TABLE 1
2018 World Symposium of Pulmonary Hypertension (NICE) Classification System.

1 Pulmonary Arterial Hypertension
**1.1 Idiopathic PAH**
**1.2 Heritable PAH**
1.3 Drug- and toxin-induced PAH (table 3)
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 (PVOD/PCH) involvement**
**1.7 Persistent PH of the newborn syndrome**

2 PH due to left heart disease
2.1 PH due to heart failure with preserved LVEF
2.2 PH due to heart failure with reduced LVEF
2.3 Valvular heart disease
**2.4 Congenital/acquired cardiovascular conditions leading to post-capillary PH, including pulmonary vein stenosis**

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, including pulmonary growth abnormalities, congenital diaphragmatic hernia, bronchopulmonary dysplasia and Down syndrome**

4 PH due to pulmonary artery obstructions
4.1 Chronic thromboembolic PH
4.2 Other pulmonary artery obstructions

5 PH with unclear and/or multifactorial mechanisms
**5.1 Hematological disorders, including sickle cell disease**
5.2 Systemic and metabolic disorders
5.3 Others
5.4 Complex congenital heart disease

Adapted from reference 1