OUTCOME OF LUNG DISEASE-RELATED PULMONARY HYPERTENSION AND IMPACT OF PULMONARY VASOREACTIVITY

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Background: The clinical characteristics, hemodynamic perturbations and outcomes of lung disease-related pulmonary hypertension (LD-PH) are poorly defined.

Methods: From a two-center prospective cohort of PH patients referred for initial hemodynamics, we identified 70 patients with LD-PH. Causes included obstructive lung disease in 22, interstitial lung disease in 18 and sarcoidosis in 23. Baseline characteristics and long-term survival were compared with 88 patients with idiopathic PH (iPH). Vasoreactivity was defined by 30% drop in mean pulmonary artery pressure (mPAP) during inhalation of 40 ppm nitric oxide.

Results: Mean age of LD-PH patients was 59±13 years, 61% were women and WHO function class distribution was 24/54/21 (% II/III/IV). Compared to iPH there were fewer women (61% vs. 76%, p=0.035). LD-PH patients had smaller right ventricular (RV) sizes by echo (p=0.005) and less tricuspid regurgitation (p<0.001). mPAP was lower in LD-PH (39±12 vs. 47±16 mmHg, p<0.001) but other hemodynamics were similar. Survival was equally poor in both groups (Figure). In the presence of vasoreactivity, survival was dramatically improved in LD-PH patients. Proportional hazards analysis revealed that function class (p=0.017), cardiac index (p=0.010) and vasoreactivity (p=0.041) independently predicted survival.

Conclusions: Despite lower mPAP and less RV enlargement than iPH, LD-PH has equally poor clinical outcome. Vasoreactivity independently predicts survival in this patient population.

![Survival: IPH vs. LD-PH](image1.png)

![Effect of Vasoreactivity in LD-PH](image2.png)