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Clinical Medicine

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Pleuroparenchymal Fibroelastosis: Obstacles and Challenges

Guest Editor:

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Deadline for manuscript submissions:
closed (30 June 2021)

Message from the Guest Editor

The field of pleuroparenchymal fibroelastosisis (PPFE) research is growing rapidly. PPFE is a rare interstitial pneumonia consisting of alveolar septal elastosis and intra-alveolar collagenosis, predominantly located in the upper lobes. The prognosis may be related to the development of fibrosing interstitial pneumonia in the lower lobes. However, there is marked variation in the pathogenesis and clinical features of PPFE. Although the diagnostic criteria for PPFE are based on histological findings by surgical lung biopsy (SLB), we have limited chances to perform an SLB for the diagnosis of PPFE. A proposal concerning the diagnostic criteria for idiopathic PPFE with and without an SLB was recently published. Mechanisms of the occurrence and evolution of fibroelastosis in PPFE are still unknown. In addition, treatment strategies for PPFE, targeting the inhibition of elastosis, have not been established. The present Special Issue aims to deepen our understanding of the characteristics of PPFE to increase our knowledge of a, so far, poorly explored research area, and to seek innovative treatment of PPFE.

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Message from the Editor-in-Chief

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