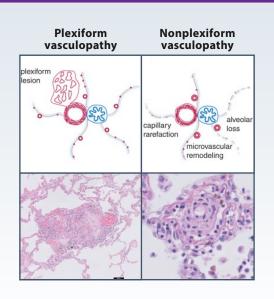
What Are the Histologic Patterns and Their Clinical Correlates in Patients With a Diagnosis of Idiopathic PAH or Hereditary PAH?



STUDY DESIGN

- Cross-sectional registry study of 50 patients with idiopathic pulmonary arterial hypertension (PAH)
- Lung histologic examination by both qualitative and quantitative analysis by two experienced pathologists
- Histopathologic characteristics were correlated with clinical and hemodynamic parameters

RESULTS



	Plexiform Vasculopathy (n=26)	Nonplexiform Vasculopathy (n=24)
Age, y	44 ± 11	60 ± 16
Sex, male	5 (20%)	16 (67%)
Smoking, active or history	7 (48%)	14 (75%)
DLCO, % pred	66 ± 16	41 ± 17
Mutation carriers out of patients who were tested	9/15 (60%)	0/11 (0)

Potential differences in pathobiological mechanisms between patients with plexiform and nonplexiform microvascular disease should be taken into account in future research strategies unravelling the pathophysiologic features of PAH and developing biology-targeted treatment approaches.