Pulmonary Arterial Hypertension

Hipertensión arterial pulmonar

WHAT IS PULMONARY ARTERIAL HYPERTENSION (PAH)?

PAH, usually called "pulmonary hypertension", is a rare disease affecting 10-20 per 1,000,000 individuals.
To lead a normal life, blood must be oxygenated, and this process takes place in the lungs (see Figures).
When blood pressure in the pulmonary artery rises, the right side of the heart has to work harder, causing its enlargement and, finally, its lack of force to pump enough blood through the lungs.
As a result, patients have shortness of breath and fatigue on exertion.

WHAT CAUSES PAH?

The walls of the small arteries carrying blood through the lungs become thick and narrow, increasing resistance to blood flow.
In many cases, the cause is unknown, and is called "idiopathic pulmonary hypertension"; in others, it is hereditary and associated with a specific gene mutation.

WHAT DISEASES OR MEDICAL CONDITIONS PREDISPOSE TO THE DEVELOPMENT OF PAH?

- Connective tissue disorders: Scleroderma, lupus, rheumatoid arthritis, and others.
- Liver diseases, such as cirrhosis.
- Human immunodeficiency virus (HIV) infection.
- Heart and lung diseases that cause high pulmonary pressure without this being strictly considered PAH.
- Blood diseases: Myeloproliferative syndromes or sickle cell anemia.
- Drugs: Amphetamines, methamphetamines, and cocaine.

WHAT IS CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION?

When a blood clot blocks the pulmonary arteries, and despite anticoagulant therapy the patient continues with high pressure in those arteries, a disease known as "chronic thromboembolic pulmonary hypertension" can develop.
It is important to identify this form of pulmonary hypertension because it has a specific treatment, which can be curative in many patients.

WHAT ARE THE SYMPTOMS OF PAH?

- Fatigue - shortness of breath.
- Fainting.
- Swelling of the ankles.
- Chest pain.
- Bluish color of the lips and skin.

HOW IS PAH DIAGNOSED?

If the doctor suspects PAH, he will order a Doppler-echocardiography to estimate pulmonary arterial pressure and determine if there are signs of PAH.
Depending on the results, he may order a cardiac catheterization to measure the pressure within the pulmonary artery.

HOW IS PAH TREATED?

There are general guidelines for all patients.
- A salt-free diet.
- Pneumonia and flu vaccines.
- Women with PAH are advised to avoid pregnancy.
- Avoid strenuous exercise.
- Patients with severe PAH and low blood oxygen level may need oxygen supply if traveling by plane.

Treatment depends on the cause of the underlying condition and severity of the disease, and varies from treating the condition causing high pulmonary pressure to specific treatments to reduce it.
PAH patients should be treated by multidisciplinary teams in specialized centers for the treatment of this disease.

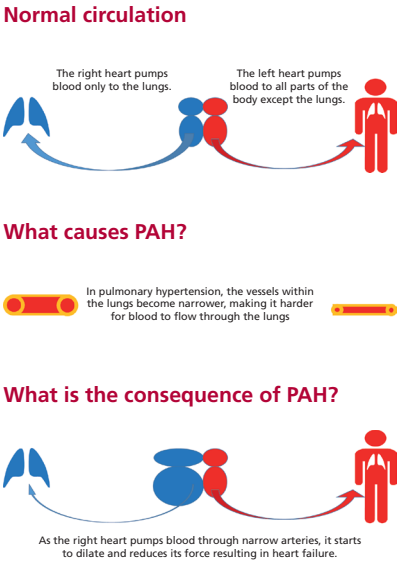

Diagram: Ignacio Bluro, M.D.

Fig. 1. PAH mechanisms and consequences.

Author: Ignacio Martín Bluro, M.D.
Staff Physician - Department of Cardiology,
Hospital Italiano de Buenos Aires

Editor: Julio Manuel Lewkowicz, MD[MTSAC]
Sanatorio Güemes, Buenos Aires