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# CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION (CTEPH)

Chronic thromboembolic pulmonary hypertension (CTEPH) is a form of pulmonary hypertension, or high blood pressure, affecting the lungs. It is caused by blood clots.

## Learn About CTEPH

There are many causes of pulmonary hypertension, or high blood pressure, affecting the arteries of the lungs. In some patients, pulmonary embolism, or blood clots, in these arteries can form scarlike tissue that can block or narrow the arteries, leading to a form of pulmonary hypertension called CTEPH. This condition is difficult to diagnose, often leading to delayed or missed diagnoses. However, once diagnosed, there are treatment options available to patients at medical centers that specialize in this disease.

### Key Facts

- CTEPH is caused by chronic blood clots in the lungs that become scarlike tissue blocking or narrowing the pulmonary arteries.
- CTEPH can be a cause of persistent shortness of breath that is otherwise unexplained.
- Many patients can be cured with surgery. In those patients who are not candidates for surgery, medications, and other procedures can improve symptoms.

### What Is CTEPH?

After a blood clot in the lungs (called a pulmonary embolism), the majority of patients will regain normal blood flow in the affected arteries after a few months of taking blood-thinning medications. In a minority of patients (about 1 in 25) the clot becomes scarlike tissue despite the proper use of bloodthinners. As these "scars" block blood flow to the pulmonary arteries, the blood pressure increases in these vessels. The normal pressure in the pulmonary arteries is very low (about 25/10 mm Hg). The right side of the heart that pumps the blood to the lungs to get oxygen is normally thin-walled. But in CTEPH, as the pressure in the pulmonary arteries increases, the right side of the heart enlarges, and eventually begins to fail as the heart becomes overworked. This strain on the right side of the heart leads to the symptoms of CTEPH.

### How CTEPH Affects Your Body

The main complaints of patients with CTEPH are primarily due to the right heart straining to pump blood against the high pressures in the pulmonary arteries. The symptoms begin with shortness of breath, especially with exercise like climbing stairs or walking up inclines. As the disease progresses, fatigue (or feeling excessively tired) and possibly light-headedness or passing out with exercise may develop as the heart fails. The difficulty of pumping blood to the high-pressure pulmonary arteries may cause fluid to back up into the abdomen or cause the legs to swell.

### How Serious Is CTEPH?

If untreated, CTEPH is a chronic and progressive disease that leads to heart failure and can result in death. The good news is that many patients can be cured with pulmonary thromboendarterectomy (PTE) surgery. In those that are not surgical candidates, symptoms can be managed with a medication to treat pulmonary hypertension.

## CTEPH Symptoms, Causes, and Risk Factors

### What Are the Symptoms of CTEPH?

The first symptom of CTEPH is typically shortness of breath with exertion. Fatigue, a decline in exercise capabilities, palpitations, and chest heaviness with exertion can also occur. This can progress to light-headedness and even passing out with exercise. Some patients have swelling of their legs. Rarely, coughing up blood can occur.

Symptoms can start very mild and progress with time and are similar to other diseases. This makes CTEPH difficult to diagnose, especially in its early stages.

### What Causes CTEPH? What Are the Risk Factors?

CTEPH is caused by chronic blood clots in the lungs that become scarlike tissue blocking or narrowing the pulmonary vessels. About 75% of patients with CTEPH have a history of a prior blood clot in their lungs called a pulmonary embolism. Recurring blood clots may increase the risk of developing CTEPH. Some medical conditions such blood disorders, inflammatory diseases, or a history of cancer can also increase the risk of developing CTEPH.

### When to See Your Doctor

If you have had a blood clot or pulmonary embolism and are still experiencing symptoms after 3 months of treatment, you should discuss the possibility that chronic blood clots are causing your shortness of breath. In fact, any shortness of breath that is otherwise not well explained should explored with your doctor.

## Diagnosing and Treating CTEPH

### What to Expect

The diagnosis of CTEPH involves using a lung ventilation-perfusion scan and an echocardiogram (an ultrasound of the heart) to look for the disease. If these tests are normal, then CTEPH is ruled out. However, if they are abnormal, more tests may be done to confirm the diagnosis and locate the blood clots.

### How Is CTEPH Diagnosed?

A ventilation-perfusion (V/Q) scan is the next test often used to diagnose CTEPH. In this test, a radioactive protein is injected into your bloodstream to trace how blood circulates through the lungs. Blockages to blood flow in the lungs may suggest the presence of blood clots in the lungs.

To establish the diagnosis of chronic blood clots, and to accurately locate the clots, a pulmonary angiogram, CT angiography or magnetic resonance imaging (MRI) of the pulmonary arteries is necessary. For each study, contrast is injected into the pulmonary arteries so that blockages in these vessels can be identified. A physician familiar with CTEPH, often in conjunction with a surgeon experienced in the procedure, determines if these clots are in locations that allow for surgical removal. Ideally, patients should be evaluated by medical centers that specialize in the care and management of patients with pulmonary hypertension, and in particular, CTEPH.

Signs of strain on the heart caused by pulmonary hypertension may be measured using an ultrasound called an echocardiogram and a right -sided heart catheterization.

### How Is CTEPH Treated?

The definitive treatment for CTEPH is a delicate surgery called a pulmonary thromboendarterectomy (PTE), also referred to as a pulmonary endarterectomy. This surgery is done while the heart is stopped, and a heart-lung

bypass machine does the work of the heart and the lungs. The surgeon carefully removes the clots from the pulmonary arteries. This surgery if highly specialized, and should be done by medical centers with CTEPH expertise. PTE surgery is curative in over 90% of patients.

For patients who are not good candidates for PTE, either due to chronic clots that are not easily accessible to the surgeon or who have other medical conditions that make PTE too risky, there are other options. A newer procedure called balloon pulmonary angioplasty uses a tiny balloon which is inflated inside the pulmonary artery to reduce the narrowing, and open the vessel. This procedure can reduce pulmonary artery pressure and help relieve right heart strain. There are also medications for inoperable CTEPH or residual pulmonary hypertension after PTE surgery.

## Living With CTEPH

It is important to learn about CTEPH, the medications used to prevent blood clots, and the treatment options including surgery and medications to improve pulmonary hypertension.

### What to Expect

All patients with CTEPH need to be involved with their care. All CTEPH patients, including those who successfully undergo PTE surgery, should remain on blood thinners for the rest of their lives to prevent new blood clots from forming. There are many options for anticoagulation medications available and you and your doctor can discuss which option is best for you. No matter which anticoagulation medication a patient is prescribed, there are risks of bleeding. Your doctor will go through these risks prior to starting these medications.

If you are deemed to be a candidate for PTE surgery, then make sure your doctor sends you to a center with experience doing this operation. Though the needs of each individual may be different, most patients who are candidates for PTE surgery should expect to be in the hospital for about 2 weeks after surgery. The majority of patients feel their shortness of breath is improved immediately after surgery, but many have pain from the large incision over the breastbone. It takes about 2 months for the bone to heal after it is wired back together. During this time, patients should not drive, lift anything heavier than 10 pounds, or submerge the wound in water (ie, no swimming or baths).

### Managing CTEPH

As mentioned, lifelong anticoagulation is recommended to prevent worsening CTEPH from recurrent blood clots. You may be prescribed a low salt diet to help control blood pressure and the strain on your heart. Talk to your doctor about any special monitoring or changes in diet that might be required because of the blood thinner you are prescribed. Finally, exercise is important. While most patients with CTEPH have some limitations due to shortness of breath, you should be able to exercise at a low intensity like walking. Your body will tell you if you are overexerting yourself. If you develop severe shortness of breath or light-headedness, you should stop to rest.

### Finding Support

There are many resources for patients with CTEPH. The Pulmonary Hypertension Association assists patients in finding CTEPH specialists and provides access to support groups for others with CTEPH, including patients who have undergone PTE surgery.

## Questions to Ask Your Doctor About CTEPH

Making notes before your visit, as well as taking along a trusted family member or friend, can help you through the first appointment with your doctor.

- Do I need a ventilation/perfusion scan (VQ scan) to screen for chronic blood clots and an echocardiogram to screen for pulmonary hypertension?
- Do I need a heart catheterization to measure the pressures in the pulmonary arteries or other diagnostic tests to confirm the presence of chronic blood clots?
- Am I a candidate for pulmonary thromboendarterectomy (PTE)?
- What are my options for anticoagulation? What are the risks and benefits of each of these medications? How are these medications monitored?

### Questions for Surgical Candidates

- What are the risks of PTE surgery?
- What is the experience of the center and the surgeon?
- How many cases do they perform/year and what are their outcomes?
- What are the possible complications from PTE surgery and am I at increased risk for any of these complications?
- How long is the expected hospital stay and what will the recovery be like?
- Are there any alternatives to PTE surgery?
- Will I need medications for pulmonary hypertension after surgery?

### Questions for Patients Deemed Not to Be Surgical Candidates

Get a second opinion at another center to determine if you are a candidate for PTE surgery there. Because of the specialized nature and risks of the surgery, it is very common to seek a second opinion, preferably from another specialized center if possible.

### Other Questions for Your Doctor

- Am I a candidate for balloon pulmonary angioplasty?
- What medications are available to treat my pulmonary hypertension, if I am not a surgical candidate?

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