What is Chronic Thromboembolic Pulmonary Hypertension (CTEPH)?

Information for patients and families

Read this booklet to learn:

- what CTEPH is
- about the Toronto CTEPH Program
- how CTEPH is treated
- who to call if you have any questions
What is CTEPH?

Chronic – a condition that last a long time (months to years)

Thromboembolic – blood clots (thrombi) travel from your arms and or legs through the veins into the blood vessels of the lungs and block the pulmonary arteries

Pulmonary – related to lungs

Hypertension – high blood pressure inside blood vessels

CTEPH is 1 type of pulmonary hypertension. It is caused by repeated blood clots or blood clots that don’t dissolve in the arteries of your lungs. These blood clots are called pulmonary emboli or PE.

Half of patients with CTEPH don’t have symptoms or have never been diagnosed with pulmonary emboli. It is not well understood why some people are able to dissolve the pulmonary emboli inside their pulmonary arteries and others go on to develop chronic clot and scar tissue, even while taking blood thinners.

Knowing what happens when blood flows through lungs normally can help you understand what happens when you have CTEPH.

Normal blood flow through your lungs
(see picture of Healthy heart and lungs on page 4)

Your heart and lungs work together to move blood easily through your lungs:

1. The blood that flows from the veins in your arms, legs and other body parts to your heart is low in oxygen and high in carbon dioxide. It flows into the right side of your heart.

2. The blood flows through your right atrium into your right ventricle.

3. Your right ventricle fills with blood and pumps the blood through your pulmonary arteries and through the millions of blood vessels in your lungs.

4. As the blood moves through your pulmonary arteries, it picks up oxygen and gets rid of the carbon dioxide.
5. When the blood is full of oxygen, it flows into the left atrium of your heart and into your left ventricle.

6. Your left ventricle pumps this blood to the rest of your body.

These steps happen every time your heart beats.

**What happens when you have CTEPH:**
(see picture of CTEPH on page 4)

- The blood that flows through your lungs contains blood clots that travel from veins in legs or arms. These blood clots become stuck in the pulmonary arteries, causing them to become blocked or narrowed.

- Over time, these clots become scar tissue (also called webs) inside your pulmonary arteries.

- This scar tissue restricts the blood flow through your pulmonary arteries. This causes the pressure inside your lung vessels to increase (pulmonary hypertension).

- The right side of your heart has to work harder to move blood through your narrowed pulmonary arteries.

- Over time, blood from your pulmonary arteries backs up in the right side of your heart.

- As the blood backs up, your right ventricle becomes larger, and its muscle becomes thicker (right-sided heart failure). This leads to swelling in your abdomen and ankles from water retention (fluid build-up).

- Without treatment, most patients with CTEPH die within 5 years.
Healthy heart and lungs

1. Right atrium (RA)
2. Right ventricle (RV)
3. Pulmonary artery
4. Left atrium (LA)
5. Left ventricle (LV)
6. Aorta

Chronic thromboembolic pulmonary hypertension (CTEPH)

Scar tissue
Blocked blood vessels
Right side of the heart has too much pressure
Narrowed blood vessels
What can increase the chances of having CTEPH?

- large blood clots (pulmonary emboli) or repeated pulmonary emboli in your pulmonary arteries
- presence of pulmonary emboli with no other risk factors (also called idiopathic pulmonary emboli)
- surgery to remove your spleen
- health problems that cause inflammation, such as osteomyelitis (bone inflammation) or inflammatory bowel disease

What are the symptoms of CTEPH?

At first, people with CTEPH do not have any symptoms. As the disease gets worse, most people will feel:

- short of breath, especially when they are active
- very tired
- a tightness or pain in their chest or abdomen
- light headed

Who should be referred to the Toronto CTEPH Program?

Anybody with shortness of breath and an abnormal ventilation-perfusion scan (V/Q scan) should be referred to the Toronto CTEPH Program.

A V/Q scan is a medical test that looks at how air and blood move through your lungs. This test can be done in any hospital. The result of this test is often the first step to finding out if you have CTEPH.

How do you know for sure whether or not I have CTEPH?

We have you do many more medical tests. These tests include:

- Echocardiogram (ECHO) – an ultrasound of your heart
- Electrocardiogram (EKG or ECG) – a recording of the electrical activity of your heart
How is CTEPH treated?

CTEPH is the only type of pulmonary hypertension that is curable. Pulmonary Thromboendarterectomy (PEA) surgery is the best treatment for CTEPH. All patients who come to the Toronto CTEPH Clinic are evaluated for PEA surgery.

During your clinic visit, the thoracic surgeon and his team will check your health and look at all of your test results. This will help them decide if you are a candidate for PEA surgery.

The goals of PEA surgery are to:

- improve your breathing and reduce or remove your need for extra oxygen (if you are using it)
- bring the blood flow inside your lung vessels back to normal
- decrease how hard the right side of your heart has to work and prevent heart failure
- make it possible for you to return to your normal activities or exercises
You can find more information about PEA surgery in the patient brochure:

**Pulmonary Thromboendarterectomy Surgery: Information for patients with CTEPH and their families**

If surgery is not the best option for you, we will refer you to the Pulmonary Hypertension Program. The health care team there can work with you to decide the best treatment.

**Who do I call if I have questions or need more information?**

**Toronto CTEPH Program**
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