

**Pulmonary
Hypertension and
Sarcoidosis**

PH&S

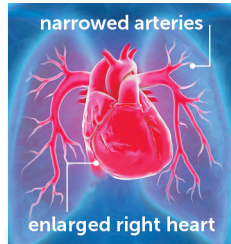


Pulmonary Hypertension Association
Empowered by hope

ABOUT

PULMONARY HYPERTENSION

Pulmonary hypertension is a complex and often misunderstood disease. PH sometimes is confused with systemic hypertension or “high blood pressure” that affects arteries throughout the body.



PH refers to pressure within the blood vessels of the lungs. The blood vessels can become stiff and narrow, which makes it more difficult for the right side of the heart to pump blood through them.

There are five types of PH based on different causes. Each form of PH is different, so it is important for newly diagnosed patients to find PH specialists who can pinpoint the cause of their PH. The specialist then develops a treatment plan specifically for the patient’s type of PH.

Every person with PH is different, and scientists continually conduct new research to improve the outlook for people living with PH. With proper care and treatment, people with PH can live many years.



NORMAL mean pulmonary artery pressure is between 8-20 mmHg at rest.

PULMONARY HYPERTENSION is defined as a resting mean pulmonary artery pressure at or above 20 mmHg.



Sarcoidosis-Related PH

Sarcoidosis is an inflammatory disease that can affect many different organs in the body. The exact cause is a mystery to doctors and researchers.

Sarcoidosis often causes problems in the lungs, which leads to shortness of breath with activity. This can happen because of problems in the:

- > Lung tissues (scarring/fibrosis or inflammation).
- > Airways (similar to asthma).
- > Blood vessels (elevated pressure or PH).

In sarcoidosis-associated PH, increased pressure in the lungs can weaken the right side of the heart and ultimately cause it to fail.

Sarcoidosis-related PH can happen for several reasons. Fibrosis, inflammation and airflow obstruction all can cause some degree of PH, since they make it harder for the lungs to transfer oxygen. Furthermore, the fibrosis itself can distort and destroy the lungs' blood vessels, elevating pressures.

Some patients have enlarged lymph nodes, which put pressure on lung vessels and cause PH. In others — when there is little or no fibrosis, inflammation or airflow obstruction — PH is thought to be the result of sarcoidosis directly affecting pulmonary vessels.

It's important to report changes in symptoms, such as unexplained shortness of breath; increased fatigue; swelling of the ankles, legs or abdomen; chest discomfort or pain; and light-headedness or fainting. All can be signs of sarcoidosis-related PH. People with sarcoidosis often have exercise limitations.

Pulmonary complications are the leading cause of death in sarcoidosis patients, so regular follow-up visits with your health care team are important. While many sarcoidosis patients go on to lead long lives without much trouble from their disease, sarcoidosis-related PH is a serious and complex condition that requires further evaluation.

Diagnosing PH

To determine whether someone with sarcoidosis has developed PH, a doctor likely will order pulmonary function tests, a six-minute walk test, blood work, a chest X-ray or chest CT scan, an echocardiogram and a right heart catheterization. A heart catheterization is the definitive test to diagnose PH.





Chest x-ray showing stage 2 sarcoidosis.

Case courtesy of Frank Gaillard, MBBS, MMed, Radiopaedia.org, rID: 6546

between the air entering the lungs and the blood circulating through the lungs (diffusion capacity of carbon monoxide).

Ventilation-perfusion (V/Q) scan: Determines whether there are undissolved clots in the blood vessels of the lungs.

Natriuretic peptide tests: Measure specific proteins in a blood sample and help screen for heart failure. Tests of the proteins NT-proBNP (N-terminal pro b-type natriuretic peptide) and BNP (brain natriuretic peptide) are two examples. Elevated levels of these proteins can signify heart failure.

Right heart catheterization: The most accurate diagnostic test for PH. It measures the pressures and flow directly in the pulmonary artery. The invasive procedure usually takes place on an outpatient basis in the hospital. Doctors guide a small tube (catheter) through the right side of the heart and into the pulmonary artery to directly measure the pulmonary artery pressure.

Declining diffusion capacity is known to be associated with PH development among people with sarcoidosis. In addition, many patients whose oxygen levels fall during a six-minute walk test also have underlying SAPH.

Echocardiogram: An ultrasound of the heart that provides a rough estimate of the pressure in the lungs. It assesses the function of how blood flows through the heart by examining the right and left ventricles, valves, etc.

Pulmonary function tests: Non-invasive tests that measure how much air the lungs can hold, how much and how quickly someone can blow air, and how oxygen and carbon dioxide are exchanged

Treatment Options

All people with sarcoidosis-related PH should have therapies to optimize their sarcoidosis-specific treatment, including immunosuppressive therapies and bronchodilators. In addition, patients should pay attention to optimal fluid balance with diuretics (water pills) and a low-sodium diet. Patients should be evaluated for low oxygen levels because oxygen levels can fall when exercising and sleeping.

Exercise is of utmost importance. Cardiopulmonary rehabilitation has been shown to improve symptoms and exercise capacity for people with various forms of PH.

There is limited information about the effectiveness of PH-specific medications for people with sarcoidosis. Several studies that could provide more information are underway. The decision whether to use PH-specific medications can be complicated and requires careful follow up by a PH specialist.





ADDITIONAL RESOURCES

Foundation for Sarcoidosis
Research
Stopsarcoidosis.org

ACKNOWLEDGMENTS

Robert P. Baughman, MD,
University of Cincinnati.

H. James Ford, MD, University of
North Carolina, Chapel Hill.

Oksana A. Shlobin, MD, Inova
Fairfax Hospital, Falls Church,
Virginia.








About the Pulmonary Hypertension Association

Headquartered in Washington, D.C., the Pulmonary Hypertension Association is the oldest and largest nonprofit patient association dedicated to the pulmonary hypertension community. PHA's mission is to extend and improve the lives of those affected by PH.

PHA engages people with PH and their families, caregivers, health care providers and researchers, who work together to advocate for the PH community, provide support to patients, caregivers and families, offer up-to-date education and information on PH, improve quality patient care, and fund and promote research. For information, visit PHAssociation.org.

PHA's mission is to extend and improve the lives of those affected by pulmonary hypertension. PHA's vision is a world without PH, empowered by hope.



-  301-565-3004
-  Support Line: 800-748-7274
-  PHAssociation.org
-  [/PulmonaryHypertensionAssociation](https://www.facebook.com/PulmonaryHypertensionAssociation)
-  [@PHAssociation](https://www.instagram.com/PHAssociation)
-  [/PHAssociation](https://www.linkedin.com/company/PHAssociation)
-  [/user/PHAssociation](https://www.youtube.com/user/PHAssociation)