



**CHRONIC THROMBOEMBOLIC PULMONARY
HYPERTENSION & PULMONARY
THROMBOENDARTERECTOMY:
A GUIDE FOR PATIENTS AND THEIR FAMILIES**



ABOUT CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

Pulmonary hypertension is high blood pressure in the pulmonary arteries—the blood vessels that carry blood through the lungs. Blood circulation into the lungs becomes restricted and high pulmonary pressure results. The heart becomes strained making you feel short of breath and tired. Without treatment, it can lead to heart failure and death.

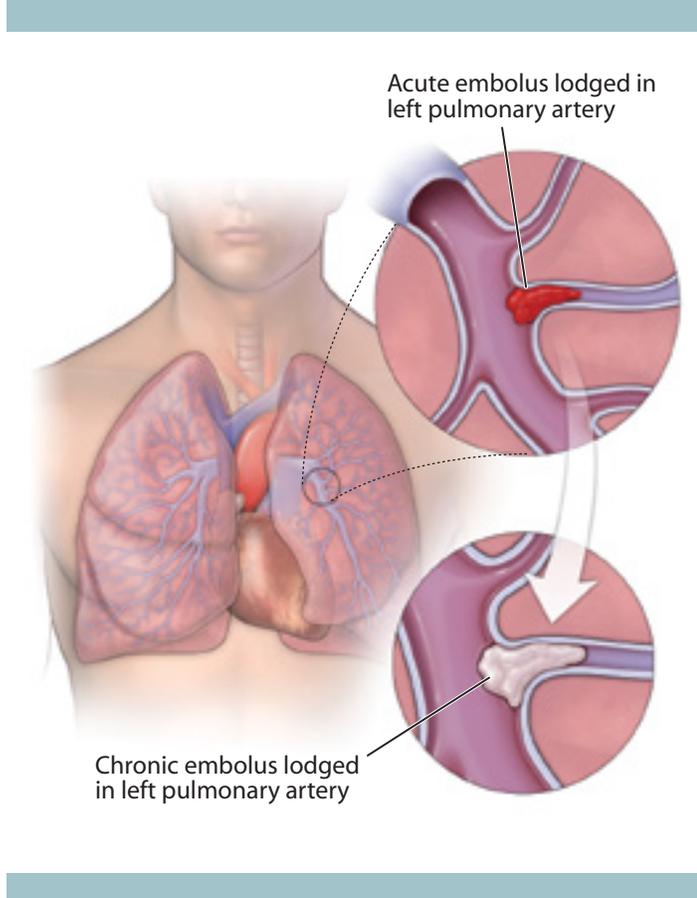
Chronic thromboembolic pulmonary hypertension (CTEPH) is a specific type of pulmonary hypertension caused by blood clots in the lung blood vessels. Other more common causes of pulmonary hypertension include chronic lung disease and chronic heart disease, such as weakened left heart or faulty heart valve.

In CTEPH, segments of the pulmonary arteries are blocked by one or more blood clots. The clots that trigger CTEPH often start elsewhere, such as in the leg, and

CTEPH CAN OFTEN BE CURED WITH SURGERY.

This complex surgery, called **pulmonary thromboendarterectomy (PTE)**, involves removal of the chronic clot from the lung vessel. The success of the surgery depends very much on a careful pre-op evaluation, selection, and preparation of patients. Surgical expertise is critical, as is the need for expert care after surgery. PTE is sometimes called pulmonary endarterectomy or PEA.

travel through the veins and heart to become lodged in the lung. This type of clot in the lung is called a pulmonary embolism. (“Thrombo” refers to a blood clot, so a “thromboembolism” is a clot that has traveled from somewhere else.) If these clots do not resolve over time with blood thinners, CTEPH can occur.



CTEPH begins when a clot lodges in a pulmonary artery. Over time, the clot totally or partially blocks blood flow through the lungs. As the heart works harder to overcome this blockage, it becomes enlarged and can fail.

Many patients have a more common type of temporary lung clot that dissolves on its own or with treatment. This is called an acute pulmonary embolism.

But when a clot does not go away, it becomes “chronic.” Over time, a chronic pulmonary thromboembolus extends within the artery, clings to the artery wall, and resembles scar tissue. This complex fibrous clot also triggers narrowing of other blood vessels throughout the lung. These chronic clots increase pulmonary pressure and they reduce blood through the lungs, leading to the array of symptoms and serious complications associated with CTEPH.

Although CTEPH symptoms are similar to those of other types of pulmonary hypertension, the treatment is quite different. That’s why it is critical that patients be evaluated at a center specializing in CTEPH.



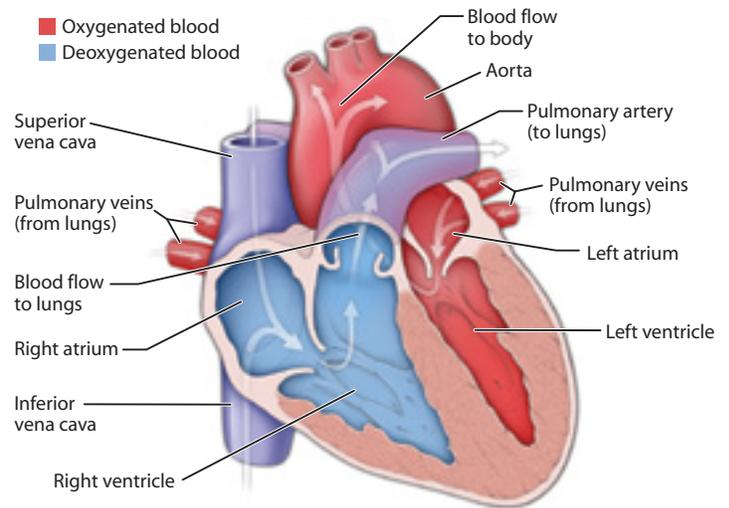
IMPORTANT FACTS ABOUT CTEPH AND PTE

- Some people have no symptoms of pulmonary hypertension until it is well advanced.
- CTEPH may be a more common form of pulmonary hypertension than once believed, owed to relative under-diagnosis of the condition.
- Pulmonary thromboendarterectomy (PTE) surgery is potentially curative for CTEPH. Only a few select hospitals in the U.S. have a high volume CTEPH/PTE Program with excellent outcomes.
- Patients with CTEPH who have PTE surgery live longer than those who only have treatment with medications.
- Not all patients with CTEPH can be helped by PTE—but all patients with CTEPH should be considered potentially operable and curable until they have been thoroughly examined by an experienced CTEPH team.
- For patients who cannot be helped by PTE, other treatments—including medications and balloon pulmonary angioplasty (BPA)—may be offered here at Temple.

A CLOSER LOOK AT THE PULMONARY ARTERIES

Your heart works like a pump with two outlets. The left side of the heart takes blood from the lungs and pumps it through the aorta out to the rest of the body—from head to toes. After this blood has delivered oxygen to the tissues throughout the body, it returns to the right side of the heart, which then pumps it back to the lungs to pick up more oxygen and start the whole cycle again.

The left pumping chamber (left ventricle) is bigger and the left-sided blood pressure (systemic) is naturally higher. The right side only needs to pump into the lungs, so the right ventricle is smaller and pulmonary blood pressure is naturally lower.



NORMAL BLOOD PRESSURE

Systemic

120/80 mmHg

Pulmonary

25/10 mmHg

Each blood pressure measurement has two numbers. The first “systolic” number is the peak pressure, measured right after the heart’s ventricles contract. The second “diastolic” number is the resting pressure, measured in between heartbeats. The mmHg stands for millimeters of mercury.

PULMONARY HYPERTENSION

Pulmonary Artery Pressure

Higher than 40 mmHg

Systemic blood pressure is measured with a blood pressure cuff. Pulmonary artery pressure can be measured directly with a catheter test or indirectly with an echocardiogram imaging test.



CAUSES OF CTEPH

CTEPH happens when clots block the lung arteries. Clots attach to the vessel lining and can become “fibrotic” (scarred or inflamed). The clots can grow larger over time. Often the clots indirectly lead to narrowing of smaller arteries throughout the lung (“arteriopathy” or “vasculopathy”)—which makes the pulmonary hypertension even worse and harder to treat.

Although many patients with CTEPH have had a recent episode of acute pulmonary embolism or a deep venous thrombosis (a blood clot in the leg), as many as half have no known history of such clotting problems.

SYMPTOMS

Symptoms of CTEPH are similar to those for other types of pulmonary hypertension. These symptoms are subtle in the early disease stages and may not show up for months or even years. As the disease progresses, symptoms become more noticeable. They are mainly due to the diminished oxygen supply, the weakened heart, and the increased lung pressures.

The most common symptom is **shortness of breath on exertion**.

Other symptoms may include:

- Lightheadedness or fainting
- Fatigue
- Chest pain
- Swelling of ankles or abdomen
- Bluish lips and skin (cyanosis)
- Racing pulse
- Palpitations (a strong feeling of a fast heartbeat)

As the disease advances, people become less active and begin to have symptoms even when resting. Eventually, extreme breathlessness occurs and patients may become completely bedridden and require hospitalization.

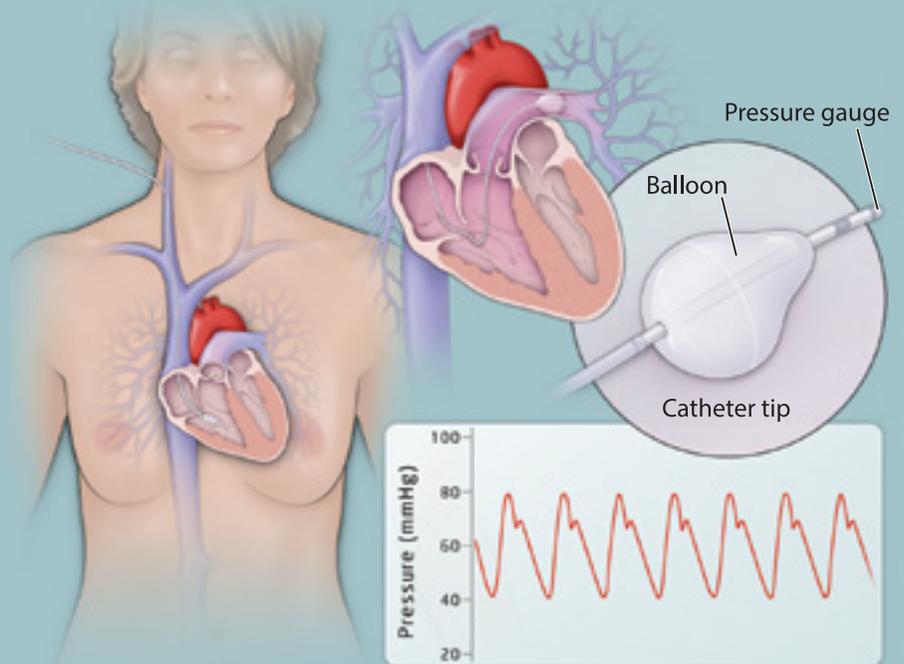


DIAGNOSIS

- **Echocardiography** is the most common way to estimate the pulmonary blood pressure. This noninvasive test uses sound waves (ultrasound) to create a moving picture of the heart. This “echo” also reveals any enlargement of the right side of the heart and helps doctors figure out the underlying cause of the pulmonary hypertension (for example, whether it is caused by a lung vessel problem or a heart problem).
- **Right heart catheterization** is the gold standard for confirming pulmonary hypertension and measuring its severity. This test provides a direct measurement of blood pressure. It is done by passing a catheter through a vein in the leg or neck into the right side of the heart to measure the blood pressure in the main vessel leading to the lung.

CARDIAC CATHETERIZATION

A catheter (long thin tube) is inserted into a vein in the neck, leg, or arm and is guided toward the pulmonary artery. With a blood pressure gauge at the catheter tip, the doctor measures blood pressure in the right atrium, the right ventricle, and within the pulmonary artery. The catheter can also detect oxygen levels (“saturation”) in the veins, heart, and arteries. It may also test patient response to treatments that relax the pulmonary arteries.





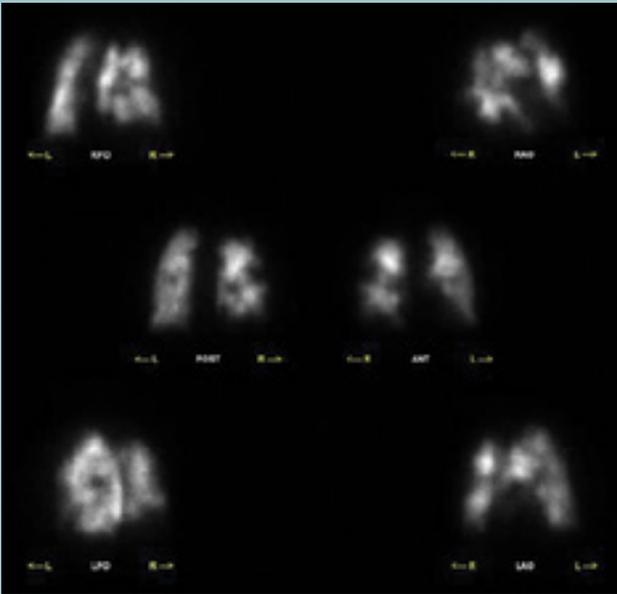
When CTEPH is suspected, other tests include:

RADIONUCLIDE VENTILATION/PERFUSION LUNG SCAN (V/Q SCAN): This important screening test for CTEPH provides a picture of air and blood circulation in the lungs. After radiation-labeled substances are injected and inhaled, V/Q images reveal any areas of the lung not receiving blood flow due to a clot (“perfusion defects”).

CT SCAN: Computed tomography with contrast can also show blood clots and image the lung tissue.

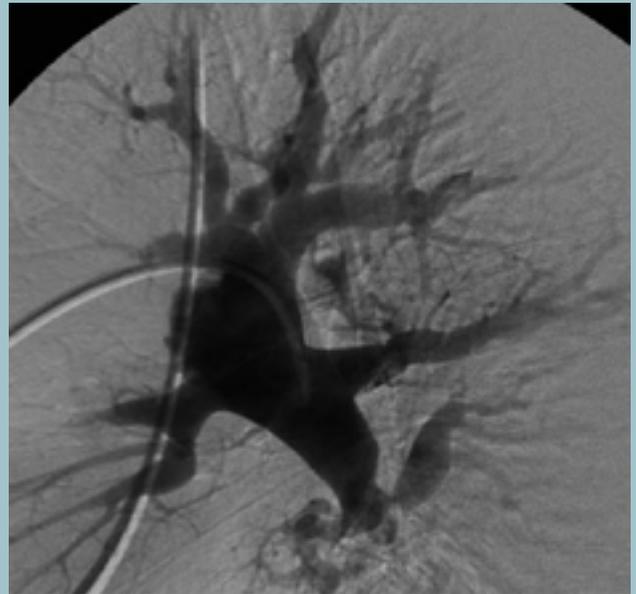
PULMONARY ANGIOGRAPHY: A catheter is inserted to reach the pulmonary artery (as in right heart catheterization) and then a dye is injected into the arteries of both lungs. This provides a very clear picture of a clot’s size and location. The result of angiography plus catheter pressure measurements allow the CTEPH team to plan for potential PTE surgery performed after referral to an experienced PTE center.

V/Q Scan



Abnormal perfusion portion of a V/Q scan showing multiple severe perfusion defects (clots).

Pulmonary Angiogram



The injected dye shows blocked pulmonary artery segments typical of CTEPH and allows the CTEPH team to assess and plan for PTE surgery.



TREATMENT OPTIONS

Many treatments are available for pulmonary hypertension. The choice of therapy depends very much on the specific type of pulmonary hypertension—its underlying causes, its severity, and the patient's other medical conditions.

For eligible patients with CTEPH, the therapy of choice is pulmonary endarterectomy (PTE).

This major surgery removes the old blood clots from the lung vessel. (Note that PTE is different and much more complex than surgery to remove just an acute pulmonary embolism (called a pulmonary embolectomy). To prevent new clots from forming, all patients with CTEPH need anticoagulation therapy (usually warfarin) beginning before surgery and continuing for the rest of their lives.

PTE can either completely reverse the PH, or lead to very dramatic improvement. Studies show that those with CTEPH who have PTE survive much longer than those who receive only medications. Those who have PTE also often avoid the need for long-term use of complex medications or supplemental oxygen and they have a better quality of life.

DO I QUALIFY FOR PULMONARY THROMBOENDARTERECTOMY (PTE)?

Your CTEPH treatment team will talk with you and perform specialized tests to determine if PTE could help you. You may qualify for PTE surgery if:

- Your clots are accessible to the surgeon in large- and medium-sized pulmonary arteries.
- You do not have other medical conditions that increase your surgical risk or limit your chances of success; although advanced age and conditions such as kidney or liver disease may slightly increase your surgical risk, they do not automatically disqualify you from surgery—talk to your CTEPH treatment team.

Patients who are truly inoperable may benefit from medicinal therapy or balloon pulmonary angioplasty to treat their PH.



HOW PULMONARY ENDARTERECTOMY (PTE) IS DONE

Pulmonary endarterectomy requires an incision through the breastbone (median sternotomy) and cardiopulmonary bypass (heart-lung machine). Your body will also be cooled and, for brief periods, your heart stopped to protect your organs and prevent excessive bleeding (this is called deep hypothermic circulatory arrest). In most cases, multiple clots will be removed from the lungs.

The surgeon opens the pulmonary artery and carefully removes the chronic clot. Often, the clot plug comes out looking like a mold of the lung anatomy. The artery is closed after all major blockages are removed.

The entire surgery can take anywhere from 4 to 8 hours.

WHAT TO EXPECT AFTER SURGERY

You will recover in the ICU initially after surgery and spend approximately a week in the hospital. Repeat pulmonary pressure measurements and echocardiography assist the CTEPH team in monitoring the early success of the surgery. Full recovery at home will take several weeks.

You will likely notice improvements in breathing within the first two weeks after surgery. Improvement often continues over the first two months following surgery. Overall, quality of life is much improved in most patients. You will need to take blood thinners for the rest of your life.

Some patients will continue to have some degree of pulmonary hypertension after PTE. These patients may require medications to dilate the smaller lung blood vessels that are still elevating the pulmonary pressure.



Extensive chronic clot removed from both lungs.

WHAT IF I CANNOT HAVE PTE?

For non-operable patients with severe CTEPH, other options may be available. Vasodilating drugs may reduce the pulmonary pressure and help your ability to exercise.

In certain situations, your team may suggest a balloon angioplasty to open up the blocked pulmonary artery.



WHY CHOOSE TEMPLE HEART & VASCULAR INSTITUTE?

Temple's cardiovascular specialists are experts in diagnostic, medical, surgical and interventional techniques. The Temple Pulmonary Hypertension, Right Heart Failure & CTEPH/PTE Program sees hundreds of patients with pulmonary arterial hypertension or CTEPH every year and we are one of the few U.S. heart centers with extensive experience in CTEPH and PTE.

From specialized diagnostic techniques, such as exercise cardiac catheterization, to technically complex operations, such as PTE or BPA, the temple PH and CTEPH team has a long record of accomplishment in improving long-term outcomes and quality of life in CTEPH patients.

The Temple Pulmonary Hypertension, Right Heart Failure & CTEPH/PTE Program is nationally recognized for helping patients with all types and subtypes of this disease. Our large multidisciplinary team includes nationally renowned cardiologists specialized in PH, cardiothoracic surgeons, interventional cardiologists, intensive care physicians, transplant physicians, lung pathologists, rehabilitation specialists, social workers, and many expert nurse practitioners.

Our team emphasizes comprehensive and supportive care throughout the entire process. From your first visit to Temple to the end of your recovery process, you will always feel our commitment to outstanding care.

The CTEPH team is available for immediate evaluations and second opinions. You can meet face-to-face with our team to discuss your suitability for PTE. If surgery is needed, they can tell you about the risks and benefits of your specific procedure.

To schedule an appointment with one of our CTEPH specialists, or for more information, call 800-TEMPLE-MED.