



Interstitial Lung Diseases: Focus on Pleuroparenchymal Fibroelastosis

Guest Editor:

Dr. Martina Bonifazi

Department of Biomedical Sciences and Public Health, Università Politecnica delle Marche, Ancona, Italy

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Message from the Guest Editor

The term "interstitial lung diseases" (ILDs) includes a wide spectrum of heterogeneous entities with different prognoses and treatment options. Pleuroparenchymal fibroelastosis (PPFE) is a specific clinicopathologic entity that affects the visceral pleura and the subpleural parenchyma with an upper-lobe predilection, characterized by elastic intra-alveolar fibrosis and scattered fibroblastic foci. Its pathogenesis is still unclear, but the heterogeneous spectrum of clinical presentations and behavior suggests that it may represent the final expression of a variable interplay between immune dysregulation, environmental exposure, and genetic predisposition. PPFE can present as an idiopathic form or in association with a variety of different conditions, including infections, lung and bone marrow transplantation, and autoimmune diseases.

The main goals of this Special Issue are to update and summarize insights and challenges of PPFE, and leaders in the different fields will be invited to provide comprehensive reviews on interstitial lung disease topics.





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Prof. Dr. Felipe Fregni

1. Neuromodulation Center and
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Massachusetts General Hospital,
Harvard Medical School, Boston,
MA 02114, USA
2. Department of Epidemiology,
Harvard T.H. Chan School of
Public Health, Boston, MA 02115,
USA

Message from the Editor-in-Chief

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Biomedicines Editorial Office
MDPI, Grosspeteranlage 5
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