Bijlage II

Clinical Classification of Pulmonary Hypertension (Nice 2013)

1. Pulmonary Arterial Hypertension (PAH)
1.1. Idiopathic PAH (iPAH)
1.2. Heritable PAH (HPAH)
1.2.1. BMPR2 mutation
1.2.2. ALK1, endoglin, SMAD9, CAV1, or KCNK3 mutation
1.2.3. Unknown mutation
1.3. Drugs and toxins induced
1.4. Associated with
1.4.1. Connective tissue disease (PAH-CTD)
1.4.2. HIV Infection (PAH-HIV)
1.4.3. Portal hypertension
1.4.4. Congenital heart diseases (PAH-CHD)
1.4.5. Schistosomiasis
1' Pulmonary veno-occlusive disease (PVOD) and/or pulmonary capillary hemangiomatosis (PCH)
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
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 disease 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
3.7.1 Congenital diaphragmatic hernia
3.7.2 Bronchopulmonary dysplasia
4. Chronic tromboembolic pulmonary hypertension (PH-CTE)
5. Pulmonary hypertension with unclear or multifactorial mechanisms
5.1. Hematologic disorders: chronic haemolytic anaemia, 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: Segmental PAH, tumoral obstruction, fibrosing mediastinitis, chronic renal failure