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FOREWORD: IPF

Idiopathic pulmonary fibrosis: recent milestones in disease management

Ulrich Costabel

Idiopathic pulmonary fibrosis (IPF) is a chronic, progressive fibrosing interstitial pneumonia of unknown cause that is limited to the lungs [1]. The disease is inevitably fatal, with an estimated median survival of 2–5 yrs following diagnosis; the mortality rate in IPF is higher than mortality rates associated with a number of malignancies [2]. Historically, there were no pharmacological treatments approved in Europe for managing patients with IPF and there was a clear need for treatments, which may modify the disease course and help preserve lung function.

This is now an exciting time for physicians, investigators and patients, as there is new hope for patients with IPF. Throughout 2011 there were important milestones, including the new guidelines from the American Thoracic Society (ATS)/European Respiratory Society (ERS)/Japanese Respiratory Society (JRS)/Latin America Thoracic Association (ALAT) Committee on how to diagnose IPF. Reliable predictors of the course of disease have now been identified that can help to predict which patients have a higher likelihood of death within the next year. Change in forced vital capacity over time has consistently been shown to be a strong predictor of mortality, and is also one of the most robust end-points that can be used in clinical trials of patients with IPF to assess the efficacy of potential treatments. Another great advance is the recent European approval of pirfenidone for the treatment of patients with mild-to-moderate IPF.

With this background of recent developments and increasing expectations, this issue of the *European Respiratory Review* includes a series of articles highlighting the substantial progress that has been made in the management of IPF, examining the pathogenesis of the disease along with the current treatment paradigm. These articles are based on presentations from a satellite symposium at the 2011 ERS Congress entitled ''IPF: new knowledge and new hope for patients''. DU BOIS [3] has reviewed the 2011 ATS/ERS/JRS/ALAT guidance on the diagnosis of IPF and discusses clinical situations that may not be fully addressed by the authors [1]. RICHELDI [4] presents data from the most recent Cochrane meta-analyses of agents in IPF, with an emphasis on the treatment effect of pirfenidone and its role as an agent in IPF management. GÜNTHER *et al.* [5] present a summary of recent and emerging data that are shaping our present understanding of the pathogenesis of IPF [1], and show that there are multiple pathways that could be targeted by future anti-fibrotic agents. Finally, COTTIN [6] summarises the most recent data from trials of pirfenidone and considers its role in treating IPF. These articles provide an outstanding opportunity for all clinicians involved in the management of patients with IPF to review the most important advances that have been achieved to date, bringing new knowledge and new hope for patients with IPF.

STATEMENT OF INTEREST

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CORRESPONDENCE: U. Costabel, Ruhrlandklinik, Universitätsklinik, Tüschener Weg 40, 45239 Essen, Germany. E-mail: ulrich.costabel@ruhrlandklinik.uk-essen.de

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