IPF, PPF: Why patient and clinician views on disease impact diverge

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Idiopathic pulmonary fibrosis (IPF) and progressive pulmonary fibrosis (PPF) remain devastating conditions, characterised by relentless decline in lung function and profound impact on quality of life. Despite advances in antifibrotic therapies, a significant gap persists between the clinical metrics used to assess disease and the lived experience of patients. This disconnect complicates management and highlights unmet needs in patient-centred care.

KEY TAKEAWAYS

- The Pivot: Patient-reported outcomes and clinician assessments often do not align in IPF and PPF, indicating a need for more holistic evaluation.
- The Data: Patients consistently report a higher burden of symptoms and functional limitations than clinicians perceive.
- The Action: Incorporate patient-reported outcome measures (PROMs) more systematically into routine clinical practice to bridge the perception gap.

Idiopathic pulmonary fibrosis is a chronic, progressive, and ultimately fatal lung disease of unknown aetiology. It is defined by a specific pattern of usual interstitial pneumonia (UIP) on high-resolution computed tomography (HRCT) or surgical lung biopsy. The disease leads to irreversible scarring of the lung tissue, impairing gas exchange and causing progressive dyspnoea, cough, and fatigue. The median survival from diagnosis is typically short, often comparable to aggressive malignancies, underscoring the urgent need for effective interventions and comprehensive care.
Progressive pulmonary fibrosis, on the other hand, describes a phenotype of various interstitial lung diseases (ILDs) that share a common trajectory of worsening fibrosis, declining lung function, and increased mortality, despite treatment for the underlying ILD. This progressive phenotype can affect patients with conditions such as chronic hypersensitivity pneumonitis, unclassifiable ILD, and connective tissue disease-associated ILD. Recognising PPF is critical, as it allows for the consideration of antifibrotic therapies that have demonstrated efficacy in slowing disease progression.

The Patient Experience: A Daily Struggle

Patients with IPF and PPF face a daily battle against debilitating symptoms that profoundly affect their physical and mental well-being. Dyspnoea is almost universal, often progressing from exertional to rest dyspnoea, severely limiting daily activities. Chronic cough, frequently dry and intractable, can lead to exhaustion, social embarrassment, and sleep disturbance. Fatigue is another pervasive symptom, often underestimated by clinicians, but reported by patients as one of the most impactful aspects of their disease. These symptoms collectively erode quality of life, leading to social isolation,

anxiety, and depression. Many patients report a significant loss of independence, relying on family members for tasks they once performed effortlessly. This often goes unrecognised in routine clinical encounters, where the focus remains on lung function tests and imaging. For a deeper dive into these challenges, consider our previous coverage on IPF/PPF: Patient and Pulmonologist Perspectives Highlight Care Gaps.

The Pulmonologist's Lens: Clinical Metrics and Guidelines

Pulmonologists typically assess disease progression and treatment efficacy using objective measures. Forced vital capacity (FVC) is the cornerstone, with a decline of ≥10% over 6-12 months often considered a clinically meaningful progression. Other metrics include diffusing capacity of the lung for carbon monoxide (DLCO), six-minute walk distance (6MWD), and changes on HRCT scans. These parameters are essential for monitoring disease and guiding therapeutic decisions, but they do not fully capture the patient's subjective experience. Guidelines from professional bodies, such as the European Respiratory Society (ERS) and the American Thoracic Society (ATS), emphasize these objective measures for diagnosis and management. But these guidelines, while robust in their clinical recommendations, sometimes fall short in integrating the patient's voice into the assessment of treatment success or failure. Clinicians, often pressed for time, may inadvertently prioritise these quantifiable markers over detailed discussions about symptom burden and functional limitations.

Bridging the Perception Gap

The disconnect between patient and pulmonologist perspectives is a persistent challenge. Patients consistently report a higher burden of symptoms and functional impairment than their clinicians perceive. This discrepancy can lead to undertreatment of symptoms, missed opportunities for supportive care, and a general feeling among patients that their concerns are not fully understood. For example, a patient might report severe fatigue, but if their FVC remains stable, a clinician might not fully appreciate the impact of that fatigue on daily life. This gap is not due to a lack of empathy, but rather a systemic reliance on objective measures that do not always correlate directly with subjective well-being. Incorporating patient-reported outcome measures (PROMs) more systematically into routine clinical practice could help bridge this gap. Tools like the St. George's Respiratory Questionnaire (SGRQ) or the King's Brief Interstitial Lung Disease (K-BILD) questionnaire offer structured ways to capture patient perspectives on symptoms, activity, and quality of life. These instruments provide valuable complementary information to traditional physiological measures, offering a more holistic view of the patient's condition. The Oxford Handbook of Respiratory Medicine provides a concise overview of these assessment tools and their application in practice.

The Role of Antifibrotic Therapies

Antifibrotic therapies, such as pirfenidone and nintedanib, have demonstrated efficacy in slowing the rate of FVC decline in patients with IPF and, more recently, in those with PPF. These drugs represent a significant advance, offering the first real hope of modifying the disease course. But while these agents slow physiological decline, their impact on patient-reported symptoms and quality of life can be less dramatic or take longer to manifest. Patients may still experience progressive dyspnoea and fatigue, even with stable FVC, leading to frustration and a feeling that the treatment is not working. This highlights the need for realistic expectations management and a focus on comprehensive supportive care alongside antifibrotic treatment. Understanding the nuances of ILD diagnosis and management is crucial for patient outcomes, and our practical ATS 2026 update on ILD provides further context on evolving strategies.

Beyond the Clinic: Multidisciplinary Care and Support

Effective management of IPF and PPF extends beyond pharmacological interventions. A multidisciplinary approach involving pulmonologists, specialist nurses, physiotherapists, occupational therapists, and palliative care specialists is essential. Pulmonary rehabilitation programs can significantly improve exercise capacity and reduce dyspnoea, even in advanced disease. Oxygen therapy, when indicated, can alleviate hypoxemia and improve functional status. Palliative care, often introduced late in the disease trajectory, should be integrated early to address symptom burden, psychological distress, and end-of-life planning. These supportive measures, while not directly targeting the underlying fibrosis, have a profound impact on the patient's daily life and overall well-being. They represent areas where patient and clinician perspectives can more readily align, as the benefits are often immediately felt by the patient. The challenge lies in ensuring equitable access to these services across different healthcare systems. For instance, the management of other complex conditions also benefits from a multidisciplinary approach, as seen in ITP management, where bridging clinician and patient perspectives is equally vital.

The Unanswered Questions

The field still grapples with how to best integrate patient values and preferences into treatment decisions. While objective measures are indispensable, the subjective experience of living with a progressive fibrotic lung disease demands equal attention. Future research needs to focus not only on novel antifibrotic agents but also on interventions that directly address symptoms and improve quality of life. Developing and validating PROMs that are sensitive to changes in IPF and PPF, and ensuring their routine use in clinical trials and practice, will be critical. The ultimate goal is to move towards a truly patient-centred model of care, where both the numbers and the narrative contribute to a complete understanding of disease impact and treatment success. This requires a shift in mindset, recognising that a stable FVC does not always equate to a stable patient experience.

CLINICAL IMPLICATIONS

The persistent divergence between what clinicians measure and what patients experience in IPF and PPF is not merely an academic point; it has direct consequences for patient care. If we are not asking the right questions about symptoms and daily function, we are missing opportunities to intervene with supportive therapies that genuinely improve quality of life, even when lung function continues its inevitable decline. Relying solely on FVC and DLCO, while clinically expedient, creates a blind spot. Antifibrotic drugs slow progression, but they do not eliminate symptoms. Clinicians must actively solicit patient-reported outcomes, perhaps through validated questionnaires, to gain a more complete picture. This requires dedicated time and a willingness to look beyond the spirometry report.

Industry, too, has a role. Future clinical trials should integrate PROMs as co-primary or key secondary endpoints, not just exploratory measures. Demonstrating an impact on quality of life, alongside physiological benefits, would provide a more compelling and patient-relevant evidence base for new therapies.

A truly patient-centred approach means acknowledging that the patient's subjective experience is as valid a measure of disease burden and treatment success as any objective physiological parameter. It is a shift from treating a lung disease to caring for a person living with a lung disease.

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REFERENCES

1. 1. Muri J, Durcová B, Slivka R, et al. Idiopathic Pulmonary Fibrosis: Review of Current Knowledge. Physiol Res. 2024;73(4):487-497. doi:10.33549/physiolres.935322
2. 2. Moss BJ, Ryter SW, Rosas IO. Pathogenic Mechanisms Underlying Idiopathic Pulmonary Fibrosis. Annu Rev Pathol. 2022;17:515-546. doi:10.1146/annurev-pathol-042320-030240
3. 3. Richeldi L, Collard HR, Jones MG. Idiopathic pulmonary fibrosis. Lancet. 2017;389(10082):1941-1952. doi:10.1016/S0140-6736(17)30866-8
4. 4. Koudstaal T, Wijsenbeek MS. Idiopathic pulmonary fibrosis. Presse Med. 2023;52(3):104166. doi:10.1016/j.lpm.2023.104166
5. 5. King TE Jr, Pardo A, Selman M. Idiopathic pulmonary fibrosis. Lancet. 2011;378(9807):1949-61. doi:10.1016/S0140-6736(11)60052-4
6. 6. Benegas Urteaga M, Ramírez Ruz J, Sánchez González M. Idiopathic pulmonary fibrosis. Radiologia (Engl Ed). 2022;64 Suppl 3:227-239. doi:10.1016/j.rxeng.2022.10.009

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