

**Pulmonary  
Hypertension and  
Liver Disease**

**PH&L**

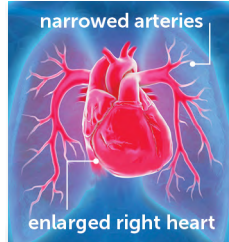


**Pulmonary Hypertension Association**  
Empowered by hope

ABOUT

# PULMONARY HYPERTENSION

**P**ulmonary 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.



## **Liver Disease-Associated PH (Portopulmonary Hypertension)**

The liver is the largest solid organ in the body, and it has many jobs crucial for survival. It produces a substance called bile that breaks down fats for digestion; stores vitamins; makes proteins necessary for blood clotting and other important functions; and ensures stable levels of sugar in the blood.

Most importantly for this topic, the liver filters and removes harmful substances from the blood.

If the liver becomes damaged or diseased, it can create problems throughout the rest of the body.

Damage to the liver over time can lead to a condition called cirrhosis, which refers to scarring of the liver tissue. The scarring impedes how much blood flows through the liver and impairs its function.

Cirrhosis is the most common cause of portal pulmonary hypertension, which is increased blood pressure in the portal veins that transport blood from the intestines to the liver.

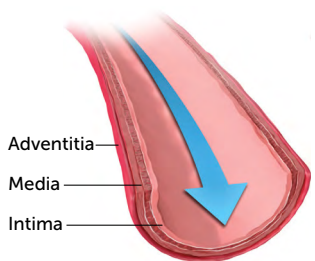
This increased pressure causes blood to bypass the liver. As a result, the blood is not filtered by the liver, and chemicals and toxins are not removed from the blood. The blood vessels of the lungs are then exposed to potentially toxic substances that can damage the small arteries of the lungs and cause Group 1 PH (pulmonary arterial hypertension, or PAH).

Portopulmonary hypertension is a type of PAH that occurs as a result of portal hypertension and advanced liver disease. Patients with PoPH experience similar symptoms as patients with other forms of PAH not associated with liver disease.

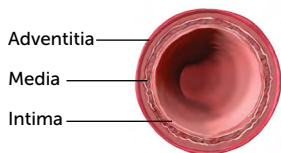
These symptoms can include shortness of breath with activity; increased fatigue; swelling of the ankles, legs or abdomen; chest discomfort or pain; and light-headedness and fainting.

PoPH is different from hepatopulmonary syndrome, another type of liver disease-associated abnormality of the pulmonary vessels. In contrast to PoPH, hepatopulmonary syndrome is characterized by widening of the pulmonary blood vessels and low oxygen levels in the blood.

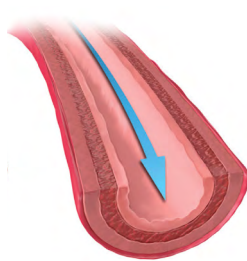
**A Healthy Pulmonary Artery**



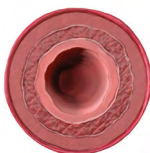
Arteries are healthy and flexible, blood flows easily through the vessels.



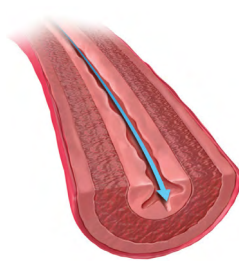
**B Mild PAH**



Thick and stiff artery walls limit blood flow and increase the resistance.



**C Moderate PAH**



Blood flow is restricted as the artery narrows.



The reason PoPH develops is unclear, but research has given us some clues. Patients with PoPH have a deficiency of prostacyclin (a substance that causes the blood vessels to relax), and an excess of endothelin1 (a substance that causes the blood vessels to constrict and the vessel walls to thicken).

In addition, women and those with autoimmune-related cirrhosis may be at increased risk of developing PoPH. Most patients diagnosed with PoPH are adults.

## Diagnosing PoPH

A specialist can diagnose PoPH when a patient has high blood pressure in the liver in addition to the usual signs of PAH. Doctors also can identify PoPH when people undergo tests to determine whether they are candidates for liver transplant.

Any patients with chronic liver disease and shortness of breath, even if they aren't considering transplant, should discuss with a specialist the possibility of getting an echocardiogram to screen for PH.

**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

Researchers are still studying treatment options for people with PoPH. Because people with PoPH have been excluded from most clinical trials for PH-specific drugs, doctors follow PAH treatment recommendations for PoPH gleaned from other PAH studies.

There is hope that existing PH-specific treatments may be helpful. Small studies have described improvement in PoPH with drugs such as epoprostenol, treprostinil, bosentan, ambrisentan, macitentan and sildenafil.

The outcome for people with PoPH after liver transplant is unpredictable. The risk of complications and death from liver transplants increases with higher pressures in the pulmonary arteries.

For this reason, it is important to lower the pressure in the pulmonary arteries with PH-specific treatments before liver transplantation. If the pressure is high in the lungs during transplant, mortality during and after surgery is more likely.





However, limited but encouraging data suggest that carefully selected patients who respond to PH-specific therapy can undergo liver transplantation with excellent survival rates and in some cases improvement or complete resolution of PoPH.

In all cases, the sooner a patient is tested, the sooner doctors can address or rule out problems like PoPH.

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### **ADDITIONAL RESOURCES**

American Liver Foundation:  
[Liverfoundation.org](http://Liverfoundation.org)

### **REFERENCES**

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Hilary DuBrock, MD, MMSc, Mayo Clinic, Rochester, Minnesota.

Kathryn del Valle, MD, Mayo Clinic, Rochester, Minnesota.

## 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.

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**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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