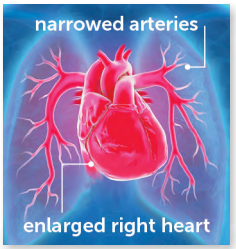
Pulmonary Hypertension and Congenital Heart Disease

PH&

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.

Congenital Heart Disease-Associated PH

Congenital heart disease can result from various heart and blood vessel abnormalities that develop before a person is born. These abnormalities often fall into two categories: simple (a single abnormality) or complex (multiple abnormalities).

CHD occurs in about one in 100 children, making it one of the most common congenital conditions. People with CHD live much longer today because of recent improvements in surgery, treatments and medications. There are now more adults — about 1.4 million in the U.S. — than children with CHD.

Three to 10 percent of people with CHD will develop PH. PH with CHD can significantly worsen symptoms and lead to a shorter lifespan, so it's important to secure proper diagnosis and treatment for PH and CHD.

Many types of CHD can cause PH. When CHD leads to PH, it's often because of increased blood flow and pressure into the blood vessels in the

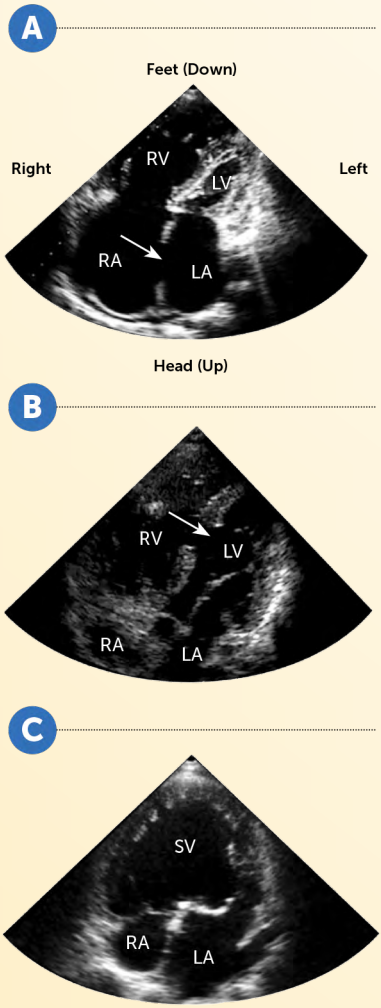
lungs. The increased flow results from defects or shunts in the heart or the major vessels. Common examples include holes between the upper heart chambers (atrial septal defects) or the lower heart chambers (ventricular septal defects). In those cases, blood that has already gone through the lungs flows from the left heart into the right heart and then back into the lung vessels. If there is enough extra flow for a long enough time, the lung vessels may become thickened and rigid, and the pressure within them will increase.

If the pressure in the blood vessels in the lungs increases enough, the flow across the defect in the heart will start to move in the opposite direction. As a result, poorly oxygenated blood flows back to the rest of the body, which lowers the overall blood oxygen level and causes cyanosis (blue fingers and lips). This condition is called Eisenmenger Syndrome. It is important to detect heart defects and shunts as early as possible so doctors can more easily correct them, which can help prevent PH. Recent evidence suggests current PH medications combined with other therapies can help people who have developed CHD-associated PH.

Echocardiogram images showing different congenital heart diseases — spaces between the chambers of the heart marked with white arrows — leading to PAH.

Images show the heart upside down, so the left and right ventricles and atrium are reversed.

RV: *Right ventricle,* **LV:** *Left ventricle,* **SV:** *Single ventricle,* **RA:** *Right atrium,* **LA:** *Left atrium.*



Diagnosing PH

Doctors who suspect PH in addition to congenital heart disease can order any of the following diagnostic tests:

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

People with CHD-associated PH should be followed closely and regularly by doctors in centers with experience managing the condition. This is especially true for people with Eisenmenger Syndrome.

In general, people with CHD-associated PH benefit from a team-based approach to care with input from a CHD specialist and a PH specialist. There also are several specific treatment options that should be considered:

Oxygen supplementation: Many people with CHD-associated PH have low oxygen levels in the blood. Some also have varying degrees of lung disease. Oxygen therapy hasn't definitively shown that it improves exercise tolerance or survival, but it might improve symptoms.

Exercise: Most people with CHD-associated PH benefit from regular, low- to moderate-intensity exercise such as walking. In most cases, patients should avoid high-intensity exercise because it's usually not well tolerated.

Anticoagulation: Anticoagulants are blood thinners used to prevent clots. Many people with CHD-associated PH have increased risk of bleeding, so the benefits and risks of anticoagulation must be carefully balanced. Aspirin may be preferable to a direct-acting anticoagulant for many patients.

Diuretics: People with CHD-associated PH can develop fluid buildup in their abdomen or legs. When this happens, diuretics (water pills) can help remove excess fluid. Doctors will adjust the medications to prevent dehydration, which can lead to kidney damage, low blood pressure and a dangerous thickening of the blood.

Treatment of iron deficiency: The low blood-oxygen levels in people with Eisenmenger Syndrome can cause the body's blood cell count to increase. Drawing blood generally isn't recommended because it can cause blood to thicken, particularly if fluid isn't adequately replaced. Iron deficiency is common and should be addressed.

Pregnancy avoidance: CHD-associated PH carries an increased risk for mother and fetus during pregnancy. People with Eisenmenger Syndrome have up to a 50 percent risk of maternal death and a 40 percent risk of miscarriage. Women with CHD-associated PH generally should avoid pregnancy and discuss birth control options with their doctor.

Advanced medical therapy: People with CHD-associated PH should be considered for PH-specific medicines. There are several medical trials available for people with this.

Transplant: If symptoms worsen despite medical therapy, patients may be considered for lung or combined heart and lung transplantations. Those undergoing lung transplantation may need their CHD to be repaired surgically at the same time, if not before. People with CHD-associated PH should be referred early for transplantation consideration to allow adequate time for evaluation.

ADDITIONAL RESOURCES

Adult Congenital Heart Association | Achaheart.org

American Heart Association | Heart.org

Mended Little Hearts | Mendedhearts.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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