Pulmonary Hypertension Classification (WHO)

The World Health Organization (WHO) classifies pulmonary hypertension into 5 main groups based on etiology. Group 1 includes patients with pulmonary arterial hypertension (PAH), which can be due to drugs, associated diseases, idiopathic, or genetic. Hereditary PAH is often due to a BMPR2 mutation. Group 2 includes patients whose pulmonary hypertension is due to left heart disease. Group 3 includes patients who pulmonary hypertension is due to chronic lung diseases or hypoxia such as from COPD or obstructive sleep apnea. Group 4 includes patients with chronic pulmonary embolisms while Group 5 patients have multifactorial etiologies.

PLAY PICMONIC

Group 1

Pulmonary Arterial Hypertension
Lungs Artery and BP
Group 1 patients are those with pulmonary arterial hypertension (PAH). This is due to increased pulmonary vascular resistance. PAH may be idiopathic, hereditary, drug-induced, or associated with other diseases.

Idiopathic
Idiot-path
Idiopathic pulmonary arterial hypertension (IPAH) is the most common type. It occurs sporadically.

Hereditary
Hair-red-kid
Hereditary (or familial) PAH is often associated with a mutation in bone morphogenetic protein, receptor type 2 (BMPR2). This subtype of PAH is inherited in an autosomal dominant fashion.

BMPR2 Mutation
BuMPeR-car (2) Tutu Mutant
Bone morphogenetic protein receptor type 2 (BMPR2) normally inhibits vascular smooth muscle proliferation. The loss-of-function mutation seen in hereditary PAH causes increased cellular proliferation thus increasing pulmonary vascular resistance.

Group 2

Left Heart Disease
Left Heart Diseased
Group 2 patients are those with left heart disease. Left heart disease can cause pulmonary hypertension due to pulmonary vessel congestion or increased pulmonary blood floow. Left heart disease includes valvular heart disease, systolic/diastolic dysfunction, and left-to-right shunts. Treatment focuses on improving comorbid conditions.

Group 3

Lung Diseases or Hypoxia
Lungs Diseased and Hippo-O2
Patients are classified in group 3 if their pulmonary hypertension is due to chronic lung disease or hypoxia. Over time, hypoxic pulmonary vasoconstriction causes descruction of the vascular bed and smooth muscle hypertrophy. Since the total cross-sectional area of pulmonary capillaries decreases, pulmonary pressures increase.

COPD

COPD-Cop

Chronic obstructive pulmonary disease (COPD) is a common cause of PH.

Obstructive Sleep Apnea

Obstructed Sleeping Ape-needs-air

Obstructive sleep apnea (OSA) is another cause of PH. Repetitive nocturnal oxygen desaturation results in the same hypoxic vasoconstriction as seen in COPD that over time will increase pulmonary vascular resistance.

Group 4

Chronic Pulmonary Emboli

Crone with Lungs Elmo

Group 4 patients have chronic or recurrent pulmonary embolisms. The persisent obstruction of pulmonary vessels causes increased pulmonary vascular resistance.

Group 5

Multifactorial

Multiple-factories

Group 5 PH patients have multifactorial or unique etiologies such as metabolic systemic, or hematologic disease. End-stage renal disease or extraluminal compression from a mass can also result in PH.