Did you know that blood clots can cause pulmonary hypertension?
PULMONARY HYPERTENSION, OR PH, is complex and often misunderstood. PH means high blood pressure in the lungs. PH is different from regular hypertension. In PH, the blood vessels in the lungs become damaged and/or narrowed and the heart has to work harder to pump blood through them. PH can be caused by many different problems such as heart failure, diseases that damage the lung or multiple hardened blood clots, to name a few. It is important to understand that not all PH is the same.

Pulmonary arterial hypertension (PAH, Group 1 PH) is caused by a tightening or thickening of the walls of the pulmonary arteries, leading to elevated pulmonary arterial pressures. In patients with PAH, the right ventricle has to work harder to push blood through narrowed arteries in the lungs. Eventually, this extra stress causes the heart to enlarge and become less flexible, compromising its ability to push blood out of the right ventricle, through the lungs, and into the rest of the body, and ultimately resulting in right heart failure.

- In idiopathic pulmonary arterial hypertension, the cause of PAH is unknown.
- Heritable PAH, formerly known as familial or genetic PAH, is relatively uncommon. Of the small percentage of people who do carry the PH gene, only a small number of carriers will develop the disease.
- Associated PAH is most common. In these patients, PAH is associated with another underlying disease or condition. PAH is commonly associated with connective tissue disease (such as diffuse scleroderma, limited scleroderma [CREST syndrome] and systemic lupus erythematosus), liver disease, HIV, methamphetamine use, schistosomiasis, to name a few.
Pulmonary hypertension due to left heart disease (Group 2 PH) is believed to be the most common form of pulmonary hypertension. PH due to left heart disease can occur due to left ventricular dysfunction or valvular anemia.

Pulmonary hypertension due to chronic lung diseases and hypoxemia (Group 3 PH) occurs as a result of other, underlying lung diseases, including COPD, idiopathic pulmonary fibrosis, combined pulmonary fibrosis and emphysema, sleep disordered breathing, chronic exposure to high altitudes, and developmental abnormalities.

Chronic thromboembolic pulmonary hypertension (CTEPH, Group 4 PH) is a form of pulmonary hypertension caused by old, organized blood clots in the lungs (pulmonary emboli). In these patients, chronic clots form a physical barrier within the pulmonary arteries, leading to the onset of the pulmonary hypertension.

Pulmonary hypertension is a serious condition, and without treatment, symptoms can only become worse, leading to heart failure and even death. PH affects people of all ages and ethnic backgrounds. The most common symptoms are shortness of breath with physical activity, fatigue, lightheadedness and sometimes fainting. Because these symptoms can be caused by any number of other medical problems, diagnosis is often delayed. Identifying a case of PH can be difficult and may require a specialist. Once the type of PH is diagnosed, however, treatment can begin immediately.

Proper diagnosis and therapy from a doctor who understands PH is essential. Every patient is different. The choice of treatment is based on how sick a patient is and the risks and benefits of any particular therapy. Regardless of risk, it is important that patients and their healthcare providers engage in frequent follow-ups with ongoing discussions about the management of their condition. Current guidelines suggest that changes in therapy should be considered for patients not reaching their treatment goals.
My doctor says I have pulmonary hypertension from chronic blood clots. What does that mean?

Between 1 percent and 5 percent of people who have experienced an acute pulmonary embolism (a fresh blood clot in the lungs) may develop chronic thromboembolic pulmonary hypertension (CTEPH). This is a type of pulmonary hypertension that is caused by 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 mature into scar tissue — a process known as “organization of the clot” — which will either obstruct or narrow the pulmonary vessels. If there is a large amount of organized clot, there is the risk of developing pulmonary hypertension, which in turn can put strain on the right side of the heart.

It is important to note that it can be months to years after the acute pulmonary embolism before symptoms of CTEPH develop, even if the clot has been treated with blood thinners or anticoagulants. It is also 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 of 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), episodic lightheadedness with exertion, palpitations and, rarely, coughing up blood.

My doctor says I can’t have CTEPH because I’ve never had a pulmonary embolism or a blood clot in my leg. Is that true?

CTEPH can occur even if you don’t have a history of pulmonary embolism or blood clots in your legs or arms (deep venous thrombosis (DVT)). 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. While increased risk does not guarantee that you will develop CTEPH, your healthcare team should consider chronic thromboembolic disease if symptoms of exercise shortness-of-breath, or exercise limitations unexpectedly become a problem. You should be aware that these complaints are not specific to CTEPH, which is why many CTEPH patients are often misdiagnosed with other conditions such as asthma, COPD, or heart failure. So, if you are not responding to a treatment plan, pulmonary hypertension needs to be considered. And if you’ve been diag-
nosed with pulmonary hypertension, it is essential that CTEPH be ruled out as a possibility.

**How do doctors test for this disease?**

The recommended test to look for CTEPH is the **lung ventilation-perfusion scan (V/Q scan)**, and this test should be performed in all patients with pulmonary hypertension. This diagnostic tool tests for blood clots in the lungs by producing a picture of air and blood flow to the lungs. A small dose of radioactive material is breathed in and another small dose is injected via a blood vessel into the lungs. When your lungs are scanned, the lung vessels that are blocked with clot can be detected. If the blood flow to your lungs is normal, you don’t have CTEPH. The V/Q scan is safe, even in patients with severe pulmonary hypertension. If there are problems with blood flow to your lungs, however, then other studies such as a CT scan of the lungs, a pulmonary angiogram, or a Magnetic Resonance (MR) of the chest will confirm the diagnosis of CTEPH and help to establish whether or not the clots can be removed.

**What can be done if I’m diagnosed with CTEPH?**

The reason why it’s so important to establish whether you have CTEPH is because it can be cured in many patients. Surgery to carefully remove the scar tissue from the pulmonary arteries, called a pulmonary thromboendarterectomy (PTE)\(^1\), has been shown to alleviate the pulmonary hypertension and right heart strain in the majority of patients undergoing 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 surgery. The PTE is a technically challenging surgery, available at a limited number of centers within the United States experienced in the diagnosis and management of CTEPH. If a patient’s clots are considered inoperable, then pulmonary hypertension-targeted medical therapy has been shown to have some benefit in reducing the pulmonary pressures and improving functional status, and there is medication which 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 indicated for patients who are not good candidates for PTE or for patients who still have pulmonary hypertension after PTE surgery. If you fall into one of these categories, your PH team will work with you to choose a medication based on your disease severity, side effects and other relevant personal factors.

**Footnotes**

1. Pulmonary thromboendarterectomy (PTE) is sometimes referred to as pulmonary endarterectomy (PEA)
The Pulmonary Hypertension Association (PHA) was founded by and for PH patients. The organization has led the way in bringing pulmonary hypertension into the national and international consciousness. PHA is constantly increasing its services to the medical community through educational programming, membership sections for healthcare professionals, and much more:

**Website:**
PHA’s website is a comprehensive source of information for patients, caregivers and healthcare professionals. Please visit us at [www.PHAssociation.org](http://www.PHAssociation.org).

**Find a Doctor:**
The “Find a Doctor” section of PHA’s website allows patients and referring physicians to search for PH-treating physicians by state at: [www.PHAssociation.org/Patients/FindADoctor](http://www.PHAssociation.org/Patients/FindADoctor). While PHA does not endorse any of these physicians, PHA strongly recommends that all PH patients see a PH specialist who will be able to provide them with the best PH care.

**Pulmonary Hypertension: A Patient’s Survival Guide:**
This extraordinary 300+ page book was written by a patient and medically reviewed. It presents the illness in a very human and readable way. It is available from PHA at minimal cost, at a discount for PHA members, and one complimentary copy is provided to members of PHAs professional membership bodies. The *Survival Guide* is available for purchase online at [www.PHAssociation.org/SurvivalGuide](http://www.PHAssociation.org/SurvivalGuide).

**Online information about PH:**
For information on PH diagnosis, symptoms, treatments and more, visit [www.PHAssociation.org/Patients/AboutPH](http://www.PHAssociation.org/Patients/AboutPH).

**Support Groups:**
From the first support group started in 1991 around a kitchen table in Florida, PHA grew to 45 in 2001, and to more than 245 in 2015. In many places, patients have the opportunity to meet, learn from, and find common understanding with others in similar circumstances. Find a support group in your area at [www.PHAssociation.org/LocalSupportGroups](http://www.PHAssociation.org/LocalSupportGroups).
The mission of the Pulmonary Hypertension Association is to find ways to prevent and cure pulmonary hypertension, and to provide hope for the pulmonary hypertension community through support, education, research, advocacy and awareness. PHA’s members stand as part of a community that is fighting back against this terrible illness.

**PHA fulfills its mission through:**

- Funding for research
- Quarterly medical journal *Advances in Pulmonary Hypertension*
- *PHA Online University* offering free CME credits and the latest information on pulmonary hypertension ([www.PHAOnlineUniv.org](http://www.PHAOnlineUniv.org))
- Professional membership sections:
  - PH Clinicians and Researchers (PHCR)—for physicians and doctorate-level researchers
  - PH Professional Network (PHPN)—for nurses and allied health professionals
- Educational conferences and materials for medical professionals and patients
- A wealth of information in the *Survival Guide*
- PH patient support groups
- Quarterly magazine *Pathlight*
- Advocacy and awareness campaigns
- Toll-free Patient-to-Patient Support Line (1-800-748-7274)
- PHA’s online community for patients and caregivers at [www.MYPHAAssociation.org](http://www.MYPHAAssociation.org)

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