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Title: Baseline characteristics of idiopathic PAH patients with severely reduced diffusion capacity

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Body: Introduction: Pulmonary arterial hypertension (PAH) is a disease affecting the vasculature of the lung and is known to decrease carbon monoxide diffusion capacity (DLCO). Although most patients have a moderately decreased DLCO, a group of patients has severely reduced DLCO. The reason for this low DLCO is unclear. Therefore, the aim of this study is to describe baseline characteristics of idiopathic PAH patients with a low DLCO and compare this group with a group of idiopathic PAH patients with moderate low or normal DLCO. Methods: Patients with idiopathic or familial PAH were included. Patients were grouped based on the finding of a bimodal distribution in DLCO-values. Results: A total of 170 patients were included. DLCO groups were DLCO <45% (N=58) and ≥45% of predicted (N=112). Group characteristics are shown in table 1.

Conclusions: Compared with IPAH patients with moderate low or normal DLCO, IPAH patients with a severely reduced DLCO were characterized at baseline by a higher age, increased number of males, more often a history of smoking and coronary disease, and a lower exercise performance. No differences however were found in hemodynamic parameters.