

Clinical classification of pulmonary hypertension⁵

Group 1. Pulmonary arterial hypertension (PAH)

- 1.1 Idiopathic PAH
- 1.2 Heritable PAH
- 1.3 Drug- and toxin-induced PAH including aminorex, cocaine, fenfluramine, dexfenfluramine, benfluorex, methamphetamines, dasatinib, and toxic rapeseed oil; some possible toxins include cocaine and St John's wort (for a full list see original reference)
- 1.4 PAH associated with:
 - 1.4.1 Connective tissue disease
 - 1.4.2 HIV infection
 - 1.4.3 Portal hypertension
 - 1.4.4 Congenital heart disease
 - 1.4.5 Schistosomiasis
- 1.5 PAH long-term responders to calcium channel blockers
- 1.6 PAH with overt features of venous/capillaries (pulmonary veno-occlusive disease/pulmonary capillary haemangiomatosis) involvement
- 1.7 Persistent pulmonary hypertension of the newborn syndrome

Group 2. Pulmonary hypertension due to left heart disease

- 2.1 Heart failure with preserved left ventricular ejection fraction (LVEF)
- 2.2 Heart failure with reduced LVEF
- 2.3 Valvular heart disease
- 2.4 Congenital/acquired cardiovascular conditions leading to postcapillary PH

Group 3. PH due to lung diseases and/or hypoxia

- 3.1 Obstructive lung disease
- 3.2 Restrictive lung disease
- 3.3 Other lung disease with mixed restrictive/obstructive pattern
- 3.4 Hypoxia without lung disease
- 3.5 Developmental lung disorders

Group 4. PH due to pulmonary artery obstructions

- 4.1 Chronic thromboembolic pulmonary hypertension (CTEPH)
- 4.2 Other pulmonary artery obstructions including tumours, arteritis without connective tissue disease, congenital pulmonary artery stenosis, and parasites

Group 5. PH with unclear and/or multifactorial mechanisms

- 5.1 Haematological disorders including chronic haemolytic anaemia, and myeloproliferative disorders
- 5.2 Systemic and metabolic disorders including pulmonary Langerhan's cell histiocytosis, Gaucher's disease, glycogen storage disease, neurofibromatosis, and sarcoidosis
- 5.3 Others including chronic renal failure with or without haemodialysis, and fibrosing mediastinitis
- 5.4 Complex congenital heart disease