PH&

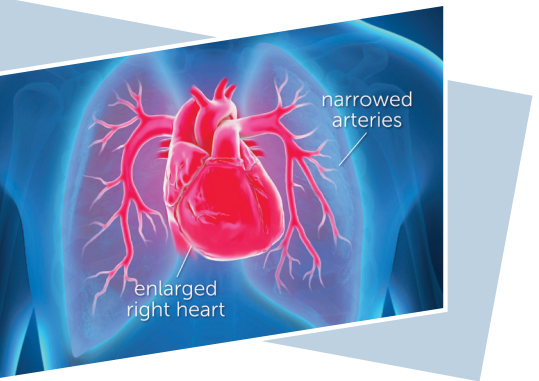
Pulmonary Hypertension & CHRONIC THROMBOEMBOLIC DISEASE

PHA Pulmonary Hypertension Association
Empowered by hope

ABOUT PULMONARY HYPERTENSION

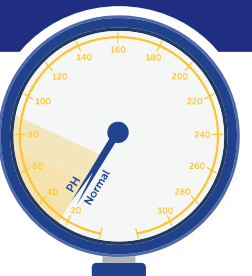
Pulmonary hypertension (PH) is a complex and often misunderstood disease. The term PH simply means high blood pressure in the arteries of the lungs. In regular hypertension — also known as high blood pressure or systemic hypertension, which you can check with a blood pressure cuff — the pressure in the 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.



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.

*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**.*



CHRONIC THROMBOEMBOLIC DISEASE AND PH

Between 0.5 and 5 percent of people who have experienced an acute pulmonary embolism (a blood clot in the lungs) may develop World Health Organization Group 4 PH (chronic thromboembolic pulmonary hypertension, or CTEPH).[1,2] This type of PH is caused by physical blockage of the pulmonary arteries by old, organized blood clots. The reasons why someone develops chronic blood clots are not known. Most who suffer an acute pulmonary embolism will naturally dissolve the clot while on blood thinners. In some patients, however, the clot will not dissolve and instead becomes scar tissue — a process known as organization of the clots, which either will obstruct or narrow the pulmonary vessels. If there is a large amount of organized clots, there is a risk of developing PH.

It is important to note that it can be months to years after an acute pulmonary embolism before symptoms of CTEPH develop, even if the clot has been treated with blood thinners or anticoagulants. It also is possible for CTEPH to develop from multiple small clots over a longer period of time without an identifiable acute episode of pulmonary embolism. The most common complaints from patients with CTEPH are shortness of breath with activity or a noticeable decline in exercise capacity. Less common symptoms include unusual chest discomfort (especially when taking a deep breath), lightheadedness with exertion, palpitations (irregular or strong heartbeat) and, rarely, coughing up blood.

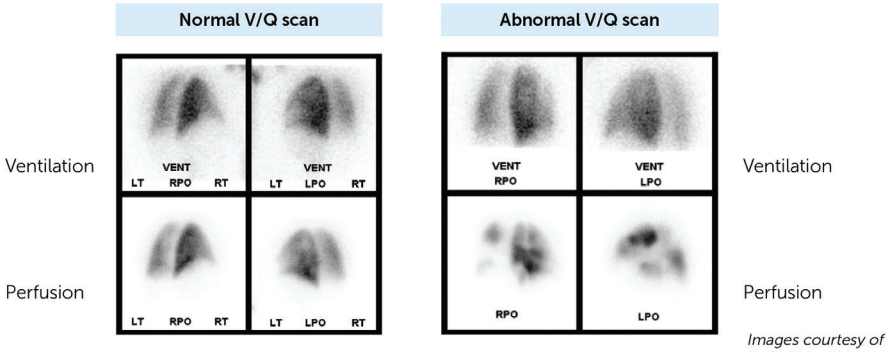
CTEPH can occur even if someone doesn't have a known history of pulmonary embolism or blood clots in the legs or arms, a condition known as deep vein thrombosis. About half of CTEPH patients do not

[1] Ende-Verhaar YM, et al. Eur Respir J. 2017;49(2).
[2] Pengo V, et al. N Engl J Med. 2004;350(22):2257-64.

remember an acute pulmonary embolism before being diagnosed with CTEPH.[3] People at increased risk of developing CTEPH include those with catheters placed in their central veins for a prolonged period of time (especially if these have been infected), patients with a history of an infected pacemaker, those who have had their spleen removed and individuals with a known clotting tendency. Health care teams should consider CTEPH if symptoms of shortness of breath with activity or exercise limitations unexpectedly become a problem. These complaints, however, are not specific to CTEPH, which is why many CTEPH patients often are misdiagnosed with other conditions such as asthma, COPD, heart failure or another form of PH.

DIAGNOSING CTEPH

The recommended test to screen for CTEPH is a lung ventilation-perfusion scan (V/Q scan). This test should be performed in all patients with PH. In it, patients get two different scans that are analyzed together. During the ventilation scan, patients breathe in a medication (benign radionuclide gas) mixed with oxygen through a mask. The scanner will take pictures as the patient breathes, and a health care provider will be able to see the sections of the lungs that are able to



Images courtesy of Kelly Chin, M.D.

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

receive air. During the perfusion scan, another medication (benign radionuclide labelled albumin) is injected into a vein in the arm. The scanner then looks at which sections of the lungs are receiving blood flow. If there is a blockage in one of the pulmonary arteries because of a blood clot, that section will not appear as dark on the image, but the ventilation scan could still look normal. The V/Q scan is safe, even in patients with severe PH. If there are problems with blood flow to the lungs, however, then other studies such as a CT scan of the lungs, a pulmonary angiogram, or a magnetic resonance imaging (MRI) of the chest will confirm the diagnosis of CTEPH and help to establish whether or not the clots can be removed.

TREATMENT OPTIONS FOR CTEPH

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

The reason why it is so important to screen for CTEPH in all patients with PH is because it is the only form of PH that can potentially be cured by surgically removing the clots. Surgery to carefully remove the scar tissue from the pulmonary arteries, called a pulmonary thromboendarterectomy (PTE) or pulmonary endarterectomy (PEA), has been shown to reduce PH and right heart strain in the majority of patients who undergo this procedure. Every patient diagnosed with CTEPH should be evaluated by an expert CTEPH team, including CTEPH physicians and PTE surgeons, to assess their candidacy for PTE, since it is the first recommended treatment. PTE is a technically challenging surgery and there are a limited number of centers within the U.S. experienced in the diagnosis and management of CTEPH.

If a patient's clots are considered inoperable, or if a patient still has some PH after the surgery, then PH-specific medical therapy has been shown to have some benefit in reducing the pulmonary pressures and improving functional status; there is medication that has received FDA approval specifically for this patient population. However, medical therapy should not be used in place of a careful evaluation for PTE by an expert center and is only recommended for patients who are not good candidates for PTE or for patients who still have PH after PTE surgery.

For some patients with inoperable disease, a procedure called balloon pulmonary angioplasty (BPA) has become available at select CTEPH centers in the U.S. in the last few years. During this procedure, a 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. This can open this section of the blood vessel up and help blood flow through better. Though frequently on PH-specific medical therapy as well, early results indicate that patients who are not candidates for surgery may benefit from this procedure. However, more research needs to be completed to understand the long-term outcomes of some of the emerging treatment options and which types of patients would benefit most from a PTE, a BPA, oral drugs or a combination.

ABOUT THE PULMONARY HYPERTENSION ASSOCIATION

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's mission is to extend and improve the lives of those affected by PH. PHA's vision is a world without PH, empowered by hope.

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.

PHA would like to thank the following member of the PH medical community for his work in the writing, review and development of this brochure:

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Cover photo courtesy of Angela Michelle.

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