

CTEPH - Group 4 Pulmonary Hypertension

What is Group 4 pulmonary hypertension (CTEPH)?

Chronic thromboembolic pulmonary hypertension (CTEPH) is a type of pulmonary hypertension (Group 4 PH), which is high blood pressure in the lungs. CTEPH 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 (PE). After a pulmonary embolism, up to 4% of patients may develop CTEPH within two years. CTEPH is the only type of PH that may be curable.

Here is a breakdown of the name:

- 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 the lungs
- Hypertension - high blood pressure inside blood vessels.

What causes CTEPH?

About 75% of patients diagnosed with chronic thromboembolic pulmonary hypertension have had one or more blood clots in the lungs. Other risk factors include:

- Having a high risk for blood clots
- Chronic inflammatory diseases, such as osteomyelitis or inflammatory bowel disease
- Having your spleen removed
- Thyroid replacement therapy
- Cancer
- Family history of blood clots
- Being female
- Blood clotting disorders (hypercoagulability)

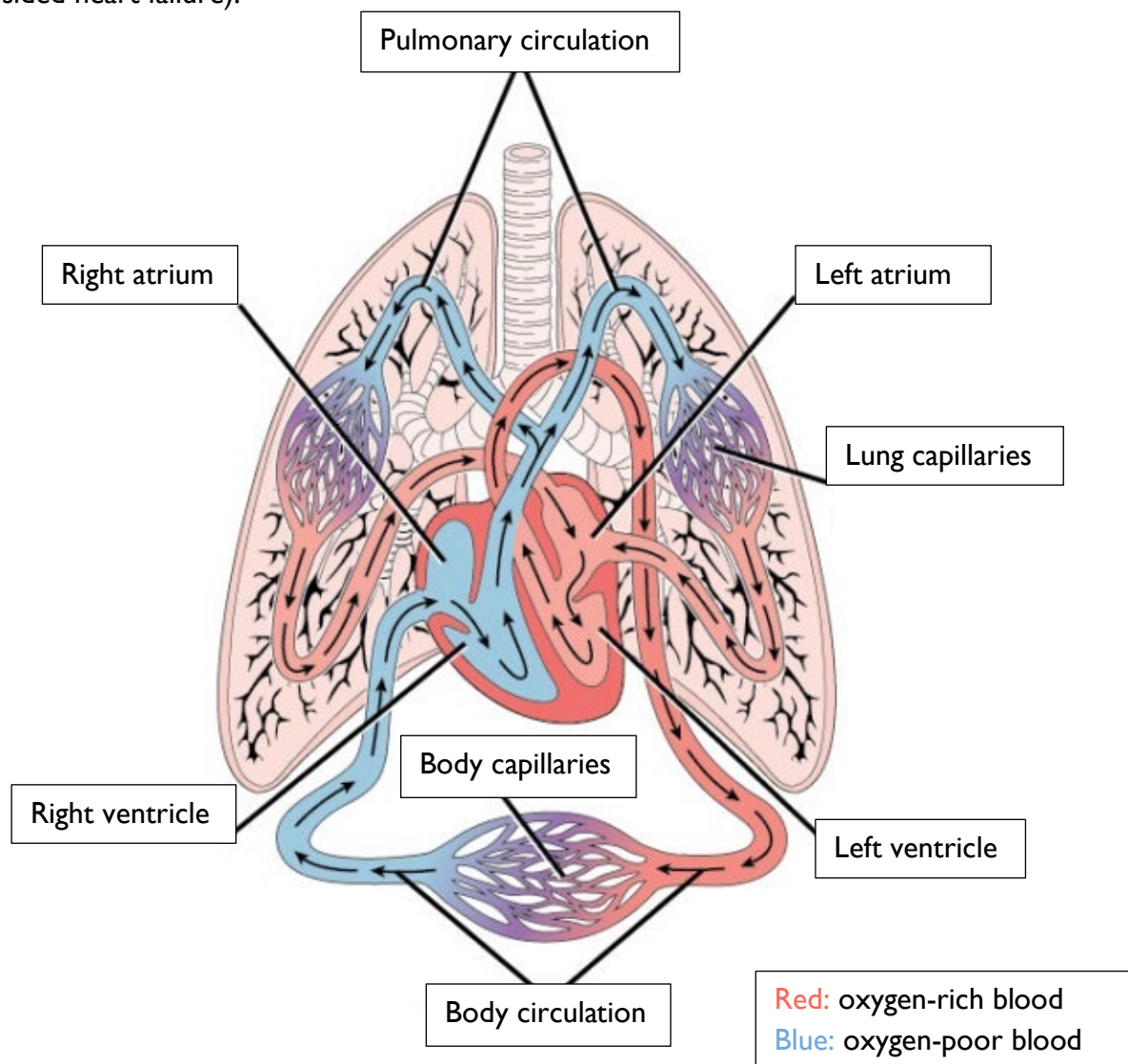
It is not well understood why some people are able to dissolve the pulmonary emboli inside their pulmonary arteries and others go on to develop chronic clot and scar tissue, even while taking blood thinners.



What happens when you have CTEPH?

In CTEPH, the blood that flows through your lungs contains blood clots that travel from veins in your legs or arms. These blood clots get stuck in the pulmonary arteries, blocking or narrowing them. The clots become scar tissue (also called webs), which 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).



Diagnosis

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

At a CTEPH clinic, you will do more medical tests. These tests may include:

- **Echocardiogram (ECHO):** an ultrasound of your heart.
- **Electrocardiogram (EKG or ECG):** a recording of the electrical activity of your heart.
- **CT scan** of the chest: a special type of x-ray that looks at your lungs and blood vessels.
- **Pulmonary angiogram:** intravenous (IV) dye is injected into your vein and travels to the vessels of your lungs. The dye helps us see blood flow and check for any blockages.
- **Femoral and carotid Doppler:** an ultrasound of your leg and neck arteries.
- **Pulmonary function test (PFT):** breathing tests.
- **Right heart catheterization:** a flexible tube (catheter) is put into your body through a vein in your groin or wrist to measure the pressures of the right side of your heart and pressure in the pulmonary artery/lungs.
- **Left heart catheterization:** a catheter is inserted through an artery, usually wrist or groin, and dye is injected to look for blockages in the arteries of the heart.
- **Exercise right heart catheterization:** the same as a regular right heart catheterization except they will exercise you while you are lying back to see whether the pressure in your pulmonary artery/lungs increases with exercise. This is done when you do not have pulmonary hypertension at rest, but experience shortness of breath with any activity. This is currently only done in Toronto at the CTEPH surgical centre. Some of the referring provinces are working on offering this test, but it requires a cardiologist with specialty training to interpret and conduct.

These tests confirm whether you have CTEPH and help your health care team decide if surgery is the best treatment for you.



Treatment

Surgery

Pulmonary thromboendarterectomy (PEA) is the treatment of choice for CTEPH. The surgery leads to major clinical improvements and can be curative in many patients. Most patients experience significant functional recovery and can return to their baseline physical activity level without supplemental oxygen.

CTEPH patients are evaluated for PEA surgery at a CTEPH clinic. During your clinic visit, the thoracic surgeon and their team will check your health and look at all your test results. This will help them decide if you are a candidate for pulmonary thromboendarterectomy.

Other treatments

If PEA surgery is not the best option for you, the CTEPH team will assess you to find other treatment options. These may include:

Medication

The pulmonary hypertension medication riociguat (Adempas®) is approved in Canada for the treatment of non-operative or residual post-operative CTEPH.

Balloon pulmonary angioplasty

Balloon pulmonary angioplasty (BPA) is a procedure that is done by an interventional radiologist, in which a catheter is inserted into your blood vessels and a balloon is inflated to open up the vessel and increase blood flow throughout your lungs. BPA improves the blood flow in your lungs to help improve your shortness of breath, prevent the right side of your heart from getting weaker, and decrease your pulmonary hypertension.

Oxygen therapy

Patients may require supplemental oxygen to maintain adequate oxygen levels. Typically, the target is an oxygen saturation above 90%, but this will vary depending on the condition.

Cardiopulmonary rehabilitation

Cardiopulmonary rehabilitation can help symptoms and increase exercise capacity for people with many types of pulmonary hypertension.

Transplant

Depending on the severity of pulmonary hypertension, heart or lung transplantation might also be an option.

Pulmonary circulation image credit

Rye C, Wise R, Jurukovski V, DeSaix J, Choi J, & Avissar Y. (2016). *Biology*. OpenStax. https://openstax.org/books/biology/pages/40-1-overview-of-the-circulatory-system#fig-ch40_01_03 used under a Creative Commons Attribution license





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