Table 1. WHO Clinical Classification of PH[3]

Group	Clinical Classifications

1
PAH
	• Idiopathic
	• Heritable
	• Drug-induced and toxin-induced
	• Associated with
	  - Connective tissue disease
	  - Portal hypertension
	  - Congenital heart disease
	  - Schistosomiasis
	• PAH long-term responders to calcium channel blockers
	• PAH with overt features of pulmonary veno-occlusive disease or pulmonary capillary hemangiomatosis
	• Persistent PH of the newborn syndrome

2 PH associated with left heart disease	• Due to heart failure with preserved left ventricular ejection fraction • Due to heart failure with reduced left ventricular ejection fraction • Valvular heart disease • Congenital or acquired cardiovascular conditions leading to postcapillary PH
3 PH associated with lung disease or hypoxia	• Obstructive pulmonary disease • Restrictive pulmonary disease • Other lung diseases with a mixed restrictive or obstructive pattern • Hypoxia without lung disease • Developmental lung disorders

4
PH due to pulmonary arterial obstruction
	• Chronic thromboembolic PH
	• Other pulmonary artery obstruction
	  - Sarcoma medium or high grade) or angiosarcoma
	  - Other malignant tumors (renal, uterine, germ cell tumor of the testis, other tumors)
	  - Nonmalignant tumors (uterine leiomyoma)
	  - Arteritis without connective tissue disease
	  - Congenital pulmonary artery stenosis
	• Parasites (hydatidosis)

5
PH with an unclear or multifactorial mechanism
	• Hematologic disorders
	  - Chronic hemolytic anemia
	  - Myeloproliferative disorders
	• Systemic and metabolic disorders
	  - Pulmonary Langerhans cell histiocytosis
	  - Gaucher disease
	  - Glycogen storage disease
	  - Neurofibromatosis
	  - Sarcoidosis
	• Other
	  - Fibrosing mediastinitis
	  - Chronic renal failure (with or without dialysis)
	  - Pulmonary tumorous thrombotic microangiopathy
	  - HIV infection

PAH, pulmonary arterial hypertension; PH, pulmonary hypertension.