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Clinical phenotypes and outcomes of precapillary pulmonary hypertension of sickle cell disease

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Clinical phenotype of precapillary pulmonary hypertension of sickle cell disease is influenced by the genotype. Thrombotic lesions appear as a major component of PH related to SCD, more frequently in SC patients. http://bit.ly/32b2nEx

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ABSTRACT

Rationale: Precapillary pulmonary hypertension (PH) is a devastating complication of sickle cell disease (SCD). Little is known about the influence of the SCD genotype on PH characteristics.

Objectives: To describe clinical phenotypes and outcomes of precapillary PH due to SCD according to disease genotype.

Methods: A nationwide multicentre retrospective study including all patients with SCD-related precapillary PH from the French PH Registry was conducted. Clinical characteristics and outcomes according to SCD genotype were analysed.

Results: 58 consecutive SCD patients with precapillary PH were identified, of whom 41 had homozygous for haemoglobin S (SS) SCD, three had S-β_0 thalassaemia (S-β_0 thal) and 14 had haemoglobin SC disease

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(SC). Compared to SC patients, SS/S-β_0 thal patients were characterised by lower 6-min walk distance (p=0.01) and lower pulmonary vascular resistance (p=0.04). Mismatched segmental perfusion defects on lung scintigraphy were detected in 85% of SC patients and 9% of SS/S-β_0 thal patients, respectively, and 50% of SS/S-β_0 thal patients had heterogeneous lung perfusion without segmental defects. After PH diagnosis, 31 patients (53%) received medical therapies approved for pulmonary arterial hypertension, and chronic red blood cell exchange was initiated in 23 patients (40%). Four patients were managed for chronic thromboembolic PH by pulmonary endarterectomy (n=1) or balloon pulmonary angioplasty (n=3). Overall survival was 91%, 80% and 60% at 1, 3 and 5 years, respectively, without influence of genotype on prognosis.

Conclusions: Patients with precapillary PH related to SCD have a poor prognosis. Thrombotic lesions appear as a major component of PH related to SCD, more frequently in SC patients.