

## Interstitial Lung Diseases: A Major Threat for Cardiovascular System

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### Editorial

The interstitial pulmonary diseases (ILDs) comprise a huge group of more than 200 entities, on pathological as well as specific clinico-radiological ground [1]. Moreover idiopathic pulmonary fibrosis is considered as commonest entity among them characterised by chronic progressive fibrosis, associated with hidden etiology, showing a motif of usual interstitial pneumonia (UIP). Addition to this, it is attributed by heterogeneous disease pattern associated with proliferative fibroblasts at different strata of genesis. Moreover pulmonary vascular as well as cardiac association is growingly highlighted as an essential component of increased morbidity as well as mortality in interstitial lung disease [2,3]. Researchers proposed that connective tissue disorders associated interstitial fibrosis, sarcoidosis, as well as pulmonary Langerhans cell histiocytosis are frequently complicated with secondary pulmonary hypertension, diagnosed by mean PAP >25 mmHg measured at rest during right heart catheterization [4]. Somehow Pulmonary hypertension is an under explored co morbidity in patients with interstitial lung ailment and have potential to deteriorate the function of lung as well survival. Pulmonary hypertension originate in patients with interstitial fibrosis through various mechanisms, including vascular inflammation, thrombotic angiopathy, pulmonary vasoconstriction and vascular remodeling, which may lead to vascular destruction associated with progressive lung parenchymal fibrosis. Doppler echocardiography recognized as an indispensable device in the screening of suspected pulmonary hypertension, however Right heart catheterization is needed to confirm the presence as well as, assess its severity to suggest therapy [5].

Although the prevalence of pulmonary hypertension in Interstitial lung diseases is unpredictable, Leuchte et al found it 31.8% among 88 consecutive patients with various ILDs [6]. In advanced lung disease, PH generally results from chronic hypoxic pulmonary vasoconstriction and vascular remodeling [7-10]. This vascular remodeling involves all layers of the pulmonary arterial wall and includes intimal thickening and medial hypertrophy. Factually, pulmonary hypertension not dependent on the severity of the diseases and can be precipitated even in absence of hypoxemia in patients with ILDs [11]. The pulmonary endothelial cell produces several important vasoactive mediators (eg, nitric oxide, prostacyclin, and endothelin) that modulate pulmonary vasomotor tone, vascular smooth muscle cell proliferation, and vascular remodeling [12]. Hypoxia increases plasma endothelin 1 levels [13]. The titer of circulating endothelin 1 has been reported to be elevated in patients with ILD, particularly in those with pulmonary hypertension [14,15]. According to few eminent researchers vascular smooth muscle dysfunction has been accused in the pathogenesis of pulmonary arterial hypertension. Pulmonary arterial smooth muscle cells from patients with this disorder exhibit abnormal proliferative response to growth factors such as transforming growth factor  $\beta$ , bone

morphogenetic protein, and platelet-derived growth factor [8]. Additionally, these smooth muscle cells exhibit abnormal migration and extracellular matrix formation as well as dysfunctional ion channels [16-18]. Though various hypothesis may contribute to the development of pulmonary hypertension among various ILDs while, Panagiotou et al carried out a comprehensive study and promulgate that the increased vascular resistance in the pre-capillary pulmonary circulation leads to increased pulmonary arterial pressure (PAP) which may be responsible for the development of pulmonary hypertension in ILD [19,20]. However hypoxic pulmonary vasoconstriction and obliteration of the vascular bed by progressive parenchymal fibrosis are considered major culprits for the increased pulmonary vascular resistance in ILD [21,22]. The development of pulmonary hypertension, further deteriorate the condition of moribund patient and increase the frequency of re admission in intensive care setup resulting in increased mortality however further research in this dimension to explore the underlying mechanisms of this complex phenomenon will enable a better understanding of the natural history of ILD, which may be helpful to generate the remedial tool to protect them from further deterioration and to take early preventive measures.

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