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Mild parenchymal lung disease and/or low diffusion capacity impacts survival and treatment response in patients diagnosed with idiopathic pulmonary arterial hypertension

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Patients with IPAH who have mild parenchymal lung disease have significantly worse outcomes, in terms of survival and treatment response, when compared to patients with IPAH who do not have evidence of parenchymal lung disease <http://bit.ly/3agkYn0>

Cite this article as: Lewis RA, Thompson AAR, Billings CG, *et al.* Mild parenchymal lung disease and/or low diffusion capacity impacts survival and treatment response in patients diagnosed with idiopathic pulmonary arterial hypertension. *Eur Respir J* 2020; 55: 2000041 [<https://doi.org/10.1183/13993003.00041-2020>].

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ABSTRACT There are limited published data defining survival and treatment response in patients with mild lung disease and/or reduced gas transfer who fulfil diagnostic criteria for idiopathic pulmonary arterial hypertension (IPAH).

Patients diagnosed with IPAH between 2001 and 2019 were identified in the ASPIRE (Assessing the Spectrum of Pulmonary Hypertension Identified at a Referral Centre) registry. Using prespecified criteria based on computed tomography (CT) imaging and spirometry, patients with a diagnosis of IPAH and no lung disease were termed IPAH_{no-LD} (n=303), and those with minor/mild emphysema or fibrosis were described as IPAH_{mild-LD} (n=190).

Survival was significantly better in IPAH_{no-LD} than in IPAH_{mild-LD} (1- and 5-year survival 95% and 70% versus 78% and 22%, respectively; p<0.0001). In the combined group of IPAH_{no-LD} and IPAH_{mild-LD}, independent predictors of higher mortality were increasing age, lower diffusing capacity of the lung for carbon monoxide (D_{LCO}), lower exercise capacity and a diagnosis of IPAH_{mild-LD} (all p<0.05). Exercise capacity and quality of life improved (both p<0.0001) following treatment in patients with IPAH_{no-LD}, but not IPAH_{mild-LD}. A proportion of patients with IPAH_{no-LD} had a D_{LCO} <45%; these patients had poorer survival than patients with D_{LCO} ≥45%, although they demonstrated improved exercise capacity following treatment.

The presence of even mild parenchymal lung disease in patients who would be classified as IPAH according to current recommendations has a significant adverse effect on outcomes. This phenotype can be identified using lung function testing and clinical CT reports. Patients with IPAH, no lung disease and severely reduced D_{LCO} may represent a further distinct phenotype. These data suggest that randomised controlled trials of targeted therapies in patients with these phenotypes are required.