

PH group	Histopathological Features	Animal model
Group 1 PAH	<u>Early phase:</u> Medial hypertrophy Intima proliferation Muscularization of normally non-muscular arteries	Hypoxia/SU5416 mouse
	<u>Late phase:</u> Intima fibrosis Loss of luminal vascular volume Plexiform lesions Recanalization of arteries Fibrinoid necrosis	Hypoxia/SU5416 rat
Group 1' Pulmonary venoocclusive disease	Congestion of pulmonary parenchyma Hemosiderosis Fibrosis of small veins/venules	
Group 2 PH with left heart disease	Arterialization of large or middle-sized pulmonary veins Interstitial edema and fibrosis Hemosiderosis Medial hypertrophy/adventitial thickening of pulmonary arteries	
Group 3 PH associated with lung disease and/or hypoxemia	<u>3.1 and 3.3–3.5. Hypoxic pulmonary vasculopathy</u> Muscularization of arterioles Medial hypertrophy of muscular pulmonary arteries	Hypoxia/SU5416 rat
	<u>3.2. Pulmonary vasculopathy associated with interstitial lung disease</u> Features of hypoxic pulmonary vasculopathy Intimal fibrosis of arteries	and Hypoxia/SU5416 mouse
Group 4 PH due to chronic thrombotic/embolic disease	Thromboembolic obstruction of distal pulmonary arteries Eccentric intimal fibrosis	
Group 5 Miscellaneous	Heterogeneous group of disorders: some showing features of congestive vasculopathy some post-thrombotic vasculopathy	