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Title: Baseline characteristics and survival of patients with pulmonary hypertension in interstitial lung disease in the “HYPID” study

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Body: Background. Pulmonary hypertension (PH) in patients with interstitial lung disease (ILD) is frequent and associated with a shorter survival. Objective. To study the clinical, pulmonary function, haemodynamic characteristics, and survival in a large cohort of patients with ILD-PH at right heart catheterization. Methods. A prospective multicenter observational study (HYPID) was initiated in July 2010 in French expert centers for rare pulmonary diseases and/or PH (NCT01443598). Results. Out of 220 patients included (mean age

63 ±10 years; 135 males), ILD was idiopathic pulmonary fibrosis (n=37), combined pulmonary fibrosis and emphysema syndrome (n=51), systemic sclerosis with ILD (n=38), sarcoidosis (n=33), and other ILD (n=61). NYHA class was I-II in 19% of patients, III in 59%, IV in 20%, N/A in 2%. Six-min walk distance was 282 ± 147m. Hemodynamic characteristics were mean pulmonary arterial pressure (mPAP) 39 ± 10 mmHg, cardiac index (CI) 2.7 ± 0.7 L/min/m², and pulmonary vascular resistance (PVR) 542 ± 265 dyn.s.cm-5. mPAP was ≥35 mmHg in 59% of the cases and ≥40 mmHg in 40% of the cases. PVR poorly correlated with FVC (r=0.516, p=0.028) or PaO2 (r=0.593, P=0.007). mPAP poorly correlated with FVC (r=0.587, p=0.007) and PaO2 (r=0.593, P=0.007). The median time to death or transplant was 3.85 years, with no significant difference between patients with mPAP < or ≥ 35 or 40 mmHg. Conclusion. Hemodynamic characteristics do not correlate with pulmonary function in ILD-PH. Preliminary data suggest that the level of mPAP (< or ≥ 35 or 40 mmHg) does not impact survival.