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# Physiological predictors of survival in patients with sarcoidosis-associated pulmonary hypertension: results from an international registry

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**Decreased 6-min walk distance and reduced diffusion capacity are associated with decreased survival in patients with sarcoidosis-associated pulmonary hypertension** <http://bit.ly/2UQfWJh>

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## ABSTRACT

**Introduction:** Sarcoidosis-associated pulmonary hypertension (SAPH) is associated with reduced survival in single-centre studies. The international Registry for SAPH (ReSAPH) with long-term follow-up was established to enrich our knowledge of this complication of sarcoidosis. This analysis aims to elucidate factors associated with reduced transplant-free survival in SAPH patients.

**Methods:** ReSAPH contains prospectively collected outcomes of SAPH patients since the time of registry enrolment. Information analysed includes right heart catheterisation data, pulmonary function testing, chest radiography, Scadding stage and 6-min walk distance (6MWD), among others. Cox regression models were used to identify independent predictors of transplant-free survival.

**Results:** Data from 215 patients followed for a mean±SD 2.5±1.9 years were available for analysis. In the 159 precapillary patients, the Kaplan–Meier-adjusted 1-, 3- and 5-year transplant-free survival was 89.2%, 71.7% and 62.0%, respectively. Kaplan–Meier-adjusted 1-, 3- and 5-year transplant-free survival in the incident group was 83.5%, 70.3% and 58.3%, respectively, and in the prevalent group was 94.7%, 72.2% and 66.3%, respectively. Patients with reduced diffusing capacity of the lung for carbon monoxide ( $D_{LCO}$ ) (<35% predicted) and 6MWD <300 m in the precapillary cohort had significantly worse transplant-free survival. Reduced 6MWD and preserved forced expiratory volume (FEV<sub>1</sub>)/forced vital capacity (FVC) ratio were identified as independent risk factors for reduced transplant-free survival in the precapillary cohort.

**Conclusion:** Reduced  $D_{LCO}$  (<35% pred) and 6MWD (<300 m) at the time of registry enrolment were

associated with reduced transplant-free survival in the overall precapillary cohort. Preserved FEV<sub>1</sub>/FVC ratio was identified as an independent risk factor for worsened outcomes.