Chronic Thromboembolic Pulmonary Hypertension

Chronic thromboembolic pulmonary hypertension (CTEPH) is a condition that causes high blood pressure in the arteries (blood vessels) in the lungs. A pulmonary embolus (PE) is a blood clot in the lung. CTEPH is caused by PEs that are new or do not get better and go away. These blood clots often come from blood clots in the legs that travel to the lung.

Figure 1. Blood vessels in the lungs

If you have any questions, please ask your physician or nurse.

When caught early and treated with blood thinning (anticoagulant) medications, the body tends to get rid of PEs on its own within 3 to 6 months. However, sometimes these clots do not go away and may clog the arteries in the lungs. This causes high blood pressure in these arteries. When this happens, the heart may work harder to pump blood through the lungs. Over time, the right side of the heart will start to enlarge and weaken.
Symptoms
CTEPH may cause:
- Shortness of breath during exercise
- Dizziness
- Swelling in the legs or abdomen
- Chest pain
- Fainting

Testing
It is important to learn the cause of pulmonary hypertension to find the right treatment. Your care team may diagnose CTEPH based on symptoms that do not go away after an acute PE. In some cases, CTEPH is diagnosed without any history of a PE. Since symptoms of CTEPH are similar to symptoms of other conditions, your physician may do the following tests. This lets them make the right diagnosis.

Echocardiogram: This test is an ultrasound of the heart. It can show the size and shape of the heart chambers, how well the valves work and how the blood moves inside heart.

Right heart catheterization: This is often an outpatient procedure. A clinician inserts a catheter (thin tube) into a large vein (usually in the neck). Then, they guide it into the chambers of the heart. This will allow the physician to measure the blood pressure inside the heart. This is the most accurate test for CTEPH.

Pulmonary angiogram: This test is often done at the same time as the right heart catheterization. A clinician will inject contrast dye into the small catheter. Then, they take X-rays of the lungs. This shows the location, size, shape and number of clots.

Ventilation-perfusion (VQ) lung scan: This is a 2-part test. During the ventilation part of the test, you will inhale a gas through a mask. At the same time, your clinician will take pictures of the lungs. This will show the airflow in all areas of the lungs. During the perfusion part of the test, you will receive a special injection through an IV (into the vein) line. This will show how well blood moves through the arteries in the lungs.

Computed tomography (CT) scan: This test uses X-ray images and computer processing to show images of the lungs. It can spot blockages in the arteries of the lungs. It can also show the health of the lung tissue.

Treatment
CTEPH can lead to complications if left untreated. These can include heart failure and pulmonary hypertension. However, you can manage CTEPH with proper treatment. Your physician will discuss the right treatment for you. Treatment may include:

Medications
Anticoagulant medications help prevent new blood clots from forming in the lungs. These medications also get rid of clots in other parts of the body.
The physician may also prescribe medications to reduce the blood pressure within pulmonary arteries.

**Pulmonary thromboendarterectomy (PTE) surgery**
PTE is a type of surgery to remove blood clots in the lungs and restore normal blood flow. Your surgeon does it through an incision in the chest. Removing the clots in surgery may cure CTEPH. But, not everyone is a candidate for this surgery.

**Balloon angioplasty (BPA)**
This is a minimally invasive, catheter-based procedure. BPA moves blood clots out of the way to help restore normal blood flow to the lung.

**Northwestern Medicine Bluhm Cardiovascular Institute CTEPH Program**
We are committed to improving the quality of life in our patients through our quest for excellence in compassionate and innovative healthcare. Patients with CTEPH have access to our multidisciplinary CTEPH program. This is the largest program of its kind in Illinois. We are also one of the few programs in the nation to offer both of the leading treatment options for CTEPH:

- Pulmonary thromboendarterectomy surgery (PTE)
- Balloon pulmonary angioplasty (BPA).

Our goal is to advance healthcare and achieve better health outcomes. Our team is on the forefront of research. They have published several peer-reviewed articles. We are also enrolling patients in ongoing clinical research studies and trials.

**To make an appointment**
Please call the Northwestern Medicine CTEPH Program at 312.695.4965 (TTY: 711). For your first appointment, we must have your most recent physician visit notes and images of your test results on a disc. These may include:

- Demographics sheet
- Health insurance card
- History and physical
- Consultation notes (pulmonary hypertension specialist, cardiologist, pulmonologist, hematologist)
- Laboratory results
- Chest CT/CT angiogram (report and images)
- VQ scan (report and images)
- Chest X-ray (report and images)
- Pulmonary function test (report)
- Echocardiogram (report and images)
- Stress test (report)
- Right and/or left heart catheterizations (reports)
- Coronary and/or pulmonary angiograms (reports and images)
If you have had these tests, please request the results from your primary care physician. Please send reports and images by FedEx delivery services to:

Northwestern Medicine Bluhm Cardiovascular Institute CTEPH Program
Galter Pavilion
675 North Saint Clair Street
Suite 19-100
Chicago, Illinois 60611

Contact information
To learn more, please go to northwesternmedicine.nm.org/CTEPH

If you have any questions, contact us at:
Phone: 312.695.4965 (TTY: 711)
Fax: 312.926.3668
Email: pulmhypertension_rn@nm.org