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State of the Art in Idiopathic Pulmonary Fibrosis

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Message from the Guest Editors

Idiopathic pulmonary fibrosis (IPF) is a lethal disease of unknown etiology, elusive pathogenesis, and very limited therapeutic options. The onset and progression of IPF are influenced by multiple environmental and intrinsic factors, such as exposure to harmful substances, aging and genetic predisposition; however, the magnitude of the contribution of these factors to IPF and the chronological order of downstream pathogenic events remain uncertain. The main hallmarks of IPF are the abnormal activation of lung epithelial cells and the accumulation of fibroblasts/myofibroblasts along with the excessive deposition of extracellular matrix proteins. Recent technological advances and interdisciplinary approaches unmasked the involvement of a broad spectrum of molecular and cellular mediators in the pathogenesis of IPF. By critically evaluating the complexity of the disease and the translational value of pre-clinical studies, we would like to provide here a platform for conceptual and technological innovation in the field of IPF and shed light on new therapeutic strategies that may become a part of future treatment options.



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Special Issue



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