PH Simplified: The Basics of PH and WHO Classification Groups

Session Description
Panelists will review the basics of PH: how the heart is affected by high blood pressure in the lungs, the difference between high blood pressure in the lungs and high blood pressure throughout the rest of the body, as well as common PH symptoms. Panelists will also discuss the different types of PH and the reasons behind the extensive testing that is required for accurate diagnosis and choosing the best treatment plan.

Speakers
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Learning Objectives
Attendees will be able to:
- Understand the interaction of the pulmonary circulation, heart and lungs
- Understand the different problems that can lead to Pulmonary Hypertension (PH)
- Understand the current PH classification system
- Understand why diagnostic tests are ordered and why they are important
- Understand how management can be different for different types of PH

Overview of PH
PH is high blood pressure in the blood vessels in the lungs, which is very different from systemic hypertension (e.g., blood pressure measured in your arm). PH can cause the right side of your heart to work harder; the goals of PH treatment are to make you feel better, to help you to do more and to prevent progression to right heart failure. Understanding the common diagnostic tests performed in the evaluation of PH and why they are performed is helpful to understand how different types of PH may be treated differently.

Common Symptoms of PH
Many of the symptoms of PH are common and can be seen with other problems of the heart or lungs
- Shortness of breath: Some people experience a mild form of this, or only when they exercise
- Heart palpitations
- Swelling of the legs and weight gain
- Passing out or a feeling that you might pass out, particularly with exercise

Common Tests
- History and Physical
- Blood tests
- Chest x-ray and/or chest CT scan
- Echocardiogram (sonogram of the heart)
- Electrocardiogram (EKG)
- 6-minute walk test
- Right heart catheterization
Other Tests

- Pulmonary function testing
- Ventilation/perfusion scan
- Overnight oximetry or sleep study (overnight polysomnography)
- Cardiac stress test
- Left heart catheterization

Why so much testing is necessary

- It is important to be sure that your symptoms are from PH and not another cause, so your doctor may request some tests to rule out other causes, such as asthma or problems with how your heart squeezes
- PH can only be definitively diagnosed with a right heart catheterization
- It is important to understand what type of PH someone has as treatment varies depending on the cause

Types of PH

- Updated 2013 World Health Organization (WHO) PH Classification System
  - Group 1: Pulmonary arterial hypertension (PAH)
    - Often associated with autoimmune diseases like scleroderma, use of certain drugs/toxins, congenital heart disease, liver disease, HIV infection; sometimes the cause is unknown (called idiopathic)
    - Treatment is with PH-specific medication(s) to lower the blood pressure in the veins in the lungs
  - Group 2: PH due to left heart disease
    - This is the most common type of PH
  - Group 3: PH due to lung diseases and/or hypoxia
  - Group 4: Chronic thromboembolic pulmonary hypertension (CTEPH):
    - This is the only type of PH that is curable without transplant
  - Group 5: PH with unclear multifactorial mechanisms

- PH vs. PAH
  - PH is defined as an elevated mean (or “average,” which is different than the upper [systolic] and lower [diastolic] blood pressure readings) pulmonary arterial pressure (≥ 25 mmHg) as directly measured by right heart catheterization (RHC) at rest; all five groups have PH
  - PAH (Group 1) is defined by:
    - Elevated mean (average) pulmonary arterial pressure (≥ 25 mm Hg) plus elevated pulmonary vascular resistance plus normal pressures on the left side of the heart (pulmonary artery “wedge” pressure)

No evidence of significant lung disease, heart disease or chronic thromboembolic disease