Special Issue

Advances in Pulmonary Hypertension and Interstitial Lung Diseases: Mechanisms and Treatments

Message from the Guest Editors

Pulmonary hypertension (PH) is a common complication of interstitial lung diseases (ILDs). The pathophysiology of PH associated with ILDs (PH-ILDs) is complex and includes pulmonary endothelium disfunction and pre-capillary vessel remodeling similar to that seen in pulmonary arteria hypertension (PAH). In the majority of ILDs, the patient's PH remains mild; however, it can escalate severely in a small proportion of patients. Worsening PH significantly degrades patients' symptoms, quality of life, and survival. Unfortunately, treatment options for PH-ILDs are limited, with only one medication targeting pulmonary vasculature approved for use in this group of patients—inhaled Treprostinil. A better understanding of PH-ILDs is necessary to improve patient management and outcomes, including alterations in pathways on molecular and cellular level, the identification of patients at risk of severe PH, the role of PAH-targeted medication and the real risk of hypoxemia with their use, or the role of antifibrotic treatment. We invite researchers focusing on both basic and clinical sciences to share their thoughts and results from conducted studies.

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