Inhaled treprostinil for interstitial lung disease-associated pulmonary hypertension: a silver lining on a very dark cloud

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Pulmonary hypertension associated with ILD is a very severe condition. The open label extension of the INCREASE trial demonstrates acceptable tolerability of inhaled treprostinil, maintained exercise capacity at week 40, and increase in FVC. https://bit.ly/3qxHvsB


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Pulmonary hypertension (PH) associated with interstitial lung disease (ILD-PH) is classified as group 3 PH, i.e. PH associated with chronic lung disease and/or hypoxia [1]. PH is relatively frequent in patients with ILD, depending on the definition of PH, the diagnostic assessment of PH, and the type of underlying ILD [2]. In patients with idiopathic pulmonary fibrosis (IPF), PH is a frequent complication at an advanced stage of the disease [3], or when emphysema is associated, as in the syndrome of combined pulmonary fibrosis and emphysema [4, 5]. While about 8% to 15% of patients with IPF may already have precapillary PH at diagnosis [6], the frequency (and severity) of PH increases as the disease progresses [7], reaching 30% to 50% of patients with PH at the time of evaluation for lung transplantation [8–11]. The presence of comorbidities such as obstructive sleep apnoea, thromboembolism or left heart diastolic dysfunction [12] may also increase the risk of PH in patients with fibrotic ILDs. Although usually of mild or moderate haemodynamic severity, ILD-PH is associated with dramatic worsening in shortness of breath, oxygen requirements, exercise limitation, quality of life and survival [2, 13].