Table

Classification of pulmonary hypertension

Category 1: Pulmonary arterial hypertension
- Idiopathic/familial
- Related to another identifiable process: Collagen vascular disease (especially scleroderma and lupus)
- Portal hypertension
- HIV infection
- Congenital heart disease with left-to-right shunting
- Drugs, toxins, anorexigens, cocaine
- Other, including hemoglobinopathies, hereditary hemorrhagic telangiectasia, myeloproliferative disorders, splenectomy, hemolytic anemia, Gaucher disease
- Persistent PH of the newborn

Category 2: Pulmonary venous hypertension
- Left-sided heart disease
- Veno-occlusive diseases

Category 3: Disorders of the respiratory system
- Chronic obstructive pulmonary disease
- Interstitial lung disease
- Sleep-disordered breathing
- Alveolar hypoventilation disorders
- Chronic high altitude exposure
- Neonatal lung disease, alveolar capillary dysplasia

Category 4: Chronic thrombotic/embolic disease
- Thromboembolic obstruction of pulmonary arteries
- Nonthrombotic pulmonary emboli (e.g., parasites, schistosomiasis, tumor)
- In situ thrombotic diseases, sickle cell disease

Category 5: Miscellaneous
- Sarcoidosis, histiocytosis X, lymphangioleiomyomatosis

Source: Simonneau G, et al. Clinical classification of pulmonary hypertension. *J Am Coll Cardiol* 2004;43(12 Suppl S):5S-12S.