WHAT'S YOUR

WORLD SYMPOSIA ON PULMONARY HYPERTENSION

Pulmonary Hypertension Group Classifications

There are five distinct classifications of pulmonary hypertension (PH).¹ Across these groups, there are more than 30 different, specific causes of PH and people can have more than one type of PH.



Pulmonary Arterial Hypertension



PH Due to Left Heart Disease



PH Due to Lung Disease



Chronic Thromboembolic Pulmonary Hypertension (CTEPH)



PH Due to Unclear Mechanisms

CAUSES

Hereditary

Idiopathic/Unknown

Drug and toxin induced

(e.g., methamphetamines, amphetamines.)

Veno-occlusive disease

Diseases associated with PAH:

Connective tissue (e.g., scleroderma, lupus, etc.); HIV; portal hypertension; certain congenital conditions (e.g., congenital heart disease, liver disease); persistent pulmonary hypertension of the newborn

Left heart failure (heart failure with preserved or reduced ejection fraction)

Heart valve disease

Chronic lung disease conditions

(e.g., obstructive lung disease, chronic obstructive pulmonary disease, emphysema)

Restrictive lung disease

(interstitial lung disease, pulmonary fibrosis, sarcoidosis)

Obstructive sleep apnea

Alveolar hypoventilation disorders

Developmental lung diseases

Exposure to high altitudes for extended periods of time

Pulmonary embolisms

(blood clotting disorders in the lungs)

Hematologic disorders:

(Sickle cell anemia, chronic hemolytic anemia, myeloproliferative disorders, splenectomy)

Systemic disorders:

(Sarcoidosis, pulmonary histiocytosis, lymphangioleiomyomatosis, neurofibromatosis and vasculitis)

Metabolic disorders:

(e.g., Gaucher disease, thyroid disorders)

Other diseases:

(e.g., chronic renal failure with AV fistula, fibrosing mediastinitis)

TREATMENT OPTIONS²

Medications may include pills, inhaled therapies, continuous infusions and supplementary oxygen indicated for a specific type of PH.

Depending on the severity of the PH, lung transplantation may be an option. Managed by treating the underlying heart conditions.

Depending on the severity of the PH, a heart and/or lung transplantation may be an option. Managed by treating the underlying lung conditions.

In some instances, specialized therapy may be appropriate.

Depending on the severity of the PH, lung transplantation may be an option Surgery such as balloon pulmonary angioplasty or pulmonary thromboendarterectomy surgery can remove blood clots and eliminate PH.

For CTEPH patients who are not eligible for surgery. PH-targeted therapy can be prescribed.

Depending on the severity of the PH, lung transplantation may be an option.

Managed by therapies treating underlying conditions.

DIAGNOSTIC TESTS TO DETERMINE YOUR SPECIFIC PH GROUP³

Multiple diagnostic studies are conducted including: echocardiogram, electrocardiogram, right heart catheterization, pulmonary function tests, lung ventilation/perfusion scan, blood tests — HIV, antinuclear antibody test and liver function studies.

Additional testing may be required if the above tests fail to identify your PH type. Other testing may include cardiac stress tests, cardiopulmonary exercise tests, CT scans, pulmonary angiogram, cardiac MRI, sleep study or nighttime oximetry, immunology and abdominal ultrasound scans.

Produced by the Pulmonary Hypertension Association | Reference: Sysol, J. and Machado, R., 2018. Classification and pathophysiology of pulmonary hypertension. Continuing Cardiology Education, [online] 4(1), pp.2-12. Available at: https://doi.org/10.1002/cce2.71.

- 1 PH is a general term used to describe high blood pressure in the lungs from any cause.
- 2 Treatments are determined based upon the underlying cause.
- 3 Before treatment is prescribed, your type of PH must be determined. Therefore, a number of diagnostic tests are conducted. These tests are important because therapies that treat one group of PH may be harmful to another group.

