Survey to Understand the Management of Progressive Pulmonary Fibrosis in the Real-World Setting – The PROFIBRO Survey

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Introduction

Progressive pulmonary fibrosis (PPF) constitutes about 13-40% of fibrotic interstitial lung diseases (F-ILD). Current treatment strategies for PPF across the globe differ depending on the underlying disease diagnosis. There is no data on how PPF is diagnosed & treated in India.

Methods

This survey was conducted in 2 phases. In phase 1, the questionnaire was administered to clinicians attending CME programmes on respiratory diseases during the period January-December 2024; in phase 2, the survey was conducted digitally from January 2025 – August 2025. Results are expressed as percentages based on the number of responses obtained.

Results

300 clinicians across 24 Indian states participated in the survey; 93% were pulmonologists and the average years in practice was 14.3 ± 9.3 years. 55% of clinicians reported that 10–30% of their F-ILD patients develop PPF, whereas 27% clinicians reported < 10% patients. 51% of clinicians reported diagnosing PPF based on 2022 ATS/ERS/ALAT/JRS criteria, while 34% relied on clinical judgement, radiology & lung function. 38% of clinicians reported that they labelled F-ILD as PPF when progression occurred over a 12-month period, while 26% and 23% of clinicians labelled F-ILD as PPF regardless of the duration of fibrosis progression & progression over 6 months, respectively. F-HP (48%) & RA-ILD (16%) were the most reported ILDs underlying PPF diagnosis. The immunosuppressant of choice in >50% of non-IPF ILD patients before establishment of progression was prednisone (50%), mycophenolate (49%) and mycophenolate with low dose prednisone (49%). Once progression was established, antifibrotics (75% nintedanib, 40% pirfenidone) were the most frequently reported additional treatments of choice for >50% of non-IPF ILD patients. In addition, 32–35% of clinicians reported increasing the dose of prednisone and/or mycophenolate in this setting. 41% of clinicians reported considering antifibrotic combination in those who progressed on antifibrotic monotherapy.

Conclusion

Management of PPF have evolved over the years. Both steroid and steroid sparing agents are the preferred therapies before establishment of progression. Nintedanib was the preferred antifibrotic in established PPF, while dual antifibrotics was preferred in those who progressed on monotherapy.