Insight Brief

Beyond IPF: The Evolution of Progressive Pulmonary Fibrosis

How evolving disease classifications are reshaping clinical development, investment, and commercial opportunity

PRERNA KUNDARA, Associate Consultant, IQVIA Consulting Services
PRITI GIROTRA, PhD, Associate Principal, IQVIA Consulting Services
ANIMESH JHA, Principal, IQVIA Consulting Services

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From diagnosis-defined disease to a progression-defined opportunity

Interstitial Lung Disease (ILD) comprises more than 200 disorders with varying causes and clinical courses. Across this heterogeneous group, some patients develop a common pattern of progressive fibrosis marked by worsening respiratory symptoms, declining lung function, and increasing fibrosis on imaging.[1,2] Recognizing this shared disease behavior is changing pulmonary fibrosis from a set of separate disease markets into a broader, progression-defined opportunity.

Historically, this pattern was described as Progressive Fibrosing Interstitial Lung Disease (PF-ILD). The concept established that patients with different underlying diagnoses can follow a similarly progressive course associated with irreversible lung damage, functional decline, and increased mortality.[3]

The 2022 ATS/ERS/JRS/ALAT Clinical Practice Guideline introduced Progressive Pulmonary Fibrosis (PPF) as a standardized framework for identifying this phenotype. PPF is not a distinct disease. It describes progressive fibrosis in patients with fibrotic ILDs other than Idiopathic Pulmonary Fibrosis (IPF). The shift is strategically important: patients are increasingly identified by progressive disease behavior, not only by the original diagnosis.[4]

Under the guideline, PPF is diagnosed in patients with non-IPF fibrotic ILD when at least two of three domains show progression within one year, with no alternative explanation: worsening respiratory symptoms, physiological deterioration, and/or radiological progression. This multidimensional definition provides a more consistent basis for clinical practice, research, and patient identification.[4]

Table 1: Diagnostic criteria for Progressive Pulmonary Fibrosis (PPF) according to the ATS/ERS/JRS/ALAT Clinical Practice Guideline

EVIDENCE DOMAIN	CRITERIA
Clinical evidence	Worsening respiratory symptoms
Physiological evidence	Physiological evidence of disease progression, including one or both of the following: • Absolute decline in FVC ≥ 5 percentage points predicted • Absolute decline in DLCO ≥ 10 percentage points predicted
Radiological evidence	Radiological evidence of disease progression, at least one or more of the following: • Increased extent or severity of traction bronchiectasis and bronchiolectasis • New ground-glass opacity with traction bronchiectasis • New fine reticulation • Increased extent or increased coarseness of reticulations • New or increased honeycombing • Increased lobar volume loss

Note: Forced Vital Capacity (FVC) is the maximum volume of air that can be forcibly exhaled after maximal inhalation.
Diffusing Capacity of the Lung for Carbon Monoxide (DLCO) measures the lung's ability to transfer gas from the alveoli into the bloodstream and is assessed using carbon monoxide as a tracer gas.

The significance of PPF extends beyond terminology and clinical management. By recognizing progressive fibrosis across multiple underlying ILDs, the framework broadens the population relevant to therapeutic development and creates a more consistent basis for patient identification and market evaluation. For biopharmaceutical companies, this affects clinical program design, evidence generation, investment priorities, and commercial planning.[4]

A broader view of the progressive fibrosis population

IPF remains the most extensively studied fibrotic lung disease. However, recognition of progression across multiple fibrotic ILDs has expanded epidemiological interest beyond a single disease entity. Recent studies provide an early indication of the broader burden of progressive fibrosis across the ILD spectrum. Because the available estimates use different methodologies and case definitions, they should be read as indicators of scale rather than as a direct comparison of market size.

Table 2: Published incidence and prevalence estimates for IPF and PPF in the US and Europe

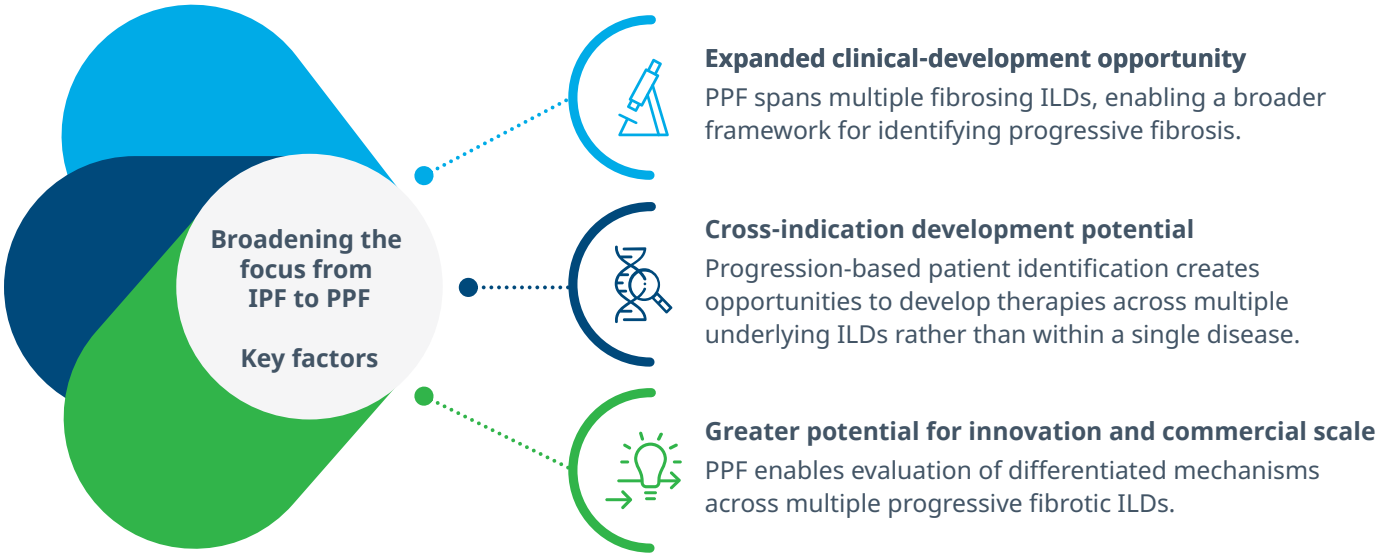
DISEASE	GEOGRAPHY	INCIDENCE (PER 100,000 PERSON-YEARS)	PREVALENCE (PER 100,000 PERSONS)	ESTIMATED PREVALENT CASES*
Idiopathic Pulmonary Fibrosis (IPF)	US[5]	9.0-18.4	32.8-67.1	174K
	Europe[6]	5.1	14.6	109K
Progressive Pulmonary Fibrosis (PPF)	US[7]	27.6	60.7	212K
	Europe[7]	6.7	30.2	225K

*Note: *Absolute prevalent cases are illustrative estimates derived by applying published prevalence rates to the corresponding population. Estimates for IPF and PPF originate from separate studies and should not be interpreted as directly comparable because of differences in study methodologies, case definitions, observation periods, and geographic coverage.*

Why is innovation expanding beyond IPF?

PPF creates a development framework that reaches beyond a single underlying diagnosis. Although fibrotic ILDs have diverse etiologies, they share pathobiological features including fibroblast activation, extracellular matrix deposition, and progressive tissue remodeling. This provides a rationale for targeting fibrotic progression across disease subtypes. Clinical evidence for nintedanib (Ofev) across progressive fibrosing ILDs, together with formal recognition of PPF in the 2022 guideline,[4] has reinforced this progression-based paradigm. The strategic opportunity is therefore broader than a larger addressable population: PPF supports cross-indication development, evaluation of differentiated mechanisms, and investment in the capabilities required to identify and manage progressing patients.

Figure 1: Key factors driving expansion of pulmonary fibrosis innovation beyond IPF



Current treatment landscape for PPF

The PPF treatment landscape is expanding. Antifibrotic therapy complements management of the underlying ILD, with the current goal of slowing progression and preserving lung function.

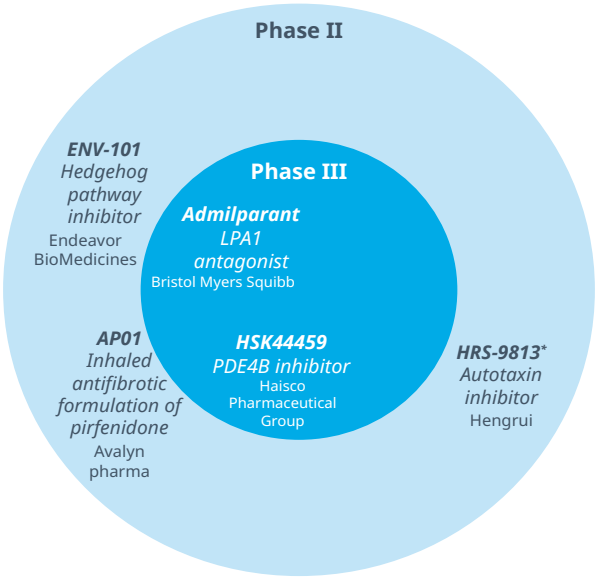
Table 3: Approved treatments for Progressive Pulmonary Fibrosis (PPF)

ASSET (BRAND NAME)	CLASS/MECHANISM	APPROVAL STATUS	KEY CLINICAL EVIDENCE
Nintedanib (Ofev)	**Multi-targeted tyrosine kinase inhibitor**	• US – March 2020[8] • EU – July 2020[9] • UK – October 2021[10]	**The Phase III INBUILD trial** demonstrated a significant reduction in the annual rate of FVC decline versus placebo in patients with progressive fibrosing ILD.[11] The **2022 ATS/ERS/JRS/ALAT guideline** conditionally recommends nintedanib for PPF following appropriate management of the underlying ILD.[4]
Nerandomilast (Jascayd)	**Preferential PDE4B inhibitor**	• US – December 2025[12] • EU – July 2026[13] • UK – July 2026[14]	**The Phase III FIBRONEER-ILD** trial demonstrated a significantly smaller decline in FVC versus placebo at 52 weeks in patients with PPF.[15]

How is the pipeline changing?

The emerging pipeline is diversifying beyond established antifibrotic approaches. Programs are evaluating differentiated mechanisms and delivery strategies intended to act on the biological drivers of fibrosis, improve pulmonary targeting, or address tolerability limitations.

Figure 2: Key emerging therapies under clinical development for PPF



Key emerging therapies targeting PPF

Note: Pipeline status as of 13 November 2026; selected assets, not exhaustive;
*HRS-9813 is a China-only trial (NCT07192939)
Source: Clinicaltrials.gov

Beyond established antifibrotics: The pipeline is diversifying toward differentiated mechanisms, including LPA1, integrin, CTGF, inflammatory, and Hedgehog pathways.

Optimizing efficacy and tolerability: Emerging approaches are exploring differentiated delivery strategies, including inhaled formulations designed to enhance pulmonary targeting while potentially reducing systemic exposure. For example, AP01 is an inhaled formulation of pirfenidone designed to deliver the therapy directly to the lungs, with the potential to reduce systemic exposure and associated tolerability challenges. Admilparant is among the leading emerging therapies in PPF, with Phase II data showing meaningful efficacy (rate of change in ppFVC over 26 weeks in patients with PPF was –4.3% with placebo versus –2.9% and –1.1% with the 30-mg and 60-mg admilparant arms, respectively), and a favorable safety and tolerability profile, supporting its advancement into the Phase III ALOFT program.[16]

Phenotype-based development: Clinical programs are increasingly enrolling patients based on progressive fibrotic disease behavior across non-IPF ILDs, reinforcing the shift toward phenotype-based development.

What will determine future success in PPF?

Recent advances have expanded treatment options, but PPF care still primarily focuses on slowing decline rather than reversing established fibrosis. Future success will depend on three connected priorities: identifying progressors earlier, demonstrating clinically meaningful differentiation, and optimizing the broader care pathway.

Figure 3: Key unmet needs shaping the next phase of innovation in PPF

1. FROM SLOWING DECLINE TO MODIFYING DISEASE	2. IDENTIFYING PROGRESSORS EARLIER	3. OPTIMISING THE BROADER CARE PATHWAY
• Nerandomilast[17] and nintedanib[11] slow FVC decline but do not reverse established fibrosis or restore lost lung capacity • Continued disease progression and symptoms impact quality of life despite treatment • Gastrointestinal adverse events can affect long-term tolerability and adherence • The need for upstream disease modification is driving emerging LPA1 antagonists, such as BMS admilparant that target pro-fibrotic signaling[18]	• Heterogeneous disease trajectories across fibrosing ILDs make early identification of progressors challenging • Variability in imaging interpretation and progression assessment can delay recognition[19] • No predictive biomarkers are currently validated for routine clinical use • Fibrosis-related markers (IGFBP2, PTX3, LGALS1, LGALS9, MMP2) show potential but require further validation[20]	• Physicians face uncertainty around the timing of treatment initiation and the use of immunomodulators versus antifibrotics[4] • Limited guidance exists on combination strategies, escalation, and monitoring approaches[21] • Many patients retain substantial functional limitations despite pharmacologic treatment[19] • Key enablers include multidisciplinary ILD teams, pulmonary rehabilitation, oxygen therapy, education, and adherence support[4]



Investment is broadening beyond therapeutic assets

Pulmonary fibrosis continues to attract pharmaceutical deal activity, with IPF accounting for most identified transactions between 2022 and 2026. However, the emerging PPF landscape points to a broader strategic requirement. Activity is extending beyond novel therapeutics to diagnostics, clinical development, patient assessment, and technology-enabled approaches.

This distinction matters. In PPF, value creation may depend not only on acquiring or licensing an asset, but also on building the ecosystem required to find progressing patients, generate evidence across heterogeneous ILDs, and support treatment decisions. Investment evaluation should therefore consider diagnostic, biomarker, monitoring, and care-pathway capabilities alongside therapeutic innovation.

Figure 4: Illustrative examples of recent deal activity relevant to PPF showcasing an expanding investment landscape

2022	2023	2024	2025
Expanding therapeutic options	**Earlier disease identification**	**Novel therapeutic innovation**	**Technology-enabled development**
Ferrer x United Therapeutics for inhaled treprostinil (Distribution/marketing)	**ChromX Health Boehringer Ingelheim** for lung-fibrosis diagnostics (Co-development)	**Novomedix x NHLBI** Novel oral small molecules (Funding)	**Qureight x Avalyn Pharma** | Phase 2b clinical development of AP01 (Contract: Research)

PPF remains an emerging area within a more established IPF investment ecosystem. The breadth of IPF transactions demonstrates the depth and maturity of pulmonary fibrosis investment, while early PPF-related activity suggests that the opportunity is becoming more diverse.

IPF: An established and diverse deal landscape

The 15 key IPF transactions identified between 2022 and 2026 span multiple deal types and development stages, involving a diverse set of pharmaceutical, biotechnology, academic, and technology partners.

DEVELOPMENT STAGE	DEAL ACTIVITY	COMPANIES/EXAMPLES
Discovery	Collaborative R&D, licensing, funding, research contract, manufacture contract	• AGC Biologics x Jikei University • Aston University x Isterian Biotech • GPCR Therapeutics x Bridge Biotherapeutics • Surrozen x TCGF • Samyang Biopharmaceuticals x Korea Drug Development Fund • Simcere Pharmaceutical Group x Stanford Medicine
Preclinical	Funding, licensing, contract: manufacture, collaborative R&D	• Senisca x Innovate UK • Pluri x Remedy cell • Shengrui Zehua x Shenzhen Pharmacin • Henry Ford Health and Michigan State University Health Sciences x NHLBI
Clinical development	Co-development, licensing	• Mediar Therapeutics x Eli Lilly • HK inno.N x Nextgen Bioscience • Daewoong Pharmaceutical x CS Pharmaceuticals • Strados Labs x Boehringer Ingelheim
Registered/approved	Rights	• Gyre Pharmaceuticals x Jiangsu Wangao Pharmaceutical

Note: Selected key transactions identified through IQVIA PharmaDeals database (search period: 2022-2026); list not exhaustive.

The strategic question is no longer only which mechanism can slow fibrosis. It is which development and commercial model can identify the right patients earlier, generate credible evidence across heterogeneous ILDs, and support adoption in a complex care pathway.

Strategic implications for biopharma

The shift toward PPF broadens the pulmonary fibrosis opportunity, but it also increases complexity. Companies must develop across heterogeneous underlying ILDs, identify progressing patients consistently, and demonstrate value within evolving treatment pathways.

For clinical development teams, the priority is to build evidence strategies that account for heterogeneity across underlying ILDs while showing benefits that are meaningful to physicians and patients. For commercial teams, earlier investment in patient identification, referral pathways, disease education, and market shaping will be critical. For business development teams, opportunity assessment should extend beyond therapeutic assets to the diagnostics, biomarkers, monitoring tools, and care-pathway capabilities needed to make a progression-based market work. The strongest opportunities may therefore come not simply from the next antifibrotic mechanism, but from connecting differentiated evidence with earlier patient identification and a practical care pathway.

List of abbreviations

ABBREVIATION	FULL FORM/MEANING
ALAT	Latin American Thoracic Association
ATS	American Thoracic Society
CTGF	Connective tissue growth factor
DLCO	Diffusing capacity of the lung for carbon monoxide
ERS	European Respiratory Society
EU	European Union
FVC	Forced vital capacity
IGFBP2	Insulin-like growth factor-binding protein 2
ILD	Interstitial lung disease
IPF	Idiopathic pulmonary fibrosis
JRS	Japanese Respiratory Society
LPA1	Lysophosphatidic acid receptor 1
LGALS1	Galectin-1
LGALS9	Galectin-9
MMP2	Matrix metalloproteinase-2
NHLBI	National Heart, Lung, and Blood Institute
PDE4B	Phosphodiesterase 4B
ppFVC	Percent predicted forced vital capacity
PTX3	Pentraxin 3
PF-ILD	Progressive fibrosing interstitial lung disease
PPF	Progressive pulmonary fibrosis
R&D	Research and development
UK	United Kingdom
US	United States

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About the authors



PRERNA KUNDARA
Associate Consultant,
IQVIA Consulting Services

Prerna Kundara is an Associate Consultant within IQVIA Consulting Services with over 4 years of experience in secondary research, competitive intelligence, market assessment, market segmentation, asset evaluation, and market landscape analysis. She has supported a range of strategic engagements by delivering actionable insights on treatment trends, innovation, and evolving market dynamics. Prerna also has experience developing Account Level Hospital Profiles (ALHPs) using HPA Audit data to support account prioritization and commercial opportunity assessment.



PRITI GIROTRA, PHD
Associate Principal,
IQVIA Consulting Services

Priti Girotra is an Associate Principal within IQVIA Consulting Services with more than 10 years of experience supporting biopharmaceutical companies across commercial strategy, market assessment, asset and portfolio prioritization, and brand planning. Her expertise spans oncology and rare diseases, with experience supporting strategic decision-making for clients across the US and EMEA through indication evaluation, commercial due diligence, and opportunity assessment.



ANIMESH JHA
Principal,
IQVIA Consulting Services

Animesh Jha is a Principal within IQVIA Consulting Services with over 16 years of experience supporting biopharmaceutical and healthcare organizations across commercial excellence, growth strategy, and go-to-market planning and implementation. He has led global consulting engagements for small to large biopharmaceutical clients across the US, EMEA, and APAC, helping organizations evaluate growth opportunities, prioritize investments, and navigate critical business decisions.

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