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Title: Does lung function predict response to therapy in PAH associated with connective tissue disease?

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Body: Introduction PAH specific therapies have been trialled in several lung diseases without success. It has also been shown that a low FVC is associated with a poor prognosis in CTD-PAH. Methods From the Royal Free Hospital pulmonary hypertension associated with connective tissue disease database we identified patients with lung function tests within 6 months of right heart catheterisation performed to confirm pulmonary hypertenson. Patients who had repeat haemodynamic studies within a year of diagnosis on first line therapy (bosentan) were included. Results There were no significant haemodynamic differences between groups at baseline (ANOVA). Follow up catheter studies demonstrated a good haemodynamic response with improvements in mean PA pressure (FVC >94%) and pulmonary vascular resistance (FVC >70%).

	FVC <70, n=23	FVC 70-94, n=28	FVC >94, n=30
baseline RAP mmHg	8.3	7.8	6.6
baseline mPAP mmHg	39.3	44.3	39.8
baseline PCWP mmHg	10.2	9.9	9.8
baseline PVR dynes.cm/5	553	670	565
baseline CI L/min/m2	2.6	2.6	2.7
f/u RAP	7.9	8.2	6.5
f/u mPAP	37.4	43.0	36.4*
f/u PCWP	9.3	10.5	10.3
f/u PVR	496	590*	476*
f/u CI	2.6	2.7	2.8
One year survival	91%	93%	100%

RAP = right atrial pressure, mPAP = mean pulmonary artery pressure, PCWP = pulmonary capillary wedge pressure, PVR = pulmonary vascular resistance, CI = cardiac index. *p<0.05 compared with baseline paired students' t-test

The changes seen in the FVC <70% group did not reach significance (e.g. p<0.09 for PVR). Conclusion A low FVC does not preclude a good treatment response, suggesting that PAH can co-exist with lung fibrosis in connective tissue diseases. Treatment of both pathologies may be needed to improve outcomes in this difficult group.