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Title: Treatment of severe pulmonary hypertension in patients with interstitial lung disease: Results in 72 patients from the “HYPID” prospective study

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Body: Background. Data regarding treatment of severe pulmonary hypertension (PH) in patients with interstitial lung disease (ILD) are scarce. Methods. The outcome of patients with ILD-PH at right heart catheterization (RHC) who received PH-specific therapy at the discretion of physicians was studied in a prospective multicenter observational study (HYPID, NCT01443598) in French expert centers. Results. Out of 220 patients included in the prospective cohort, 72 (46 males, mean age 64 ± 10 yrs) had a mean pulmonary artery pressure (mPAP) ≥35 mmHg, received PH-specific therapy, had a follow-up RHC, and were included in the analysis. 10 had idiopathic pulmonary fibrosis, 22 combined pulmonary fibrosis and emphysema, 11 systemic sclerosis, 13 sarcoidosis, 16 other ILD. NYHA class was II in 18%, III in 64%, IV in 18%. Patients received ERA in 61%, PDE5i in 42%, prostanoids in 7%, and calcium blockers in 3%. After a mean of 4.5 ±2 months, although 6-min walk distance and NYHA class were unchanged, mPAP and PVR had decreased and cardiac index had significantly increased ($p<0.0001$ for each), with no change in PaO2

and SaO2. The median time to death or transplant in the treated group was 3.24 years. Conclusion. PH-specific therapy improves hemodynamics in patients with ILD and severe PH without worsening in oxygenation.