



# Introduction

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IPF is a serious, chronic, steadily progressive and ultimately fatal disease of unknown origin, occurring predominantly in the elderly male smoker or exsmoker. Survival is worse than in many malignancies. IPF is the most dreaded and also the most frequent of the IIPs. The morphological hallmark is the UIP pattern, either on HRCT or biopsy, but it is not specific for IPF. It is crucial to differentiate this distinct entity from other ILDs with different prognoses and treatment approaches, especially from chronic extrinsic allergic alveolitis, idiopathic fibrotic NSIP and interstitial pneumonias with autoimmune features.

Recent years have seen a number of advances and changes in our understanding of the pathogenesis, diagnosis and management of IPF. It became evident that diagnostic security could be sharpened by multidisciplinary discussion and that bronchoscopic lung cryobiopsy is probably as informative as surgical lung biopsy in this setting. Many patients do not undergo surgical lung biopsy because the procedure is too risky in patients with severe disease and marked comorbidities. A number of statements and guidelines have been published but the question of how to handle a patient with probable or possible IPF compared to definite IPF has never been addressed.

After decades of therapeutic disappointment, two antifibrotic drugs are now available, which have the potential to slow disease progression by preventing ~50% of the decline in FVC. In this context, early diagnosis, which requires the recognition of velcro-like crackles on auscultation in the elderly, deserves the highest attention in the new era of IPF as a treatable disorder. There are many open questions related to antifibrotic therapy: when should we start and stop treatment? Which drug should be used first? Will the future lie in sequential or combination therapy?

This *ERS Monograph* aims to broadly describe the new achievements associated with IPF. Beginning with epidemiology, genetics and pathogenesis, the key diagnostic issues and major contributors to diagnosis, such as imaging and histopathology/BAL, are covered. This is followed by a section on how to evaluate/stage the disease for prognosis and how to monitor progression, including a discussion on the potential value of biomarkers. Several chapters are devoted to complications and comorbidities and their impact on management, such as acute exacerbation, PH, lung cancer, emphysema, cardiovascular disease and GERD. The treatment chapters cover antifibrotic drug therapy, symptom control, rehabilitation,

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palliative care and transplantation. Finally, the book considers unmet patient needs, ongoing issues in clinical trials and perspectives for the future.

As editors we believe that this new *Monograph* is timely, given the numerous developments in this field. We hope that this book will be of interest to all those who are engaged as clinicians or researchers in this evolving topic of respiratory medicine.