Table 1

Updated clinical classification of pulmonary hypertension[3]

Aetiology of PH	Treatment
1 Pulmonary arterial hypertension (PAH) **1.1** Idiopathic **1.2** Heritable **1.2.1** BMPR2 (bone morphogenetic protein receptor type 2) **1.2.2** ALK-1 (activin receptor-like kinase 1 gene), endoglin **1.2.3** Unknown **1.3** Induced by drugs and toxins **1.4** Associated with pulmonary arterial hypertension (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 **1.4.6** Chronic haemolytic anaemia **1.5** Persistent pulmonary hypertension of the newborn	PAH specific therapies (Bosentan, ambrisentan, macitentan*, sildenafil, tadalafil, riociguat*, epoprostenol, treprostinil*, iloprost) * not yet licensed in the UK
1' Pulmonary veno-occlusive disease and/or pulmonary capillary haemangiomatosis	Unclear if PAH therapies are beneficial and may cause harm
2 Pulmonary hypertension due to left heart disease **2.1** Systolic dysfunction **2.2** Diastolic dysfunction **2.3** Valvular disease	PAH therapies are contraindicated Treatment of underlying heart disease and cardiovascular risk factors
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 abnormalities	PAH therapies are contraindicated Treatment of underlying lung disease
4 Chronic thromboembolic pulmonary hypertension	Lifelong warfarin Pulmonary endarterectomy PAH therapies may be beneficial in some cases
5 Pulmonary hypertension with unclear and/or multifactorial mechanisms **5.1** Haematological disorders: myeloproliferative disorders, splenectomy **5.2** Systemic disorders: sarcoidosis, pulmonary Langerhans cell histiocytosis, lymphangioleiomyomatosis, neurofibromatosis, vasculitis **5.3** Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders **5.4** Others: tumoral obstruction, fibrosing mediastinitis, chronic renal failure on dialysis	Unclear if PAH therapies are beneficial

Adapted from Simonneau G et al. Updated clinical classification of pulmonary hypertension. *J Am Coll Cardiol* 2009;54:S43-S54