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Idiopathic pulmonary fibrosis: Clinical challenges, survival trends, and a case study from urban India

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Abstract
Idiopathic Pulmonary Fibrosis (IPF) is the most aggressive and progressive subtype of interstitial lung disease, often leading to severe functional impairment and early mortality. Despite therapeutic advances, IPF continues to surpass many malignancies in fatality, demanding prompt diagnosis and holistic management. This article reviews the clinical burden, prognostic indicators, molecular pathology, and current treatment modalities of IPF. Additionally, a real-world case of a 58-year-old male bus conductor with severe disease progression is presented to highlight the challenges in managing advanced IPF in urban India.

Keywords: Idiopathic Pulmonary Fibrosis, Antifibrotic therapy, Lung fibrosis, Interstitial Lung Disease, Pulmonary Hypertension, Case study

Introduction

Idiopathic Pulmonary Fibrosis (IPF) is a chronic, progressive fibrosing interstitial pneumonia of unknown cause, primarily occurring in older adults. Characterized histologically by usual interstitial pneumonia (UIP) pattern, IPF leads to a relentless decline in lung function. The disease carries a worse prognosis than many cancers, with a 5-year mortality rate of over 80%. Despite the availability of antifibrotic agents like pirfenidone and nintedanib, survival remains limited.

Epidemiology and Clinical Features: IPF predominantly affects males aged 60–70 years with a history of smoking or environmental exposure. An estimated 32,500 new cases occur annually in the U.S. Dyspnea on exertion, dry cough, and fatigue are early signs, progressing to resting breathlessness and hypoxemia. Quality of life significantly deteriorates, with most patients requiring supplemental oxygen within 24–36 months. Comorbidities such as pulmonary hypertension, GERD, and coronary artery disease further worsen prognosis.

Pathogenesis and Risk Factors: IPF has a multifactorial etiology involving genetic predispositions (e.g., MUC5B polymorphism, telomerase mutations), environmental exposures (e.g., wood/metal dust), GERD, and viral infections. Pathophysiologically, repetitive alveolar epithelial injury leads to aberrant wound healing, TGF-\u03b2 signaling, and fibroblast proliferation. This culminates in subpleural fibrosis and honeycombing on HRCT.

Diagnosis and Monitoring: Diagnosis relies on HRCT showing a UIP pattern and exclusion of other ILDs. Pulmonary function tests show progressive reduction in FVC and DLCO. The GAP (Gender, Age, Physiology) index helps stratify mortality risk. Patients are monitored for acute exacerbations, which occur in 5–10% annually and significantly reduce survival.

Treatment and Prognosis: Antifibrotic therapy with nintedanib or pirfenidone slows FVC decline but does not cure the disease. Corticosteroids and immunosuppressive agents are used selectively, especially in overlapping syndromes. Lung transplantation is the only intervention with a significant survival benefit but remains underutilized. Median survival without treatment is 2.8 years, extending to 4.2 years with antifibrotics. Patients with comorbidities or advanced GAP stage face worse outcomes.

Case Study

Case Report: A 58-year-old male bus conductor, a non-smoker, presented with progressive dyspnea over two years, worsening to breathlessness at rest. Initial HRCT revealed classic UIP pattern. PFTs showed FVC at 38% predicted and DLCO at 30%. He was started on pirfenidone, low-dose corticosteroids, and mycophenolate mofetil.
Despite optimal therapy, his symptoms progressed. Within 18 months, he required continuous portable oxygen (2-4 L/min). Six-minute walk distance dropped to 110 meters. He experienced severe fatigue, cough, and orthopnea. Evaluation for lung transplant was initiated but deferred due to organ unavailability and financial constraints.
His case highlights the typical trajectory of IPF: late diagnosis, limited response to therapy, and poor access to advanced care. Despite regular follow-ups, multidisciplinary support, and medication adherence, the patient remained oxygen-dependent with poor quality of life.

Discussion: This case underscores the aggressive nature of IPF and the limitations of current treatment options. Early recognition and initiation of antifibrotics are critical. Socioeconomic factors often limit access to lung transplantation, especially in low-resource settings. There is an urgent need for expanded transplant programs, public awareness, and support systems for IPF patients.

Conclusion: IPF is a devastating disease with limited survival despite medical advances. Timely diagnosis, appropriate antifibrotic therapy, and holistic care—

including psychosocial support and transplant evaluation—are essential. Real-world cases like the one described exemplify the systemic and clinical barriers that must be addressed to improve patient outcomes.

Message

Message to Health Care Workers

"Idiopathic Pulmonary Fibrosis (IPF) continues to challenge clinicians with its aggressive progression and limited treatment options. As frontline caregivers, our vigilance in early diagnosis, patient education, and multidisciplinary care—including pulmonology, palliative support, and transplant evaluation—can significantly impact outcomes. Let us advocate for holistic, compassionate care and continued research that gives hope and dignity to every IPF patient."

Message to NGOs

"Non-governmental organizations play a vital role in bridging the gap between clinical care and community support. IPF patients need access to affordable diagnostics, home oxygen support, financial aid for costly medications, and emotional counseling. NGOs can amplify awareness, enable timely referrals, and provide crucial logistical and moral support. Your efforts can transform lives where health systems may fall short."

Message to the Community

"Awareness saves lives. IPF often goes undetected until it becomes severe. If you or someone you know has persistent breathlessness or a chronic cough—especially among smokers, elderly individuals, or those with occupational dust exposure—encourage medical evaluation. The community's role is essential in supporting patients, reducing stigma, and fostering a network of care that includes family, friends, and neighbors."

Way Forward / Future Perspective

"The path ahead for IPF requires unified action—advancing public health education, improving early detection through screening programs, and investing in newer antifibrotic therapies and genetic research. Intersectoral collaboration between healthcare systems, NGOs, policymakers, and researchers is key. The ultimate goal is to shift from managing irreversible damage to intercepting disease in its early stages—preserving lung health, enhancing survival, and restoring quality of life."

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Author Biography: Dr. Rajendra Tatu Nanavare is a Chest Physician at the Group of TB Hospitals, Sewri, Mumbai, and works extensively in pulmonary medicine, TB, ILD, and pharmacovigilance.

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