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Chronic Thromboembolic Pulmonary Hypertension: Curable Type of Pulmonary Hypertension



Anastasia Bykova is the Nurse Practitioner at the Toronto CTEPH Program, Division of Thoracic Surgery, Toronto General Hospital, who follows patients diagnosed with CTEPH before and after their pulmonary endarterectomy surgery. Besides her clinical practice, Anastasia is actively involved in patient and health professional education about CTEPH, raising awareness of the diagnosis, and academic work on this topic.

Chronic thromboembolic pulmonary hypertension (CTEPH) is a type of pulmonary hypertension (PH) that is caused by chronic blood clots inside the arteries of the lungs. CTEPH is the only type of PH that is curable by a surgical procedure called pulmonary endarterectomy (PEA). The complex disease process in CTEPH poses a challenge for a newly diagnosed patient and their family to understand what is happening inside the patient's body. Therefore, I would like to talk about how CTEPH develops and how it is treated.

DIFFERENCE BETWEEN ACUTE AND CHRONIC PULMONARY EMBOLI

When a blood clot forms inside a deep vein of the legs, it can break off, travelling to the heart and settling inside the arteries of the lungs. This process is called **acute pulmonary emboli (acute PE)** (see image *Acute Pulmonary Emboli*). Our bodies have the natural ability to dissolve blood clots, and people who are diagnosed with PE will be treated with anticoagulation medications or blood thinners to help their bodies get rid of these clots. The bodies of most patients are able to dissolve pulmonary emboli by the end of treatment with anticoagulation med-

ication, with no long-term consequences to their health. However, in 3-5% of cases, the blood clots do not dissolve despite the anticoagulation treatment and create fibrous scar tissue inside the pulmonary arteries. This process is called **chronic pulmonary emboli** (see image *Chronic Pulmonary Emboli*). It is still unknown why some people are able to dissolve blood clots while others are not.



Acute Pulmonary Emboli (Acute PE)

Fresh blood clot tissue removed from the pulmonary arteries during the surgery called thrombectomy.

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Chronic Pulmonary Emboli

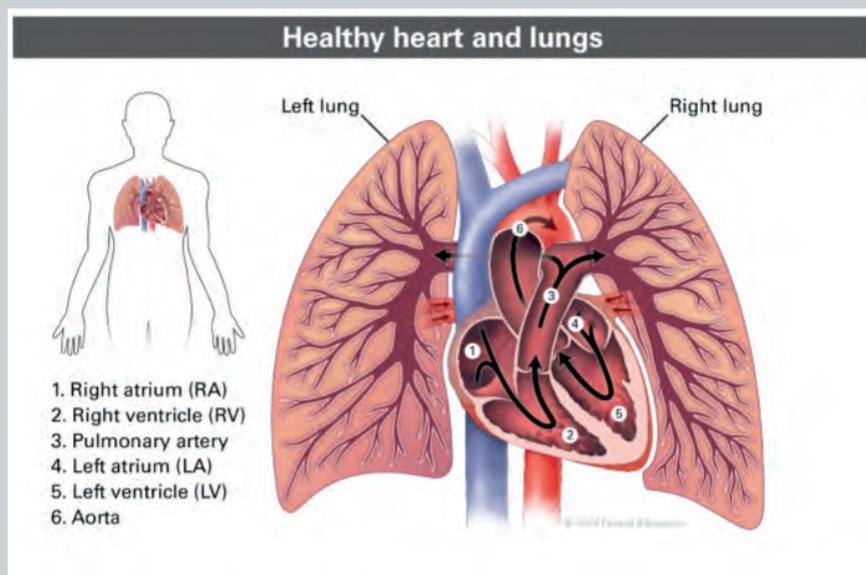
Fresh blood clot transforms into fibrous scar tissue that lines the pulmonary arteries in CTEPH. Chronic pulmonary emboli removed from right and left lungs during the surgery called pulmonary endarterectomy.

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HEALTHY HEART AND LUNGS

In order to understand what happens to the body in CTEPH, let's look first at the anatomy and function of healthy heart and lungs. The blood rich in carbon dioxide travels from the entire body, including arms, legs, and head, to return to the right atrium of the heart (see image *Healthy Heart and Lungs*). From the right atrium, the blood travels to the right ventricle and fills it up until the right ventricle contracts and pumps the blood into the pulmonary arteries. While travelling in the pulmonary arteries, the blood gives away carbon dioxide and receives oxygen. Oxygen-rich blood then returns to the heart through the left atrium and fills up the left ventricle. When the left ventricle is full and contracts, the oxygen-rich blood is pumped out to the aorta and to the rest of the body.

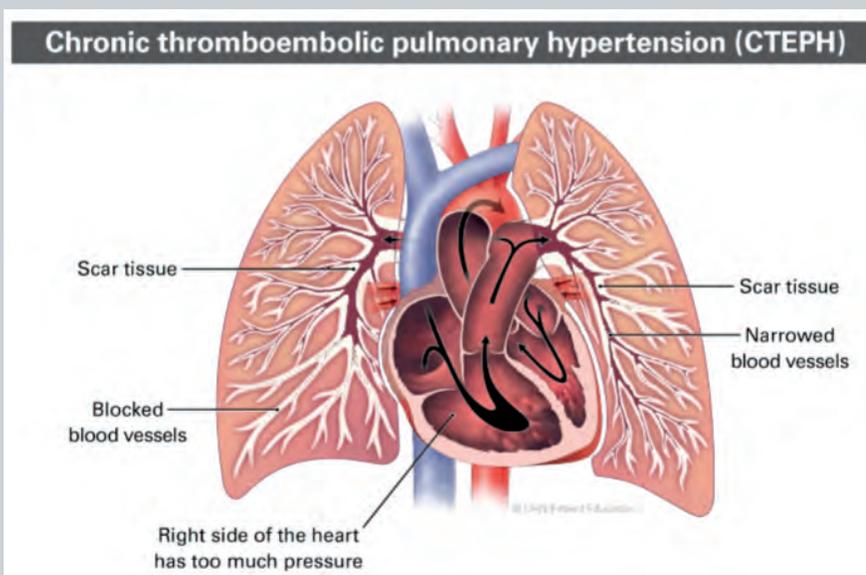


Healthy Heart and Lungs

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HEART AND LUNGS IN CTEPH

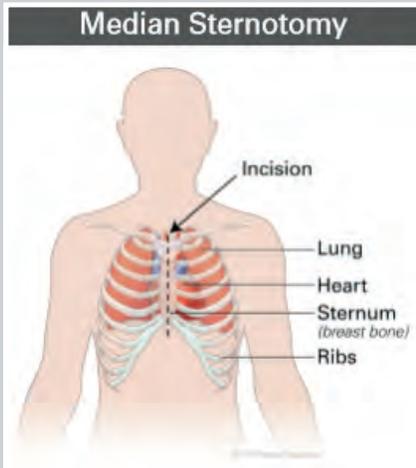
In the lungs affected by CTEPH, chronic blood clots inside the pulmonary arteries create scar tissue leading to the narrowing of the pulmonary arteries and physical obstruction of the blood flow (see image *Chronic Thromboembolic Pulmonary Hypertension*). As a result, the pressure in the lungs increases, causing PH. With the elevated pressure inside the pulmonary arteries, the right ventricle must generate more force to pump blood against blocked blood vessels. Over time, the right ventricle suffers from the high pressure it has to generate to push blood forward, which leads to thicker walls and a dilated chamber. This is called **right heart failure**. As the right side of the heart struggles to push blood forward, the fluid accumulates in the right ventricle and atrium, leading to back flow into the other parts of the body such as legs and abdomen.



Chronic Thromboembolic Pulmonary Hypertension (CTEPH)

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Median Sternotomy

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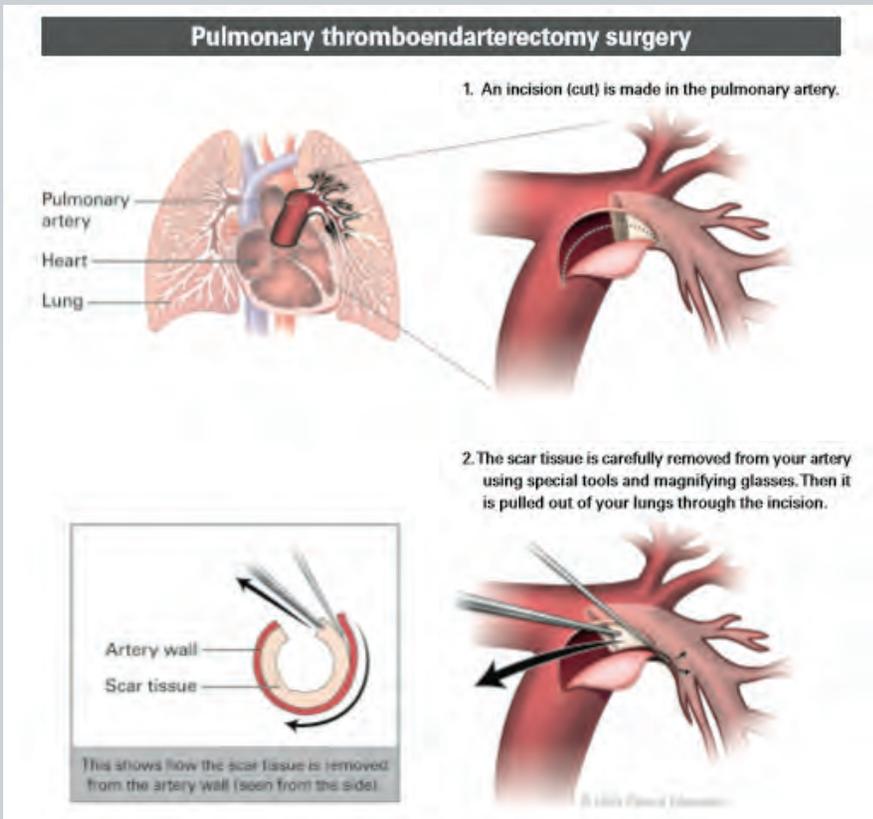
Initially, the majority of patients with CTEPH have non-specific symptoms of shortness of breath upon exertion, and fatigue from the chronic blood clots and PH. As the disease progresses, patients find that shortness of breath becomes worse, limiting their daily activities (i.e. exercising, grocery shopping, cleaning the house, mowing the lawn). Other symptoms of advanced disease are syncope, chest pain, and swelling of the legs and abdomen. Without treatment, the survival rate of a patient with CTEPH is poor.

TREATMENTS FOR CTEPH

CTEPH is unique in the way that it is the only type of PH that is potentially curable by a surgery called **pulmonary endarterectomy (PEA)**. During PEA surgery, a surgeon makes a cut through the sternum (breast bone) to expose the heart, major blood vessels including the

pulmonary artery, and parts of the lungs (see *Median Sternotomy* image). The patient goes onto the heart-lung machine and is cooled down to 18-20°C to slow down the metabolism of vital organs. The surgeon then makes a cut in the pulmonary artery to open it up (see *PEA Surgery* image). The fibrous scar tissue is located inside the pulmonary arteries and is carefully removed by the surgeon using special instruments and magnifying glasses. When all chronic clots that the surgeon can find are removed, the cut in the artery wall is closed together with sutures. The goals of PEA surgery are: to cure PH; to treat shortness of breath and improve quality of life so that the patient can return to a physically active lifestyle; and to prevent right heart failure. Most patients who wake up after the surgery report that their breathing is much better and that their shortness of breath has gone away.

Medical therapy with PH-specific medications is sometimes used to treat a select group of patients with CTEPH. Riociguat® is the only medication that has been approved by Health Canada for the treatment of CTEPH when patients are not surgical candidates for PEA or for patients with residual/recurrent PH following PEA surgery. The goal of medical therapy is to slow down the disease progression. Patients who are diagnosed with CTEPH often have significant physical limitations due to shortness of breath, causing poor quality of life. Without surgery, CTEPH is life-threatening. In the hands of an experienced CTEPH team, most patients following PEA surgery are cured of their disease, and are able to breathe normally and enjoy an active lifestyle.



Pulmonary Thomboendarterectomy Surgery

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