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Foreword

Contemporary approaches to idiopathic pulmonary fibrosis

Moisés Selman & Annie Pardo

Idiopathic pulmonary fibrosis (IPF) is a chronic progressive and lethal lung disorder of unknown etiology, with very limited therapeutic options and extremely poor prognosis [1]. Accumulation of scar tissue leads to progressive destruction of normal lung architecture, causing restricted pattern of lung volume, impaired gas exchange, exercise limitation, poor quality of life and finally death in few years after diagnosis [2]. Respiratory symptoms usually consist in progressive dyspnea and cough and the definite diagnosis is based in the presence of the usual interstitial pneumonia pattern by high resolution computed tomography and/or histopathology [1,2].

Most cases are sporadic, but in around 5–10% the disease is heritable and known as familial IPF. Linkage analysis and candidate gene approaches have identified four genes that cause the inherited form of IPF in 20% of patients with familial IPF [3]. These genes encode two surfactant proteins, SPC and SPA2, and two components of the telomerase complex, telomerase reverse transcriptase and the RNA component of telomerase [4]. The genetic architecture of susceptibility in the other 80% of familial IPF as well as in the sporadic IPF is presently unknown. Potential environmental risk factors include occupational and environmental exposures, tobacco smoking and some viral infections, primarily by herpes viruses [1,2].

IPF occurs usually in individuals over 50 years old, and the incidence and prevalence increase remarkably with advanced age (>60 years). However, the mechanisms linking aging with IPF are unclear. A number of aging associated processes [5] occur in the IPF lungs, for example, genomic instability,

telomere attrition, epithelial cell senescence and others, but these altered mechanisms also arise in others chronic respiratory diseases, such as chronic obstructive pulmonary disease, that show a very different clinical and pathological phenotype. Therefore, in addition to aging a distinctive genetic architecture and epigenetic reprogramming, likely affecting both, epithelial cells and fibroblasts, play a pivotal pathogenic role.

In this book, we aim to present new ideas and concepts regarding the pathogenesis, genetic susceptibility, epigenetic changes and aging-associated mechanisms involved in this devastating disease at the cutting edge of knowledge. The other goal is to provide to the clinician with state-of-art on clinical and physiological behavior, including an update of biomarkers for diagnosis and prognosis, as well as pharmacologic and surgical therapeutic approaches including some promising developments to deal with IPF. This dual purpose of bringing together clinical and basic information was fulfilling with the generous participation of experts of great reputation in the field.

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References

1. Raghu G, Collard HR, Egan JJ *et al.* ATS/ERS/JRS/ALAT Committee on Idiopathic Pulmonary Fibrosis. An official ATS/ERS/JRS/ALAT statement: idiopathic pulmonary fibrosis: evidence-based guidelines for diagnosis and management. *Am. J. Respir. Crit. Care Med.* 183, 788–824 (2011).
2. King TE Jr, Pardo A, Selman M. Idiopathic pulmonary fibrosis. *Lancet* 378, 1949–1961 (2011).
3. Kropski JA, Lawson WE, Young LR, Blackwell TS. Genetic studies provide clues on the pathogenesis of idiopathic pulmonary fibrosis. *Dis. Model Mech.* 6, 9–17 (2013).
4. Steele MP, Schwartz DA. Molecular mechanisms in progressive idiopathic pulmonary fibrosis. *Annu. Rev. Med.* 64, 265–276 (2013).
5. López-Otín C, Blasco MA, Partridge L, Serrano M, Kroemer G. The hallmarks of aging. *Cell* 153, 1194–1217 (2013).