Table 1

Evolution of Pulmonary Hypertension Classification: Fifth World Symposium on Pulmonary Hypertension (Nice, France; 2013)[a]

1. Pulmonary arterial hypertension
   - 1.1 Idiopathic
   - 1.2 Heritable
     - 1.2.1 *BMPR2*
     - 1.2.2 *ALK-1, ENG,* ***SMAD9, CAV1, KCNK3***
     - 1.2.3 Unknown
   - 1.3 Drug and toxin induced
   - 1.4 Associated with:
     - 1.4.1 Connective tissue disease
     - 1.4.2 Human immunodeficiency virus infection
     - 1.4.3 Portal hypertension
     - 1.4.4 Congenital heart disease
     - 1.4.5 Schistosomiasis
   - 1′[b] Pulmonary veno-occlusive disease and/or pulmonary capillary hemangiomatosis
   - **1″[c] Persistent pulmonary hypertension of the newborn**
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 disease and/or hypoxia
   - 3.1 Chronic obstructive pulmonary 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 diseases
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 disorder: sarcoidosis, pulmonary histiocytosis, lymphangioleiomyomatosis
   - 5.3 Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders
   - 5.4 Others: tumoral obstruction, fibrosis mediastinitis, chronic renal failure, **segmental pulmonary hypertension**

[a] Modifications to the previous classification are represented in bold.
[b] 1′ indicates a distinct category, but one that is not completely separated from pulmonary arterial hypertension.
[c] 1″ indicates a distinct category, but is not completely separated from pulmonary arterial hypertension.

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