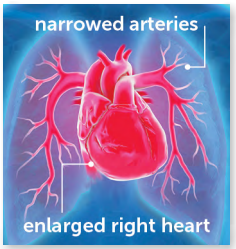
Pulmonary Hypertension and Sickle Cell Disease

PH&S

PHA
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.

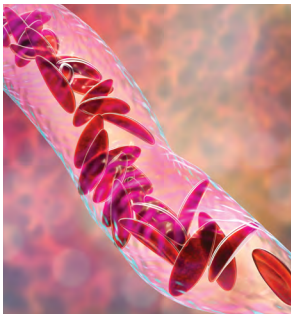
NORMAL mean pulmonary artery pressure is between 8-20 mmHg at rest.

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

Sickle Cell Disease-Related PH

Sickle cell disease, a type of inherited anemia (shortage of healthy red blood cells), is characterized by abnormally shaped red blood cells. Named for the sickle, or crescent shape, of the abnormal blood cells, the sickle cells aren't as flexible as normal blood cells. That means they easily break or get stuck in blood vessels.

The condition can cause life-long complications to many organs in the body, including the lungs. PH is an increasingly recognized complication of sickle cell disease. Studies show that 6% to 11% of adults with sickle cell disease develop PH. Other studies suggest that PH is the greatest risk factor for death among aging patients with sickle cell disease.



Red blood cells with sickle shape.

PH can occur in several ways in sickle cell disease, such as problems with the left side of the heart, obstructive sleep apnea (problems with breathing during sleep) and blood clots in the lungs.

However, the most concerning cause of PH in sickle cell disease is related to blood vessels in the lungs. Recent autopsy studies suggest that up to 75% of sickle cell patients have changes in the lung blood vessels at the time of death, signifying the presence of PH or alterations that can cause PH.

One possible explanation: The increased breakdown of red blood cells within the blood vessels in the lungs causes inflammation and reduces nitric oxide (a substance that causes blood vessels to relax). That causes small arteries in the lungs to constrict and narrow, leading to PH.

PH symptoms include shortness of breath with activity, increased fatigue, swelling of the ankles, legs or abdomen, chest discomfort or pain, and light-headedness and fainting. In sickle cell disease, those symptoms can be mistakenly attributed to other problems, particularly anemia associated with sickle cell disease.

Because PH symptoms initially can be mild, it is important for people with sickle cell disease to share any of those symptoms with their doctors.

Screening for Sickle Cell Disease-Related PH

People with sickle cell disease are more likely to develop PH, and those who suffer from PH are at higher risk of death than those without PH. Your doctor may want to perform screening tests to make sure you don't have PH, even if you don't have symptoms.

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 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.

In addition to directly measuring pressures in the heart and lungs, a right heart catheterization can determine whether patients have problems with a stiff left side of the heart. Left heart disease also can cause PH. In those cases, doctors refer patients to a cardiologist to treat the heart disease.

Treatment Options

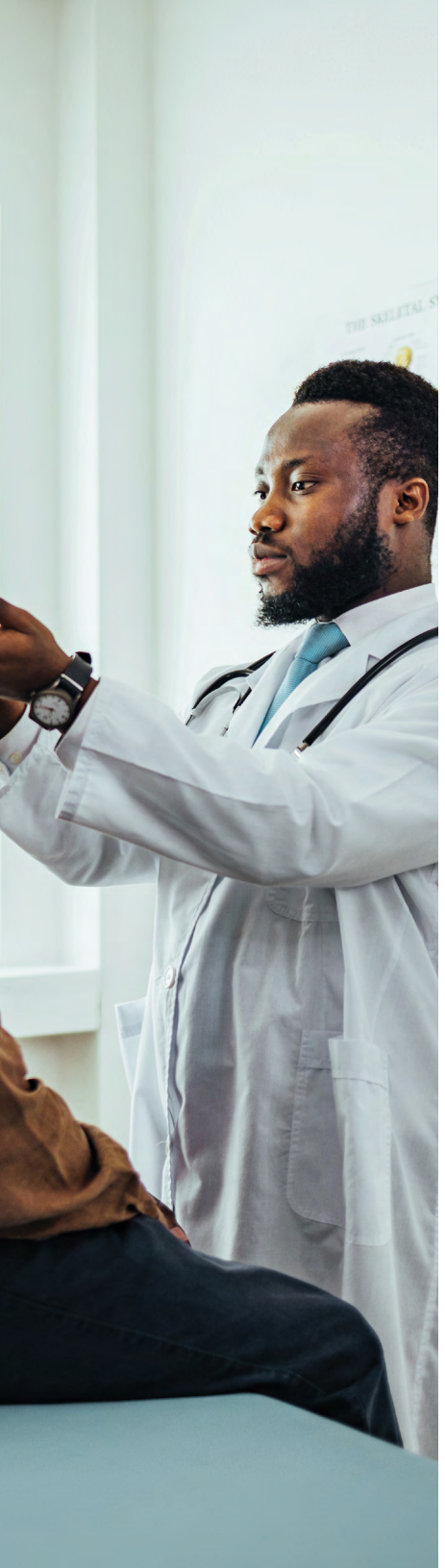
Current guidelines and the Pulmonary Hypertension Association recommend that PH specialists work with sickle cell doctors to evaluate every person whom they suspect of having PH. The patient must have a right heart catheterization, which helps physicians determine appropriate treatment as well as the cause and severity of the patient's PH.

Regardless of the type of PH you have, PHA recommends you receive the best possible treatment for your sickle cell disease. Sometimes that includes the medication hydroxyurea or chronic blood transfusions.

It is important to screen for and treat other causes of sickle cell disease-associated PH, such as blood clots in the lungs and obstructive sleep apnea. Controlling your sickle cell disease and other problems that contribute to PH can help prevent your PH from worsening.

While there are no drugs approved specifically to treat sickle cell disease-associated PH, patients with PH attributed to the blood vessels of the lungs may respond to medications that have benefitted patients with Group 1 PH (pulmonary arterial hypertension, or PAH).

No matter what treatments your doctor recommends, you should continue to follow up with your health care team to monitor your progress throughout treatment.



ADDITIONAL RESOURCES

Sickle Cell Disease Association of America
Sicklecelldisease.org

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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.

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