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Title: Response to increased load in systemic sclerosis-associated PAH and idiopathic PAH

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Body: Background: Patients with Systemic Sclerosis-associated pulmonary hypertension (SScPAH) have worse survival than patients with idiopathic PAH (IPAH). We hypothesized that the RV adapts differently in SScPAH vs IPAH. Methods: We examined 58 prospectively obtained cardiac MRIs (cMRI) in 34 SScPAH and 24 IPAH patients all diagnosed by right heart catheterization (RHC). cMRI and RHC measurements were recorded for all patients. Regression analysis was used to assess the association between diagnosis and RV measurements after adjusting for age and RV load. Results: Clinical and demographic variables are shown in Table 1. Univariate regression showed RV mass index (RVMi) varied linearly with measures of RV load. ANCOVA analysis suggested an interaction between diagnosis and RVMi adjusting for PVR and age (p=0.09). A spline model showed a significantly lower rate of increase in RVMi (0.51vs2.03g/m2*WU) in SScPAH vs IPAH at PVR>7 WU. Conclusion: RVMi varies linearly with measures of RV load. After adjusting for differences in RV load and age, SScPAH patients had a significantly decreased rate of hypertrophy with increasing PVR that was most prominent at PVR>7WU. This difference in adaptive hypertrophy may, in part, explain poorer survival in SScPAH despite lower measures of load.

Table 1: Demographic and Catheterization Data

	SScPAH (N=34)	IPAH (N=24)	p-value
Age(yrs)	62	52.5	0.001
Sex(%female)	94%	71%	0.02
mPAP(mmHg)	39.7	47.7	0.02

PVR(WU)	8.2	8.3	0.95