

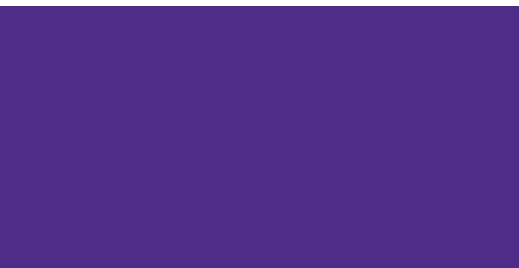


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C T E P H

Chronic Thromboembolic Pulmonary Hypertension

The Patient Journey with Chronic Thromboembolic Pulmonary Hypertension (CTEPH)



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THE BEGINNING: ONSET OF SYMPTOMS

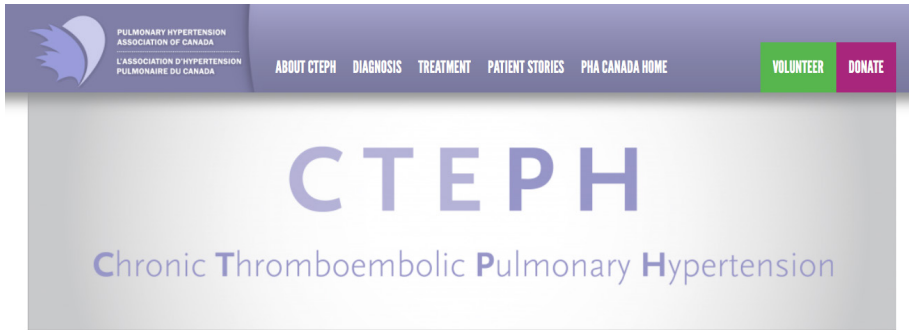
CTEPH remains an underdiagnosed disease in which chronic pulmonary emboli (blood clots) obstruct the pulmonary arteries. This process leads to elevation of blood pressure in the pulmonary arteries, causing pulmonary hypertension (PH) and stress on the right side of the heart. Initially, patients develop shortness of breath (SOB), which is mainly noticeable when navigating inclined surfaces (i.e. climbing stairs, going uphill) or with strenuous physical activity (i.e. gym, sports, mowing the lawn). As the disease

In the Spring 2016 issue of Connections, I published an article explaining how chronic thromboembolic pulmonary hypertension (CTEPH) develops and is treated. In this follow-up article, I will talk about the patient experience of being diagnosed with CTEPH and treated through pulmonary endarterectomy (PEA surgery). Given the complexity of the disease process and the time it takes for an accurate diagnosis of CTEPH to be established, patients and their families share a lengthy and emotionally challenging journey.

progresses, patients experience fatigue and breathlessness even when travelling on flat surfaces (i.e. walking in a mall or on a street). Patients will commonly presume that they are “getting out of shape” to explain these limitations. In reality, it is the underlying CTEPH that limits their exercise ability and impacts their fitness level. In an effort to get in shape, many people try to engage in physical activity while still experiencing progressive SOB and fatigue. Eventually, they will give up their efforts and over time, adapt to these symptoms by doing less exercise and adopting a more sedentary lifestyle. This adaptation process leads to weight gain and physical deconditioning, which in turn exacerbates symptoms of SOB and fatigue. With time, some people may develop additional symptoms such as chest pain, fainting, and swelling of the legs and abdomen, and may require home oxygen to maintain their oxygen level. This vicious cycle may go on until an accurate diagnosis is established.

CTEPH DIAGNOSIS AND TREATMENT

CTEPH symptoms are non-specific and can be associated with a number of more common medical conditions. As a result, the majority of CTEPH patients will be seen by multiple specialists before a definitive diagnosis is established. Patients and their families generally find the process of going from one specialist to another in search of a final diagnosis to be frustrating. The average time from the onset of symptoms to a CTEPH diagnosis is 14 months, which demonstrates the difficulty of establishing an accurate diagnosis. The process may go as follows: a respirologist may treat a patient whose CTEPH has not yet been diagnosed with antibiotics and “puffers.” When the symptoms do not improve, the patient will be referred to a cardiologist for more investigations. Eventually, a PH program will evaluate the patient and confirm a CTEPH diagnosis. Importantly, whether CTEPH is confirmed or suspected, all patients should be assessed



In collaboration with the University Health Network and the Toronto CTEPH clinic, we have published new educational resources on our redesigned CTEPH website. Visit www.phacanada.ca/cteph to access new information and resources.

How is CTEPH Treated?

There is a potential cure for CTEPH through pulmonary endarterectomy (PEA) surgery. Approved medical treatment may also slow disease progression and alleviate symptoms.

Medical Therapy

- » Riociguat (Adempas®) – approved in Canada in 2013 for treatment of non-operative or residual post-operative CTEPH (WHO class IV)

Surgical Treatment

The standard and potentially curative treatment for patients who have developed CTEPH and are suitable for surgery is a procedure called pulmonary endarterectomy (PEA). PEA is major surgery that clears the blood vessels of the lungs of clots and scar material presented in the disease. To determine whether you are a suitable candidate for surgery, you need to be thoroughly assessed at a centre specialized in the treatment of CTEPH. CTEPH specialists will assess:

- » If the blockage is reachable through surgery - if the blockage is too deep within the lungs, it may not be reachable;

for potentially curative pulmonary endarterectomy (PEA surgery) by a program specialized in the treatment of CTEPH. Early CTEPH diagnosis and referral for surgical evaluation are crucial to improve post-operative recovery and increase the chance of a cure.

BEFORE AND AFTER PULMONARY ENDARTERECTOMY (PEA SURGERY)

PEA surgery is a gold standard treatment for CTEPH. In Canada, expertise in PEA surgery is regionalized, which allows the formation of strong, multidisciplinary teams of experts who can provide highly specialized patient care. For many patients and their families, this means travelling to Toronto or Ottawa to receive a potentially curative treatment.

Before Pulmonary Endarterectomy Surgery

The Toronto CTEPH Program is the largest Pulmonary Endarterectomy Program in Canada and gets referrals from across the country. Once a CTEPH diagnosis has been confirmed and PEA surgery in Toronto is planned, the majority of patients and their families feel overwhelmed. The most common reasons are: 1) the disease process and surgical procedure are complicated to understand; and 2) the trip to Toronto takes a lot of preparation. It is important to let CTEPH patients and their families know that they are not alone in this process, and that they can reach out to their CTEPH team for guidance. The key is for the family members to help a patient prepare for

the trip by working collaboratively with the CTEPH team. Educational materials that the medical team will provide on CTEPH diagnosis and PEA surgery can alleviate some anxiety for patients and their families. An information package with clear instructions and a checklist will be sent out to ensure that patients and family members know how to prepare ahead of time. Some of the items patients and families need to plan for include: 1) identifying a support person who will accompany the patient home after their hospital discharge; 2) securing provincial health coverage/travel and accommodation assistance (if applicable); and 3) setting up home oxygen for the flight and stay in Toronto. The more prepared the patient and family will be, the less overwhelmed and stressed they will feel away from their home.

After Pulmonary Endarterectomy Surgery

The majority of patients report an immediate and ongoing improvement of SOB following PEA surgery. If the chronic clots were in the smaller pulmonary arteries, causing less SOB symptoms, the breathing will improve at a slower rate over the next few months. Families may notice initially that some patients demonstrate subtle personality changes, confusion, or even inability to focus (i.e. taking 20 minutes to write an email that would have taken two minutes to write before PEA). These mental changes are temporary and will resolve within the first few weeks as the patient recovers. One third of the patients will require supplemental home oxygen following PEA sur-

gery, even if they were not on oxygen before. This is not a reason to be alarmed that the surgery did not work! The newly unobstructed pulmonary arteries will relearn to control the amount of blood flow to match the air moving in and out of the lungs and patients will be weaned off home oxygen within a few weeks or months (depending on how sick they were before surgery).

The key is to be patient with the body and give it the necessary time to heal. As the body heals, the patient will notice significant improvement in breathing, resolution of fatigue, increased ability to be active and exercise, and sharper mental function due to the increased amount of oxygen that is available to the brain. Going through PEA surgery is a challenging process, but with proper preparation and support from a dedicated surgical team, patients and their families can confidently embark on this journey to significant quality of life improvements and a potential cure of CTEPH.

For more information on CTEPH, including contact information of clinics specialized in the treatment of CTEPH in Canada, and to read Anastasia Bykova's article, "CTEPH: Curable Type of Pulmonary Hypertension" (Connections, Spring 2016. Vol. 7, No. 1), visit www.phacanada.ca/cteph.

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