

Early Detection and Innovative Treatment is Key to Preventing Progression of Pulmonary Hypertension

Pulmonary hypertension results from increased resistance to blood flow through the pulmonary vasculature (due to progressive muscular thickening and fibrosis of the blood vessels in the lungs). The disorder is chronic and has often progressed to a late stage by the time it is accurately diagnosed.

Commonly mistaken for asthma, this disease forces the heart's right ventricle to work harder, causing it to enlarge, thicken and experience an elevation in pressure. In the early stages, the changes in the blood vessels may be reversible, but do become permanent later. If this pressure is not relieved, right ventricular failure, arrhythmias and even death may result.

What is the incidence and prognosis?

Pulmonary hypertension occurs in eight out of 100,000 people. The long-term prognosis depends on its cause, the severity of the disease at the time of diagnosis and response to therapy. Recent advances in medical therapies have had a profound impact on the outcome of pulmonary hypertension.

What are the causes of pulmonary hypertension?

Pulmonary hypertension may be idiopathic. However, secondary pulmonary hypertension may occur in many types of diseases including congenital heart disease, lung disease, various connective tissue diseases and sickle cell disease.

What are the symptoms?

In the early stages, pulmonary hypertension may be asymptomatic. However, as the disease progresses, non-specific symptoms, such as increased fatigue and occasional shortness of breath may become apparent. With further deterioration, increasing dyspnea with less exertion, dizziness, palpitations and syncope may be present. Chest pain and peripheral edema may be present when pulmonary hypertension is severe. Unfortunately, many of the symptoms may mimic other pulmonary syndromes (such as asthma), thus necessitating a higher level of suspicion for this disease.

The Pulmonary Hypertension Clinic at Cincinnati Children's provides comprehensive diagnostics and long-term treatment and follow up for patients.

Our Team

Director

Russel Hirsch, MD

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For Referrals

Call Michelle Bagby, RN, MSN, CNP at 513-803-0695.

For more information on the pulmonary hypertension team or Cincinnati Children's Heart Institute, please visit www.cincinnatichildrens.org/heart.

What may be present in a physical examination?

Early clinical presentation signs may be subtle, and may include only a prominent second heart sound. With progression, more obvious signs related to elevated right ventricular and right atrial pressure may present, such as jugular vein distention, palpable second heart sound, hepatomegaly and peripheral edema. Murmurs may or may not be present.

How is pulmonary hypertension diagnosed?

A good history and physical exam can give you clues to diagnosis along with additional testing, including electrocardiogram, chest X-ray, echocardiogram, six-minute walk test, and the gold standard for diagnosis, cardiac catheterization.

How is pulmonary hypertension treated?

While there is no cure for pulmonary hypertension, there are many treatments that have been found to relieve some of the symptoms and slow the progression of the disease. These include oxygen, inhaled nitric oxide, diuretics, calcium channel blockers, sildenafil, bosentan and prostanoids, including prostacyclin.

Pulmonary Hypertension Clinic

Part of Cincinnati Children's Heart Institute, the Pulmonary Hypertension Clinic provides comprehensive diagnostics and long-term treatment and follow up for patients with pulmonary hypertension. Russel Hirsch, MD, the clinic's director, has expertise in performing cardiac catheterizations (he is also the director of the Heart Institute's Cardiac Catheterization Laboratory) and performs all cardiac catheterizations in this population. It should also be noted that the Pulmonary Hypertension Clinic has particular interest in infants with congenital diaphragmatic hernias with resulting pulmonary hypertension.

Contact the Pulmonary Hypertension Clinic at 513-803-0695.

Call Physician Priority Link to consult with a specialist 24 hours a day, 7 days a week: 1-888-636-7997.

U.S. News & World Report ranked Cincinnati Children's the third best children's hospital in the United States in general pediatrics and in the Top 10 for heart care and heart surgery.