



Frequently Asked Questions

A diagnosis of pulmonary hypertension (PH) is life-changing and can significantly impact all aspects of your life. At first, you may feel overwhelmed by all the information you receive, but as you go along you will learn more about PH, become familiar with the language used by your medical team, and develop strategies to adjust to your new life. Here are answers to a few frequently asked questions to get you started.

What causes pulmonary hypertension?

People of all backgrounds, ages, and genders can develop PH. A small number of patients develop PH without any known cause; this type of PH is known as idiopathic pulmonary arterial hypertension (IPAH).

Pre-existing conditions can lead to the development of PH including heart disease, connective tissue disease, liver disease, HIV infection, and other lung diseases. People with a history of pulmonary emboli (blood clots in the lungs) are at greater risk of developing a type of PH called chronic thromboembolic pulmonary hypertension (CTEPH). Family history and the use of certain drugs (e.g. anorexigens) can also lead to the development of PH.

Could PH run in my family?

Yes, but it is unlikely. This type of PH, known as hereditary pulmonary arterial hypertension (HPAH), is very rare. It is caused by a mutation in genes (most commonly the BMP-R2 gene), but only about 1 in 5 people who inherit the mutated gene develop HPAH.

How does my doctor know I have PH?

PH can be difficult to diagnose because it shares many of its symptoms with other more common conditions and requires specialized tests to be confirmed. When a doctor suspects PH, they may order a series of preliminary tests to rule out other conditions before referring a patient to a specialist. These tests include: an echocardiogram, blood tests, chest X-rays, an electrocardiogram, and pulmonary function tests.

A specialist will be able to confirm a PH diagnosis after conducting one or more of these tests: exercise tolerance tests, CT scanning/imaging, ventilation/perfusion lung scan, and right heart catheterization.

To learn more about how these tests are conducted and what they reveal, visit www.livingwithph.ca.

Why is it important to see a PH specialist?

PH is a relatively rare disease and most physicians have little or no experience with its diagnosis and treatment. For this reason, referral to a centre specialized in the treatment of PH is encouraged in order to confirm diagnosis and ensure that patients receive the best care. Further, some provinces require patients to be followed by a specialist in order to receive coverage for PH-specific medications.

Centres specialized in the treatment of PH (adult and pediatric) and CTEPH are located throughout Canada. Their experienced medical teams include PH nurses and specialists such as cardiologists, respirologists, and rheumatologists.

To find the PH treatment centre nearest you, visit www.phacanada.ca/clinical-directory.

How is PH treated?

Pulmonary arterial hypertension (PAH) is treated using medical therapies approved by Health Canada. While these treatments are not a cure for PH, they can slow disease progression and alleviate symptoms. There is a potential cure for CTEPH through a surgical procedure called pulmonary endarterectomy (PEA). Approved medical treatment may also slow disease progression and alleviate symptoms in patients not eligible for PEA.

For more information on current PH treatments, visit www.phacanada.ca/learn-more.

How will my doctor determine which treatment is best for me?

The tests performed to diagnose you will allow your doctor to determine which type of PH you have and the severity of your illness. They will consider factors such as your functional class (a classification based on how PH impacts your everyday life), the pressures in your lungs, and how well your heart is functioning to determine which treatment is best suited to your needs. Your medical team will work with you to establish a personalized treatment plan and adjust it as your symptoms and lung pressures change.

How long will I live with PH?

There is no single answer for this question because each person is different. Your experience with the disease depends on many factors, including how severe your PH is at the time of diagnosis and how you respond to treatments. PH is a progressive and potentially fatal disease, but thanks to available treatments many patients are living longer and healthier lives. In the last twenty years, research has made great strides and specialists are better equipped than ever to provide patients with the best care and improve their quality of life.

How will PH affect my life?

Right after diagnosis it is hard to imagine what life with PH will be like. Over time, many people develop strategies to cope with the challenges of living with PH and lead full, happy lives. The key is to listen to your body, respect your limits, and rest when needed. Making small adjustments to your home, routine, and daily activities will allow you to conserve energy while remaining active. Support from other people who understand what it means to live with PH and who have developed strategies to cope with the disease can also be crucial.

To access resources and connect with members of the PH community, visit www.phacanada.ca.

Should I get a medical alert bracelet?

Medical alert bracelets communicate important information to health care providers in an emergency. However, not all people with PH need to wear one. A medical alert bracelet can be useful if you are on advanced therapies for PH (such as intravenous medication) or anticoagulation medication. Speak to your medical team about whether or not you should wear a medical alert bracelet and what information it should provide.

Where can I find more information?

www.phacanada.ca

Web portal for the PH community providing resources for patients and caregivers, research news, and information on community events.

- Information and resources for CTEPH patients: www.phacanada.ca/cteph
- Information and resources for families of children with PH: www.phacanada.ca/pediatrics

www.livingwithph.ca

Educational website providing comprehensive information to help patients and caregivers better understand PH, including its causes, diagnosis, and treatment.

For more information, contact PHA Canada at:

info@phacanada.ca | 1-877-774-2226