

Clinical Classification of Pulmonary Hypertension

1. **Pulmonary arterial hypertension (PAH)**

1.1 Idiopathic

1.2 Heritable

1.2.1 BMPR2

1.2.2 ALK1, ENG, SMAD9, CAV1, KCNK3

1.2.3 Unknown

1.3 Drug and toxin induced

1.4 Associated with (APAH)

1.4.1 Connective tissue diseases

1.4.2 HIV infection

1.4.3 Portal hypertension

1.4.4 Congenital heart disease

1.4.5 Schistosomiasis

1' Pulmonary veno-occlusive disease and/or pulmonary capillary hemangiomatosis

1'' Persistent pulmonary hypertension of the newborn (PPHN)

2. **Pulmonary hypertension due to left heart disease**

2.1 Left ventricular systolic dysfunction

2.2 Left ventricular diastolic dysfunction

2.3 Valvular disease

2.4 Congenital/acquired left heart inflow/outflow tract obstruction and congenital cardiomyopathies

3. **Pulmonary hypertension due to lung diseases and/or hypoxia**

3.1 Chronic obstructive lung disease

3.2 Interstitial lung disease

3.3 Other pulmonary diseases with mixed restrictive and obstructive pattern

3.4 Sleep-disordered breathing

3.5 Alveolar hypoventilation disorders

3.6 Chronic exposure to high altitude

3.7 Developmental lung disease

4. **Chronic thromboembolic pulmonary hypertension (CTEPH)**

5. **Pulmonary hypertension with unclear multifactorial mechanisms**

5.1 Hematologic disorders: chronic hemolytic anemia, myeloproliferative disorders, splenectomy

5.2 Systemic disorders: sarcoidosis, pulmonary histiocytosis, lymphangiomyomatosis

5.3 Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders

5.4 Others: tumoral obstruction, fibrosing mediastinitis, chronic renal failure, segmental PH