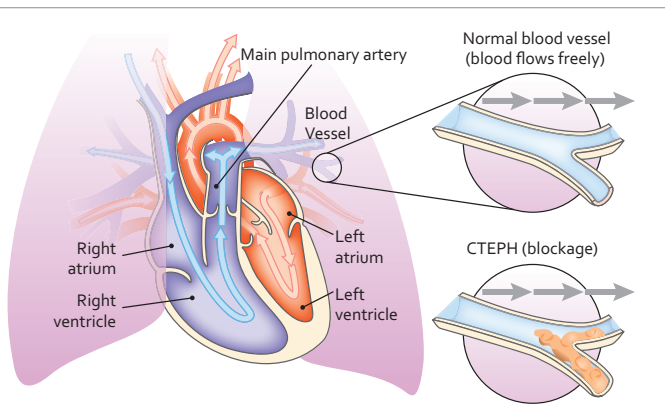


# Chronic Thromboembolic Pulmonary Hypertension

## Part 1

Chronic thromboembolic pulmonary hypertension (CTEPH) is a condition where there is elevated blood pressure in the pulmonary arteries caused by chronic blood clots (thromboembolic), which obstruct the free flow of blood through the lungs.



This is a special form of pulmonary hypertension that, unlike all the other forms, can potentially be cured with a surgical procedure. This is why it is extremely important that your health care provider makes sure that chronic blood clots are not the cause of your pulmonary hypertension.

### What Is Pulmonary Hypertension?

To understand chronic thromboembolic pulmonary hypertension, let's start by briefly reviewing what pulmonary hypertension (PH) is (see ATS Patient Series on PH at [www.thoracic.org/patients](http://www.thoracic.org/patients)). After your blood has delivered oxygen to the tissues of your body, the blood needs to come back to the lungs to get more oxygen. It does this by returning the blood to the right side of the heart, which in turn pumps the blood into your lungs. The pressure that the right side of your heart is pumping against is called your pulmonary pressure. When this pressure is too high, it is called pulmonary hypertension. The high pulmonary pressure could be caused by several medical conditions.

### What does chronic thromboembolic mean?

Thromboembolism is the medical term for blood clots. Forming blood clots is a normal defense mechanism of the body to prevent bleeding in the case of injury. However, sometimes blood clots form abnormally, typically in leg veins (so called deep venous thrombosis, or DVT), and then travel to clog the arteries in the lungs, so called pulmonary embolism (PE). With the help of blood thinners and the body's own internal clot dissolving mechanism, most pulmonary blood clots go away without any permanent damage. In a small number of people, pulmonary clots do not go away, and become scars that narrow

the size of pulmonary vessels. These clots are called chronic thromboemboli, and are the cause of increased pulmonary pressure.

### What Causes Chronic Thromboembolic Pulmonary Hypertension?

The exact cause of CTEPH is not known. One or more episodes of pulmonary embolism are the first step. Conditions that increase the chances of having blood clots include long periods of inactivity, major surgical procedures, cancer, pregnancy and the after pregnancy period, estrogen-containing oral contraceptives (birth control pills), obesity, and smoking, to name a few. Blood clots can also occur without any known cause. Conditions that seem to increase the chances of developing CTEPH include unexpected pulmonary embolism, a large PE, thyroid disease, cancer, surgical absence of the spleen, and infected pacemakers. The lupus anticoagulant/anti-phospholipid antibody syndrome is a blood coagulation disorder that is associated with CTEPH as well. CTEPH also presents without any underlying risk factor, even in people without a history of a previous blood clot in the legs or lungs. Because many patients with CTEPH have no history of PE or DVT, or do not know they had a PE, it can be overlooked or not suspected.

### What are the Symptoms of Chronic Thromboembolic Pulmonary Hypertension?

The symptoms of CTEPH are quite similar to other types of PH (see ATS Patient Information Series handout on PH at [www.thoracic.org/patients](http://www.thoracic.org/patients)). There may be no signs or symptoms of CTEPH in its early stages. You might first notice that you

become short of breath more easily. You might also notice you are more tired (fatigued) than usual. If you experience shortness of breath and/or fatigue several months after having a blood clot in the legs or lungs, your health care provider should look for CTEPH. Some patients also may feel “light headed” or even pass out. Swelling (edema) of your feet and ankles is common and may progress to swelling of your belly (ascites). Chest pain may also occur and can be mistaken for a heart attack. You may feel your heart racing or pounding (palpitations). The oxygen level in your blood may become very low, making your feet and/or fingers turn blue. Some people with CTEPH cough up blood.

### How is Chronic Thromboembolic Pulmonary Hypertension Diagnosed?

If you experience any of the above symptoms and have a history of blood clots in the legs or lungs, your health care provider should suspect CTEPH. Even without a prior history of blood clots, these symptoms should trigger a suspicion for CTEPH if they remain unexplained after basic testing. Screening for CTEPH is mandatory for everybody with pulmonary hypertension.

There are two steps to the diagnosis of CTEPH: the evaluation of pulmonary pressure, and the diagnosis of chronic pulmonary clots as the reason for the elevated pulmonary pressure.

Your health care provider will usually order an ultrasound of your heart (echocardiogram). If the echocardiogram shows the pressure on the right side of your heart may be high, they may order a cardiac catheterization. During a cardiac catheterization, a rubber tube (catheter) is placed through a blood vessel into the chambers of your heart to measure the pressure in the right side of your heart. A cardiac catheterization is the best way to measure the blood pressure in the pulmonary artery.

Your health care provider will also order tests to look for chronic pulmonary clots. The screening test of choice for CTEPH is the ventilation-perfusion scan (VQ scan). During this test, radioactive material (radioisotopes) is injected to see how well air moves through the lungs and how well the blood circulates through the lungs. A normal result from a V/Q scan means you do not have CTEPH. If the VQ scan is abnormal, you will need additional X-ray testing requiring the use of intravenous dye to confirm the diagnosis. A computed tomography (CT) pulmonary angiography is a specialized type of CT scan that shows visual images of the pulmonary arteries and gives more details about the location and extent of pulmonary blood clots. You may also have a different type of pulmonary angiography, which is done similarly to a right heart catheterization, but allows for detailed visualization of blood flow and pulmonary arteries. This test can be performed at the same time as the cardiac catheterization.

### Is There a Cure for Chronic Thromboembolic Pulmonary Hypertension?

Yes! The good news is, in most instances, CTEPH is curable with early diagnosis and surgery. This is why it is important to

recognize this disease early, and to make sure that your health care provider orders a VQ scan to look for CTEPH if you have pulmonary hypertension or unexplained shortness of breath.

### How is Chronic Thromboembolic Pulmonary Hypertension Treated?

A surgical procedure called pulmonary thromboendarterectomy (thrombow-end-arter-ectomy) (PTE, sometimes referred to as pulmonary endarterectomy, or PEA) is currently the recommended and only effective treatment for patients with CTEPH.

Although this is a cure for most people with CTEPH, some people are not good candidates for a variety of reasons. Whether you are a candidate for this treatment is determined by an expert CTEPH team. For people in whom surgery is not feasible, there are two treatment options: medical therapy to dilate pulmonary arteries, and/or a procedure called balloon pulmonary angioplasty (BPA).

Additional information about treatment of CTEPH will be provided in Part 2, Treatment of CTEPH.

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## Rx Key Points

- ✓ If you have a history of blood clots in the legs and/or lungs, and now are experiencing shortness of breath or fatigue, talk to your health care provider about CTEPH.
- ✓ If you have been diagnosed with pulmonary hypertension, make sure that your health care provider has excluded the possibility of CTEPH.
- ✓ The lung ventilation/perfusion scan (VQ scan) is the best test to look for CTEPH.
- ✓ If you have been diagnosed with CTEPH, talk to your health care provider about a referral to a specialized center. You may find CTEPH specialists at <http://www.phassociation.org/CTEPH/DoctorDirectory>.

**Healthcare Provider's Contact Number:**

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### Resources:

**Pulmonary Hypertension Association**

<http://www.phassociation.org/CTEPH>

**Pulmonary Hypertension Association Europe**

<http://www.phaeurope.org/disease-information/what-is-cteph/>

**American Thoracic Society**

<http://www.thoracic.org/patients>

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