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Title: Histopathology of idiopathic pulmonary arterial hypertension in patients with low or normal diffusion capacity

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Body: Introduction: In patients with Idiopathic Pulmonary Arterial Hypertension (IPAH) a wide range of diffusion capacity for carbon monoxide (DLCO) values can be observed. Low DLCO in IPAH is associated with worse prognosis when compared to normal DLCO[1]. Hypothesis: histopathological pattern(s) of pulmonary vasculopathy in IPAH differ between patients with low and normal DLCO. Methods: We retrospectively analyzed the histopathological (combinations of) patterns of vasculopathy in IPAH patients with low and normal DLCO. Low DLCO was defined as lowest tertile of the bimodal distribution of DLCO-value in a series of 170 IPAH patients[1] (<45% pred.). DLCO in the upper 2 tertiles (≥ 45 % pred.) was defined as normal DLCO. Results: Out of 170 IPAH patients, complete data sets were obtained in 20 patients: low DLCO N=10; normal DLCO N=10. Results are shown in table 1 (study is ongoing).

Table 1: Histopathology

	Low DLCO (N=10)	Normal DLCO (N=10)
Lung biopsies	3	7
Explant	2	-
Autopsy	5	3
Pulmonary vasculopathy pattern		
Plexogenic arteriopathy	-	5
Thrombotic arteriopathy	1	2
Hypoxic arteriopathy	1	2
PVOD*/PCH** pattern	6	1
Congestive vasculopathy	2	1
No specific pattern	1	2

* Pulmonary Veno-Occlusive Disease **Pulmonary Capillary Haemangiomatosis

Conclusion: A PVOD/PCH-like pattern is more common in IPAH with low DLCO, while plexogenic arteriopathy prevails in IPAH with normal DLCO. 1. Trip et al, abstract this meeting.