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DOI:

10.14744/ejp.2024.61960

Approaches in diagnosis, follow-up and treatment of patients with progressive pulmonary fibrosis: A questionnaire study

Ceyda Anar¹, Oğuzhan Okutan², Funda Coşkun³, Nilgün Yılmaz Demirci⁴, Berna Akıncı Özyürek⁵, Ezgi Erdem Türe⁶

ORCID:

Ceyda Anar: 0000-0002-3922-5800

Oğuzhan Okutan: 0000-0002-4660-1595

Funda Coşkun: 0000-0003-3604-8826

Nilgün Yılmaz Demirci: 0000-0001-6160-3778

Berna Akıncı Özyürek: 0000-0003-0206-7615

Ezgi Erdem Türe: 0000-0003-4285-3338

Abstract:

BACKGROUND AND AIM: Progressive pulmonary fibrosis (PPF), depending on the underlying disease, lacks a complete consensus on diagnosis, follow-up, and treatment approaches, both in our country and worldwide. This study aims to evaluate the approaches of pulmonologists and rheumatologists to PPF patients using a questionnaire.

METHODS: A web-based questionnaire consisting of 23 questions was prepared to assess the facilities of physicians' departments and their approaches to the diagnosis, follow-up, and treatment of patients with PPF. The questionnaire was sent to doctors' personal email addresses and the participants' responses were analyzed.

RESULTS: A total of 91 pulmonologists and 39 rheumatologists completed the online survey. Among the participants, 44% had less than 10 years, 35% had 10-25 years, and 21% had more than 25 years of professional experience. Multidisciplinary councils were conducted in 63% of hospitals, 71% had thoracic radiologists, and 40% collaborated with pathologists specializing in interstitial lung diseases (ILD). The most common underlying primary diseases were rheumatoid arthritis-associated ILD (46.2%) and systemic sclerosis-associated ILD (45.4%). During follow-ups, the most commonly used methods included respiratory function tests (90%), carbon monoxide diffusion tests (84%), high-resolution computed tomography (79%), and pulmonary symptoms evaluations (79%). First-line medications for the underlying disease were steroids (85%), while second-line medications were mycophenolate mofetil (58.5%). Antifibrotic drug treatment was prescribed by 85% of participants, and 78.5% of them reported that they would use a combination of antifibrotic and immunosuppressive agents. While 28% of participants reported no hesitation in the diagnosis and treatment of PPF, the absence of a multidisciplinary team (35%) and challenges in interpreting radiological findings (31.5%) were the most commonly cited obstacles.

CONCLUSIONS: This study highlights the importance of multidisciplinary councils for physicians managing patients with PPF. Although the management of PPF patients varied, the physicians' approaches to diagnosis, follow-up, and treatment of PPF patients aligned closely with recommendations in PPF guidelines.

Keywords:

Diagnosis, follow-up, progressive pulmonary fibrosis, questionnaire, treatment

How to cite this article: Anar C, Okutan O, Coşkun F, Yılmaz Demirci N, Akıncı Özyürek B, Erdem Türe E. Approaches in diagnosis, follow-up and treatment of patients with progressive pulmonary fibrosis: A questionnaire study. Eurasian J Pulmonol 2025;27:50-58.

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¹Department of Pulmonology, İzmir Katip Çelebi University, Atatürk Training and Research Hospital, İzmir, Türkiye,

²Department of Pulmonology, İstanbul Maltepe University Faculty of Medicine, İstanbul, Türkiye,

³Department of Pulmonology, Bursa Uludağ University Faculty of Medicine, Bursa, Türkiye,

⁴Department of Pulmonology, Gazi University Faculty of Medicine, Ankara, Türkiye,

⁵Department of Pulmonology, Ankara Atatürk Sanatorium Training and Research Hospital Faculty of Medicine, Ankara, Türkiye,

⁶Department of Pulmonology, Etilik City Hospital, Ankara, Türkiye

Address for correspondence:

Dr. Ezgi Erdem Türe,
Department of Pulmonology, Etilik City Hospital, Ankara, Türkiye.
E-mail: eezgjerdem@gmail.com

Received: 16-08-2024

Revised: 20-10-2024

Accepted: 03-11-2024

Published: 12-02-2025

Introduction

Interstitial lung diseases (ILDs) are characterized by inflammation and fibrosis of the lung parenchyma, with fibrotic ILD forming a specific subset of ILD.[1] There is no uniform approach to managing fibrotic ILD. Prognosis and progression are determined by the underlying disease. Progression is defined by an increase in respiratory symptoms, a decline in respiratory function (e.g., forced vital capacity [FVC], diffusing capacity of the lungs for carbon monoxide [DLCO]) and/or an increase in fibrosis on high-resolution computed tomography (HRCT). Pulmonary function tests (PFT) and HRCT are recommended for all patients at the time of diagnosis. The frequency of PFT during follow-up should be decided on a case-by-case basis.^[1,2] Evaluation of respiratory functions (e.g., FVC, DLCO) is recommended at least every 3–4 months during the first year.^[1,3] When symptom evaluation and respiratory function data are insufficient, HRCT should be utilized to assess progression.^[1,2,4] The frequency of HRCT should be tailored to the patient's clinical condition and lung function.

The precise prevalence of progressive pulmonary fibrosis (PPF) is uncertain. A recent PROGRESS study (Estimates of epidemiology, mortality and disease burden associated with progressive fibrosing interstitial lung disease in France), a real-life cohort of ILD patients, identified a progressive phenotype in nearly 25% of fibrotic ILDs other than idiopathic pulmonary fibrosis (IPF).^[5] In another real-life study, the rate of a progressive fibrotic phenotype in non-IPF ILDs was reported to range between 18% and 32%. The duration from the onset of symptoms to death ranged from 61 to 80 months. Following ILD progression, the median survival was three years.^[6]

Due to the progression of PPF varying based on the underlying disease, there is no complete consensus on its diagnosis, follow-up, or treatment approach in our country, as is the case worldwide. This study aims to evaluate the approaches to PPF management by specialists in chest diseases and rheumatology through a questionnaire.

Materials and Methods

Ethical approval for this study was obtained from the Izmir Katip Çelebi University Non-interventional Clinical Research Ethics Committee (Approval Number: 0590, Date: 26.12.2023). The study was conducted in accordance with the principles outlined in the Declaration of Helsinki.

Informed consent was obtained from all participants before administering the questionnaire. A web-based questionnaire comprising 23 questions was sent to the personal email addresses of pulmonologists and rheumatologists. Invitations to participate in the study were sent four times, at 1-month intervals starting in July 2023, through the email groups of relevant professional societies. Specialist physicians were invited to complete the questionnaire based on their personal practices and experiences.

Personal information such as participants' names, surnames, titles, and institutions was not requested. However, data regarding their areas of specialization, years of experience as specialist physicians, and the type of institution they work at (e.g., university, training hospital, or state hospital) was collected. Additionally, participants were asked whether their institution included a radiologist specializing in thoracic radiology or a pathologist specializing in the respiratory system, the number of PPF patients they encountered annually, and the areas they found most challenging during diagnosis, treatment, and follow-up. Some questions required participants to select only one answer, while others allowed for multiple choices (Appendix 1).

Statistical analysis

The number and percentage (%) of responses were used to evaluate the data provided in the questionnaire. The number of responses and percentages from descriptive statistics were used to evaluate multiple responses.

The obtained results were summarized using bar graphs. Cross-tabulations were performed to examine the distribution of opportunities and specialists in the institutions where the physicians worked. All statistical analyses were conducted using the Statistical Package for the Social Sciences (SPSS Inc., Chicago, IL, USA) version 22.0.

Artificial intelligence (AI)-assisted technologies (such as Large Language Models [LLMs], chatbots, or image creators) were not used in this study.

Results

A total of 130 specialist physicians, including pulmonologists (n=91) and rheumatologists (n=39), responded to the questionnaire. Among the physicians surveyed, 43.8% worked in university hospitals, 30.8% in general education and research hospitals, 13.1% in chest diseases hospitals, and the remaining participants worked in

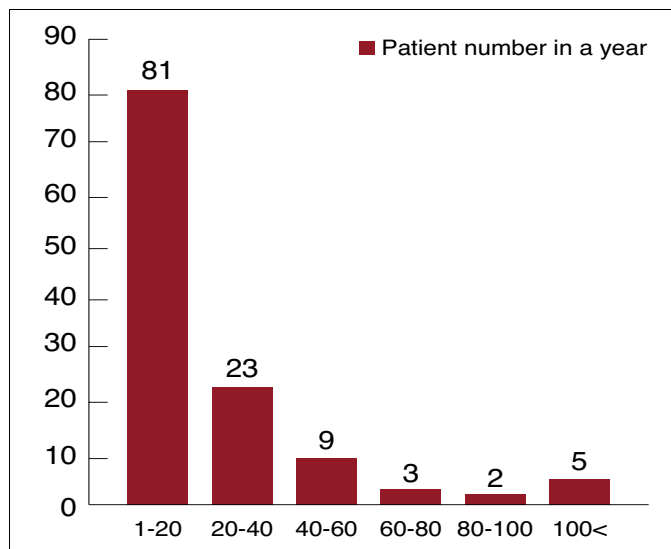


Figure 1: Number of patients examined by physicians in a year

private or state hospitals. Responses were received from 33 cities, with the majority of contributions coming from Ankara (n=28), Istanbul (n=25), and Izmir (n=24). Regarding professional experience, 23.1% (n=30) of participants had less than 5 years, 20.8% (n=27) had 5–10 years, 16.9% (n=22) had 10–15 years, 18.5% (n=24) had 15–25 years, and 20.8% (n=27) had more than 25 years of experience. Of the participants, 62.3% examined fewer than 20 patients with PPF annually, 17.7% examined 20–40 PPF patients, and 5.3% (n=7) did not follow up with any PPF patients during the year [Fig. 1]. In the centers where the physicians practiced, 75% reported the presence of both rheumatologists and pulmonologists, while 25% had only pulmonologists. The facilities available and the status of specialists in the institutions are presented in Table 1.

Rheumatoid arthritis-associated ILD (46.2%) and systemic sclerosis-associated ILD (45.4%) were the most common underlying primary diseases [Fig. 2]. Rheumatologists most commonly managed PPF patients with SSc-ILD (n=33), followed by RA-ILD (n=21) and other connective tissue disease-associated ILD (CTD-ILD) (n=13). Pulmonologists stated that they mostly follow patients with RA-ILD (42%), hypersensitivity pneumonitis (HP) (34.1%), and SSc-ILD (26%).

In patients suspected of having PPF, most physicians used at least two criteria from PPF guidelines. Among the participants, 40% preferred to assess and treat patients through a multidisciplinary council, while another 40% initiated follow-up and treatment collaboratively between pulmonologists and rheumatologists when an underlying

rheumatological disease was present. The approaches of specialists regarding follow-up, follow-up frequency, and methods used in follow-up are detailed in Table 2.

Consultation with chest disease specialists for rheumatic disease-associated ILD diagnosed by rheumatologists was requested by 83.8% of rheumatologists. The most common reason for consultation was the initiation of antifibrotic treatment (77.6%) (Table 3).

While 28% of participants reported no hesitation in the diagnosis and treatment of PPF, the most frequently cited concerns were the absence of a multidisciplinary team and difficulties in interpreting radiological findings [Fig. 3]. Among the seven participants who managed more than 80 patients per year, five reported no hesitation, while one cited the lack of a multidisciplinary team, and another expressed concerns related to the interpretation of radiological findings.

The most commonly used medications for the underlying disease were steroids (85%), mycophenolate mofetil (58.5%), and azathioprine (42%). Of the participants, 85.4% preferred to initiate antifibrotic treatment for PPF patients, and 78.5% indicated that they would use a combination of antifibrotic and immunosuppressive agents in PPF treatment. Nintedanib was selected by 50% of participants, while 43.8% preferred either nintedanib or pirfenidone (Table 4).

Discussion

The importance of PPF has grown with the publication of the 2022 American Thoracic Society (ATS)/European Respiratory Society (ETS) guideline. Physician's resources, the experience of medical centers, and the multidisciplinary approach are critical in the diagnosis and follow-up of PPF. Some findings of this study were consistent with the PPF guideline. It was concluded that pulmonologists and rheumatologists collaborate effectively in the diagnosis and treatment of PPF, with decisions often made in multidisciplinary councils.

Epidemiological data on ILD have increased in recent years; however, information on the prevalence and incidence of PPF with different etiologies remains limited. This situation may be attributed to several factors, including the heterogeneity of PPF etiologies, the small number of diagnosed patients, and the retrospective nature of patient da-

Table 1: Availability of facilities and status of specialists in institutions

	n	%
Specialty		
Pulmonologist	91	70
Rheumatologist	39	30
Work place		
University hospital	57	43.8
General education and research hospital	40	30.8
Chest diseases hospital	17	13.1
Other (private hospital, city hospital)	16	12.3
City		
Ankara	28	21.5
Istanbul	25	19.2
Izmir	24	18.4
Other cities	53	40.7
Existence of a multidisciplinary council	82	63.1
Availability of thoracic radiology	93	71.5
Presence of pathologists specializing in interstitial lung diseases	51	39.2
Rheumatologists consulting pulmonologists during the diagnostic phase of rheumatic diseases-associated ILD	109	83.2
Available tests in the hospital		
PFT	128	98.5
6MWT	111	85.3
DLCO	105	80.8
CPET	45	34.6

ILD: Interstitial lung disease, PFT: Pulmonary function test, 6MWT: 6-minute walk test, DLCO: Carbon monoxide diffusing capacity, CPET: Cardiopulmonary exercise test

tabase analyses.^[7] A study involving 36,821 patients diagnosed with ILD included cases of sarcoidosis, other fibrotic ILDs, and CTD-ILD, with respective prevalences of 24.7%, 19.5%, and 3.1%.^[8] A progressive fibrosis phenotype, other than IPF, was observed in 13–53% of patients characterized by pulmonary fibrosis.^[9] The PROGRESS study identified

a progressive phenotype in approximately 27% of fibrosing ILDs other than IPF or combined pulmonary fibrosis and emphysema. Approximately 45% of patients with a progressive fibrosis phenotype had autoimmune ILD (SSc-ILD at 26%, dermatomyositis-related ILD at 7%, and RA-ILD at 4%. Additionally, 31% were classified as unclassifiable ILD,

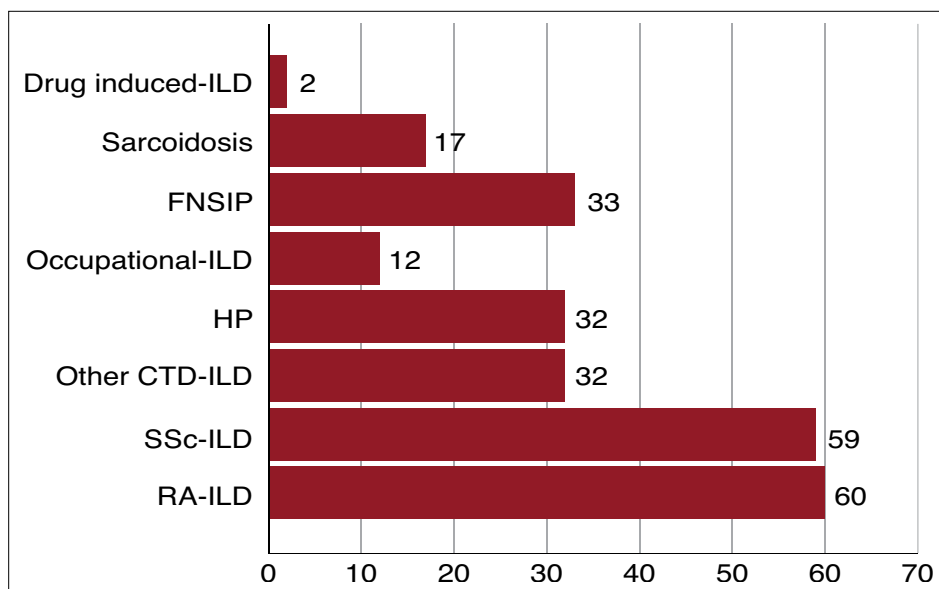


Figure 2: Most commonly encountered underlying primary disease in patients assessed as progressive pulmonary fibrosis (PPF)

ILD: Interstitial lung diseases, FNSIP: Fibrotic nonspecific interstitial pneumonia, HP: Hypersensitivity pneumonitis, CTD: Connective tissue disease-associated, SSc: Systemic sclerosis-associated, RA: Rheumatoid arthritis

Table 2: Approach to the follow-up of progressive pulmonary fibrosis (PPF)

	n	%
In the evaluation of PPF, avoid solely relying on respiratory symptoms, respiratory function tests, or radiological criteria		
Use only one criterion	16	12.3
Use at least two criteria	65	50
Use all criteria	49	37.7
Preferred options for a patient assessed or suspected to have PPF		
Referral of the patient to a multidisciplinary council for evaluation	53	40.7
For cases with an underlying rheumatological disease, conduct follow-up and treatment in collaboration with the departments of chest diseases and rheumatology	52	40
Referral to a more experienced center for further management	19	14.6
Recommend antifibrotic treatment for PPF and independently monitor the patient	6	4.6
Without medication	0	0
Frequency of follow-up for a patient being evaluated or suspected of having PPF		
Once every 3 months	64	31.2
Once every 6 months	13	6.3
Adjust follow-up frequency based on the type of underlying disease	53	25.9
Methods used during follow-ups		
PFT	117	90
DLCO	109	83.8
Assessment of pulmonary symptoms through patient inquiries	103	79.2
HRCT	102	78.5
6MWT	75	57.7
Blood tests specific to rheumatological diseases	40	32.3
Echocardiography	40	30.8
GAP score	17	13.1

PFT: Pulmonary function test, DLCO: Carbon monoxide diffusing capacity, HRCT: High-resolution computed tomography, 6MWT: 6-minute walk test, GAP: Global alignment and proportion

Table 3: Reasons for rheumatologists to request consultation with chest disease specialists for the diagnosis of rheumatic disease-associated interstitial lung disease (ILD) at your hospital

	n	%
Initiation of antifibrotic treatment	31	79.5
Evaluation of respiratory functions	15	38.5
Assessment of progressive disease despite immunosuppressive therapy	26	66.7
Others (e.g., long-term oxygen therapy, pulmonary rehabilitation etc.)	45	34.6

and 8% as chronic fibrotic HP.^[5] In a study of specialists, the most common types of the progressive fibrosis phenotype in non-IPF ILDs were idiopathic nonspecific interstitial pneumonia (iNSIP) (32%) and SSc-ILD (31%), followed by unclassified idiopathic interstitial pneumonia (IIP) and RA-ILD.^[6] In our study, the most common underlying primary disease in PPF was lung involvement of CTD, consistent with other studies, with rheumatoid arthritis being the most frequent cause. The rates of lung involvement due to systemic sclerosis and fibrotic NSIP were similar to those reported in previous studies.

According to the 2022 ATS/ERS guideline, PPF criteria include radiological evidence of pulmonary fibrosis and ILD with or without a known etiology (other than IPF), without an alternative explanation in the last year. The presence of

at least two of three criteria defines PPF: worsening respiratory symptoms, physiological evidence of disease progression, and radiological evidence of disease progression.^[1] In a study involving rheumatologists and pulmonologists, the most frequently reported reasons for assessing PPF were worsening symptom severity (27.3%), decline in lung function (e.g., FVC, DLCO), and increased fibrosis on imaging (14.2%).^[10] In our study, almost all participants reported having access to PFTs. Due to the availability of the 6-minute walk test (6MWT) and DLCO in more than 80% of cases, most participants were able to assess lung function. Consistent with guideline recommendations, half of the participants reported using at least two diagnostic criteria as outlined in the guidelines, while 37.7% stated that they evaluated patients according to all recommended criteria.

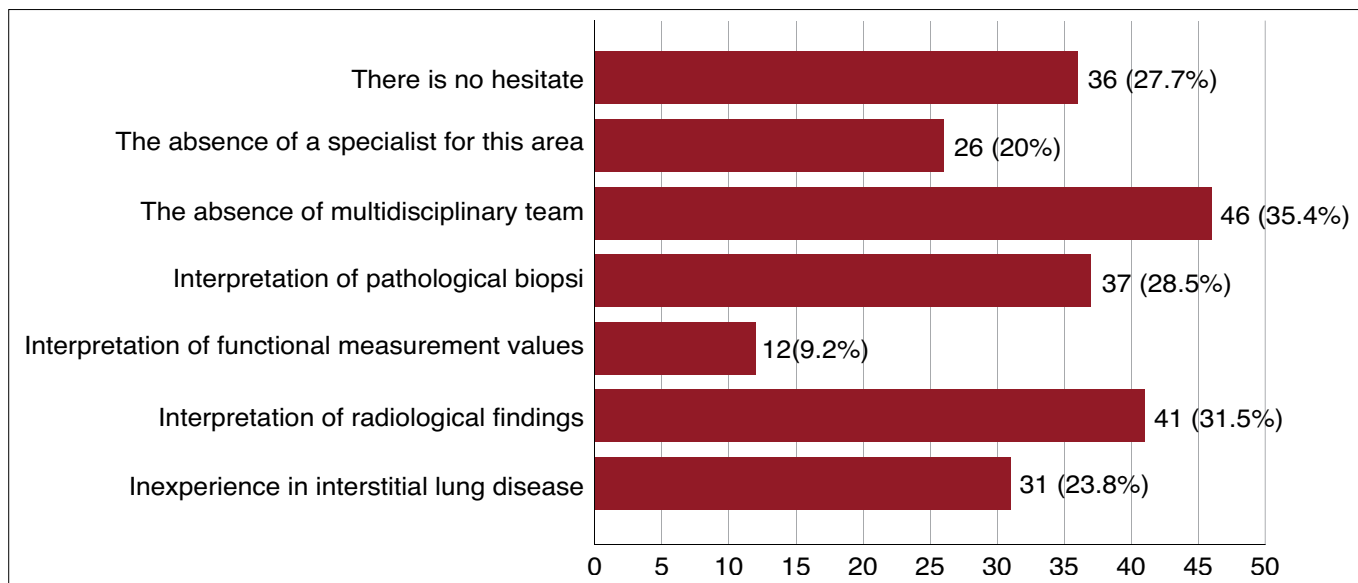


Figure 3: Situations that physicians mostly hesitate about during the diagnosis and treatment of patients with progressive pulmonary fibrosis (PPF)

Research has demonstrated that multidisciplinary approaches yield more effective results than individual assessments in the diagnosis, treatment, and monitoring of ILD.^[11,12] Multidisciplinary councils consisting of clinicians, pathologists, and radiologists play an important role in the diagnosis and management of these diseases.^[9] In our study, although most physicians worked with a pulmonary radiologist specializing in ILD, more than half did not have access to a pulmonary pathologist specializing in ILD at their institutions. In a survey conducted with physicians, 84% reported participating in multidisciplinary councils for the assessment of PPF.^[10] Similarly, in our study, the majority of physicians indicated that they could conduct multidisciplinary councils. Among 80% of the participants, the preferred approach was for the diagnosis, follow-up, and treatment processes to be evaluated in multidisciplinary councils or in collaboration with pulmonologists and rheumatologists in cases of underlying rheumatological diseases. In a multinational survey involving physicians, it was concluded that patients with non-autoimmune ILD are primarily managed by pulmonologists, while those with autoimmune ILD are often co-managed by pulmonologists and rheumatologists.^[6] Our study revealed that consultation with chest disease was frequently requested for lung involvement in rheumatic diseases diagnosed by rheumatologists. Rheumatologists reported that they most often sought assistance from pulmonologists in evaluating lung progression despite immunosuppressive treatment or in initiating antifibrotic treatment. Among participants who managed more than 80 patients annually, the majority reported no hesitation in treating PPF patients. However,

challenges included the absence of a multidisciplinary team and difficulties in interpreting radiological findings. These challenges were similar to those reported by other participants. Lack of a multidisciplinary council, difficulties in interpreting radiological findings, and challenges in the pathological evaluation of biopsies were the most frequently reported hesitations among physicians in the diagnosis and follow-up of PPF. These findings underscore the importance of a multidisciplinary approach in the diagnosis and management of PPF.

There is no uniform approach to managing fibrotic ILD. In most patients, physicians perform repeat imaging annually.^[3,6] Although a 10% reduction in FVC alone was used as an inclusion criterion in the INBUILD trial (INvestigat-

Table 4: Physicians' treatment approach for patients with progressive pulmonary fibrosis (PPF)

	n	%
Preferred agents for the underlying disease in PPF patients		
Steroids	111	85.4
Mycophenolate mofetil	76	58.5
Azathioprine	55	42.3
Methotrexate	39	30
Cyclophosphamide	37	28.5
Rituximab	31	23.8
Tocilizumab	10	7.7
Infliximab	4	3.1
Preferred antifibrotic agents		
Nintedanib	65	50
Pirfenidone	8	6.2
Nintedanib or pirfenidone	57	43.8

ing Nintedanib in Progressive Fibrosing Interstitial Lung Disease),^[13] smaller FVC reductions (5–10%) associated with symptomatic or radiological deterioration were also considered alternative inclusion criteria.^[14,15] In many cases, a comparison of serial HRCT images may be sufficient to reliably determine the degree of fibrosis progression. The interval for follow-up HRCT monitoring should be tailored to individual patient characteristics and the need for additional information about disease progression.^[2,16] In a multinational survey, most physicians reported conducting follow-ups every 2–3 months, performing respiratory function tests every 3–6 months, and HRCT scans every 6–12 months.^[6] In our study, some physicians indicated that the frequency of follow-up was determined according to the underlying disease type, consistent with literature recommendations, while most stated they routinely conducted follow-ups every three months. The majority of physicians reported using PFTs, DLCO, symptom evaluations, and HRCT in post-diagnosis follow-ups, aligning with guideline recommendations.

There is no standard pharmacological treatment regimen for all cases of PPF. Treatments are individualized based on the underlying disease. Studies have shown that, depending on the subtype of PPF, glucocorticoids are the most frequently used treatments. The most commonly used immunosuppressive agents include mycophenolate mofetil, cyclophosphamide, and azathioprine.^[1,5,6,10] Corticosteroids are commonly prescribed for various types of ILD and can lead to short-term improvements in lung function, but no randomized clinical trials have evaluated their effectiveness specifically in patients with PPF. Corticosteroids are widely used in the treatment of SSc-ILD;^[17] however, there is limited evidence to support their effectiveness in these patients.^[18,19] The recent ATS guidelines for the treatment of SSc-ILD primarily recommend mycophenolate as the first-line treatment, with cyclophosphamide, rituximab, and tocilizumab suggested as second-line options.^[20] In our study, rheumatoid arthritis (RA) and SSc were the most commonly observed conditions. In our survey, similar to findings in the literature, corticosteroids were the most preferred agents for treating the underlying disease in PPF patients. Additionally, mycophenolate mofetil and azathioprine were the most commonly preferred immunosuppressant agents.

A dilemma persists regarding the addition of antifibrotic agents to immunosuppressive therapy. There is insufficient evidence to support the combined use of immunosuppressive agents and antifibrotic drugs at the time of initial diagnosis. It is recommended to evaluate the need

for antifibrotic agents after 3–6 months of follow-up to assess disease progression.^[13] Recent studies have suggested that antifibrotic therapy may benefit selected patients with scleroderma-associated fibrotic ILD.^[20,21] However, antifibrotic drugs should not be considered first-line treatment for fibrosis-related ILDs other than IPF, SSc-ILD, and RA-ILD.^[1] Consistent with recent publications, the majority of physicians in our study indicated that they would initiate antifibrotic treatment during follow-up for PPF patients and preferred the combined use of immunosuppressive and antifibrotic therapies.

Data regarding the use of antifibrotic agents in PPF treatment are increasing. When nintedanib was administered to patients with ILD and progressive fibrosis, a slower decline in FVC was observed.^[13,21] In a survey study conducted with clinicians, 30.9% of PPF patients received nintedanib, and 66% of those receiving nintedanib were on combination therapy.^[10] In a study of patients with progressive fibrosis in unclassifiable ILD, a smaller decline in FVC was noted in patients receiving pirfenidone.^[22] However, no studies in the literature have directly compared the effectiveness of nintedanib and pirfenidone in PPF patients. Consistent with the literature, there was no clear consensus among participants regarding antifibrotic preferences. While half of the participants preferred nintedanib as an antifibrotic, approximately 44% indicated they would choose either nintedanib or pirfenidone depending on the patient.

The most significant limitation of this study was the low number of chest diseases and rheumatology physicians who participated in the survey, along with the limited participation of physicians from non-tertiary center hospitals. Additionally, we believe that differences in follow-up frequency and treatment approaches may arise due to the heterogeneous nature of PPF. We believe that the results were influenced by the majority of participants being from Ankara, Istanbul, Izmir, and university or general education research hospitals.

Conclusion

The results of this study were generally consistent with the literature. We believe that results are influenced by the fact that the majority of participants are from general education research hospitals and university hospitals. Most physicians expressed the opinion that PPF patients should be managed based on decisions made by multidisciplinary councils, including pulmonologists,

rheumatologists, radiologists, and pathologists. Due to the heterogeneous nature of PPF, physicians' approaches to follow-up and treatment decisions vary on a patient-by-patient basis. There is a need for multicenter studies involving a larger number of physicians to better understand the prevalence of the disease and physicians' approaches to diagnosis, treatment, and follow-up.

Ethics Committee Approval

The study was approved by the Izmir Katip Çelebi University Non-interventional Clinical Research Ethics Committee (No: 0590, Date: 26/12/2023).

Authorship Contributions

Concept – C.A., O.O., N.Y.D.; Design – O.O., N.Y.D.; Supervision – F.C., B.A.Ö.; Materials – O.O., C.A., E.E.T.; Data collection &/or processing – C.A., E.E.T.; Analysis and/or interpretation – C.A.; Literature search – C.A., E.E.T.; Writing – C.A., E.E.T.; Critical review – F.C., B.A.Ö.

Conflicts of Interest

There are no conflicts of interest.

Use of AI for Writing Assistance

No AI technologies utilized.

Financial Support and Sponsorship

Nil.

Peer-review

Externally peer-reviewed.

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Appendix 1: Survey questions

1. What is your specialty?
2. How long have you been practicing in your specialty?
3. What city is your workplace located in?
4. What is the name of your institution?
5. Does your hospital have specialists in rheumatology and/or chest diseases?
6. How many patients with progressive pulmonary fibrosis (PPF) have you examined in the past year?
7. Are multidisciplinary meetings conducted in your hospital to assess and discuss patients suspected of having PPF?
8. Does your hospital have radiologists specializing in thoracic radiology?
9. Does your hospital have pathologists specializing in interstitial lung diseases?
10. Which tests are available in your hospital for physiological assessment? (You may select multiple answers)
11. Do rheumatologists in your hospital request consultations with chest disease specialists for the diagnosis of rheumatic diseases-associated interstitial lung disease (ILD)?
12. If you are a rheumatologist, in which situations do you consult chest diseases specialists regarding fibrotic interstitial lung diseases? (You may select multiple answers)
13. Among the patients you evaluate as having PPF, what is the most commonly encountered underlying primary disease?
14. Which parameters do you use to evaluate PPF?
15. During the assessment of a PPF patient, which areas do you experience the most hesitation in? (You may select multiple answers)
 - a. Lack of experience with interstitial lung diseases
 - b. Interpretation of radiological findings
 - c. Interpretation of functional measurement values
 - d. Interpretation of pathological biopsy results
 - e. Absence of a multidisciplinary team
 - f. Lack of a specialist in this area
 - g. I do not experience hesitation
16. Which of the following options do you prefer for a patient you have assessed or suspected as having PPF?
 - a. I refer the patient to an experienced center.
 - b. I recommend antifibrotic treatment for PPF and follow up independently.
 - c. I monitor the patient without prescribing medication.
 - d. If there is an underlying rheumatological disease, I initiate follow-up and treatment in cooperation with chest diseases and rheumatology specialists.
 - e. If a multidisciplinary council is available, I evaluate the patient there.
17. How frequently do you follow up with a patient you have assessed or suspected as having PPF?
18. Which tests do you most commonly use while following up with a patient you have assessed or suspected as having PPF? (You may select multiple answers)
20. Which immunosuppressive agents do you most commonly use for treating the underlying disease in PPF (excluding IPF)?
21. Do you initiate antifibrotic treatment for a patient you have assessed or suspected as having PPF?
22. Do you use a combination of antifibrotic treatment and immunosuppressive therapy for a patient you have assessed or suspect as having PPF?
23. If you decide to start antifibrotic treatment, which agent do you prefer?

IPF: Idiopathic pulmonary fibrosis