 (6.6) months (Germany) to 16.2 (18.2) months (Spain)

Figure 4. Patient journey for PPF in all patients with complete diagnostic journey data (physician-reported)



IM, internal medicine; PCP, primary care physician.

Patient burden and health-related quality of life (physician- and patient-reported)

- Among all patients with PPF (N = 1771), most (88.7%) were symptomatic at the survey date. The most common symptoms experienced in the last 4 weeks before the survey date (n = 1570) were dyspnea on exertion (81.3%), cough (63.2%), dyspnea after exertion (40.1%), fatigue (32.5%), and reduced exercise tolerance (24.5%)
- Nearly a third of all patients with PPF (N = 1771) required caregiver support (29.5%). For those requiring support (n = 523), this was most commonly housecleaning and home maintenance (60.8%), shopping and meal preparation (55.1%), transportation (53.3%), managing finances (25.2%), and managing medications (24.9%)

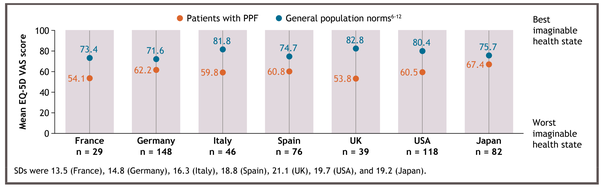
WPAI

- Among patients with PPF who completed the WPAI, median (interquartile range [IQR]) scores (where 0% represents no impairment, and 100% represents high impairment) revealed that patients reported a low level of missed work time (0% [0-12] absenteeism [n = 130]), which was expected given the mean age of the total sample was over 60 years. However, patients reported 30% (20-50) work impairment (presenteeism; n = 159), 40% (20-52) overall work impairment (n = 125), and 50% (30-70) activity impairment (n = 534)

EQ-5D VAS

- Among all patients with PPF who completed the EQ-5D VAS (n = 538; where 0 represents worst health, and 100 represents best health), a mean (SD) score of 61.2 (18.1) was reported
  - Among individual countries, patients with PPF reported lower mean EQ-5D VAS scores than the general population norms,[6-12] indicating a poorer quality of life (**Figure 5**)

Figure 5. Mean EQ-5D VAS scores by country (patient-reported)



SDs were 13.5 (France), 14.8 (Germany), 16.3 (Italy), 18.8 (Spain), 21.1 (UK), 19.7 (USA), and 19.2 (Japan).

KBILD

- Among all patients with PPF who completed the KBILD questionnaire (n = 542), a median (IQR) overall score (where 0 represents worst health, and 100 represents best health status) of 52 (47-57) was reported
  - Among individual countries, median (IQR) overall KBILD scores ranged from 48 (41-58, UK; 44-56, France) to 56 (52-64, Japan)
- Among all patients, the lowest median (IQR) KBILD domain score was breathlessness and activities (42 [36-48])
  - Among individual countries, median (IQR) KBILD scores for this domain ranged from 36 (32-44) for France to 46 (38-55) for Japan

Conclusions

- In this real-world study performed in patients with PPF between February and October 2024 across Europe, the USA, and Japan, over half of patients received nintedanib, many were prescribed a low-dose steroid or immunosuppressant only for symptom management, and many remained untreated. Nearly half of patients with PPF had concomitant rheumatologic disease, with other common comorbidities including anxiety, GERD, and depression
- Physicians and patients reported being not very satisfied with the current treatment; efficacy/long-term outcomes, safety concerns, and symptoms and quality of life were areas highlighted for improvement by physicians
- There was a high rate of misdiagnosis in the patient journey and delays in diagnosis, with a mean of ~18 months between ILD symptom onset and confirmation of PPF
- Despite treatments, many patients reported experiencing cough and dyspnea, and reduction in their quality of life
- There are several limitations with the current study, including that the participating patients may not reflect the general population of patients with PPF with other underlying ILDs. The study population may also be skewed toward those more willing to consult physicians, or with more severe disease and undergoing monitoring for treatment response, hence, untreated patients may have been underreported
- Overall, this suggests there is a critical need for improved physician awareness, earlier and more accurate diagnosis of ILD with PPF, better patient management, and more effective therapies with better safety profile to reduce PPF disease burden

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Disclosures

- Pooja Chopra reports employment by Bristol Myers Squibb at the time this study was conducted. Mark Small and Lauren Lee report employment by Adelphi Real World, which received funding from Bristol Myers Squibb as part of this research. Jeff Swigris reports payment from Bristol Myers Squibb for his work as a consultant

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