

## 1022 Effect of Hemodynamics and Functional Capacity on Survival in Patients with Sickle Cell Disease and Pulmonary Hypertension

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**Background.** Pulmonary hypertension (PH) is a common complication of sickle cell disease (SCD). The high mortality rate in this population despite mild to moderate PH raises important questions about whether PH is a marker of systemic vasculopathy and risk of death, rather than a direct cause of death.

**Methods.** We sought to evaluate the effects of hemodynamics and functional capacity on survival in patients with SCD and PH (defined as mPAP  $\geq$  25 mmHg at rest or  $\geq$  30 mmHg at exercise). We collected last available hemodynamic data from 48 patients followed for a mean of 22 months after the procedure. In 18 patients, hemodynamic data at baseline and after therapies for PH was analyzed.

**Results.** During follow up 11 patients died, and when compared to survivors these patients had higher RAP (mean $\pm$  SE mmHg, 18 $\pm$  3 vs 8 $\pm$ 1, P<0.001), mPAP (mean $\pm$  SE mmHg, 45 $\pm$ 3 vs 29 $\pm$ 1, P<0.001), PCWP (mean $\pm$  SE mmHg, 17 $\pm$ 1 vs 14 $\pm$ 1, P=0.01), PVRI (mean $\pm$  SE dyne $\cdot$ sec $\cdot$ cm<sup>-5</sup> $\cdot$ m<sup>-2</sup>, 628 $\pm$ 129 vs 281 $\pm$ 36, P<0.001), lower CI (mean $\pm$ SE L/min $\cdot$ m<sup>2</sup>, 4 $\pm$ 0.5 vs 5 $\pm$ 0.2, P=0.02) and 6MW distance (mean $\pm$ SE meters, 274 $\pm$ 30 vs 392 $\pm$ 12, P<0.001). RAP (RR 1.12, 95% CI 1.05-1.2, P<0.001), mPAP (RR 1.13, 95% CI 1.05-1.21, P<0.001), PCWP (RR 1.13, 95% CI 1.01-1.28, P=0.045), PVRI (RR 1.003, 95% CI 1.001-1.004, P<0.001), CI (RR 0.63, 95% CI 0.41-0.97, P=0.036) and 6MW distance (RR 0.988, 95% CI 0.981-0.995, P=0.01) were univariate predictors of death on proportional hazards regression analysis. Surviving patients demonstrated a more favorable hemodynamic profile during follow up RHC when compared to those who died:  $\Delta$ RAP (mean change $\pm$ SE, -1.6 $\pm$ 1 vs 5.7 $\pm$ 2, P<0.001),  $\Delta$ mPAP (mean change $\pm$ SE, -5.5 $\pm$ 2 vs 7.2 $\pm$ 5, P=0.001),  $\Delta$ CI (mean change $\pm$ SE, 0.05 $\pm$ 0.4 vs -1.1 $\pm$ 1.2, P=0.04) and  $\Delta$ PVRI (mean change $\pm$ SE, -20 $\pm$ 47 vs 400 $\pm$ 171, P=0.01).

**Conclusions.** Right and left ventricular dysfunction, high mPAP, high PVRI and decreased functional capacity are associated with mortality in patients with PH and SCD. These data strongly suggest that rather than being a marker of poor prognosis, PH is a direct cause of mortality in this population.