

Table 9-1. Diagnostic classification of pulmonary hypertension

Pulmonary arterial hypertension
Primary pulmonary hypertension
Sporadic
Familial
Related to:
Collagen vascular disease
Congenital systemic-to-pulmonary shunts
Portal hypertension
HIV infection
Drugs/toxins
Anorexigens
Other
Persistent pulmonary hypertension of the newborn
Other
Pulmonary venous hypertension
Left-sided atrial or ventricular heart disease
Left-sided valvular heart disease
Extrinsic compression of central pulmonary veins
Fibrosing mediastinitis
Adenopathy/tumors
Pulmonary veno-occlusive disease
Other
Pulmonary hypertension associated with disorders of the respiratory system and/or hypoxemia
Chronic obstructive pulmonary disease
Interstitial lung disease
Sleep-disordered breathing
Alveolar hypoventilation disorders
Chronic exposure to high altitude
Neonatal lung disease
Alveolar-capillary dysplasia
Other
Pulmonary hypertension due to chronic thrombotic and/or embolic disease
Thromboembolic obstruction of proximal arteries
Obstruction of distal pulmonary arteries
Pulmonary embolism (thrombus, tumor, ova and/or parasites, foreign material)
In situ thrombosis
Sickle cell disease
Pulmonary hypertension due to disorders that directly affect the pulmonary vasculature
Inflammatory
Schistosomiasis
Sarcoidosis
Other
Pulmonary capillary hemangiomatosis

Source: World Health Organization, 1999.