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Title: Outcomes from use of targeted therapy in pulmonary hypertension associated with sarcoidosis (PHAS)

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Body: Pulmonary hypertension is a recognised complication of sarcoidosis and may arise from several aetiological pathways. We report our experience of patients with PHAS-GroupV on targeted treatment. Retrospective study of patient outcomes with PHAS. All patients underwent right heart catheterisation satisfying criteria for diagnosis of PAH. We reviewed 16 patients, 10 of whom died in mean f/u 32m. Patients dichotomised into 2 groups (responders and non-responders) based on improvement in log ntproBNP and 6MWD within 6 months of therapy initiation.

	Responders n=8	Non-responders n=5	p value
Age y	60.8	58.8	0.71
Gender M:F	4:4	3:2	
Follow up m	54.8	17.4	0.02
Sarcoid stage III/IV	3:5	1:4	0.66
NYHA Functional Class	7:1	5:1	0.81
mPAP	52.6	45.4	0.11
CI	1.85	1.64	0.20
PVR dyn	1001	648	0.046
FVC %	61	66	0.61
KCO%	50	53	0.79
6MWDδm	+78	-52	0.004

Treatment response was associated with a reduced risk of death. Cohort phenotypes suggest that treatment response may not be predicted from sarcoid stage, functional class, FVC or presence of extrapulmonary features. However elevation in PVR may possibly be associated with a favourable response. An RCT is indicated to evaluate a larger cohort.