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Title: 'Idiopathic' pulmonary arterial hypertension with preserved lung function but co-existing parenchymal abnormalities: Response to treatment and survival

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Body: Background: FEV_1, FVC and/or TLC <60% were used in the French and Scottish registries to exclude patients with pulmonary hypertension (PH) due to lung disease. Similar criteria were used in pivotal PH trials to exclude significant lung disease. However, some patients with preserved pulmonary function who satisfy the standard criteria for idiopathic pulmonary arterial hypertension (IPAH) may nonetheless have evidence of co-existing parenchymal abnormalities on HRCT. It is unclear how response to treatment and survival is affected by the presence of modest lung disease. Aim: To compare the baseline characteristics, change in 6-minute walk distance at 3 months and survival of IPAH with and without co-existing parenchymal abnormalities on HRCT. Methods: All incident cases of IPAH with and without co-existing CT parenchymal abnormalities diagnosed between January 2001 to December 2009 in all eight PH centres in the UK and Ireland were included. All patients have FEV_1, FVC and/or TLC≥ 60% predicted. Results:

Table 1. Baseline characteristics, ∆6MWD and survival of IPAH

	with parenchymal abnormalities (n=146)	without parenchymal abnormalities (n=482)
Age, yrs	68	50
% female	42%	70%
6MWD, m	209	292
∆6MWD at 3 months, m	52	40
mPAP, mmHg	49	54
cardiac index, L/min/m2	2.1	2.1
PVRI, WU.m2	21	23
1 year survival	74%	93%
3 year survival	45%	73%

Conclusion: Despite similar baseline haemodynamics and response to treatment, survival of 'IPAH' with coexisting parenchymal abnormalities appears worse compared with IPAH without parenchymal abnormalities. Age and age related co-morbidities may account for the difference in long term outcome between the 2 groups.