Chapter 8
Idiopathic Pulmonary Fibrosis and Rare Lung Disorders

Parth Aphale
https://orcid.org/0000-0002-1004-2605
Dr. D.Y. Patil Homoeopathic Medical College and Research Centre, Dr. D.Y. Patil Vidyapeeth, Pune, India

Himanshu Shekhar
https://orcid.org/0009-0000-5383-4226
Dr. D.Y. Patil Homoeopathic Medical College and Research Centre, Dr. D.Y. Patil Vidyapeeth, Pune, India

Shashank Dokania
https://orcid.org/0009-0000-7781-9987
Dr. D.Y. Patil Homoeopathic Medical College and Research Centre, Dr. D.Y. Patil Vidyapeeth, Pune, India

ABSTRACT

Idiopathic Pulmonary Fibrosis (IPF) is a chronic, progressive, and often fatal interstitial lung disease marked by fibrotic remodeling of lung tissue due to excess extracellular matrix (ECM) deposition. Cutting-edge bioinformatics is revolutionizing our understanding of IPF through "omics" technologies, uncovering hidden mechanisms, key cellular players, and disrupted signaling pathways. Advanced tools now allow cell-by-cell mapping of diseased lung tissue, while machine learning aids in earlier diagnosis and prognosis. Despite challenges like complex data, these innovations are advancing precision medicine—offering hope for targeted therapies and improved outcomes.

DOI: 10.4018/979-8-3373-4923-7.ch008

INTRODUCTION

Imagine your lungs, which are normally soft and stretchy like sponges, slowly becoming stiff and scarred. That's what happens with Idiopathic Pulmonary Fibrosis (IPF). It's a serious and ultimately fatal lung disease where your lungs get covered in too much scar tissue, making them hard and unable to work properly.

Idiopathic Pulmonary Fibrosis (IPF) is an interstitial lung disease (ILD) which is non-specific, chronic, progressive, and fatal if not cured. It is characterized by relentless fibrotic remodeling of the lung parenchyma due to excessive extracellular matrix (ECM) deposition.

This abnormal fibrosis causes the scarring of the lung tissue beyond repair, and the patients usually come with continuous breathlessness, dry cough and progressively hypoxemia. Without the good diagnosis and treatments, IPF is still a fatal condition that affects from 13 to 20 people out of 100,000 worldwide and there are approximately 3 million people suffering from it all over the globe, predominantly older adults. The median survival following diagnosis is dismally low, at just 3 to 5 years. Disease progression is often punctuated by acute exacerbations, which significantly contribute to morbidity and mortality.

Beyond IPF, a heterogeneous group of rare interstitial lung disorders—such as lymphangioleiomyomatosis (LAM), pulmonary alveolar proteinosis (PAP), pulmonary Langerhans cell histiocytosis (PLCH), and Hermansky–Pudlak syndrome (HPS)-associated pulmonary fibrosis—presents unique clinical and research challenges. Though individually rare, these diseases are collectively significant due to their diagnostic ambiguity, diverse clinical presentations, and lack of effective curative therapies. Their low prevalence often hinders the assembly of large patient cohorts, limiting the power of clinical trials and mechanistic research.

Getting a diagnosis for IPF and these other rare ILDs is tricky. On high-resolution CT scans (HRCT), they can look very similar, showing things like hazy areas (ground-glass opacities), honeycomb-like patterns, and a web-like appearance (reticulation). These features aren't specific to just one condition. Even under the microscope, patterns like “usual interstitial pneumonia” (UIP) and “non-specific interstitial pneumonia” (NSIP) can pop up across various ILDs, making it even harder to tell them apart. So, even with better imaging and biopsy techniques, an accurate diagnosis still largely depends on a team of specialists discussing the case, and even then, different doctors might see things differently.

When it comes to treatment, our options are still pretty limited. For IPF, medications like pirfenidone and nintedanib are approved, and they do help slow the disease down a bit. But they don't actually reverse the scarring or offer a cure. For the rare ILDs, treatments often borrow from what we use for IPF or are based on just a few individual patient reports, with very little strong evidence specifically