Pulmonary Hypertension & CONGENITAL HEART DISEASE
Pulmonary hypertension (PH) is a complex and often misunderstood disease. The term PH simply means high blood pressure in the arteries of the lungs. In regular hypertension — also known as high blood pressure or systemic hypertension, which you can check with a blood pressure cuff — the pressure in the arteries of the entire body is higher than it should be. In PH, the blood vessels specifically in the lungs are affected. They can become stiff and narrow, and the right side of the heart must work harder to pump blood through them. There are five groups of PH based on different causes.

Each form of PH is different, so it is important for newly diagnosed patients to find a PH specialist who can accurately pinpoint what is causing their PH. They then can develop a treatment plan for that specific type as soon as possible after a confirmed diagnosis. Every individual with PH is different, and new research with the potential to improve the outlook for people living with this disease is conducted on a continual basis. Once in the care of a PH-treating health care team and on appropriate therapy, individuals with PH can live for 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 **25 mmHg**.*

PH, or high blood pressure in the lungs, frequently results from a narrowing of the small blood vessels in the lungs, which leads to a larger right side of the heart.
Congenital heart disease (CHD) can be a number of different heart and blood vessel abnormalities that develop before a person is born. These abnormalities are often split into two categories: simple (a single abnormality) or complex (multiple abnormalities). CHD is not a common condition. It affects less than 1 percent of people in the world.\(^1\) Today, individuals with CHD are living much longer because of recent improvements in surgery, treatments and medications. In fact, there are now more adults — more than 1.4 million in the U.S. alone — than children with CHD.\(^2\)

Many types of CHD can lead to PH. About 3 to 10 percent of people with CHD will develop PH.\(^3\) The development of PH in people with CHD may significantly worsen their symptoms and potentially shorten their lifespan, so it is important to be properly diagnosed and treated for both PH and CHD. PH usually develops in people with CHD because of increased blood flow into the blood vessels in the lungs. This flow often comes 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 these 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 period of time, the lung vessels may become thickened and stiff, 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. This results in poorly oxygenated blood flowing back to the rest of the body, which lowers the overall blood oxygen level and causes cyanosis (blue fingers and lips). This is known as Eisenmenger Syndrome. It is important to detect heart defects and shunts as early

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as possible so that they can be more easily corrected and PH can be avoided. Fortunately, recent evidence suggests that current PH medications, coupled with other therapies, can help individuals who have developed CHD-associated PH.

DIAGNOSING PH IN CHD

A cardiologist, or more specifically a CHD specialist, should be able to detect the development of PH. If someone with CHD notices an increase or worsening of symptoms (shortness of breath with activity, increased fatigue, swelling of the ankles, legs or abdomen, chest discomfort or pain, and light-headedness and fainting), this will alert the physician to the possibility of PH. A careful physical exam complemented by an echocardiogram — or cardiac ultrasound — is an important way to see if PH may be developing. A cardiac catheterization is the most definitive test to determine if PH is present. This allows your doctor to directly measure the pressure inside the blood vessels in the lungs. Right heart catheterization also helps the medical team to determine what type of PH is most likely present, which in turn will help them to select the most

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appropriate course of treatment. Finally, under certain circumstances, doctors can test how the heart and the body would respond to having the defect permanently closed to see if this would be a safe long-term treatment option.

**TREATMENT OPTIONS FOR CHD-ASSOCIATED PH**

People with CHD-associated PH should be watched 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. While oxygen therapy has not been definitively shown to improve exercise tolerance or survival, it may help to improve symptoms.

**Exercise:** Most people with CHD-associated PH benefit from regular, low- to moderate-intensity exercise such as walking. Care should be taken to avoid high-intensity exercise as it usually is not well tolerated.

**Anticoagulation:** Anticoagulants or blood thinners likely provide little benefit to most people with CHD-associated PH. Many people with CHD-associated PH are at increased risk of bleeding, so the level of anticoagulation also must be watched very carefully.

**Diuretics:** People with CHD-associated PH can develop fluid buildup in their abdomen or legs. When this happens, diuretics (water pills) can be used to help remove the excess fluid. Care must be taken to avoid overuse of these medicines because they can lead to dehydration, which can result in kidney damage, low blood pressure and a dangerous thickening of the blood.
Treatment of iron deficiency: The low blood-oxygen levels that individuals with Eisenmenger Syndrome may experience can cause the body to increase its blood cell count. Drawing blood is generally not recommended as it may lead to increased thickening of the blood, particularly if fluid is not adequately replaced. In any case, iron deficiency is fairly common and should be addressed.

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

Advanced medical therapy: Individuals with CHD-associated PH should be considered for PH-specific medicines. Persons with this condition have even participated in a number of clinical studies of these medicines.

The BREATHE-5 study specifically showed the benefit of bosentan, an endothelin receptor antagonist, in people with Eisenmenger Syndrome. The two other major classes of PH medicines (phosphodiesterase-5 inhibitors and prostacyclin receptor antagonists) also have been beneficial to individuals with CHD-associated PH. Such medicines should be used under the supervision of a PH specialist.

Transplant: If individuals with CHD-associated PH do poorly despite medical therapy, they may be considered for lung or combined heart and lung transplantations. Those undergoing isolated lung transplantation may need to have surgical repair of their CHD at the same time, if not before. Persons with CHD-associated PH should be referred early for consideration of transplantation to allow adequate time for a complete evaluation.

The Pulmonary Hypertension Association (PHA) is a community of individuals with PH, caregivers, families, medical professionals and researchers who work together to empower people battling the disease, while supporting research toward improved treatments. PHA was the first organization in the world dedicated to providing comprehensive PH patient and caregiver support, medical education, research and services that improve patients’ quality of life.

PHA facilitates more than 200 active support groups around the country and serves as a model for more than 85 international PH organizations. In addition to in-person support, PHA offers a telephone support line, an email mentorship program, educational patient programs and a variety of print and online publications. Every two years, PHA hosts its International PH Conference and Scientific Sessions, which brings together nearly 1,500 individuals with PH, caregivers and medical professionals to share information on the latest research, treatments and lifestyle coping tips.

PHA also has provided more than $8 million for PH research and offers continuing education programs for medical professionals through the PHA Online University and an in-person CME program.

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Richard A. Krasuski, M.D., Duke University Medical Center, Durham, N.C.
Michael A. Mathier, M.D., University of Pittsburgh Medical Center, Pittsburgh