

**Pulmonary
Hypertension and
Chronic
Thromboembolic
Pulmonary
Disease**

PH&

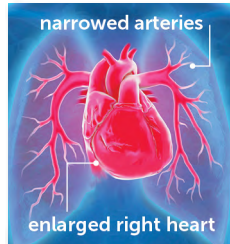


Pulmonary Hypertension Association
Empowered by hope

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.



Pulmonary Hypertension and Chronic Thromboembolic Pulmonary Disease

Chronic thromboembolic pulmonary disease with pulmonary hypertension, also known as Group 4 pulmonary hypertension, is a rare form of PH. It is a condition where there is elevated blood pressure in the pulmonary arteries caused by chronic blood clots, which block the free flow of blood through the lungs.

There is no known cause for developing chronic blood clots. Most people who develop a blood clot are prescribed a blood thinner to naturally dissolve the clot. However, clots sometimes don't dissolve and instead become scar tissue, a process known as organization of the clots. That process can obstruct or narrow the pulmonary vessels. When someone has many organized clots, they are at risk for PH.

Symptoms of CTEPH can develop months to years after an acute pulmonary embolism (blood clot in the vessels of the lungs), even if a clot has been treated with blood thinners or anticoagulants. CTEPH also can develop from multiple small clots over a longer period of time without an identifiable, acute episode of pulmonary embolism.

About 25% of patients don't recall having pulmonary embolisms before their CTEPH diagnosis. Blood clots in the arms and legs called deep vein thrombosis can also cause CTEPH.

Symptoms

CTEPH is the only form of PH that potentially can be cured by surgically removing the clots.

CTEPH often is misdiagnosed because its symptoms are similar to other conditions such as asthma, COPD, heart failure or other forms of PH. Your physician should screen for CTEPH if you experience any of these symptoms.

Most common:

- > Shortness of breath with activity.
- > Decline in ability to exercise.

Less common:

- > Unusual chest discomfort (especially with deep breaths).
- > Lightheadedness with exertion.
- > Heart palpitations (irregular or strong heartbeat).
- > Coughing up blood (rare).



Risks for Developing CTEPH

Various circumstances can increase a person's risk for CTEPH, including:

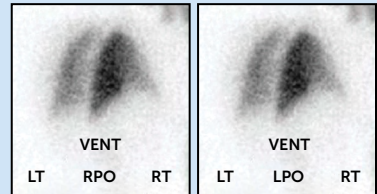
- > Catheters placed in central veins for a long time.
- > History of pacemaker-related infection.
- > Spleen removal.
- > Tendency to develop blood clots.

Diagnosing PH

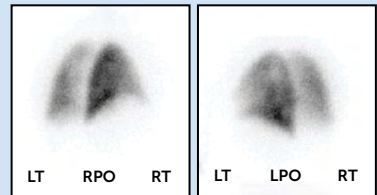
The recommended test to screen for CTEPH is a **lung ventilation-perfusion scan (V/Q scan)** to determine whether there are undissolved clots in the blood vessels of the lungs. The V/Q scan is safe, even in patients with severe PH. If there are problems with blood flow to the lungs, other diagnostic tests can confirm CTEPH and determine whether the clots can be removed. Those diagnostic tests include a CT scan of the lungs, pulmonary angiogram or a magnetic resonance imaging (MRI) of the chest.

NORMAL V/Q SCAN

VENTILATION

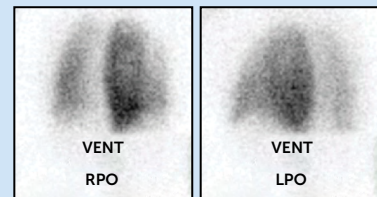


PERFUSION

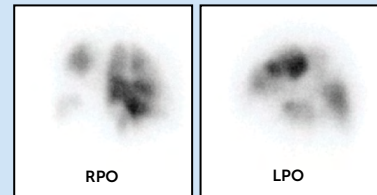


ABNORMAL V/Q SCAN

VENTILATION



PERFUSION



Images courtesy of Kelly Chin.

Treatment Options

CTEPH is the only form of PH that potentially can be cured by surgically removing the clots. An expert CTEPH team should evaluate anyone with CTEPH to determine the best treatment approach.

Pulmonary thromboendarterectomy, also referred to as pulmonary endarterectomy, is an effective approach to treat CTEPH. The procedure removes scar tissue from the pulmonary arteries and reduces PH and right heart strain for most people who undergo the procedure. PTE is a technically challenging surgery. It should be performed by an experienced medical team with expertise in managing CTEPH.

Some patients with inoperable CTEPH aren't candidates for PTE but might benefit from balloon pulmonary angioplasty. People who receive PTE but still have PH symptoms also might be candidates for BPA.

Balloon pulmonary angioplasty uses a catheter to treat blocked vessels in the heart. The catheter is inserted into a vein, usually in the neck or groin, then moved through the veins and into the blood vessels of the lungs, where the clot or scar tissue is located. A balloon at the end of the catheter is inflated for a few seconds, which pushes the scar tissue to the side of the blood vessel. The procedure can open the section of blood vessel and improve blood flow.

Additional research is needed to understand the long-term outcomes of emerging treatment options and the types of patients who would benefit most from a PTE, BPA, oral drugs or a combination of those treatments.





ADDITIONAL RESOURCES

American Thoracic Society
[Thoracic.org](https://www.thoracic.org)

American Lung Association
[Lung.org](https://www.lung.org)

REFERENCES

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Mullin CJ, Klinger JR. Heart Fail Clin. 2018; 14(3):339-351.

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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.



-  301-565-3004
-  Support Line: 800-748-7274
-  PHAssociation.org
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